GEORG-AUGUST-UNIVERSITÄT GÖTTINGEN IN PUBLICA COMMODA SEIT 1737

UNIVERSITÄTSMEDIZIN GÖTTINGEN UMG

From symptoms to slide: Decoding diagnostic dilemmas

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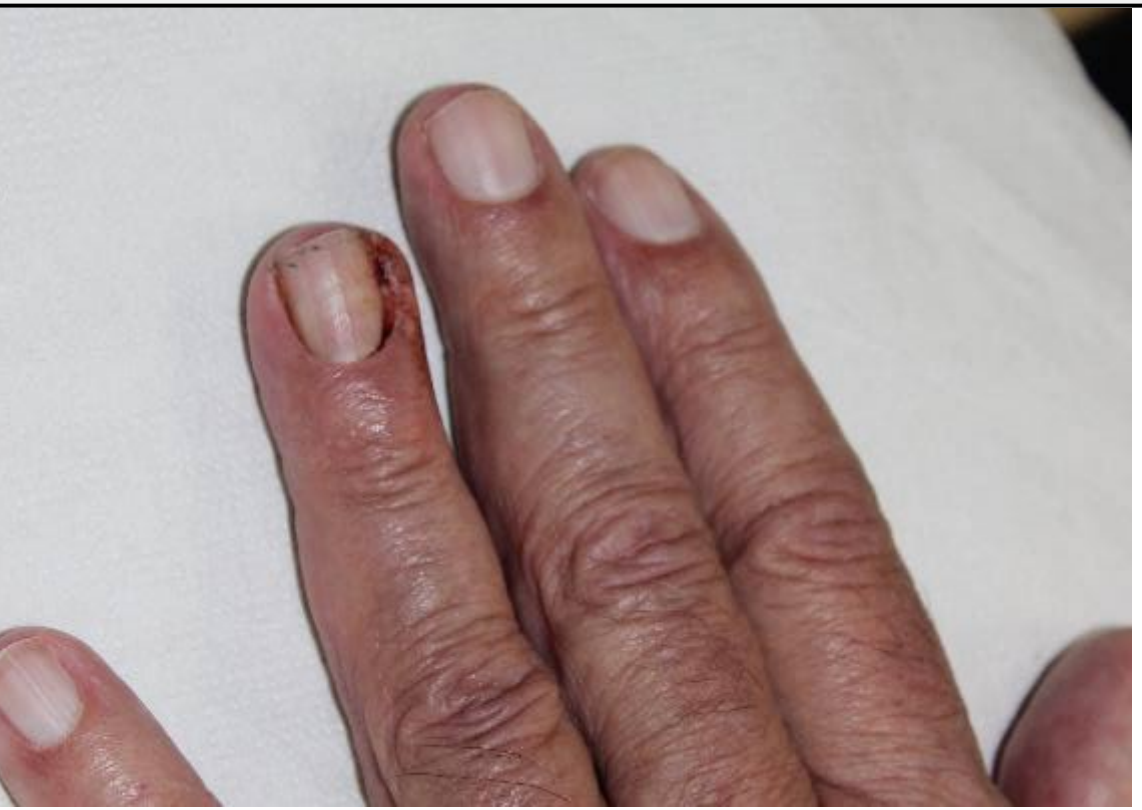
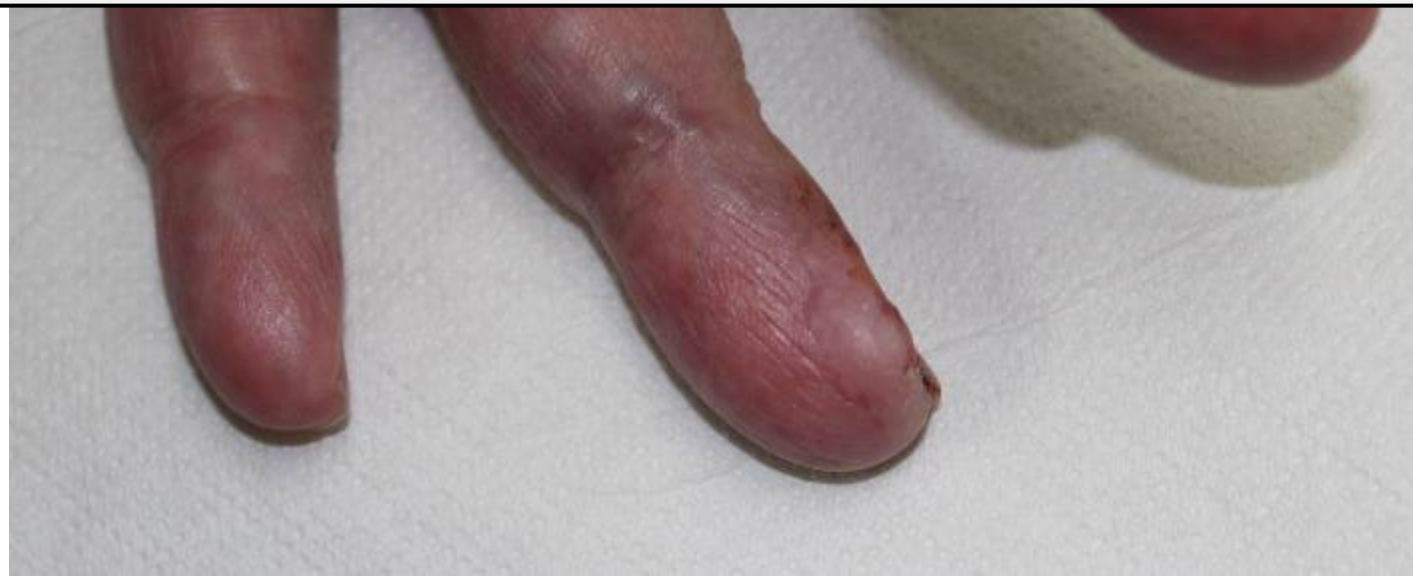
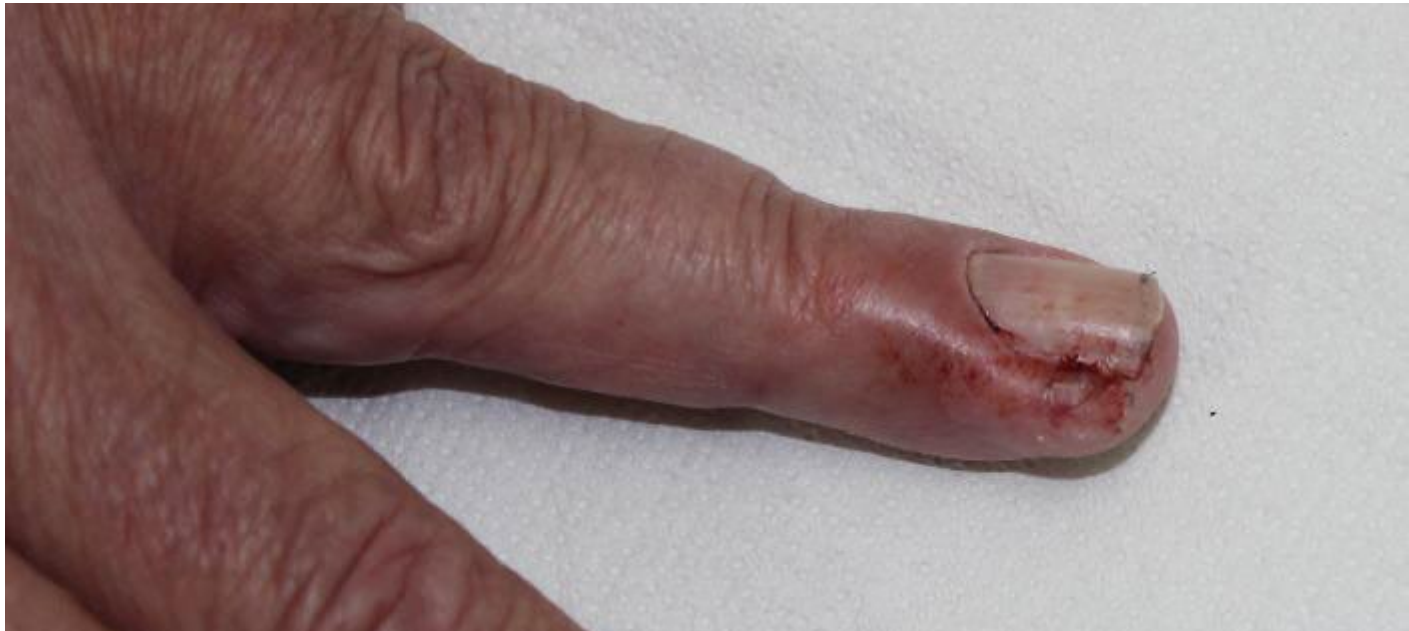
Case 1

Clinical presentation:

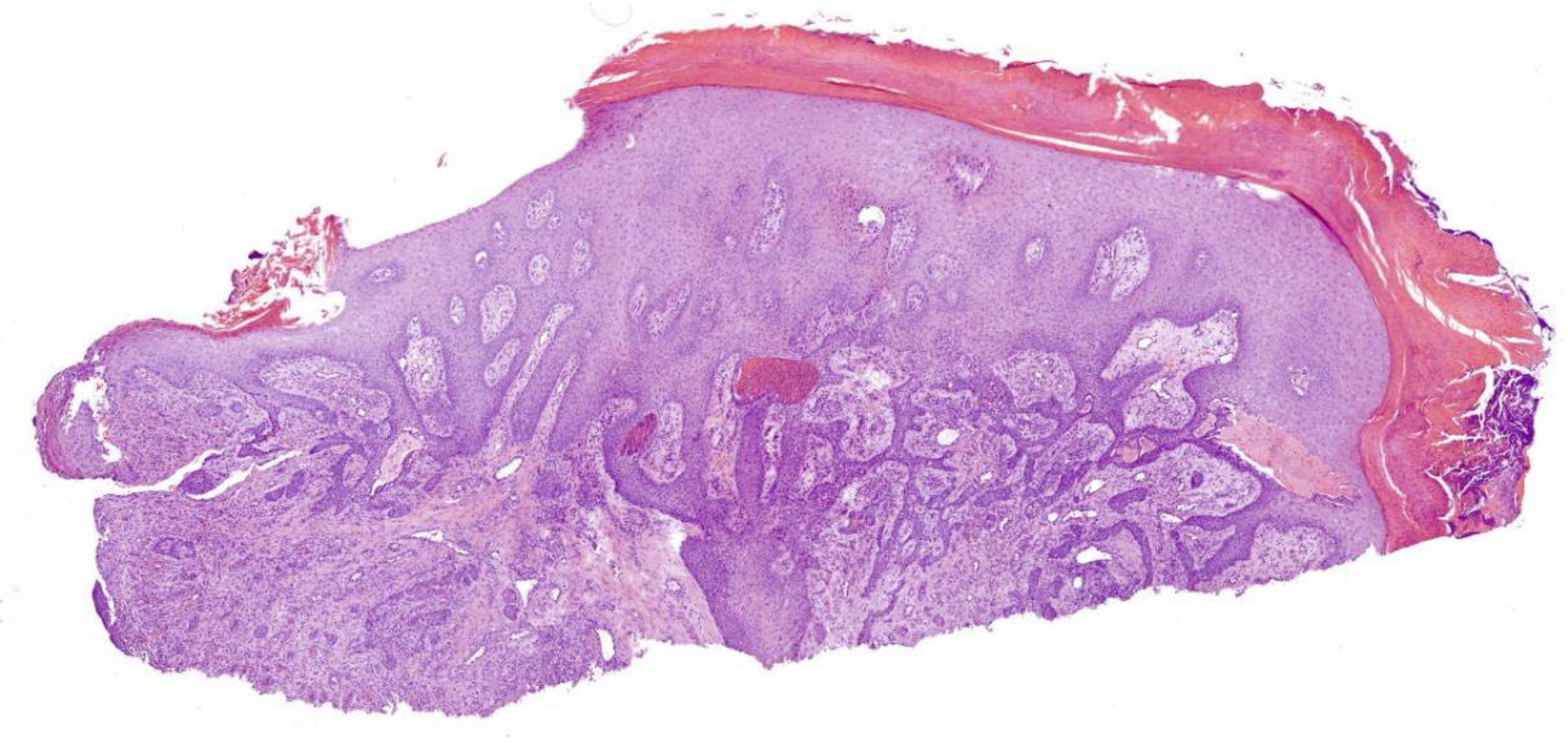


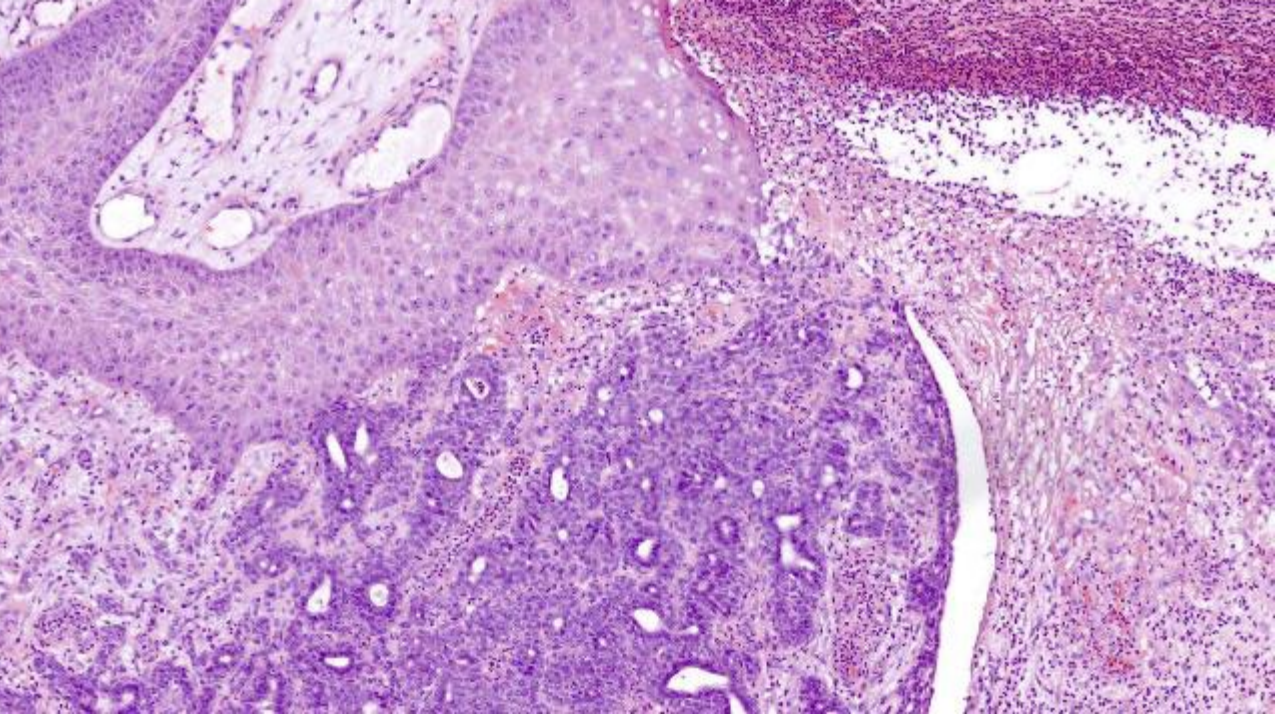


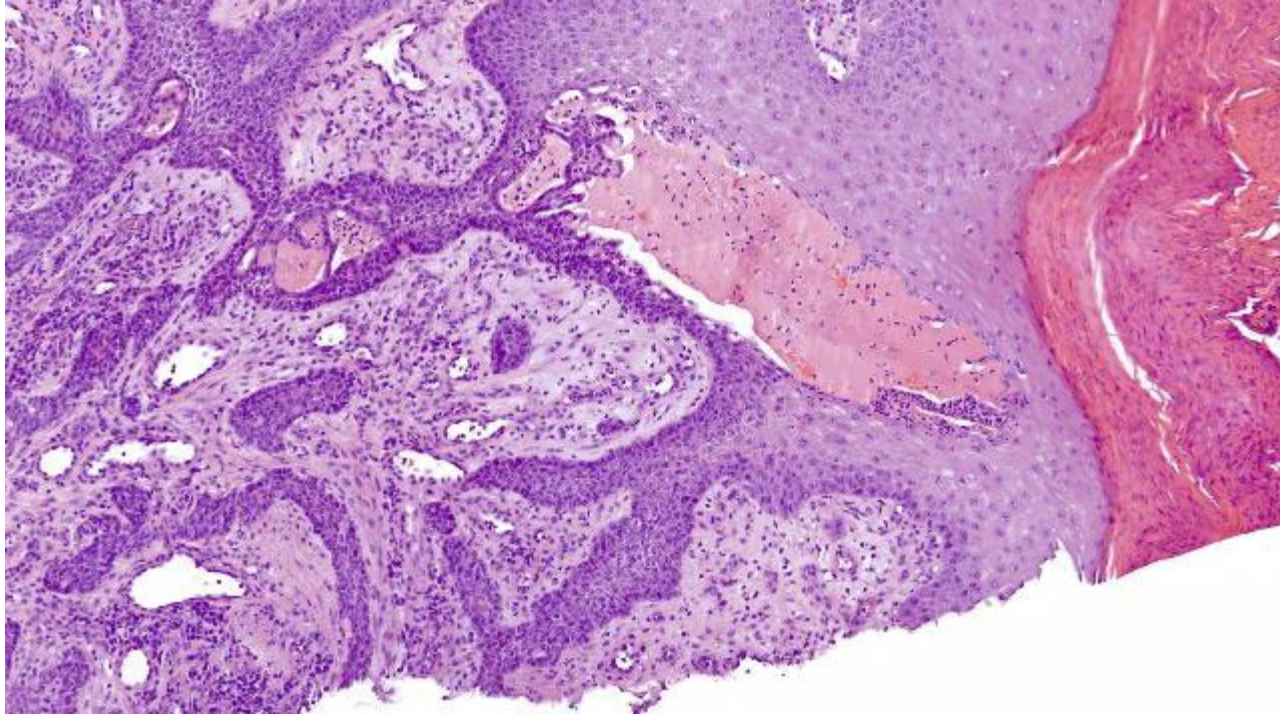


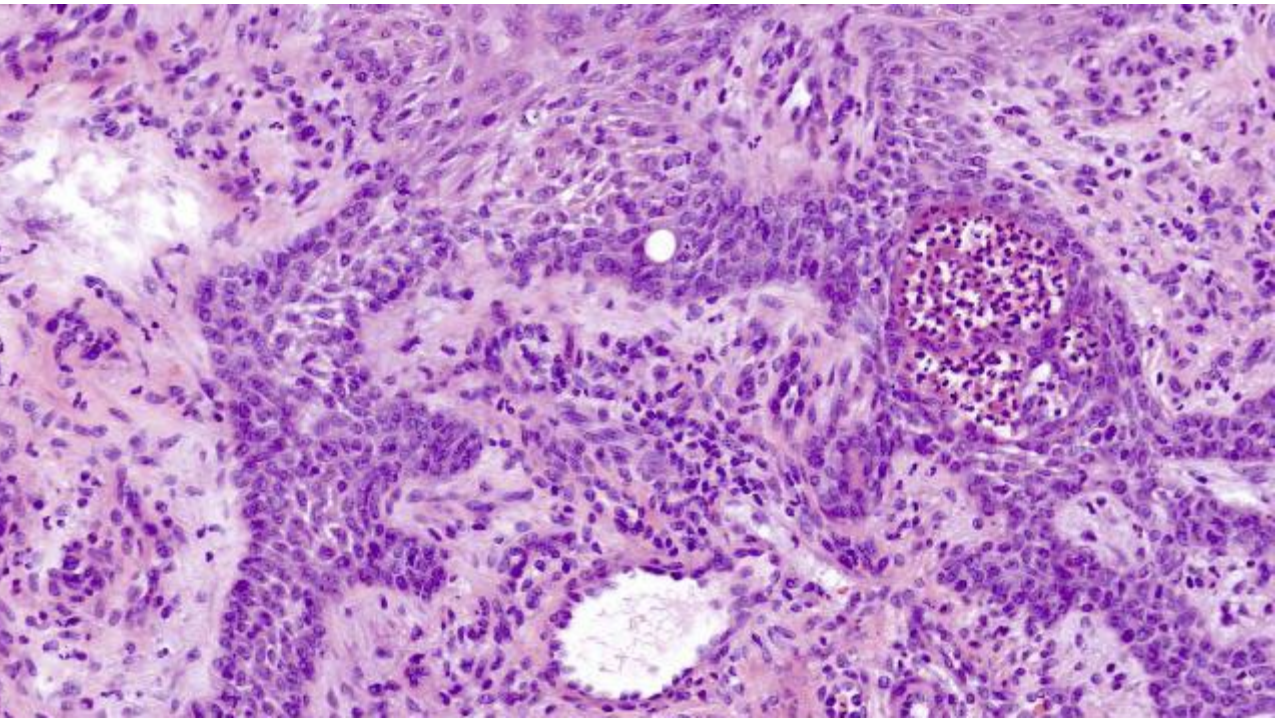
- 68-year-old male patient
- Slowly progressive swelling for one year, dig. IV left
- Occasional weeping, no pain
- Clinician suspected: chronic paronychia
- External histology: nonspecific inflammation

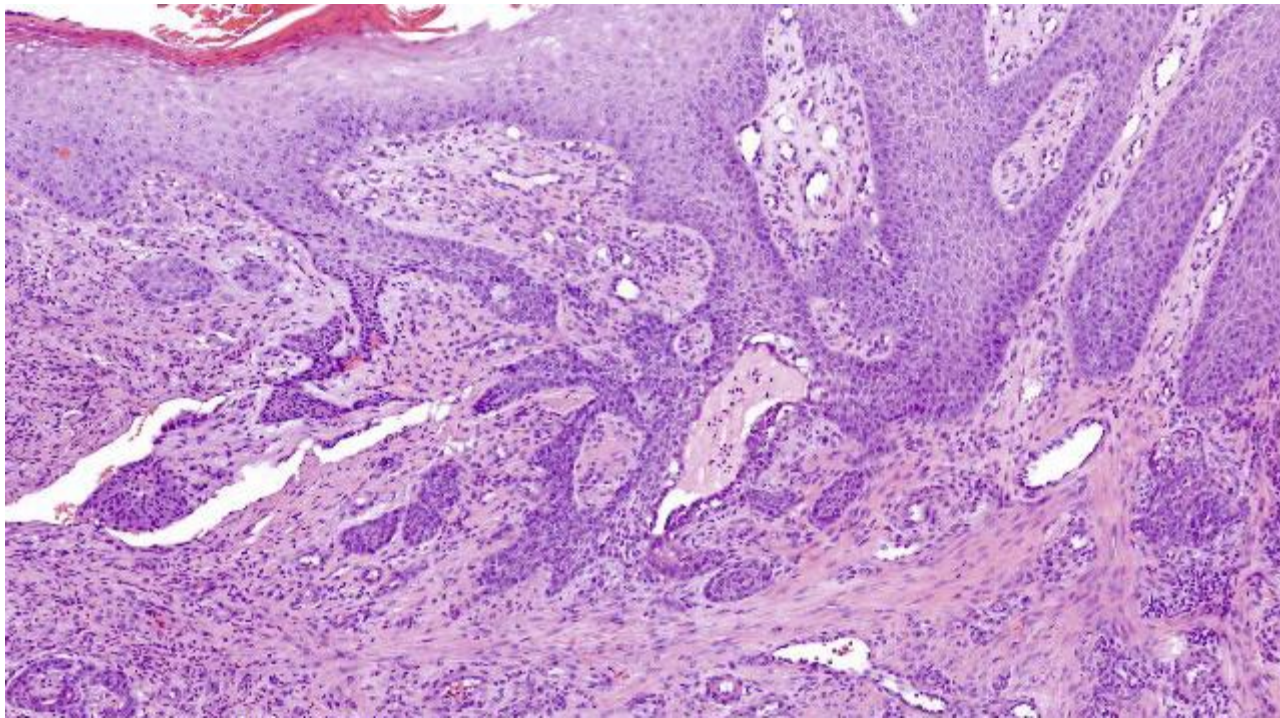
Histology:











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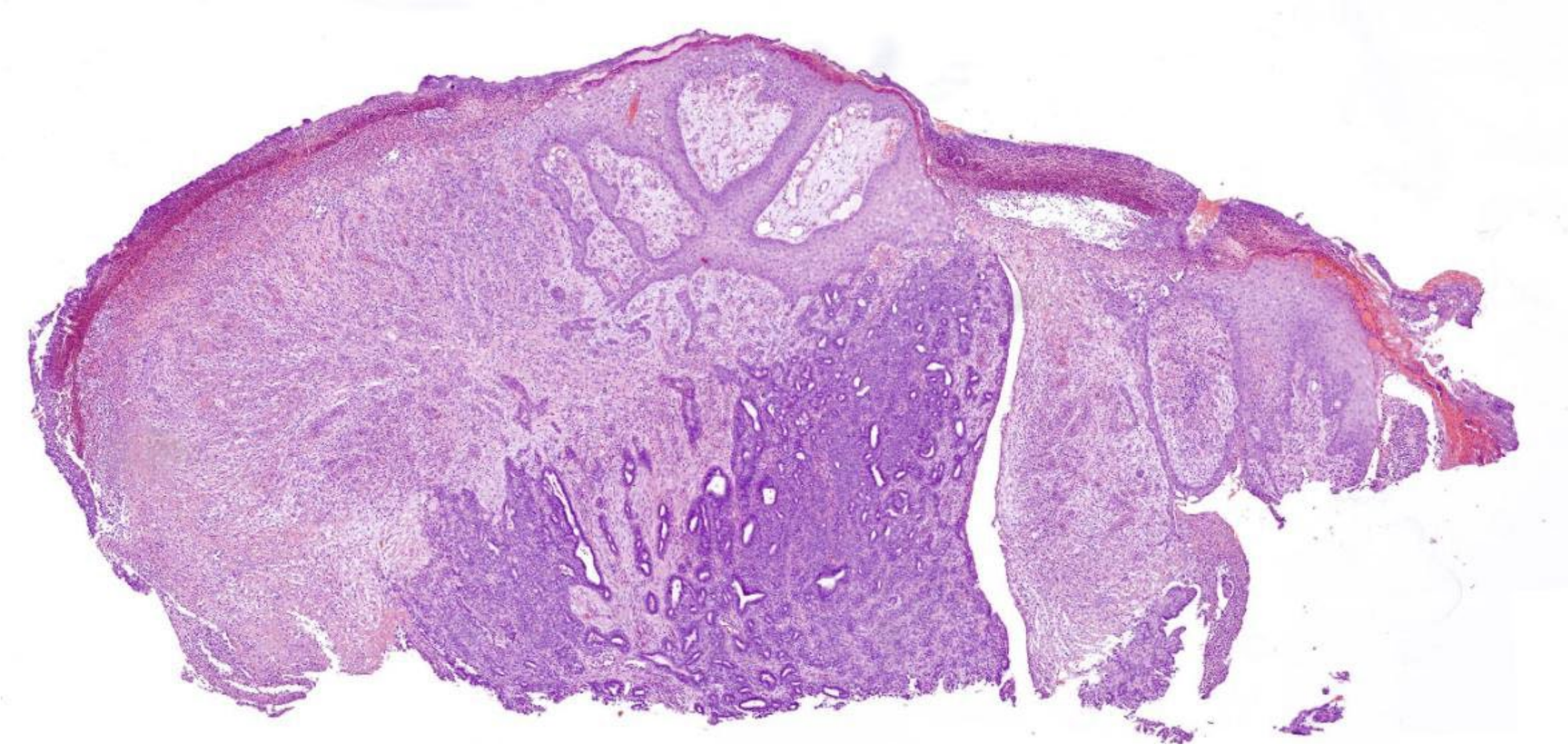

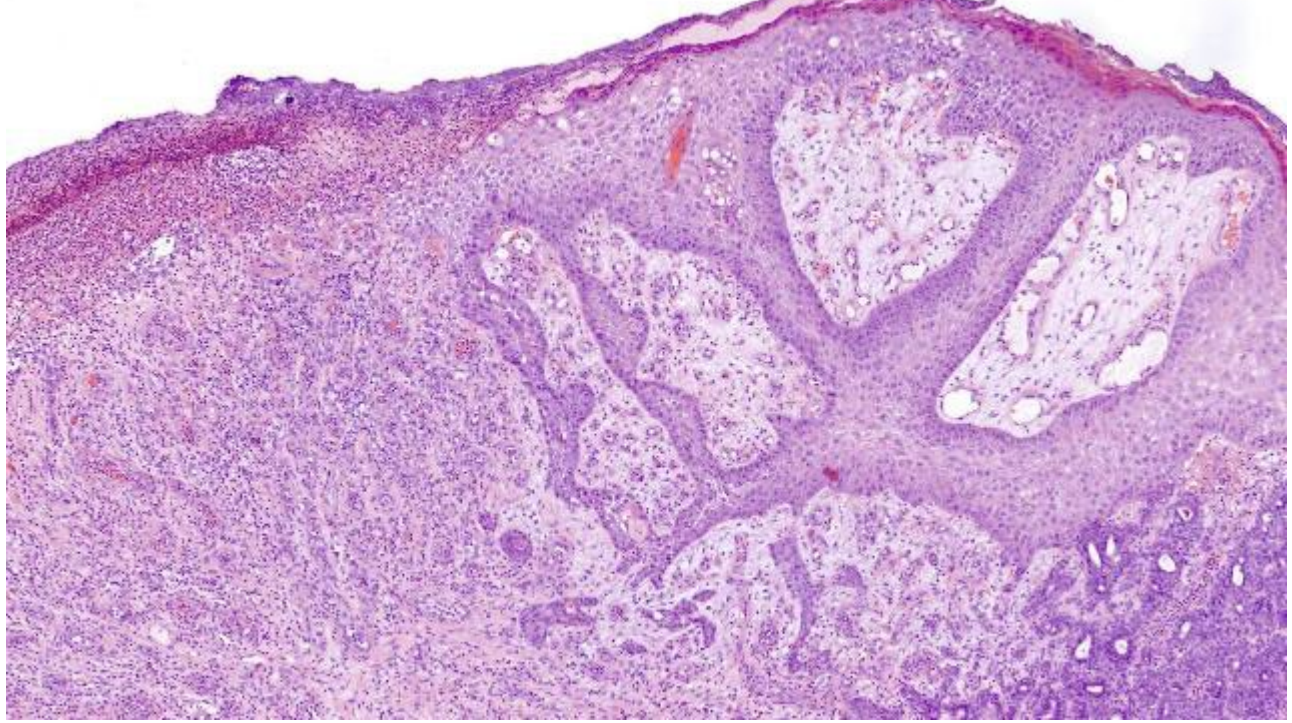
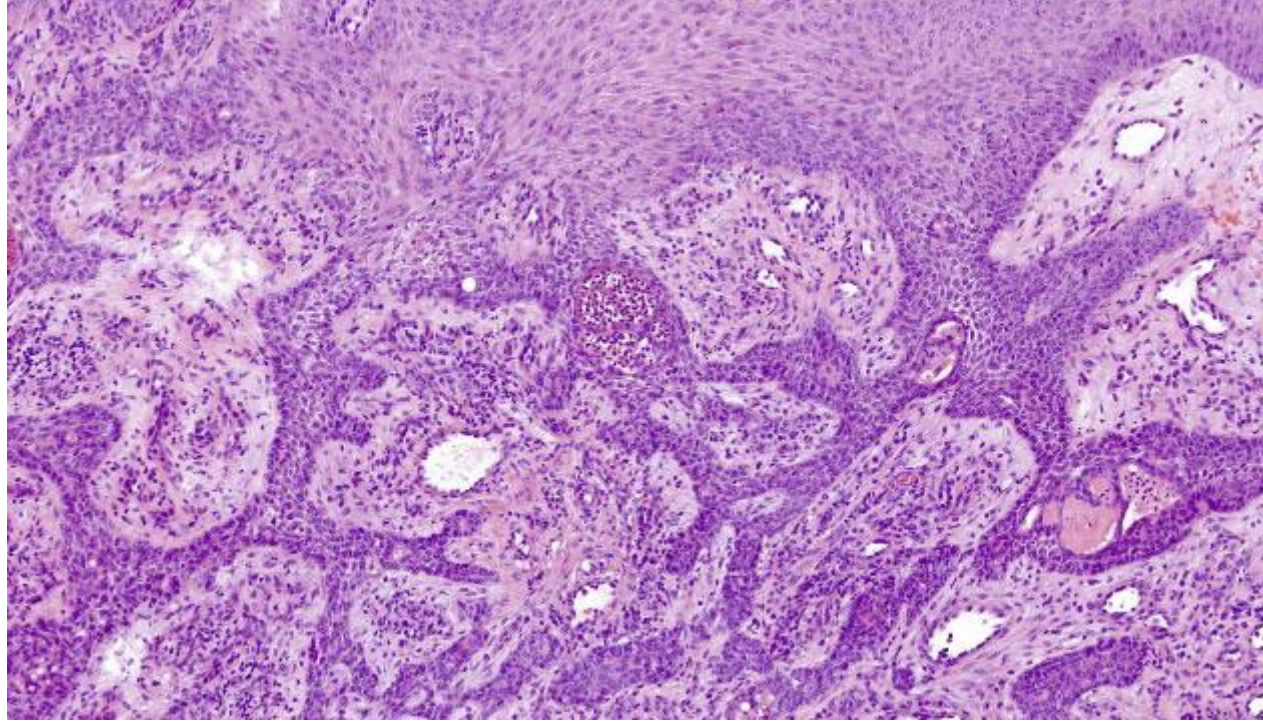
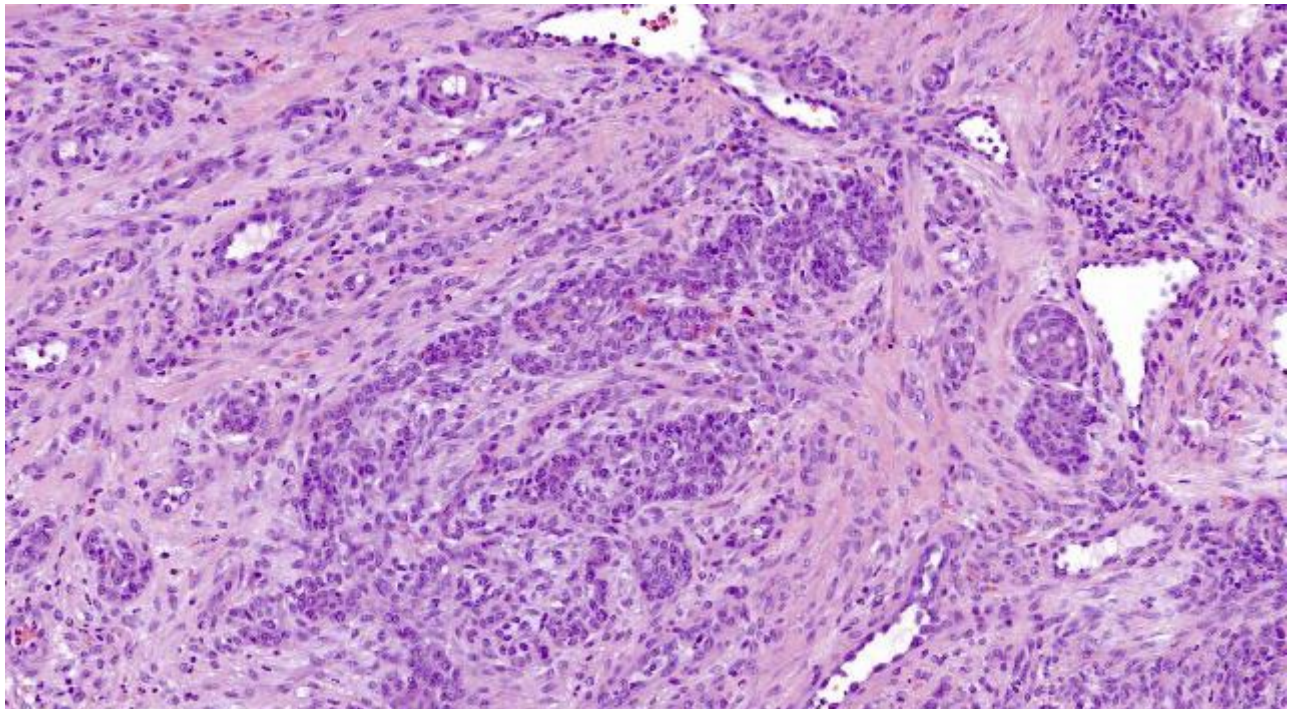
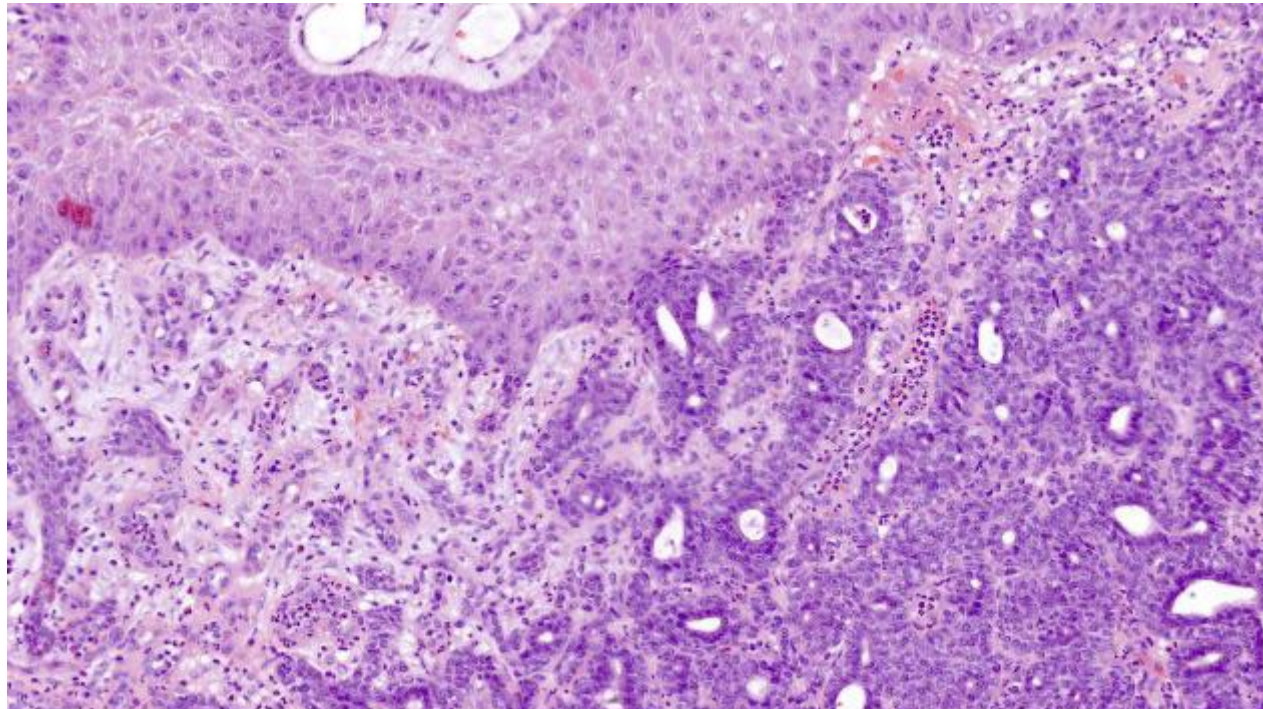

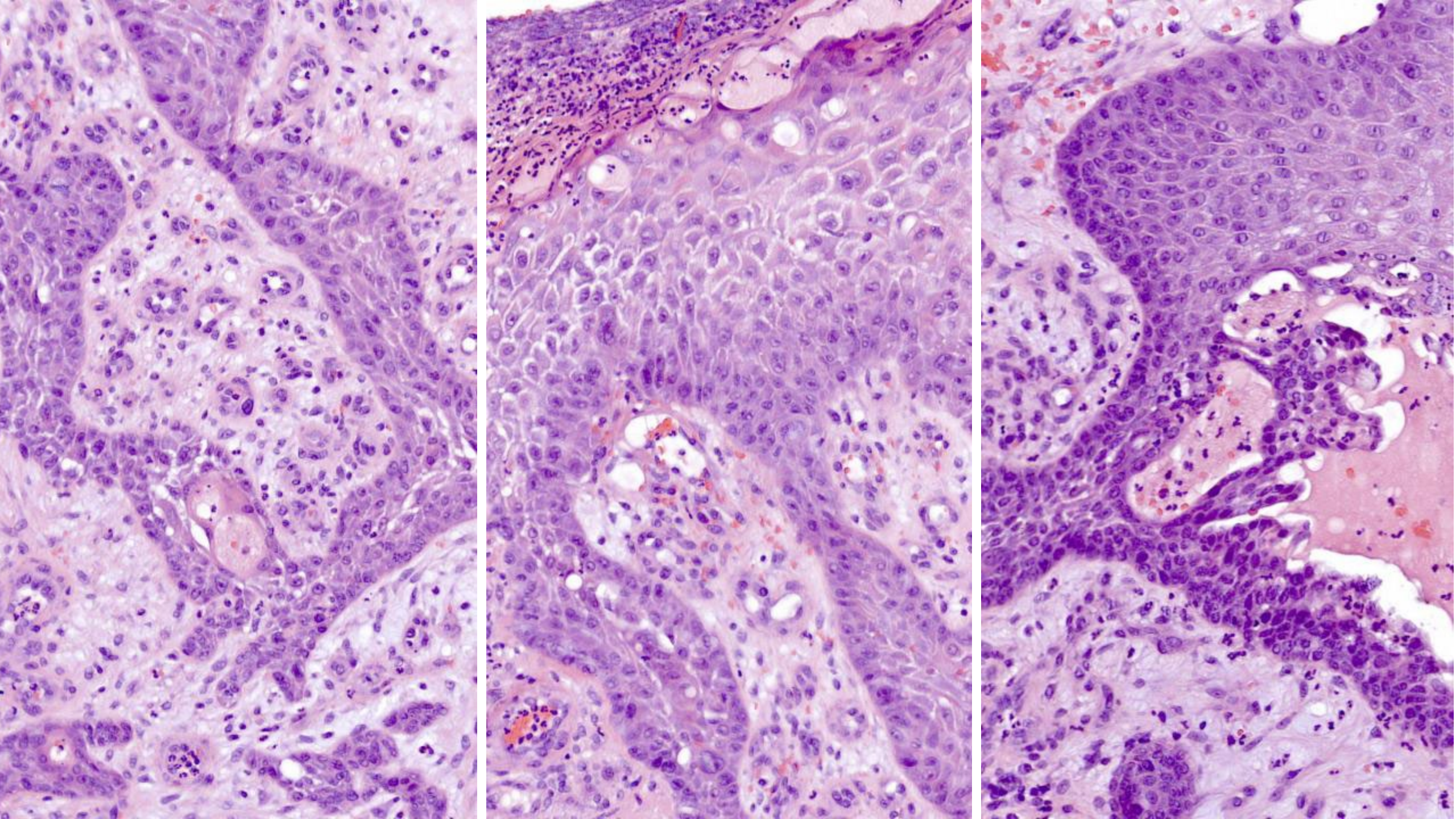

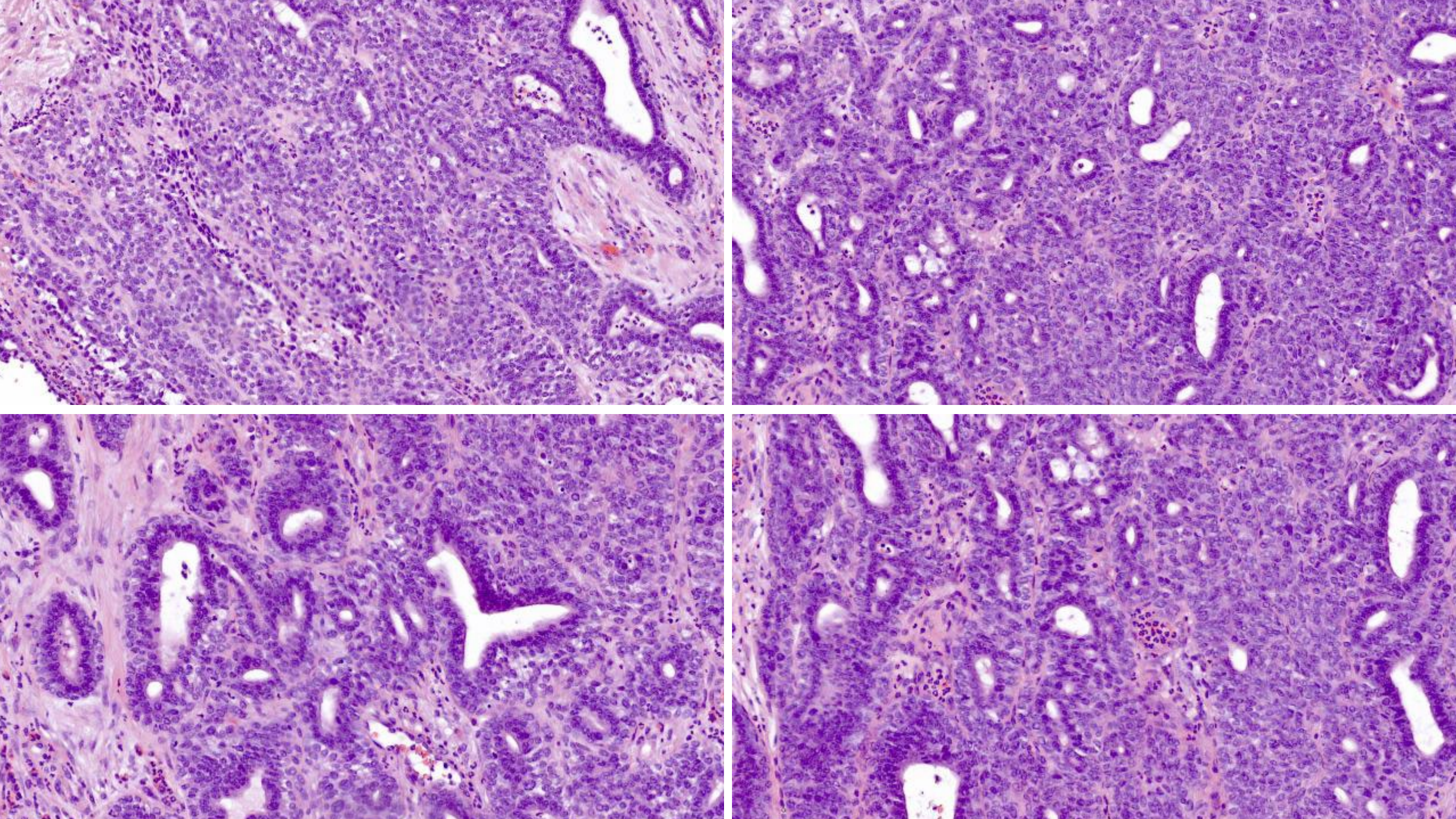

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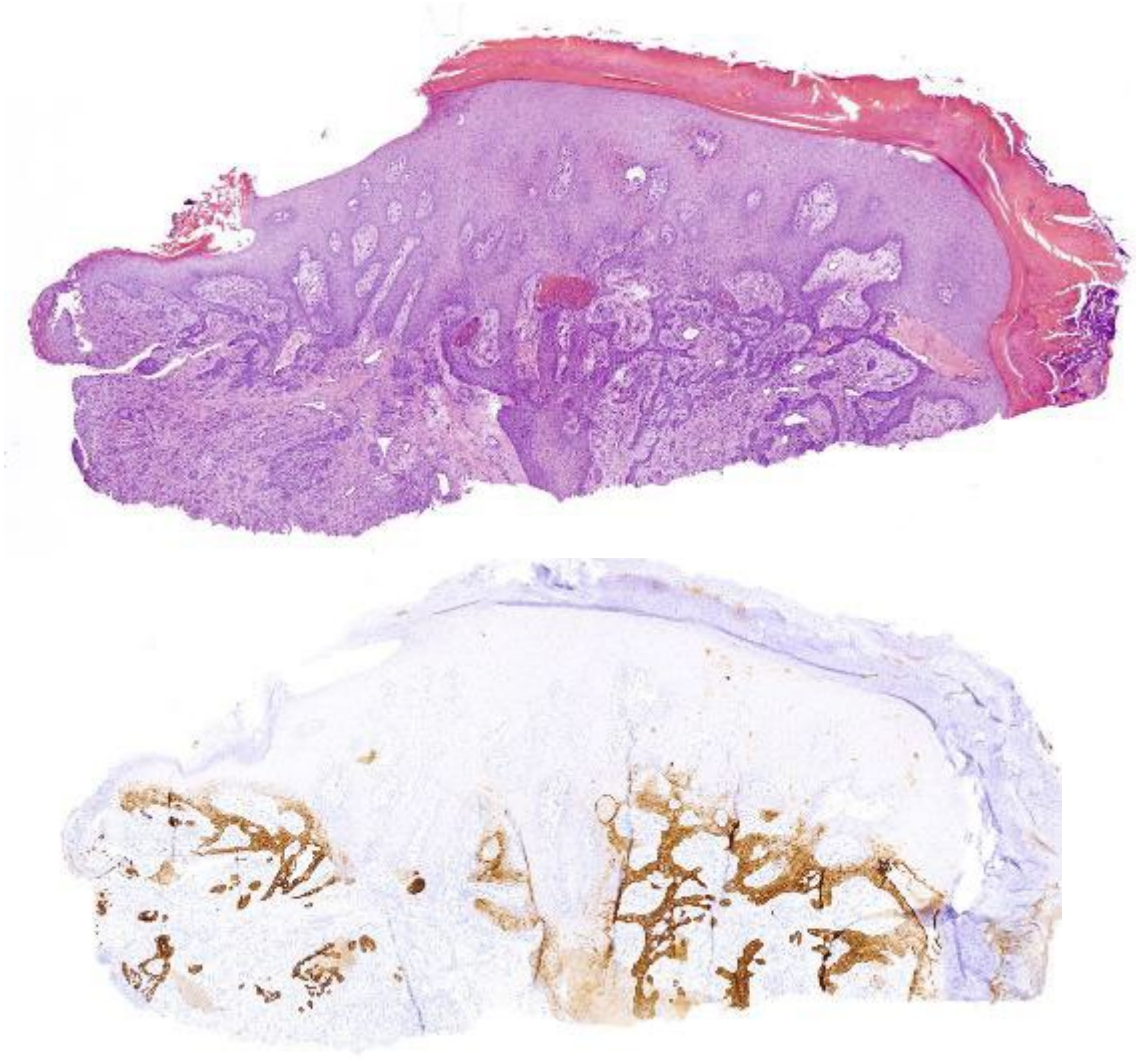
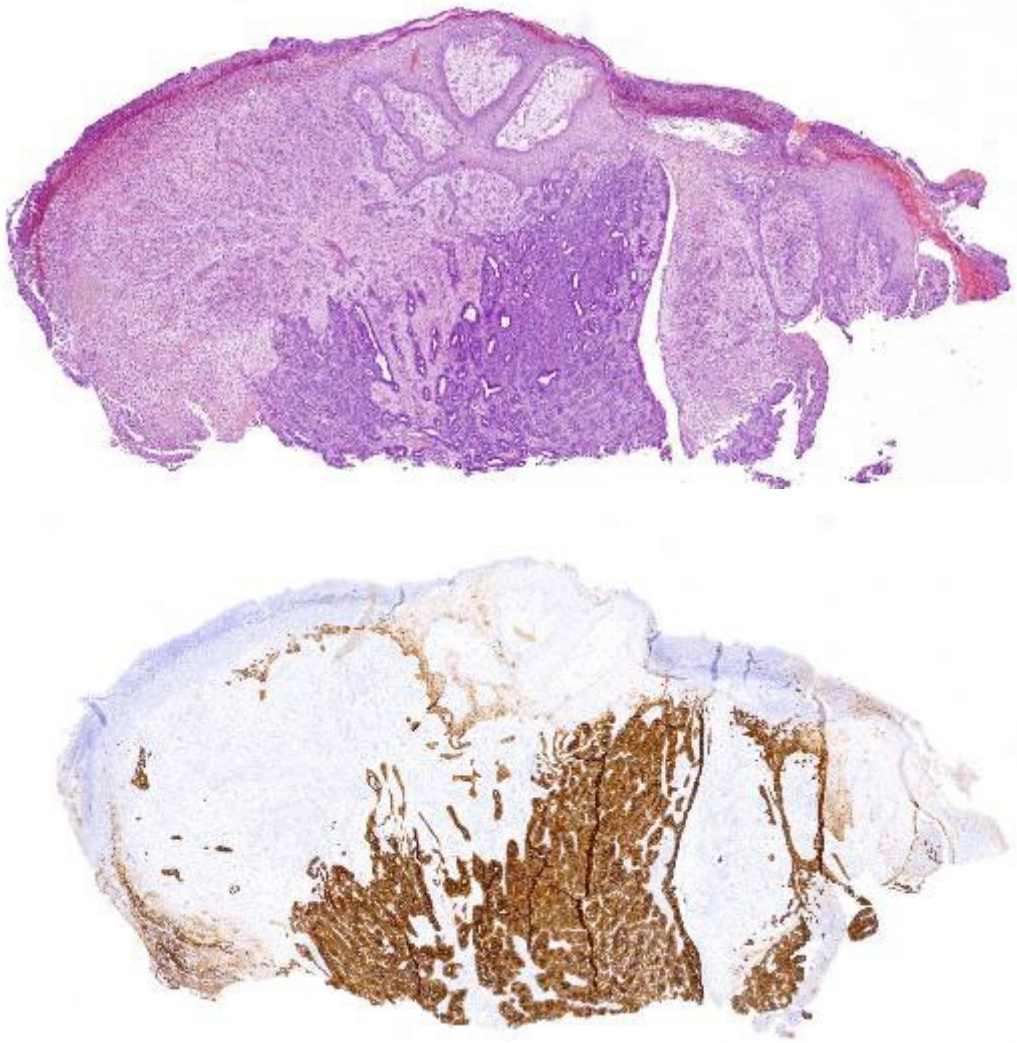


CK7



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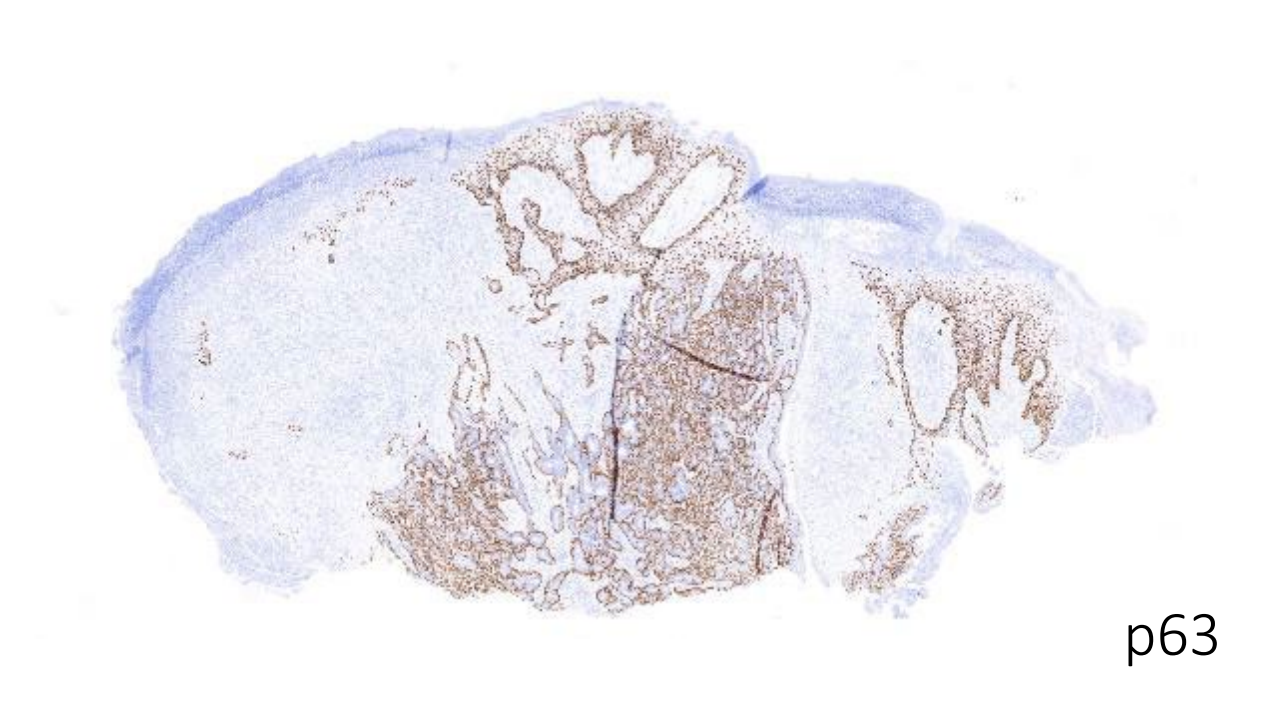
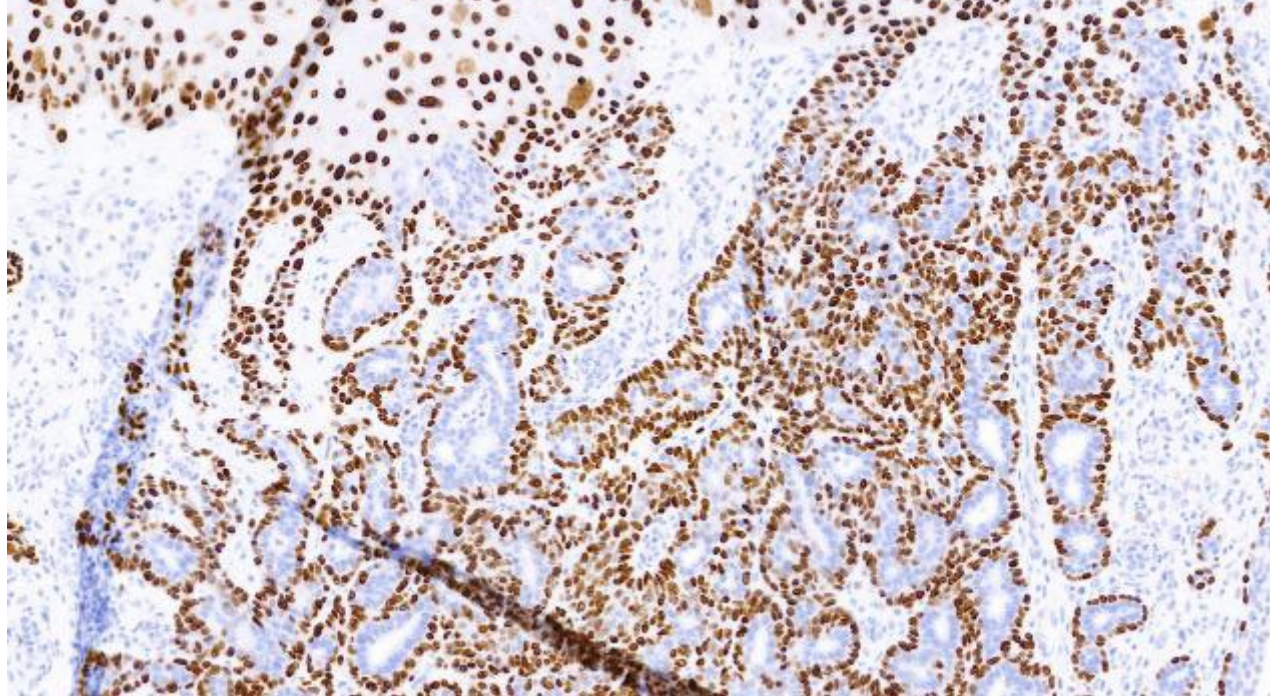
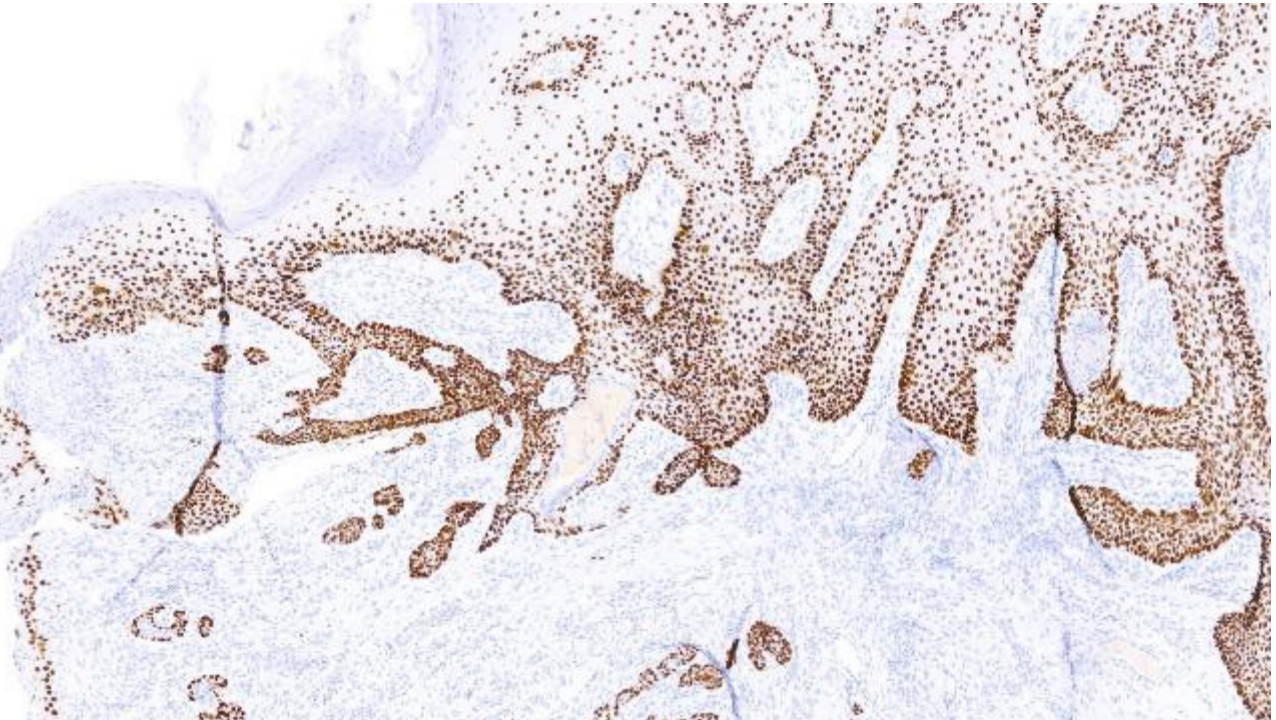
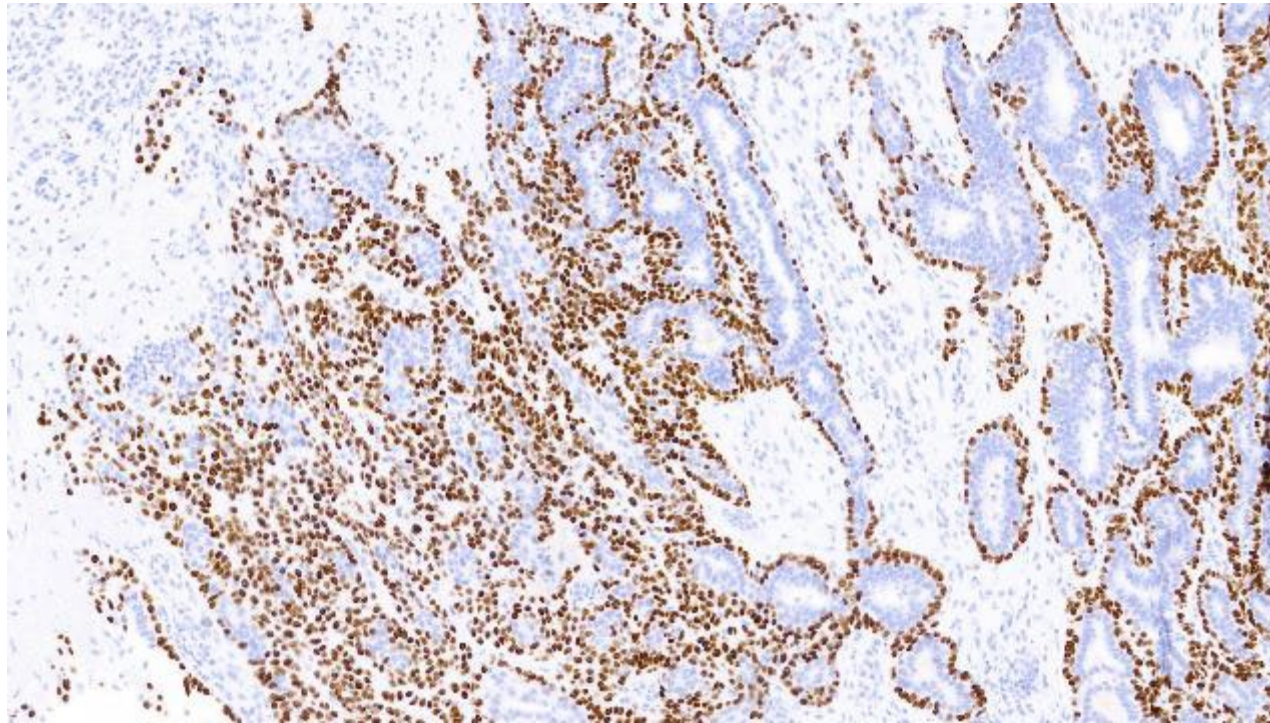


p63







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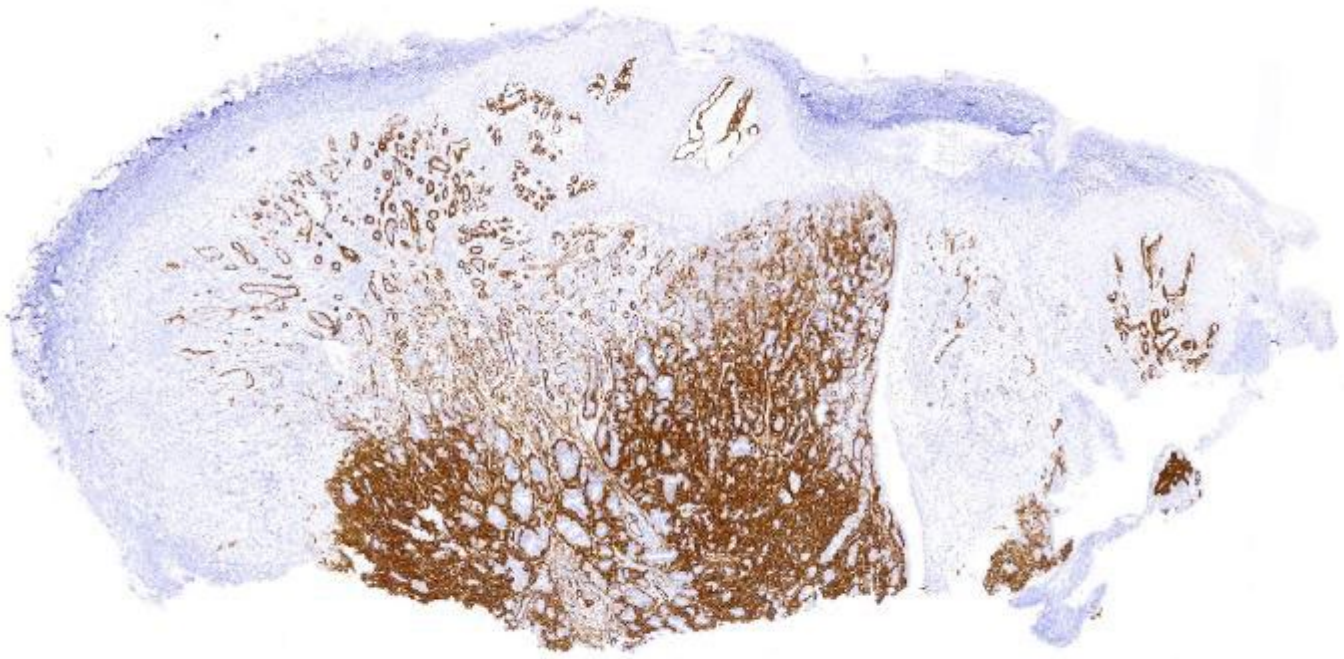
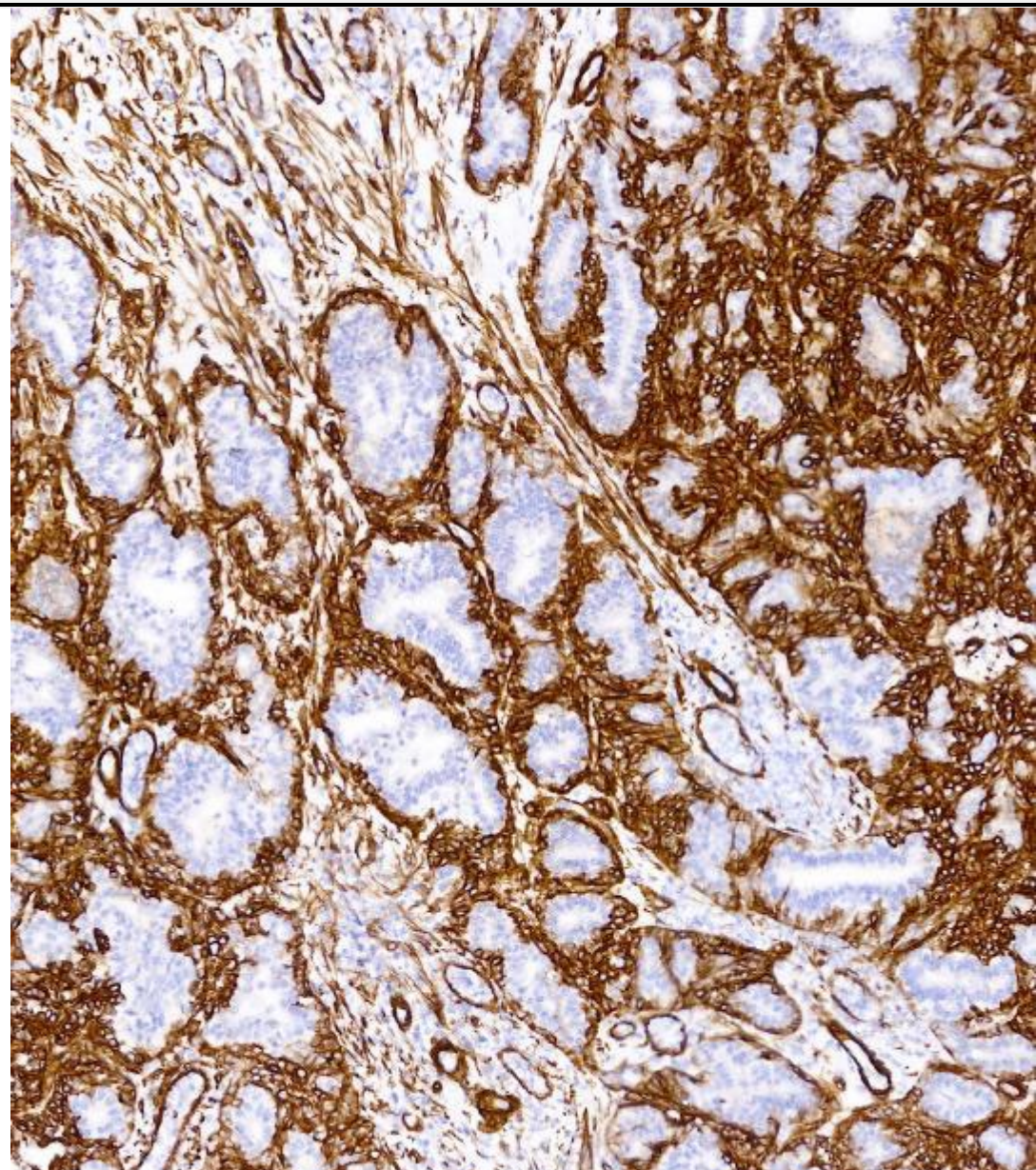
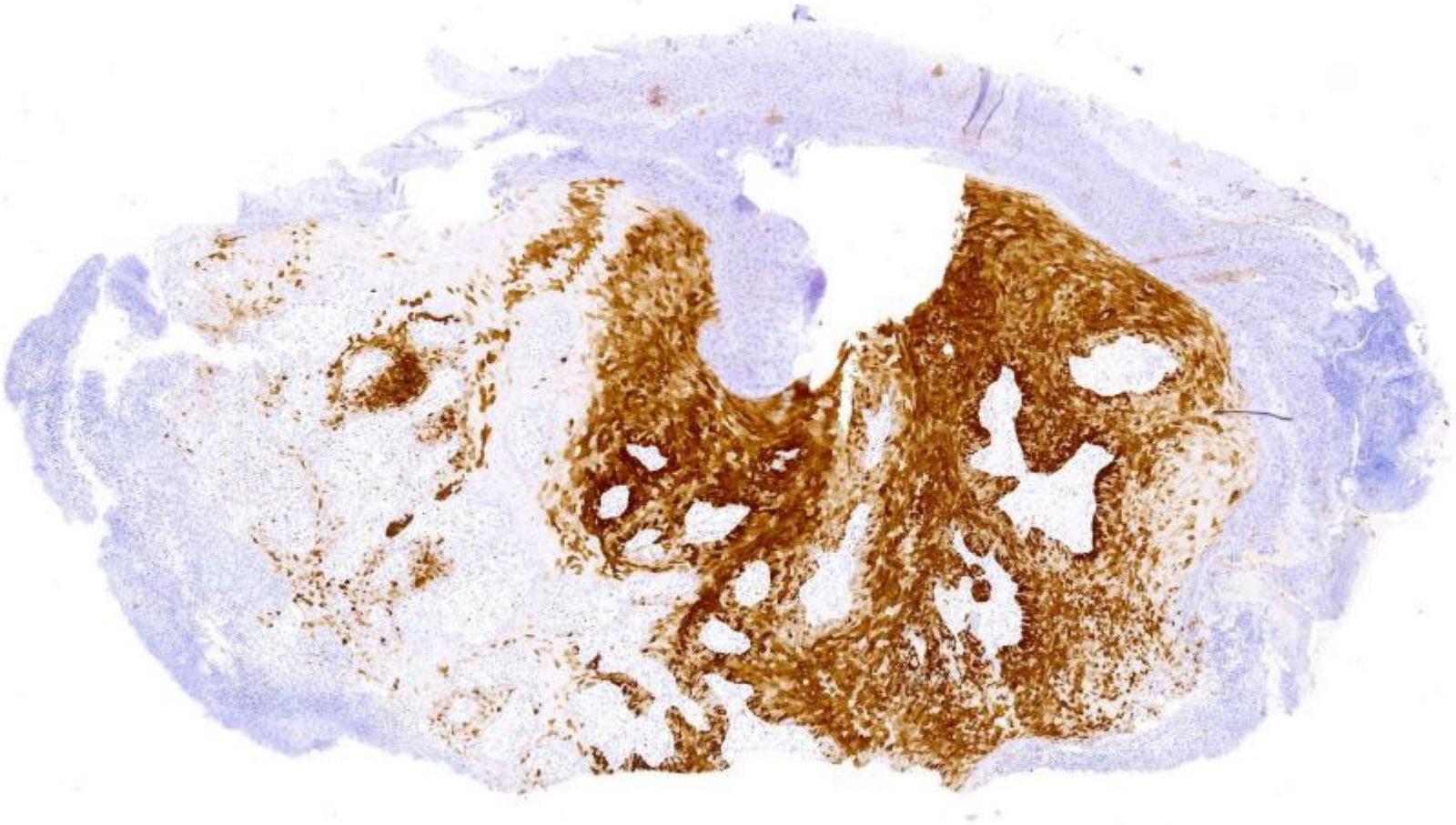


SMA

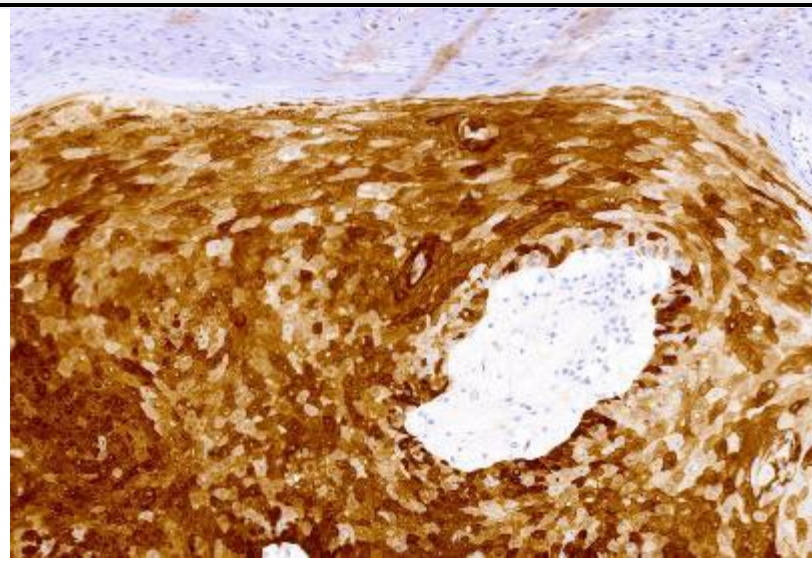


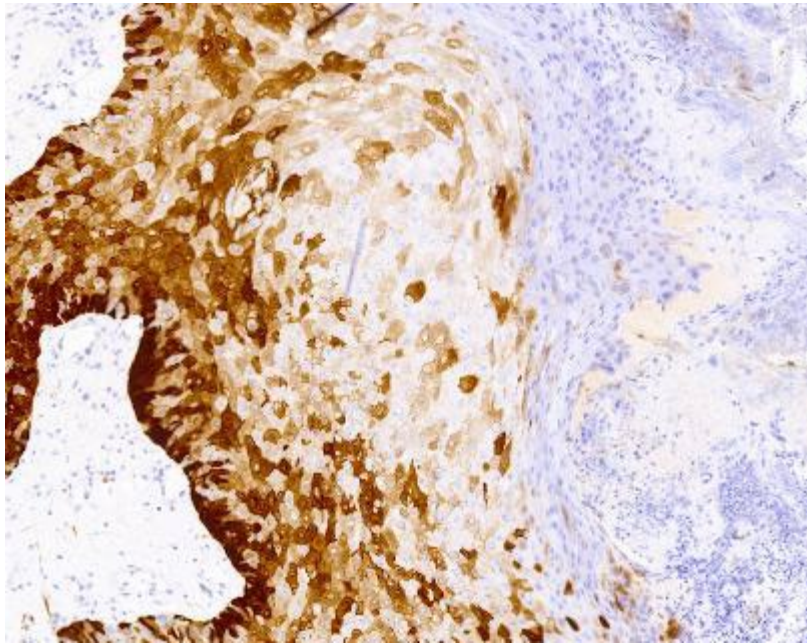
myoepithelial layer highlighted with SMA, calponin, p63 and focally podoplanin

Immunohistochemistry:

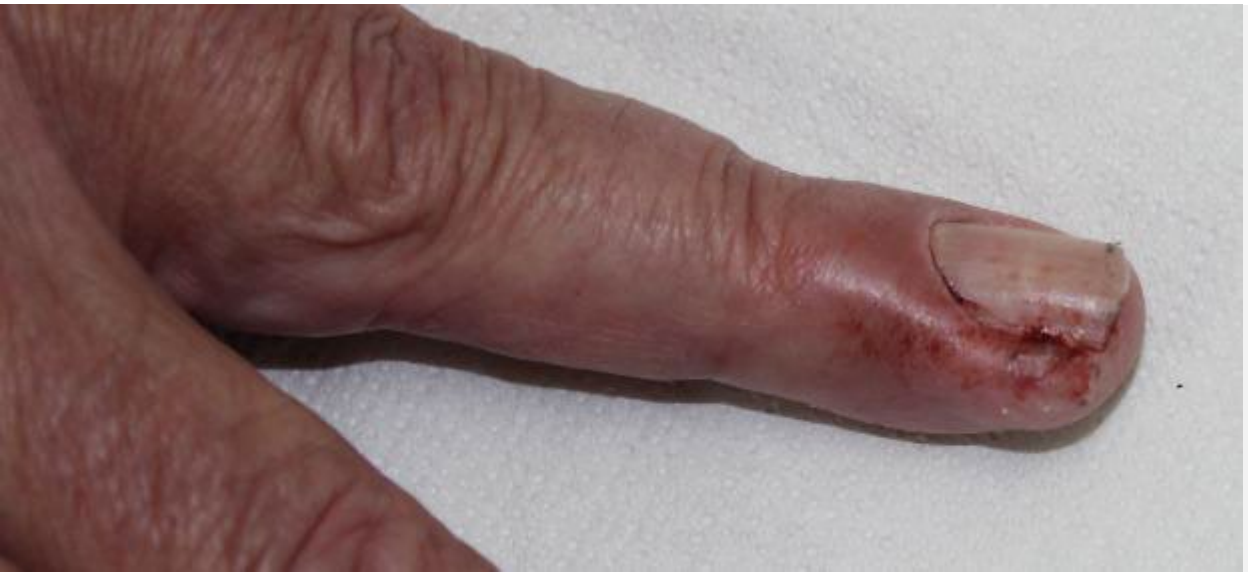


p16



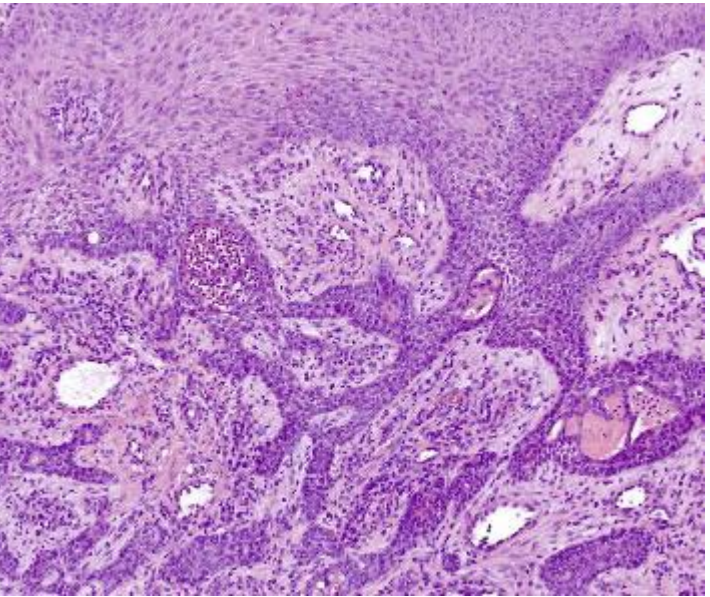


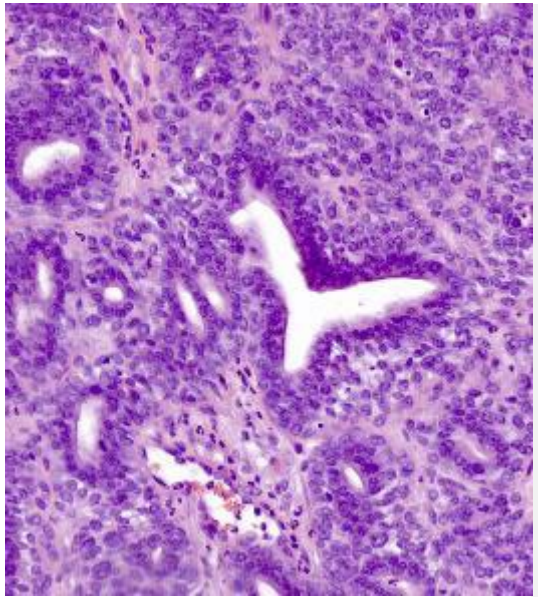
Summary of the findings:



Clinical presentation:

- Slowly progressive swelling at the lateral nail fourth for one year
- Ulceration
- Occasional weeping, no pain
- Paronychia?





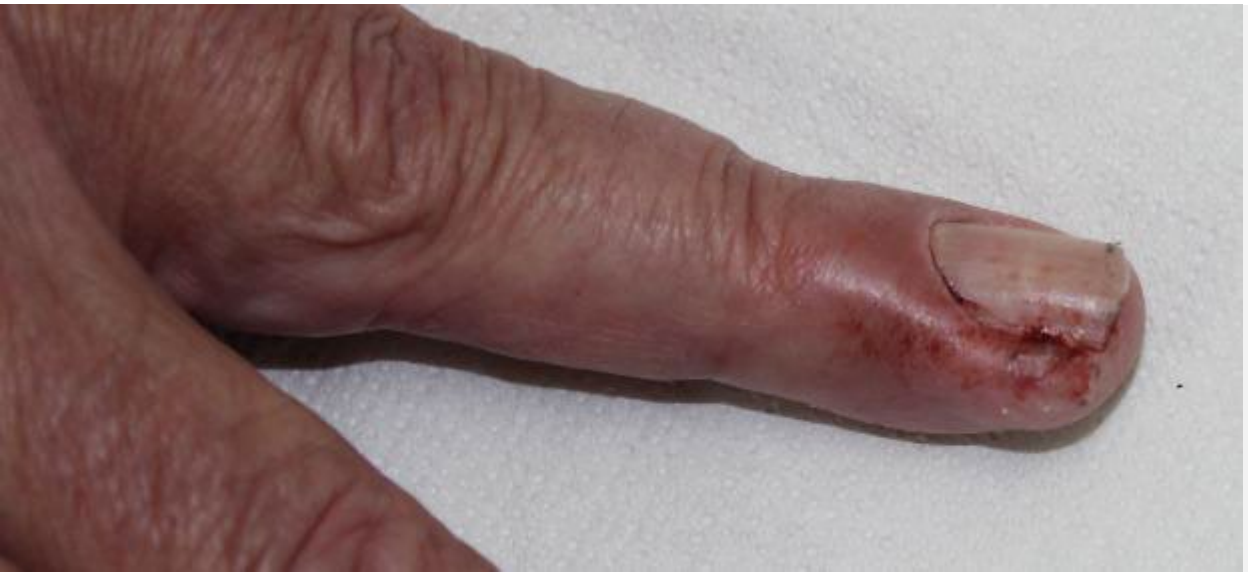
Histology:

- Dermal infiltrative adnexal npl.
- Combination of cystic, tubular/ ductal structures, papillary proliferations and solid areas
- Myoepithelial cell layer
- Mild atypia, few mitoses

diagnosis

?

Summary of the findings:



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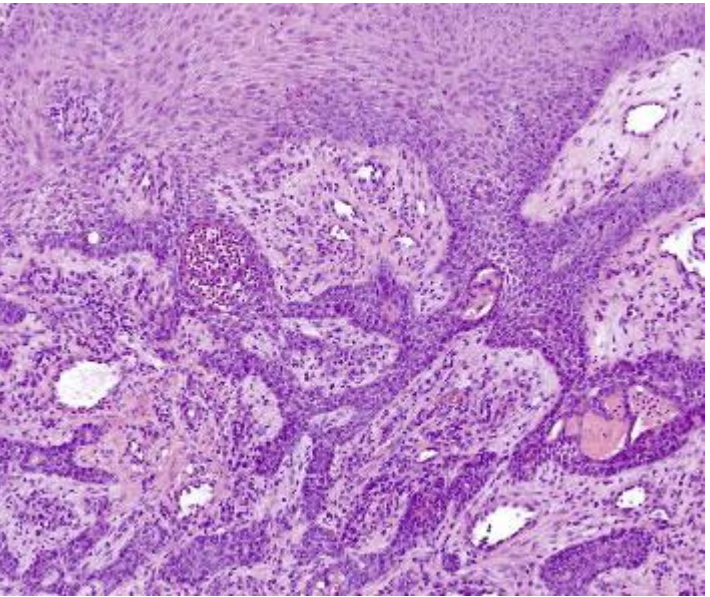

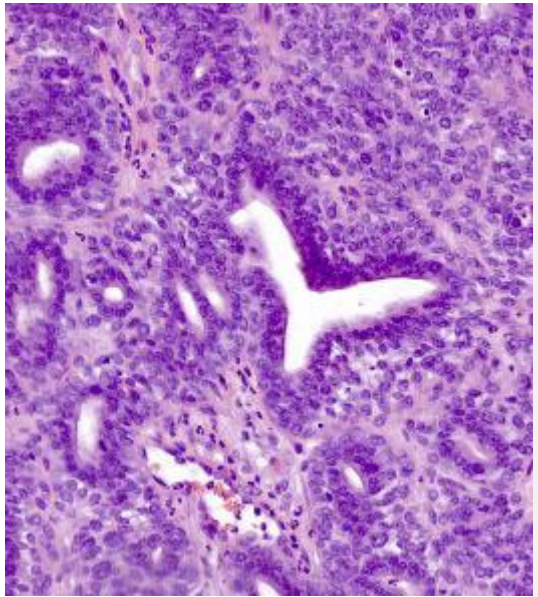

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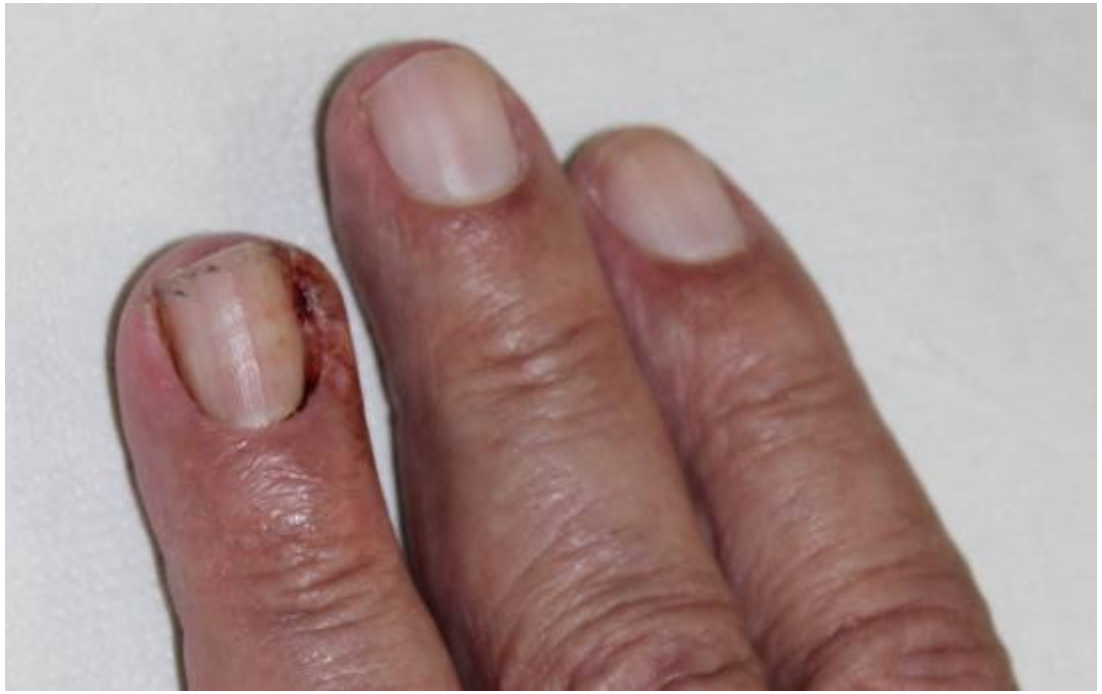
digital papillary adeno-carcinoma

HPV detection

Our patient:

Positive for HPV42 and for HPV92

Both are alpha papillomaviruses, mucosal low-risk types



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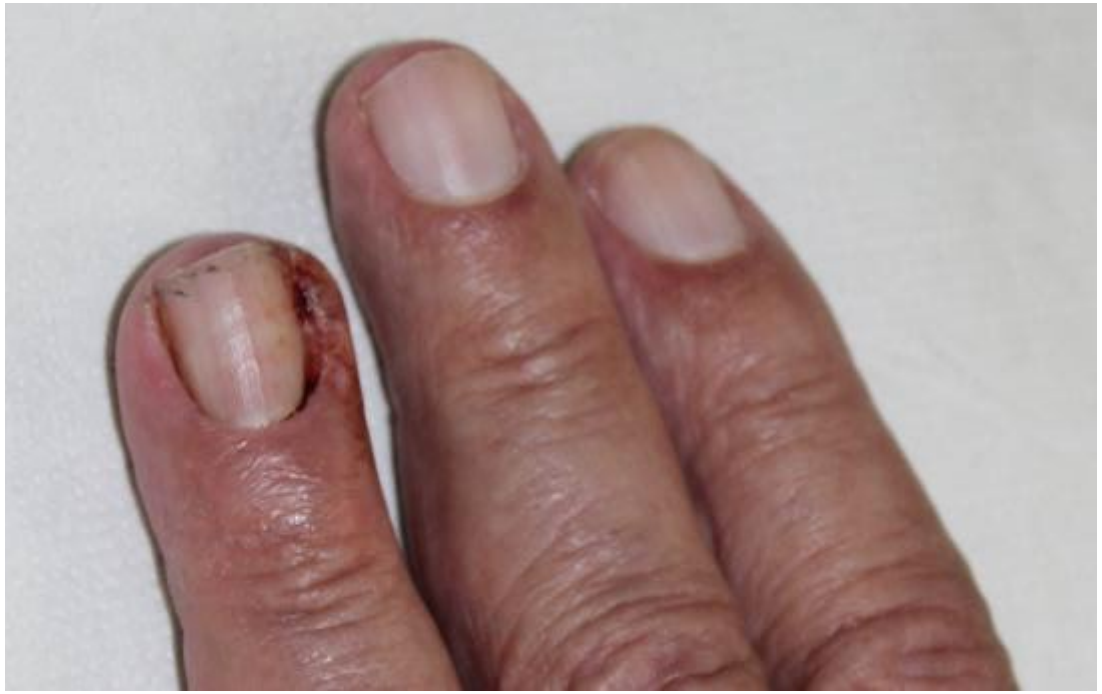
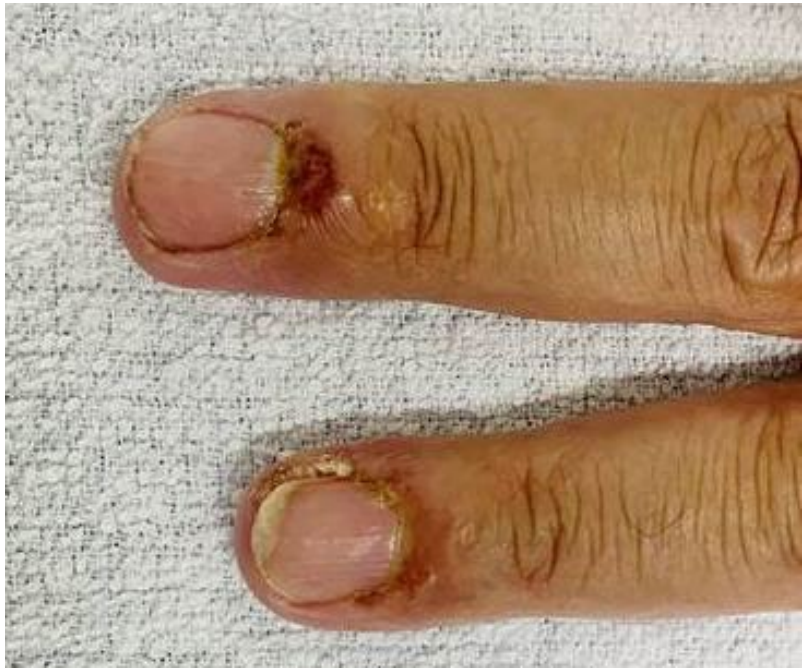


Syphilitic Paronychia:

Primary infection of syphilis

From: Shintaro K and Yukinori H Syphilitic Paronychia. Am J Med. 138, Issue 3e31-e32; 2025



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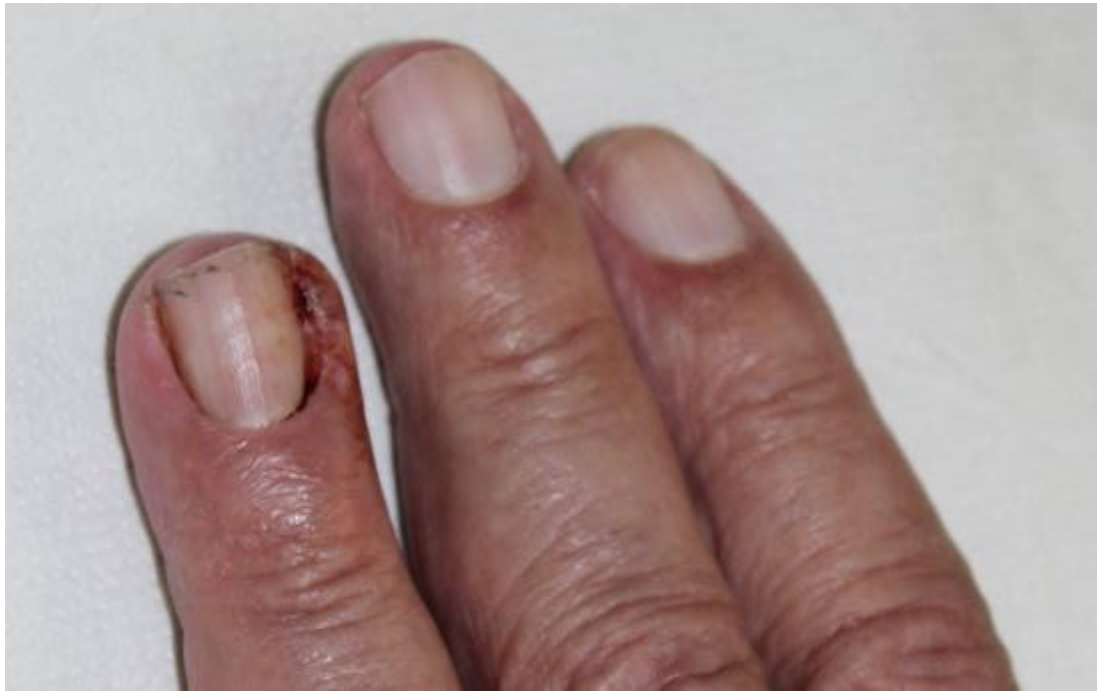
The coinfection (detection) with two mucosal HPV might lead to the hypothesis that HPV have been required through direct digital inoculation from mucosal epithelium.



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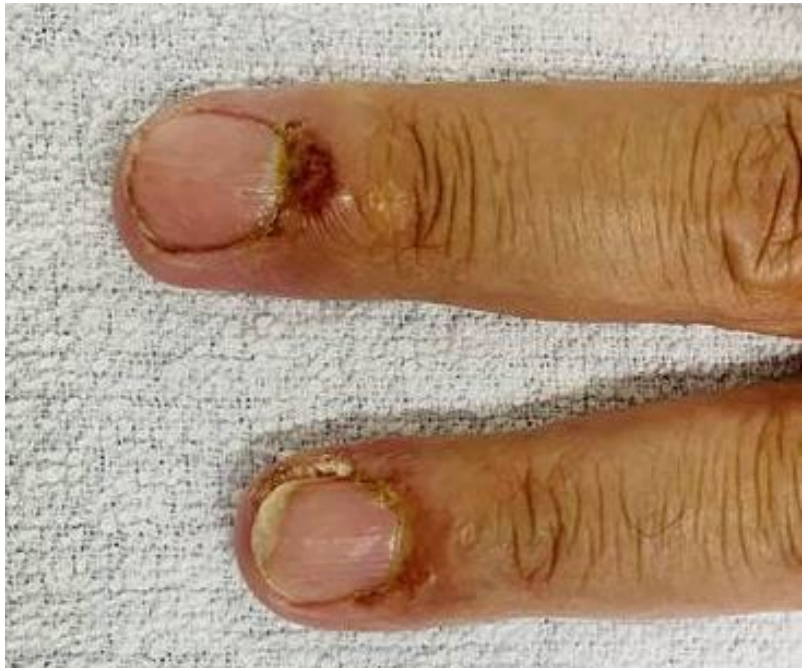
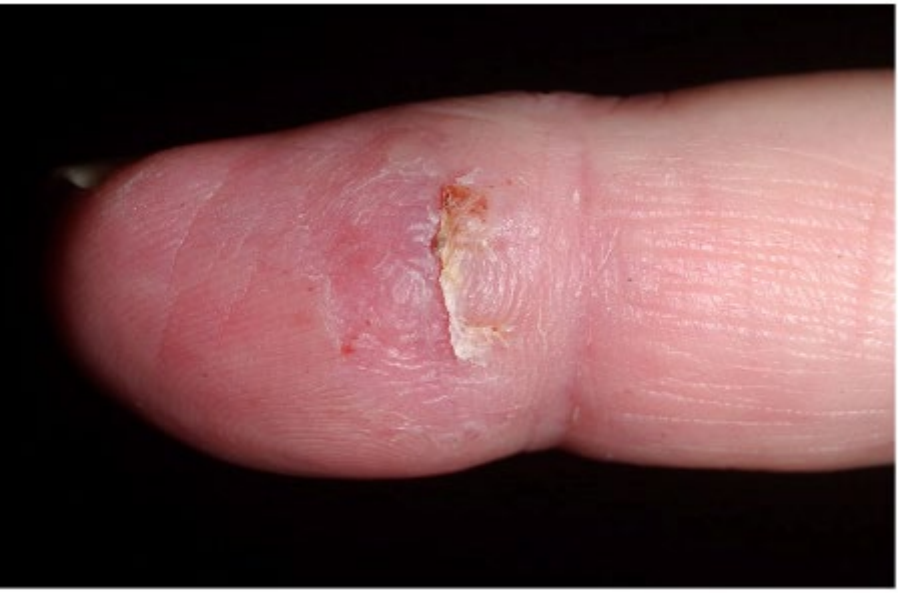
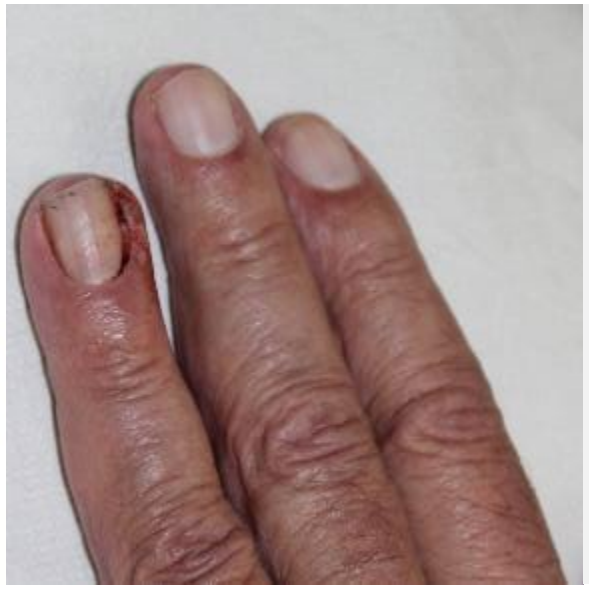


Literature: clinical features and prognosis



Mendenhall SD et al.

Bernstein E et al.



our patient

Can be misinterpreted as benign lesion:

- Ganglion (50%–80%)
- Giant cell tumors of tendon sheath (7%–12%)
- Epidermal inclusion cysts (5%–9%)
- Paronychia (rare)

- 85% hand, in 79% digital
- Less common on the feet, toes, ankle, thigh
- Male : female = 4:1
- Median age 54 years (range 16-94)
- More often in Caucasian

Local recurrence rate:	5-21%
Metastasis rate:	26-50%

Bernstein E et al. Digital Papillary Adenocarcinoma at the Site of 5 Years of Recurrent Paronychia: Case Report and Literature Review. Case Reports in Oncological Medicine. Volume 2024, Article ID 9910470. **Mendenhall** SD et al. Aggressive Digital Papillary Adenocarcinoma: A Meta-Analysis of the Presentation, Treatment, and Outcomes of a Rare Hand Tumor. HAND 2026, Vol. 21(3) 462–471

Literature: Histologic DDx of DPA

Histopathology

SHORT REPORT

Methylation analysis as a diagnostic tool for sweat gland tumours classification with emphasis on the distinction between digital papillary adenocarcinoma and mimickers

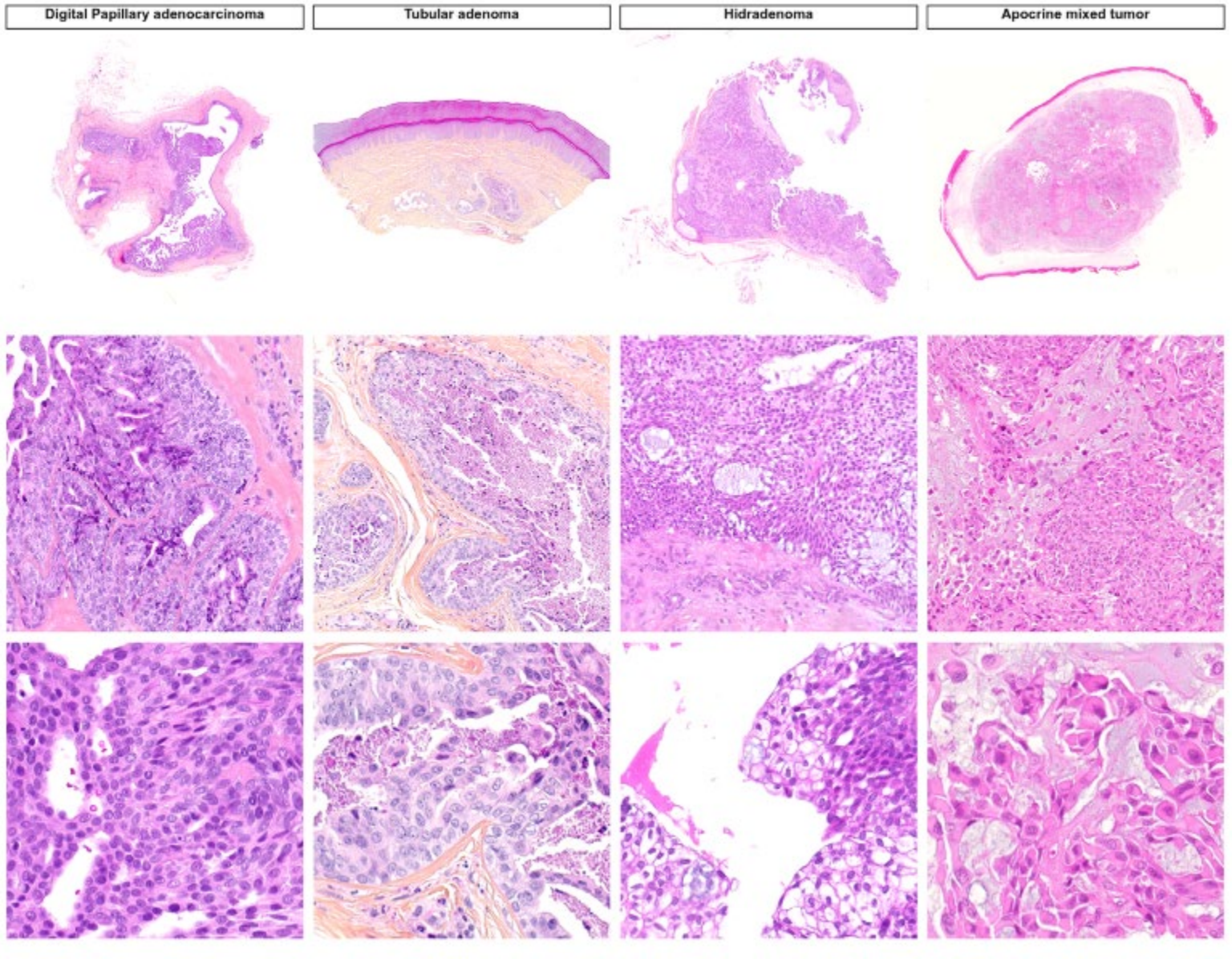
Pierre Sohier,[1,2] Nicolas Macagno,[2,3] Anne Tallet,[4] Coralie Mousset,[5] Maxime Battistella,[1,6] Eduardo Calonje,[7] Andreas Von Deimling[8] & Thibault Kervarrec[8]

Digital papillary adenocarcinoma: HPV42+ (12 out of 12)

Hidradenoma: CRTC1::MAML2 fusions (10 out of 12)

Tubular adenomas: BRAF V600E mutation (9 out of 11)

Apocrine variant of mixed tumours: TRPS1::PLAG1 fusion (5 out of 8)

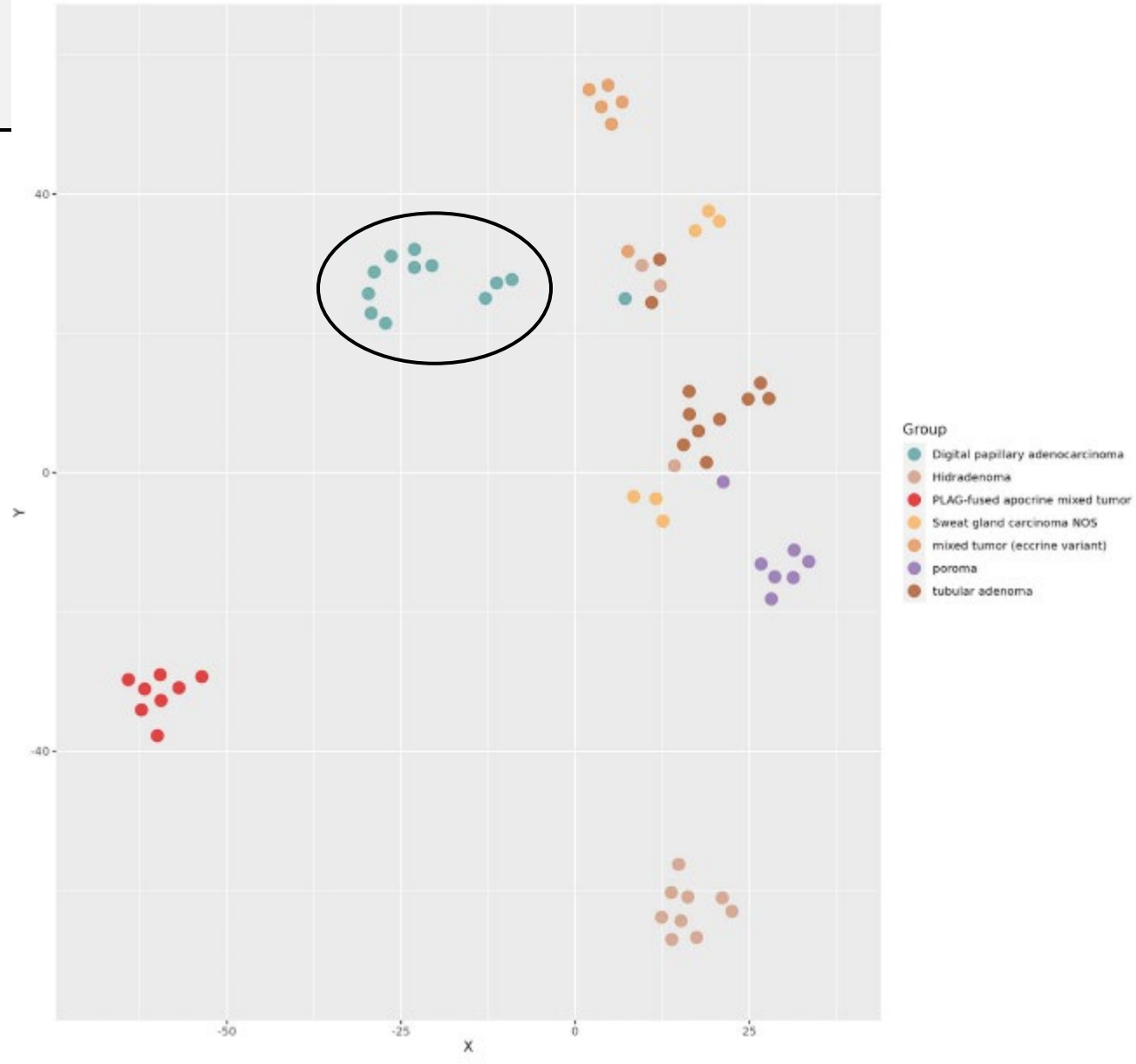


Literature

Methylation profiling of sweat gland tumours:

Most tumour types display a unique cluster, setting them apart from the others.

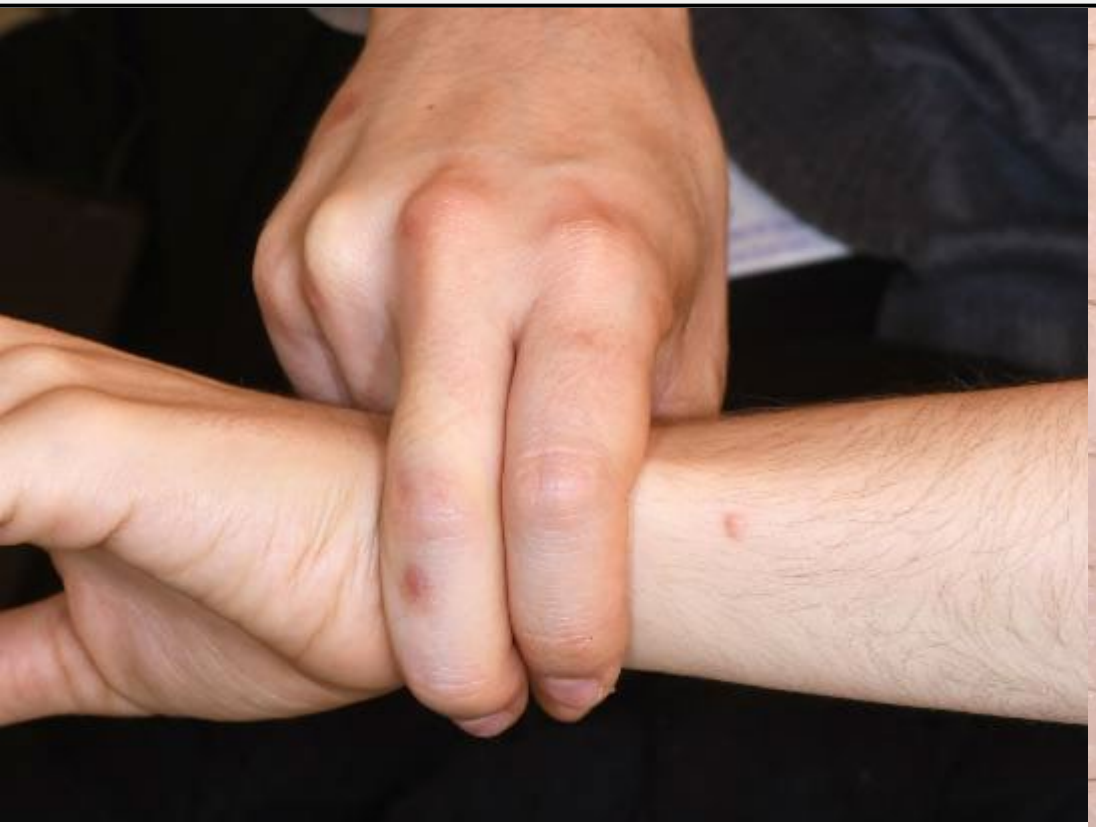
Notably, DPA cases also formed a distinct cluster distinct from tubular adenomas, hidradenomas, and mixed tumours, with the exception of one DPA case with low tumour cellularity.

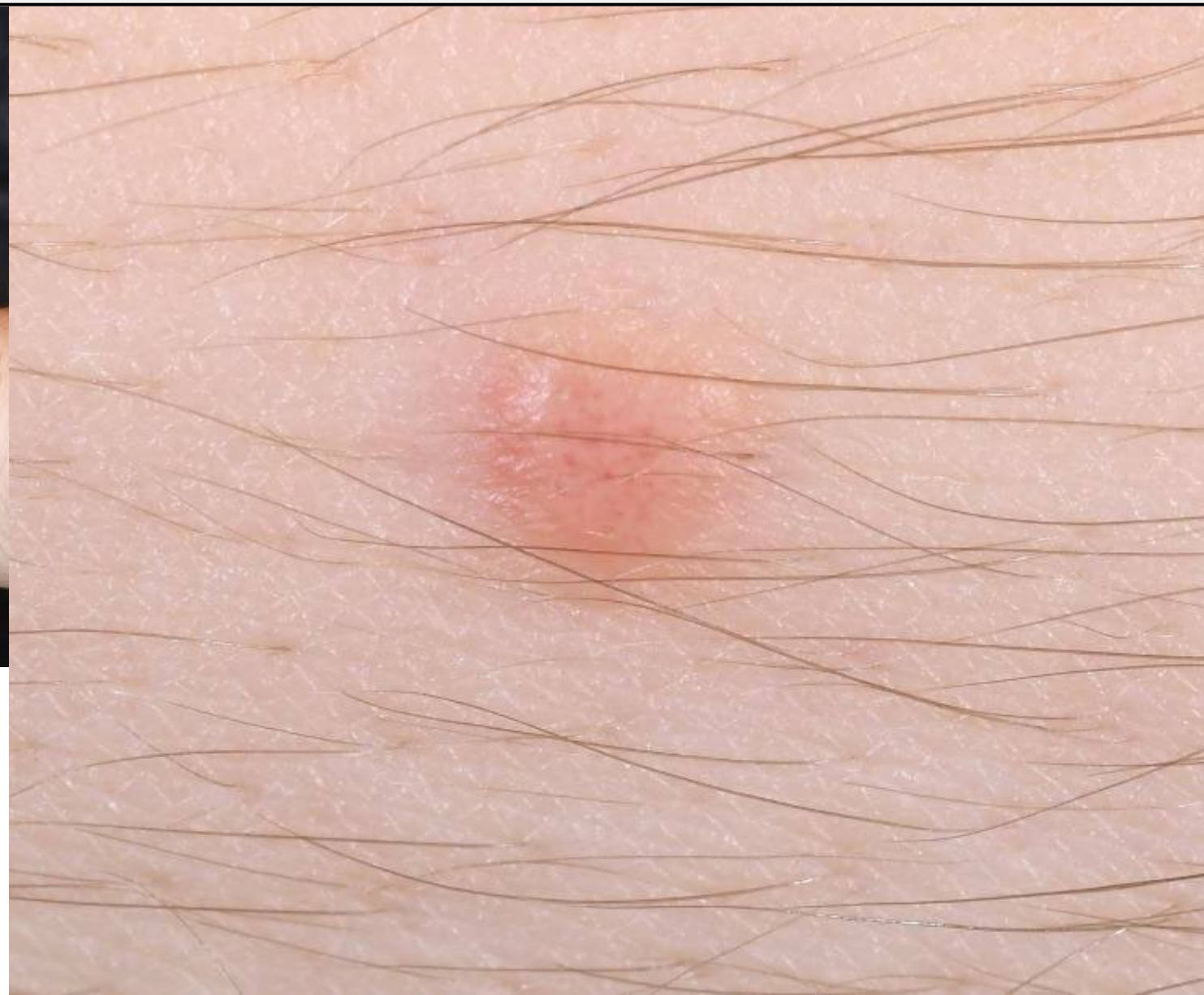


Sohier P. et al. Methylation analysis as a diagnostic tool for sweat gland tumours classification with emphasis on the distinction between digital papillary adenocarcinoma and mimickers. Histopathology 2025, 87, 618–623.

Case 2

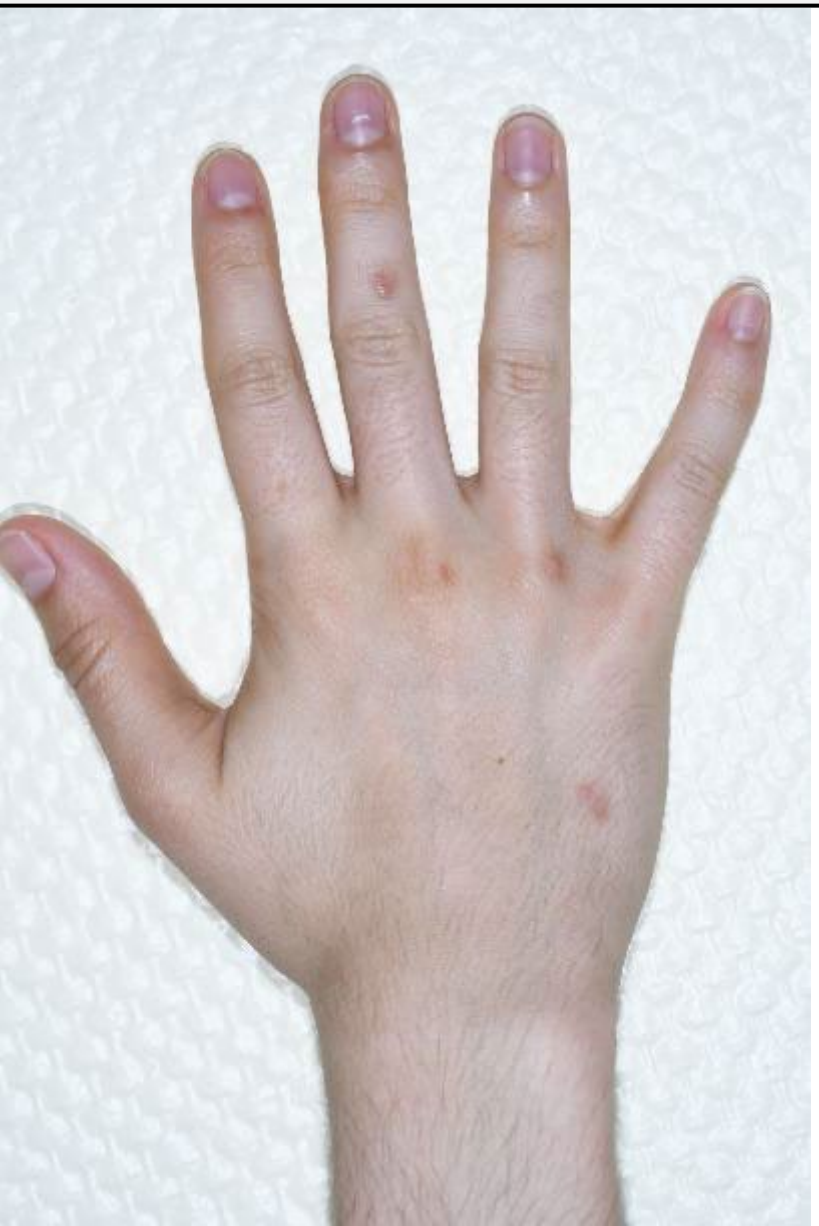
Clinical presentation:

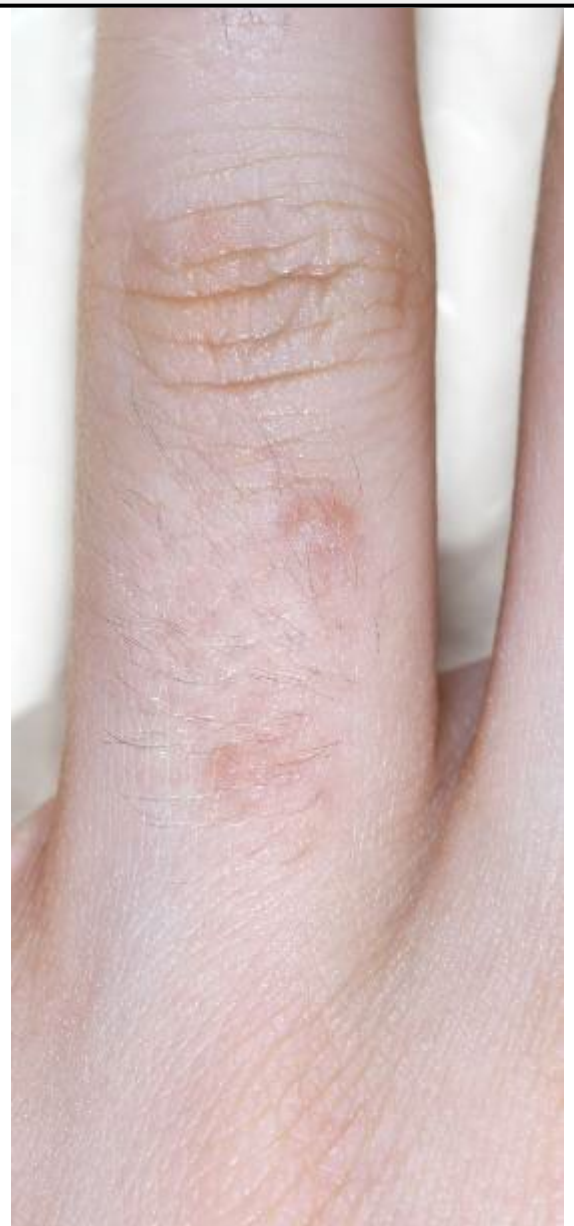


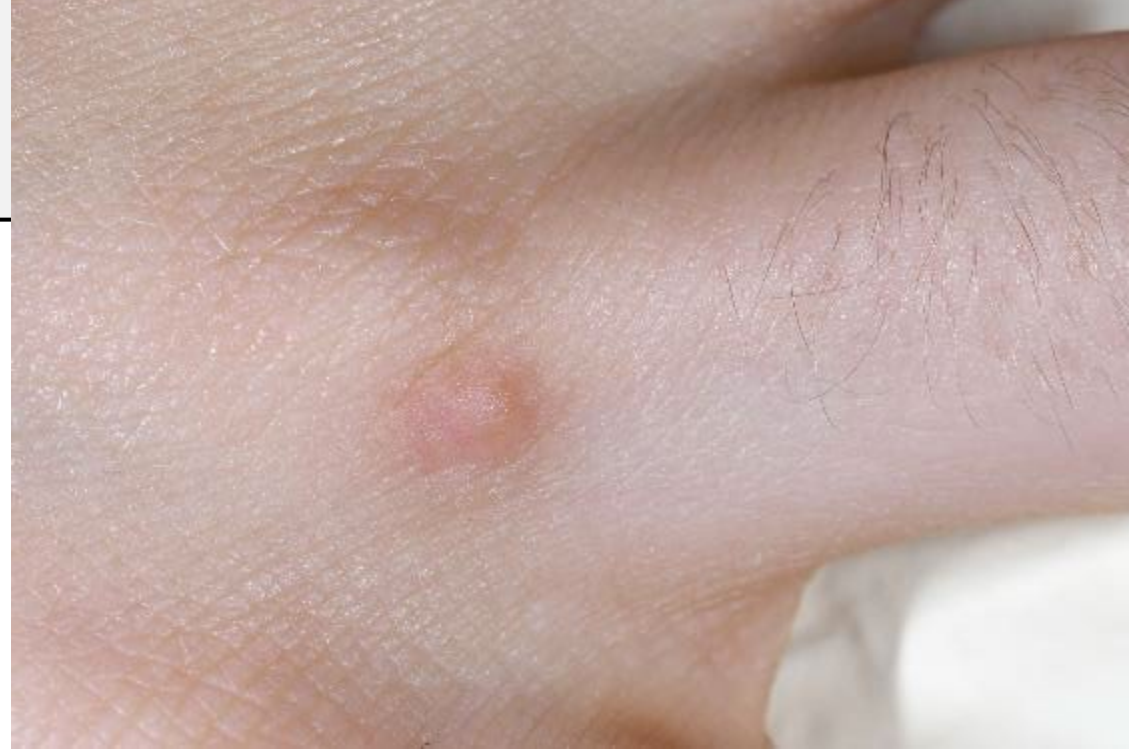


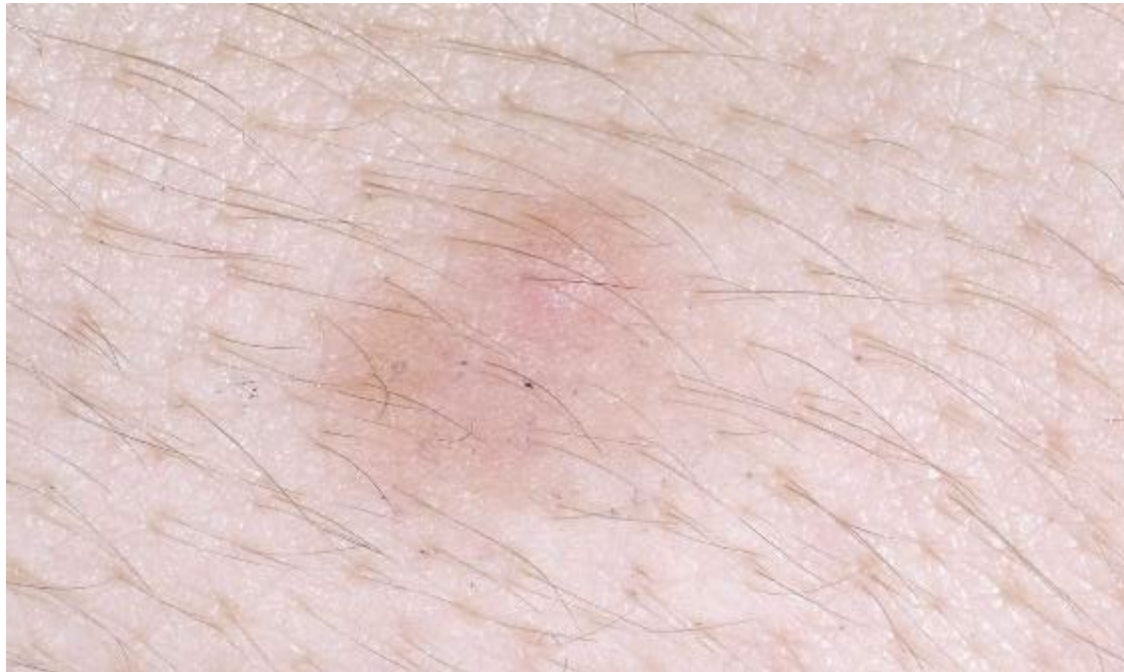
- 26-year-old female, otherwise healthy
- Development of multiple tiny nodules on both hands at the age of 14
- No family member had similar lesions
- viral warts → treatment without any effects
- Clinician suspected: dermatofibroma

Clinical presentation:









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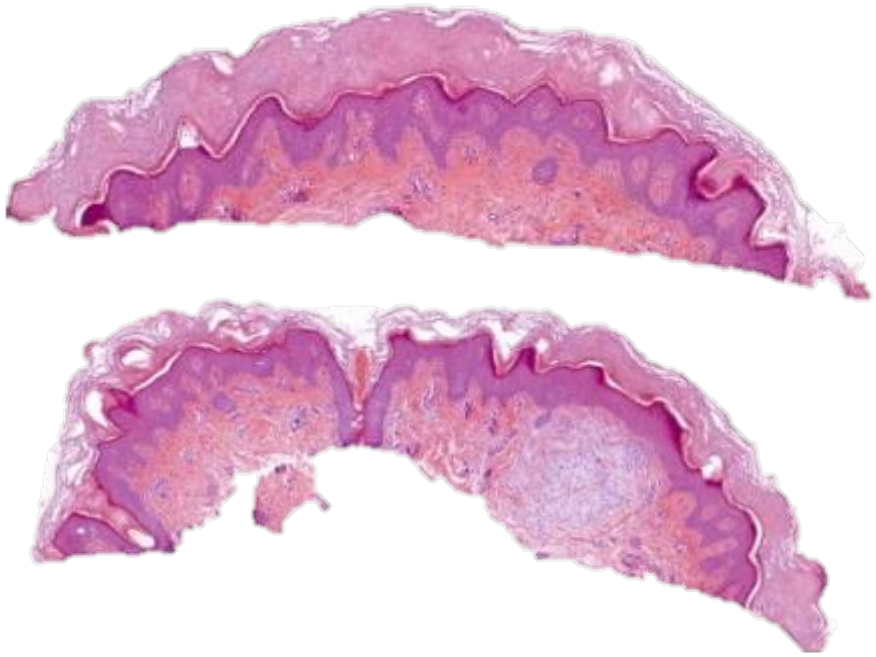

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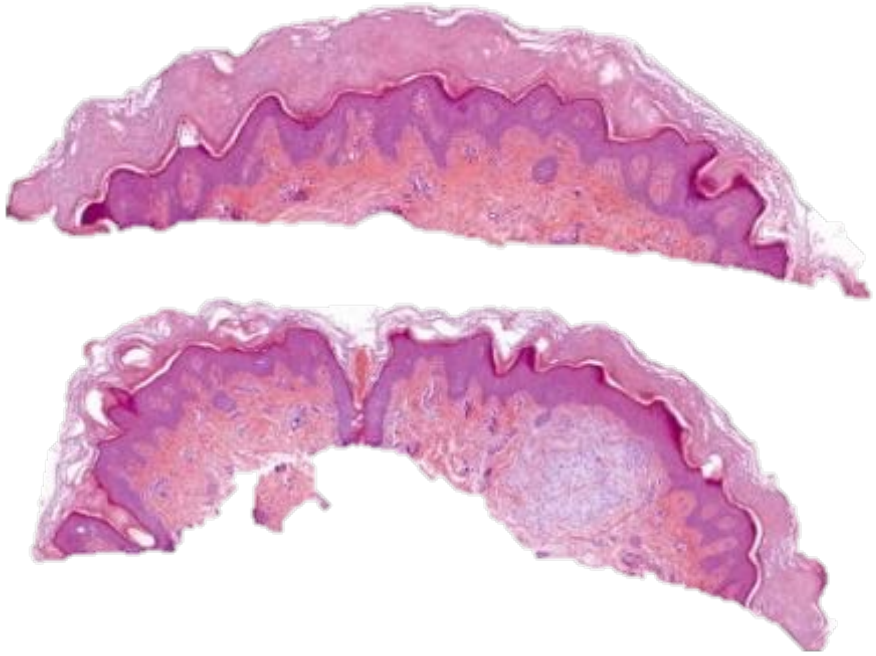
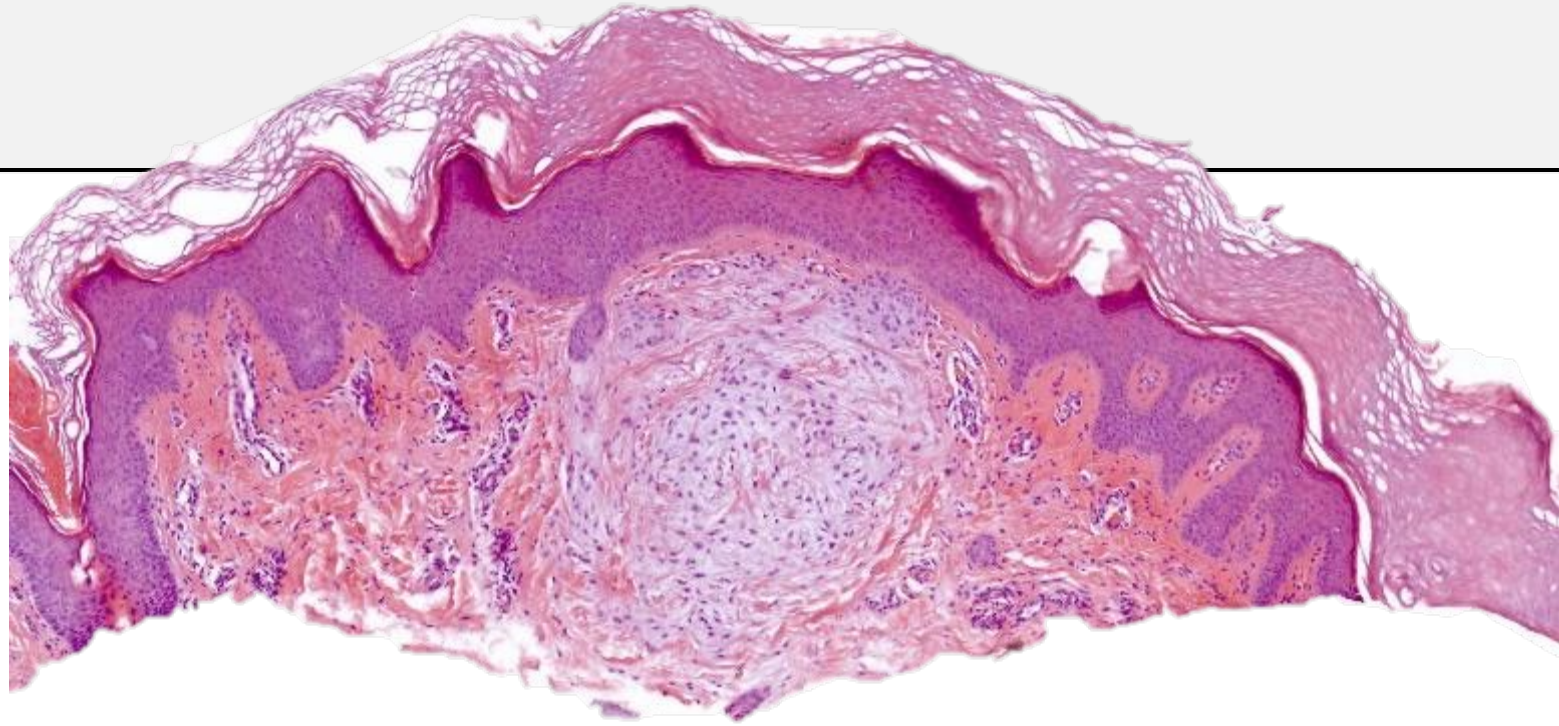
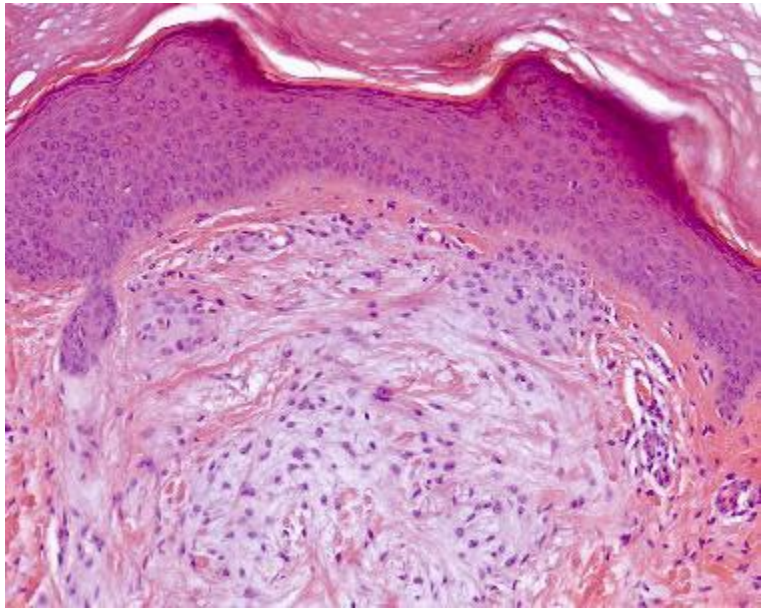
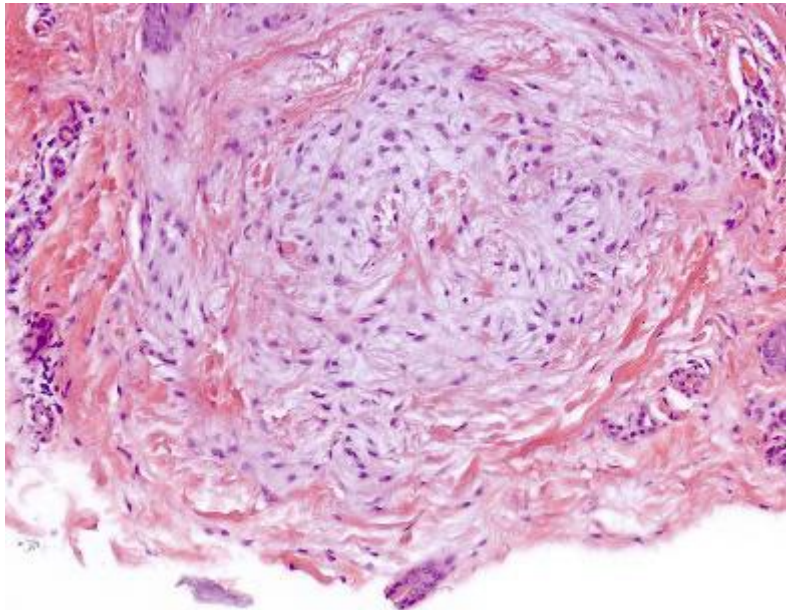


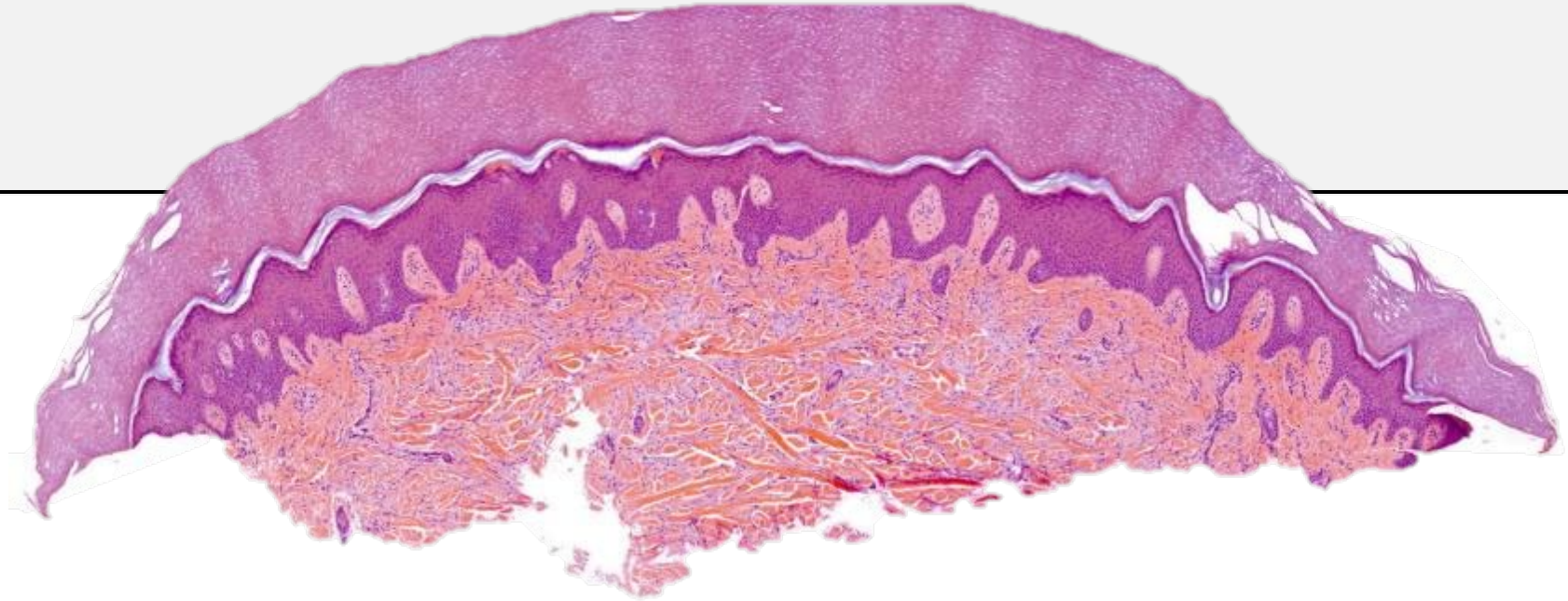


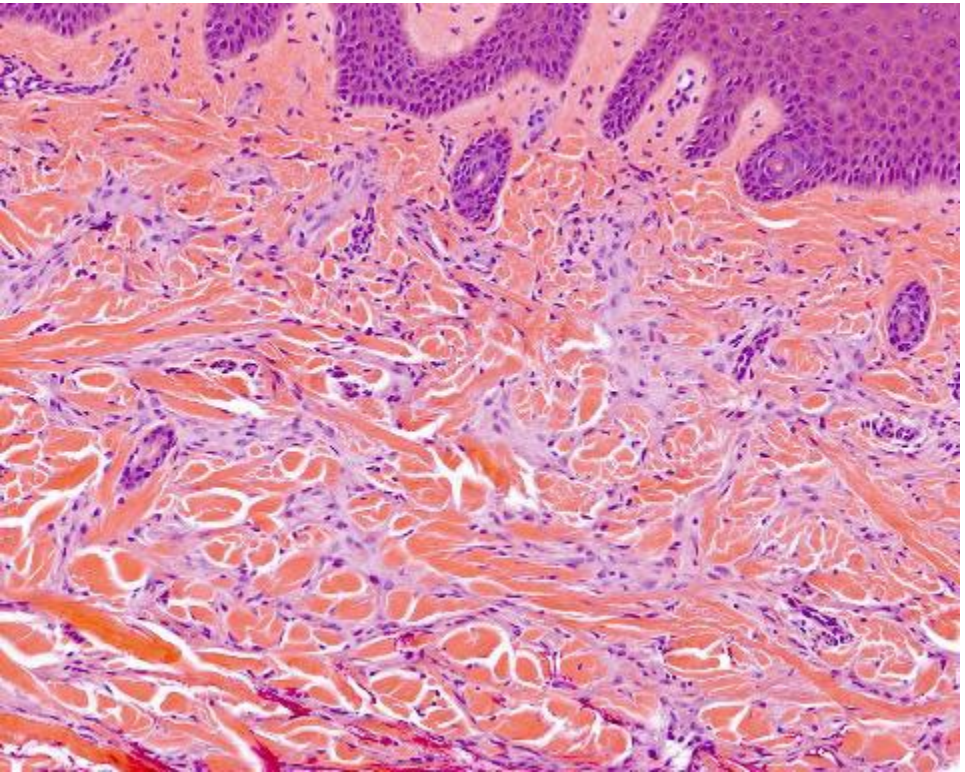


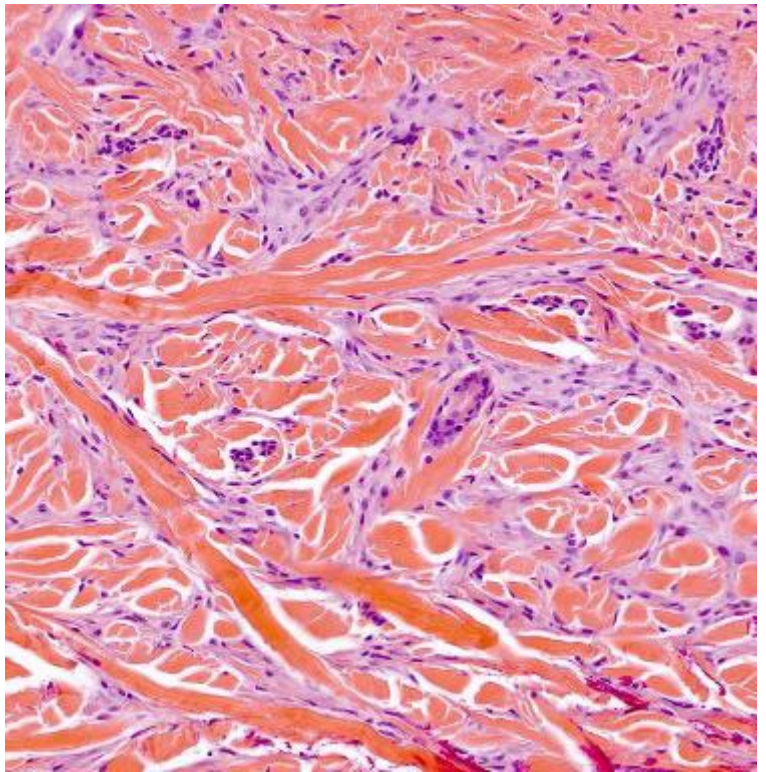


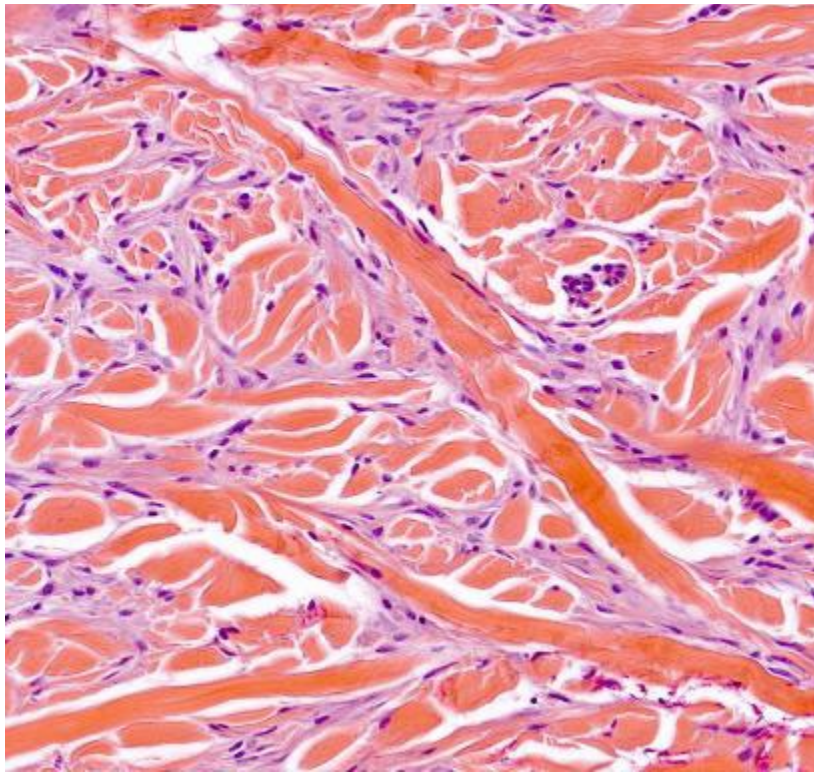
- Superficial epithelioid and spindled cells with a myxoid stroma
- Dermal nerve sheath myxoma?
- S100 and SOX-10 negative

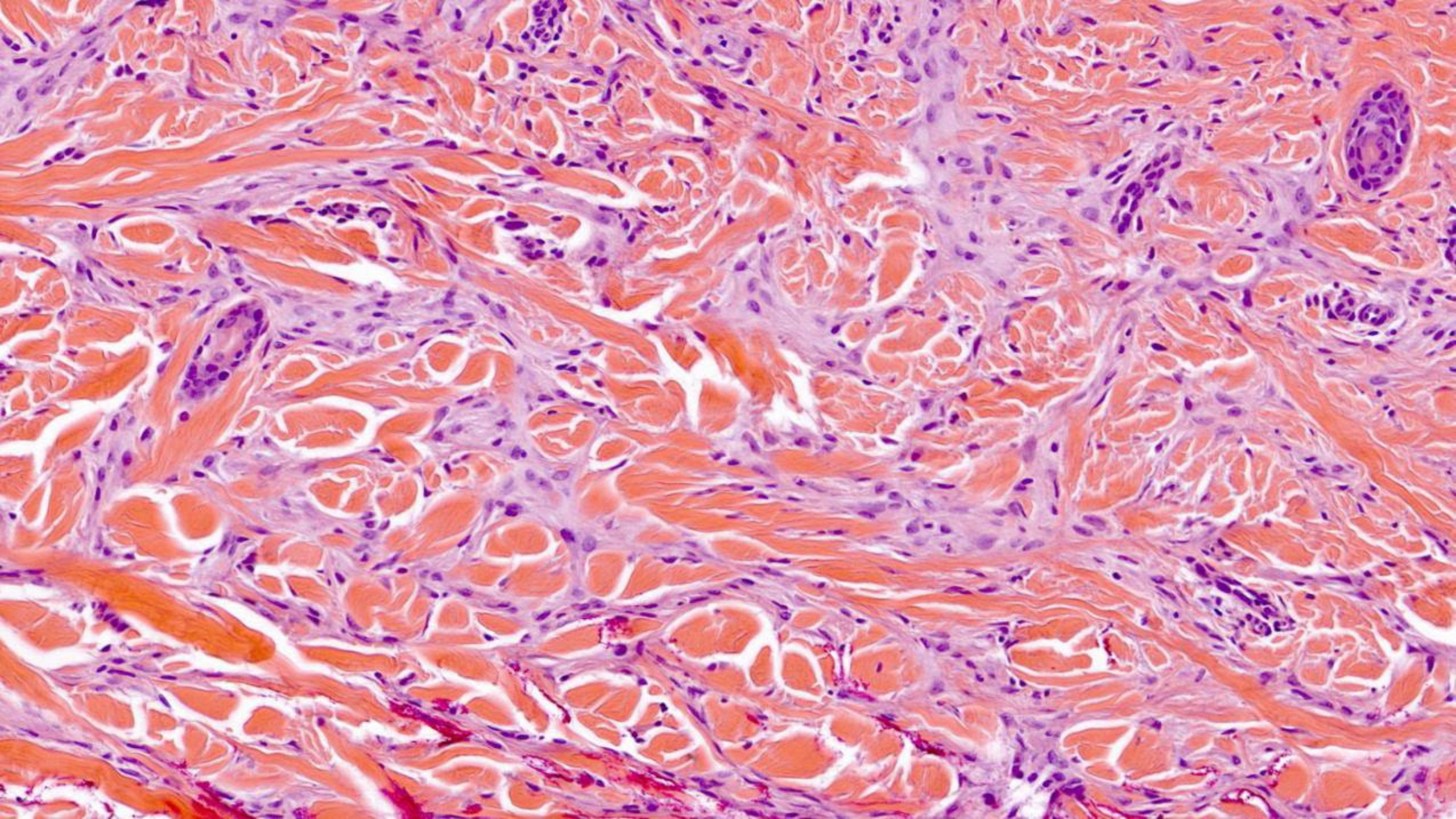
Histology:











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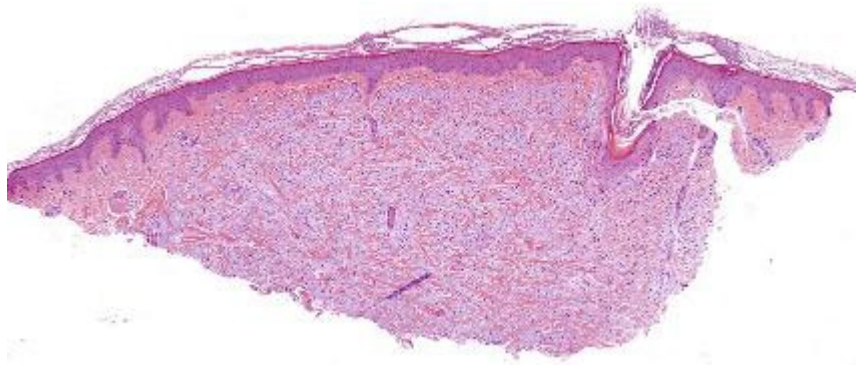
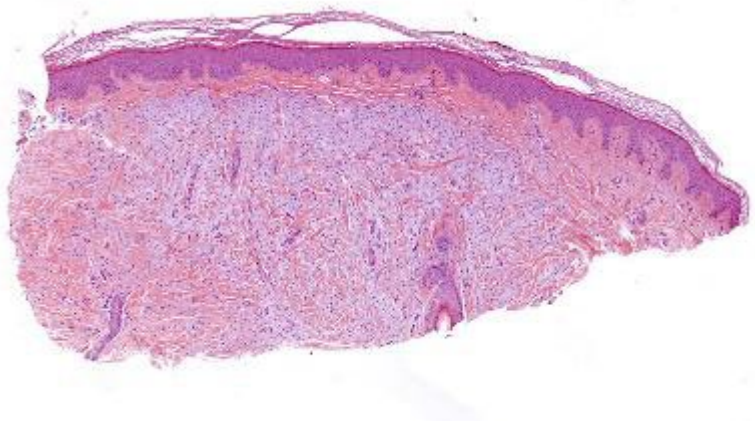
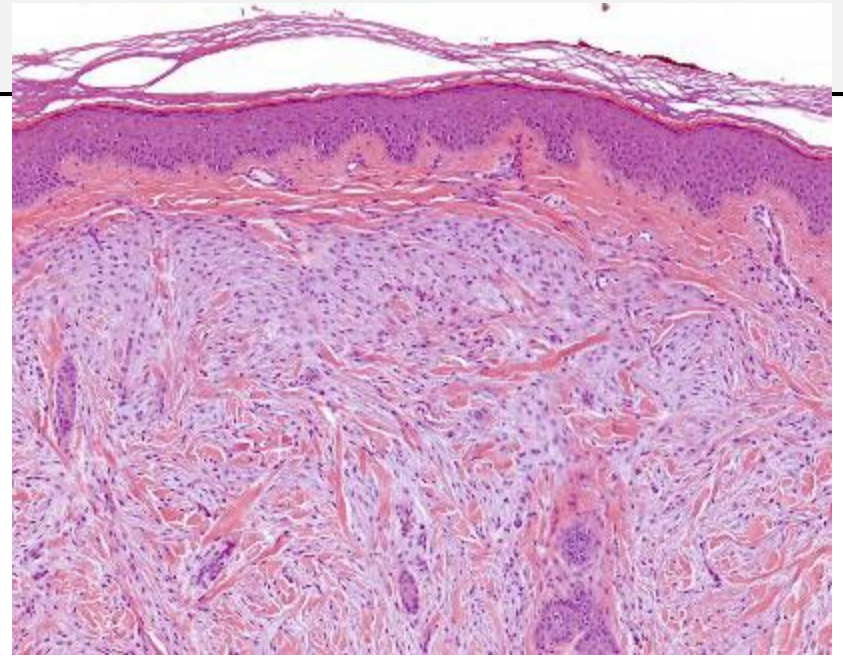
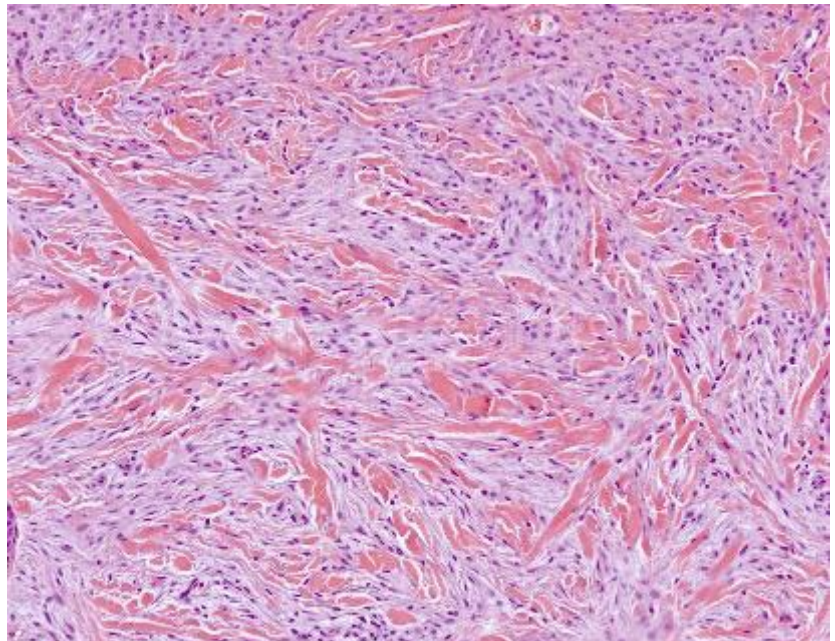
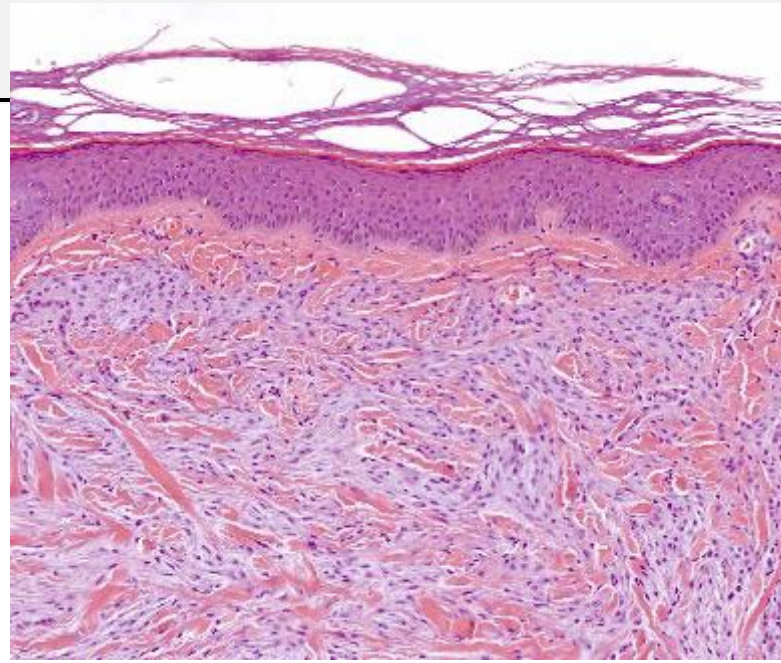
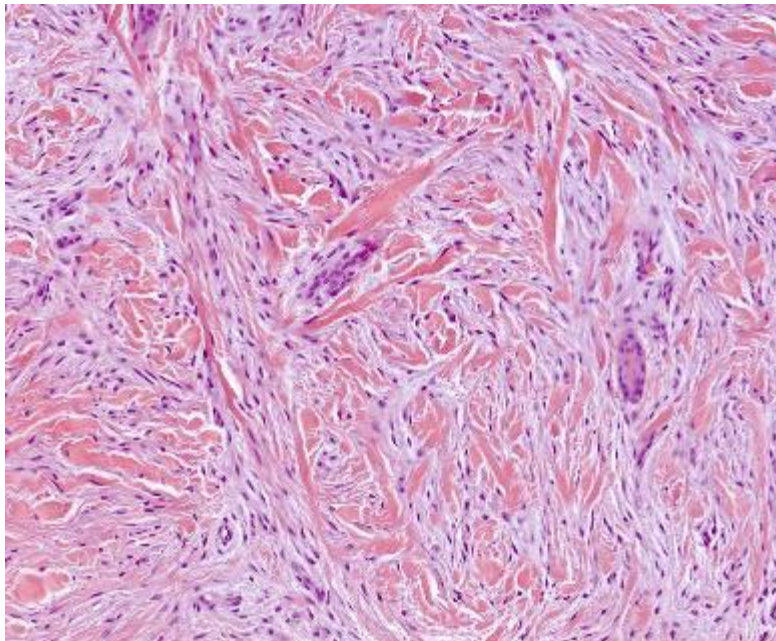


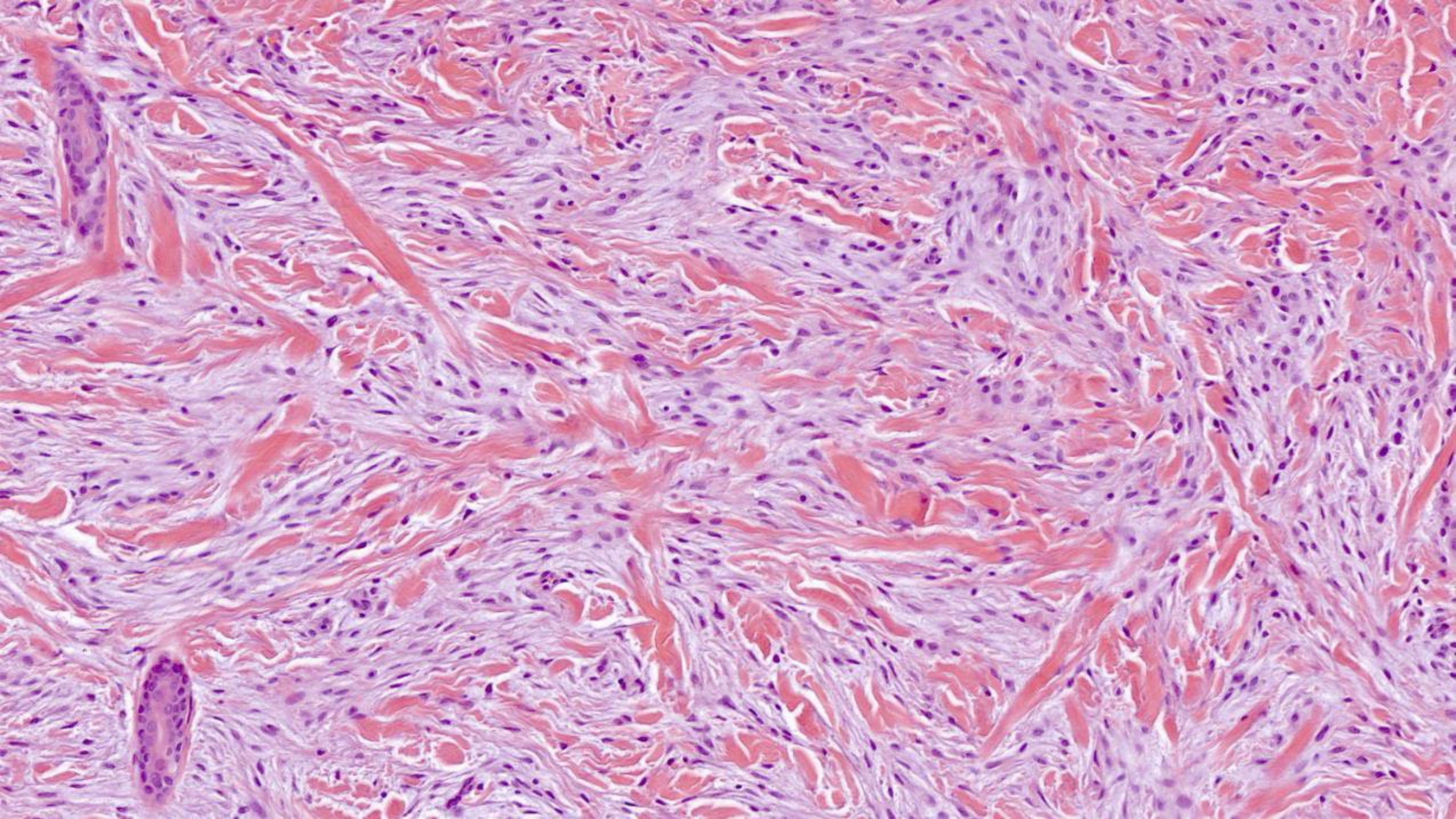




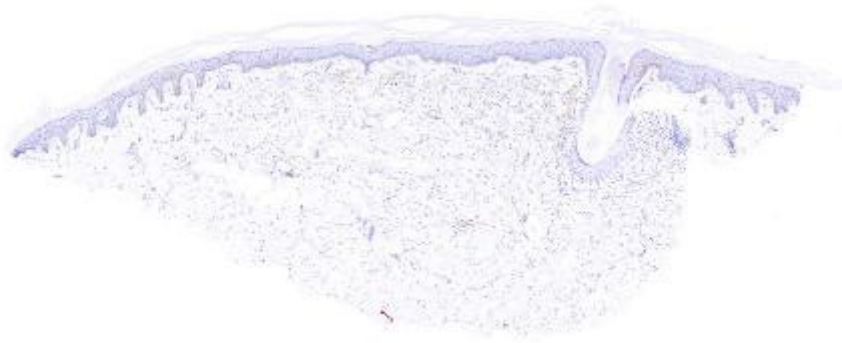




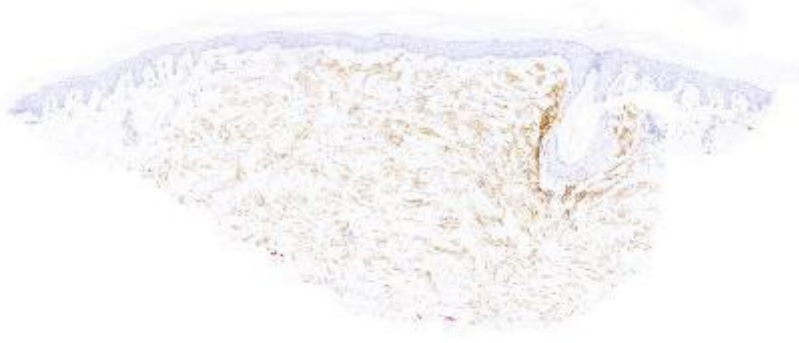




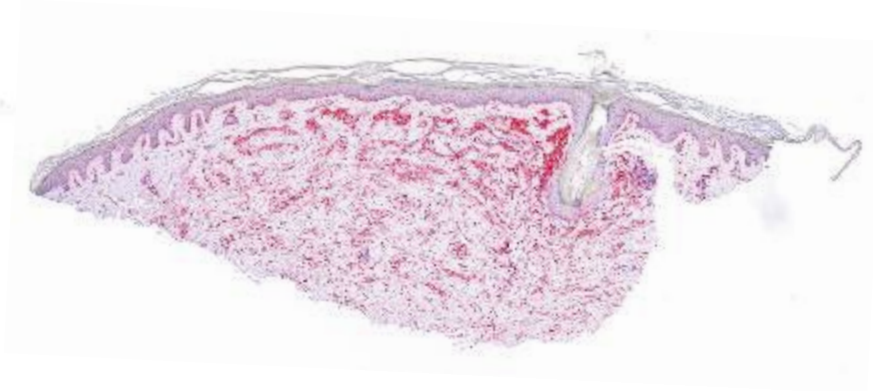
Immunohistochemistry:



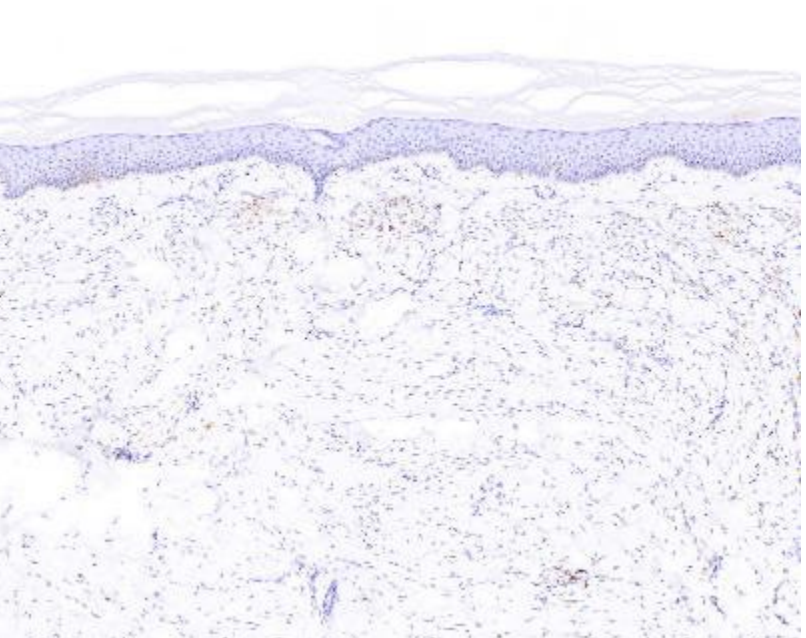
CD68

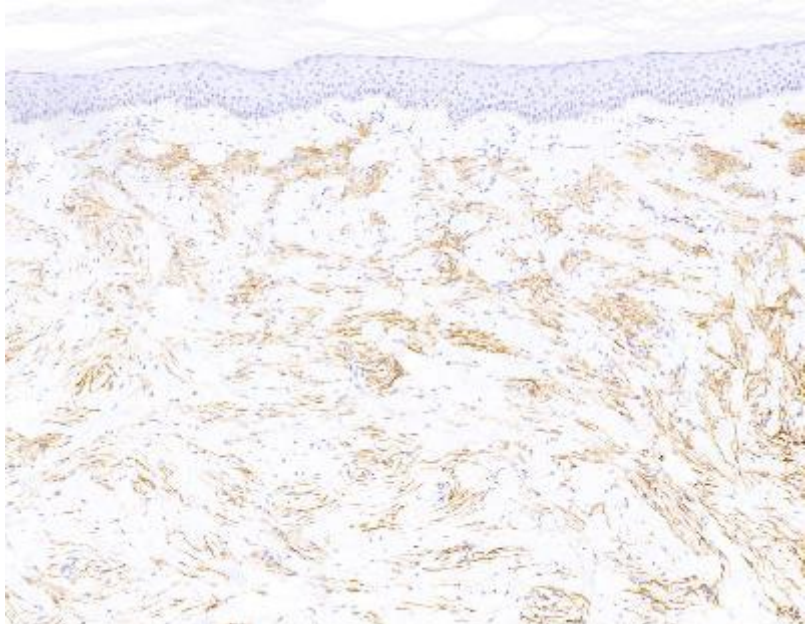


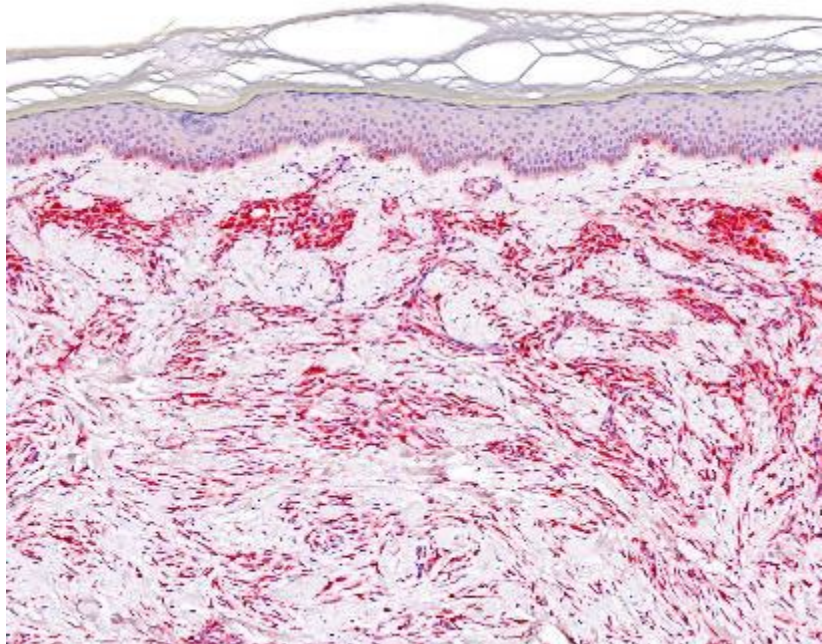
CD10



NKiC3







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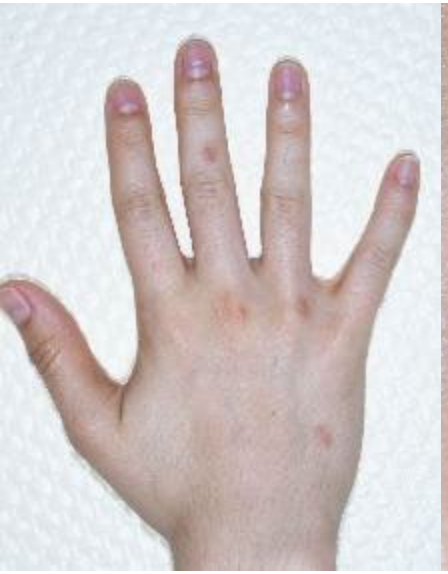
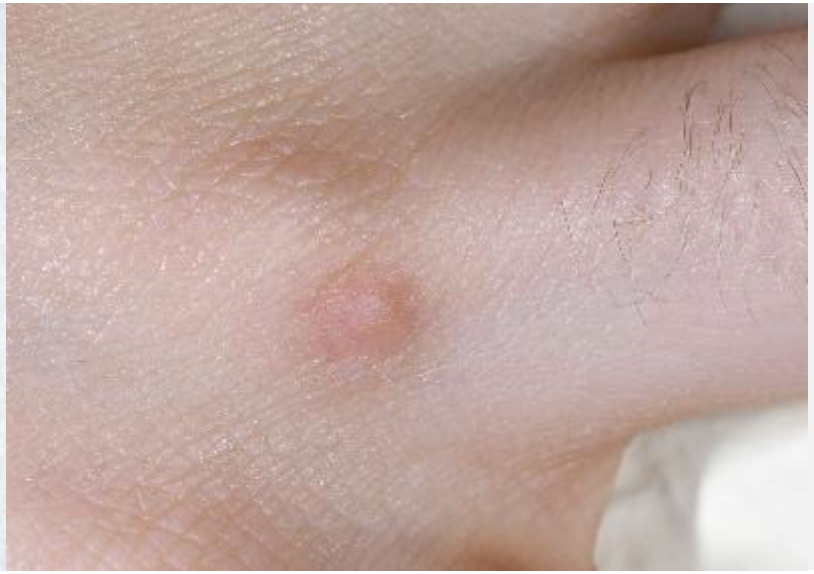


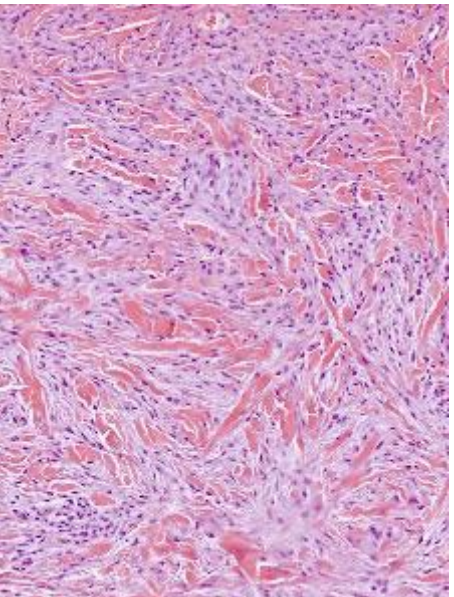


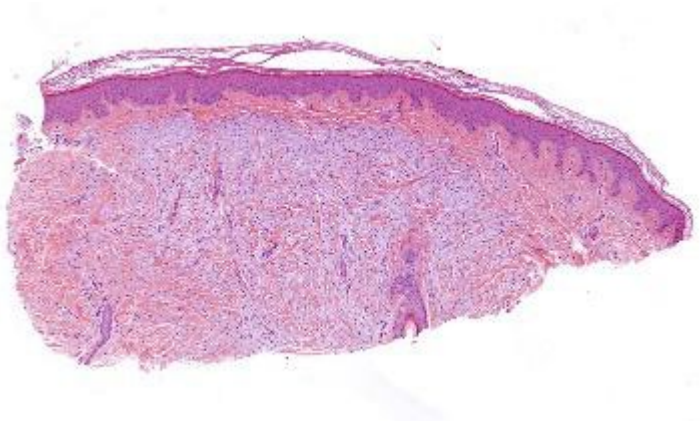
Clinical presentation:

- Multiple red-brownish papules on the back of the hand / fingers in a child
- Asymptomatic
- No family history

diagnosis

?

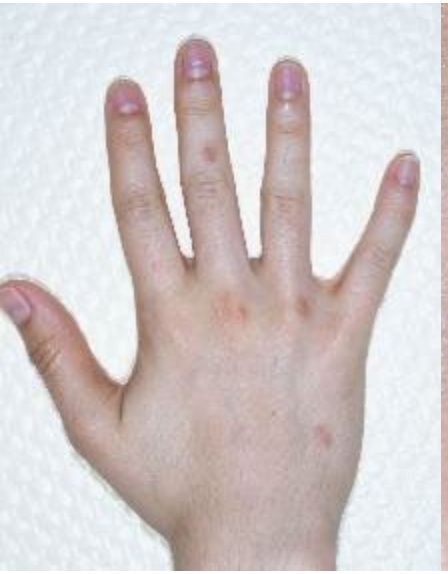


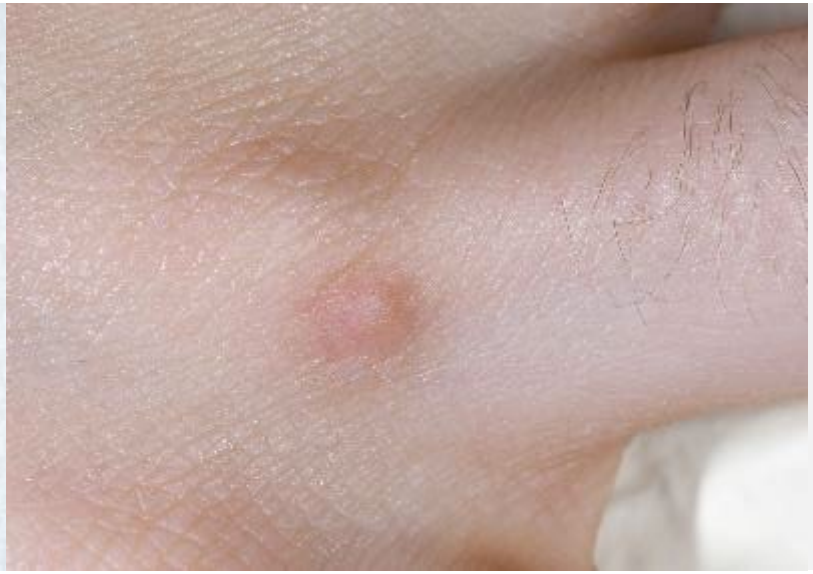


Histology:

- Dermal, well circumscribed, non-encapsulated
- Lobulated / nested epithelioid – spindle shaped cells with pale cytoplasm
- Myxoid stroma
- CD10+, NKiC3+, S100-, SOX-10-

Summary of the findings:





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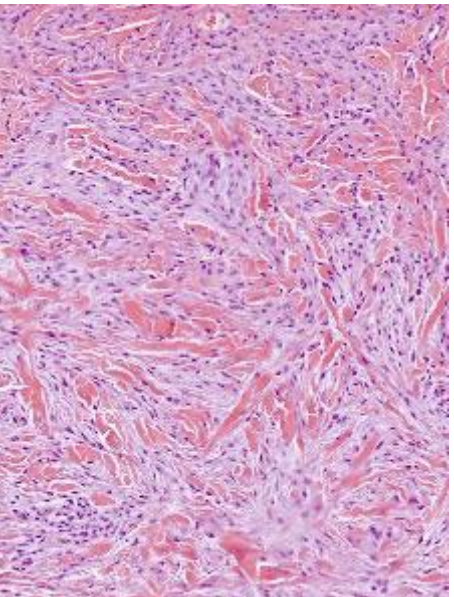
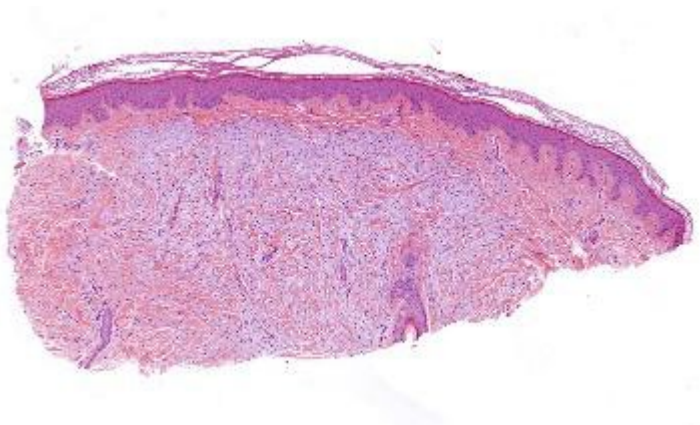
- No family history





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- CD10+, NKiC3+, S100-, SOX-10-

multiple cellular neuro-thekeomas

Literature: Multiple cellular neurothekeomas

Multiple neurothekeoma are very rare:
only eight cases
had been reported

- Children or young adults
- Slight male predominance
- Most common location: head and neck ←→
- Genetic background? In two cases family members were affected.

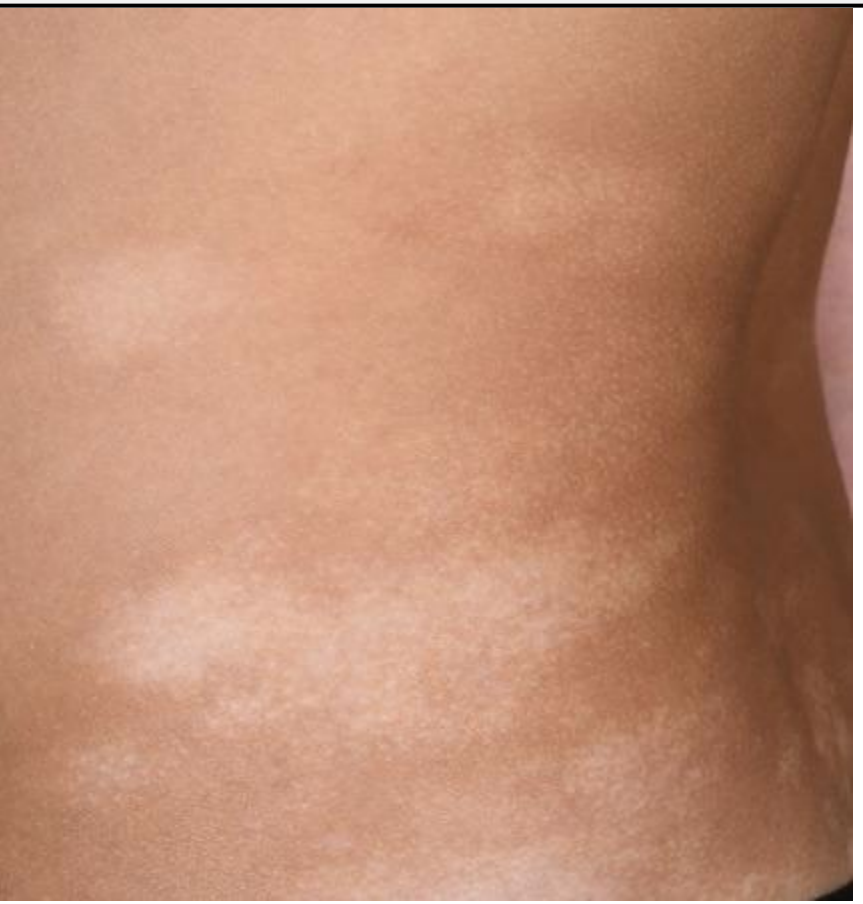
TABLE 2 Reported cases of multiple cellular neurothekeomas in a single patient

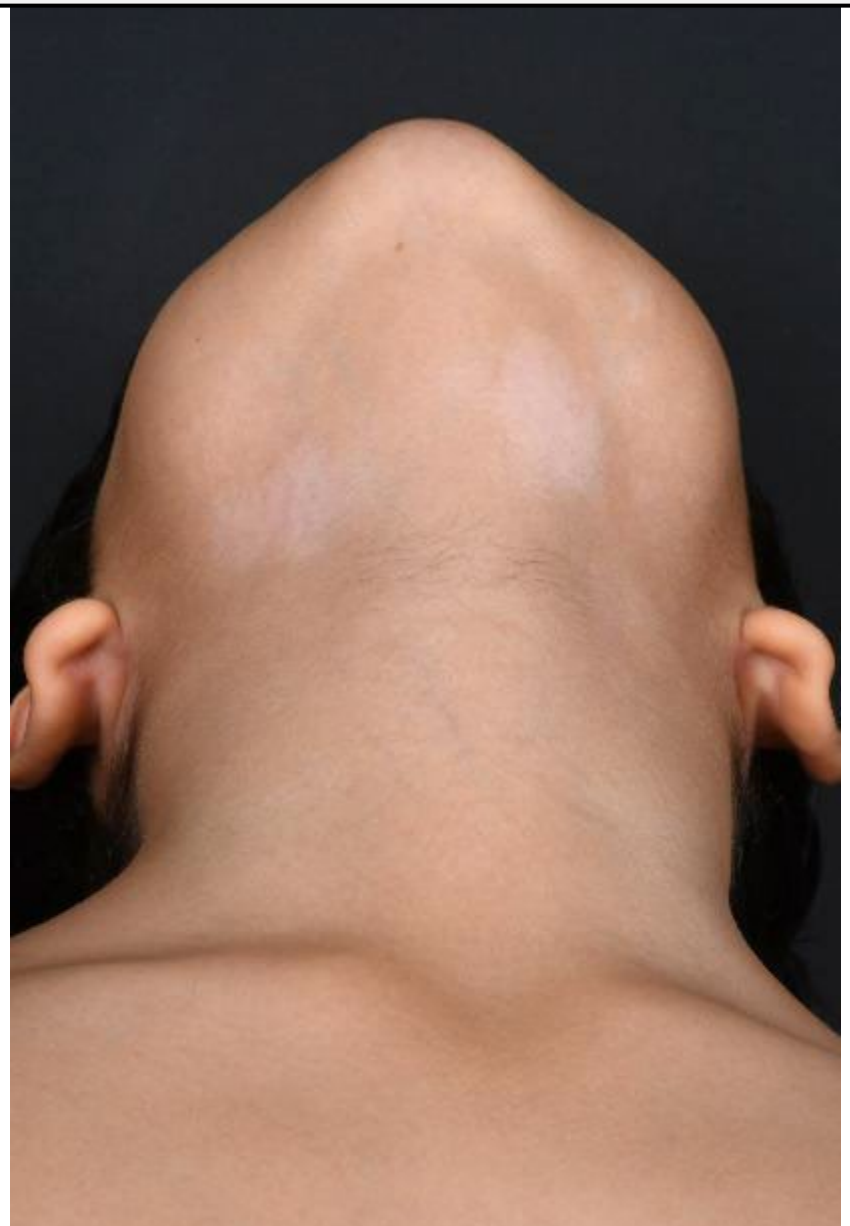
Reference	Age	Time course	Sex	Number of lesions	Locations of lesions	Clinical impression	Histologic diagnosis	Past medical history	Family history
Gill and Abi Daoud (current report)	46 years	8 years	Female	Nearly one dozen; 8 biopsy proven	Head and neck Upper extremity Lower extremity	Dermatofibroma and nevus	Cellular neurothekeoma (one initially diagnosed as dermatofibroma)	Hyperparathyroidism Left renal simple cysts Multiple uterine fibroids Epidermoid cyst of the left shoulder Fibroadenoma of the left breast	Ovarian cancer
Mahalingam et al[7]	30 years	12 years	Male	15 in total; 12 biopsy proven	Head and neck	Cyst, scar, molluscum contagiosum	Cellular neurothekoma (two initially diagnosed as angiofibroma, two initially diagnosed as intradermal Spitz nevus, and two initially diagnosed as keloidal nodules)	Not specified	Not specified
Garcia-Gutierrez et al[8]	16 years	2 years	Male	Approximately 30 (all biopsy proven)	Head and neck	Angiofibroma, adnexal neoplasms, and xanthogranulomas	Desmoplastic cellular neurothekeoma	Otherwise healthy	No similar lesions or history of syndromes
Kelly et al[9]	11 months	6 months	Male	Multiple (exact number not specified)	Head and neck Trunk	Not specified	Cellular neurothekeoma	Otherwise healthy, no pregnancy or delivery complications	Older brother with multiple neurothekeomas
Kelly et al[9]	21 years	Since childhood	Male	Multiple (exact number not specified)	Not specified	Molluscum contagiosum	Cellular neurothekeoma	Not specified	Younger brother with multiple neurothekeomas
Almeida et al[10]	9 years	3 months	Female	Multiple (exact number not specified)	Head and neck	Differential of neurofibroma, schwannoma, and myofibroma	Desmoplastic cellular neurothekeoma	Not specified	Not specified

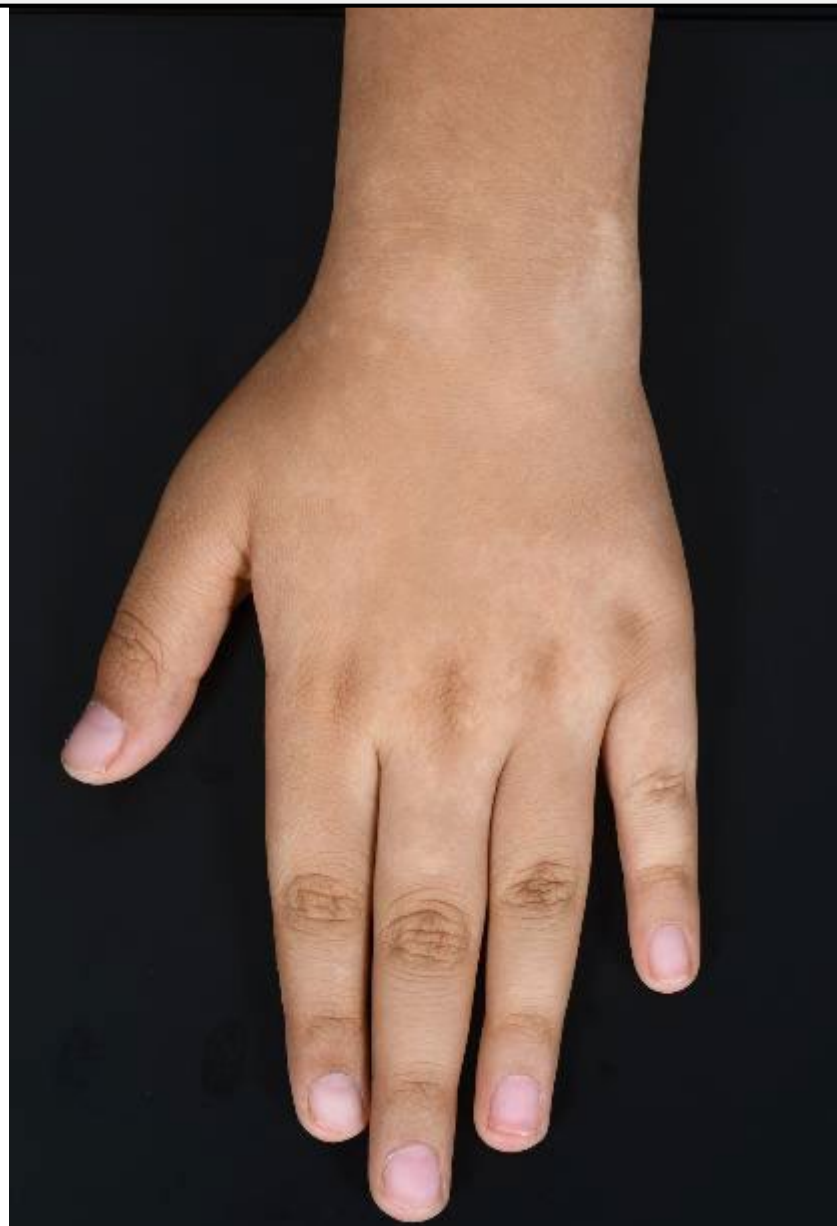
Gill P and Abi Daoud MS: Multiple cellular neurothekeomas in a middle-aged woman including the lower extremity: A case report and review of the current literature. J Cutan Pathol. 2019;46:67–73.

Case 3

Clinical presentation:

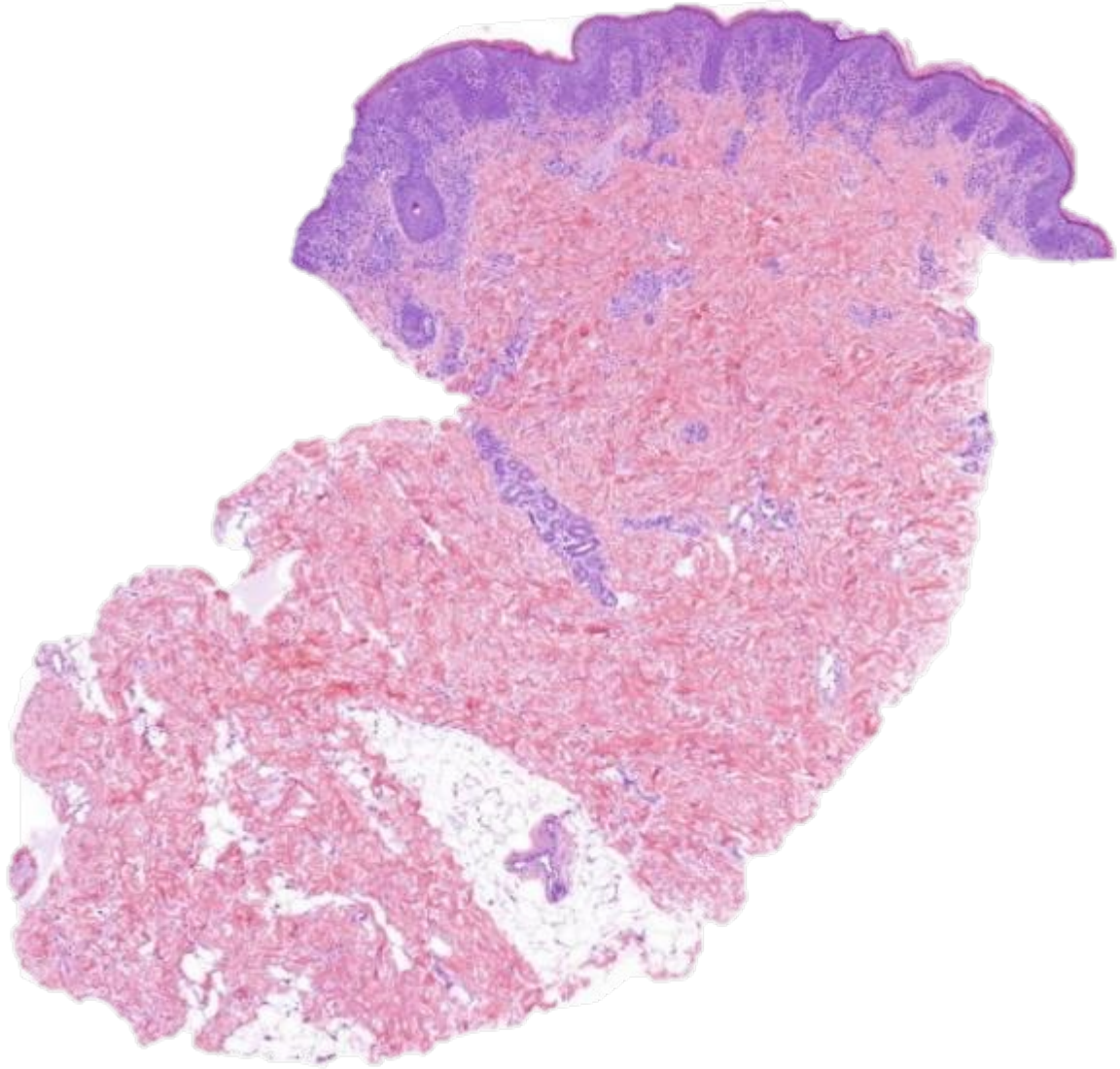


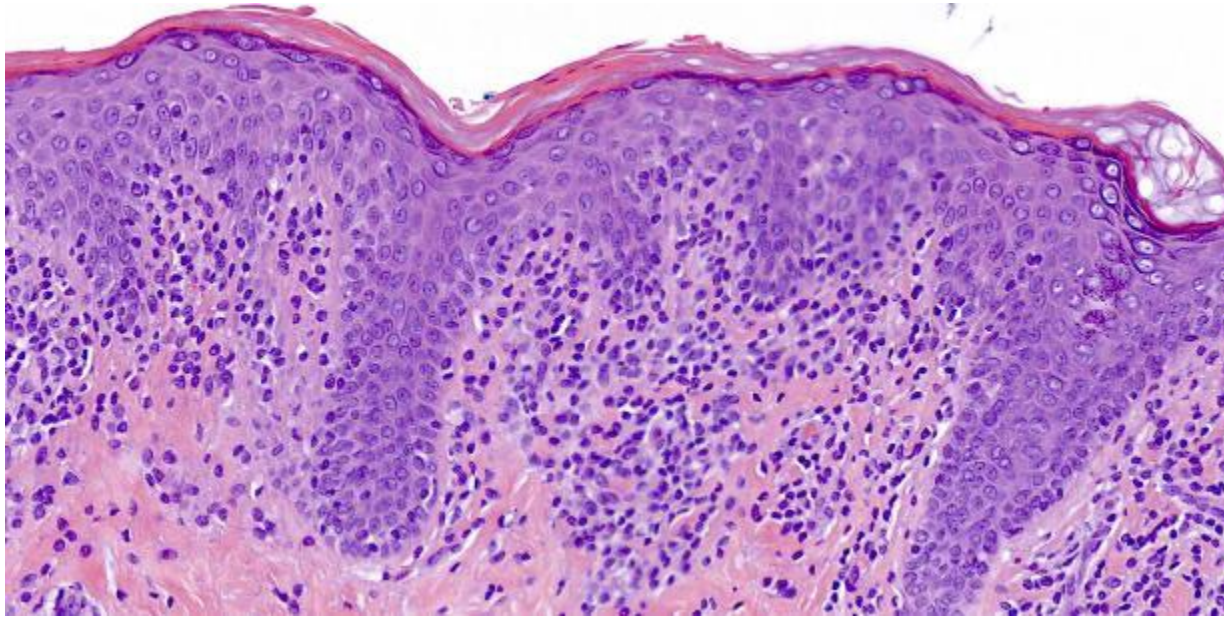


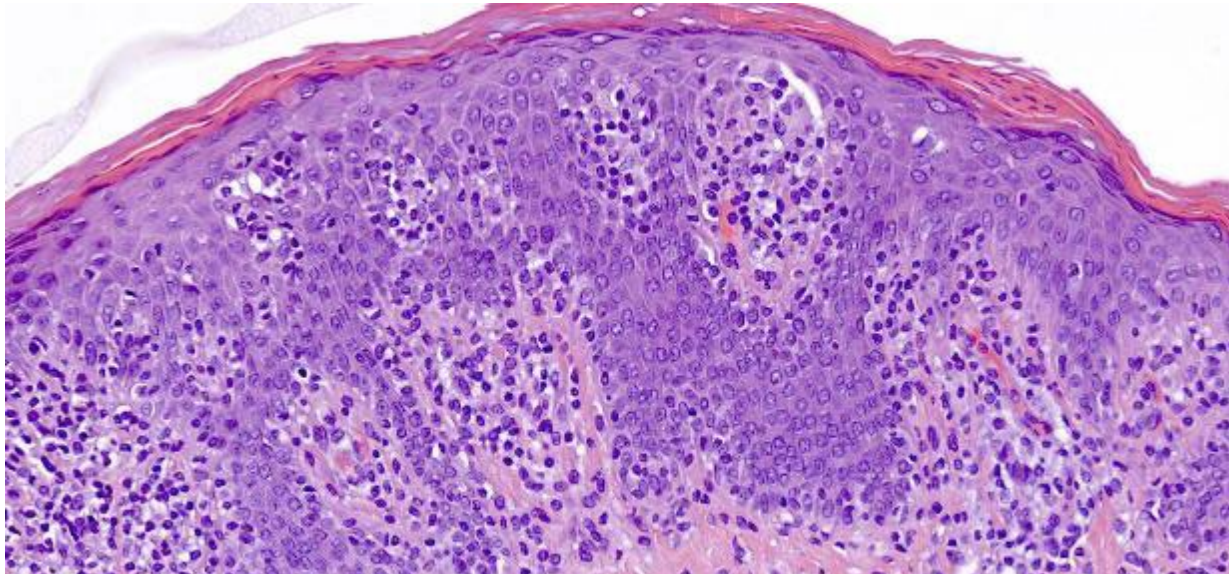


- 7-year-old boy
- Fitzpatrick skin type IV
- Since a few months progressive hypopigmented maculae

Histology:

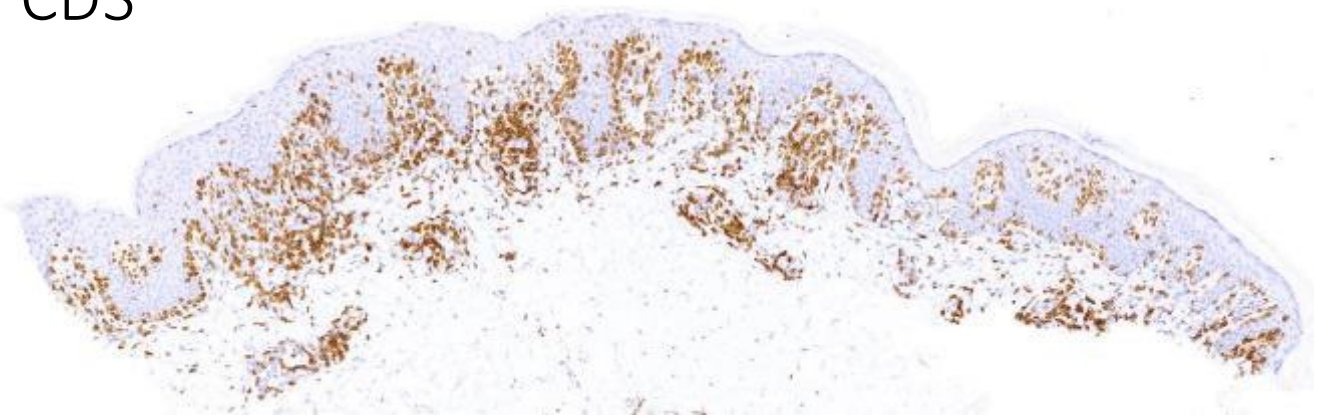




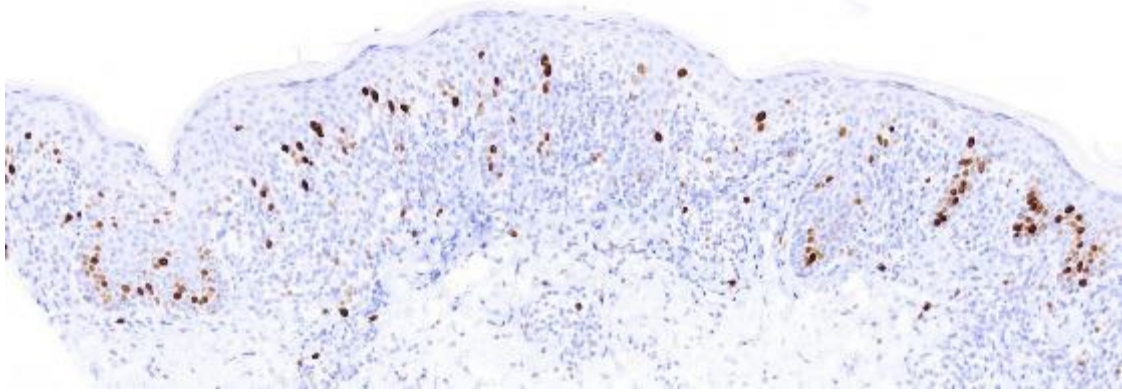


Immunohistochemistry:

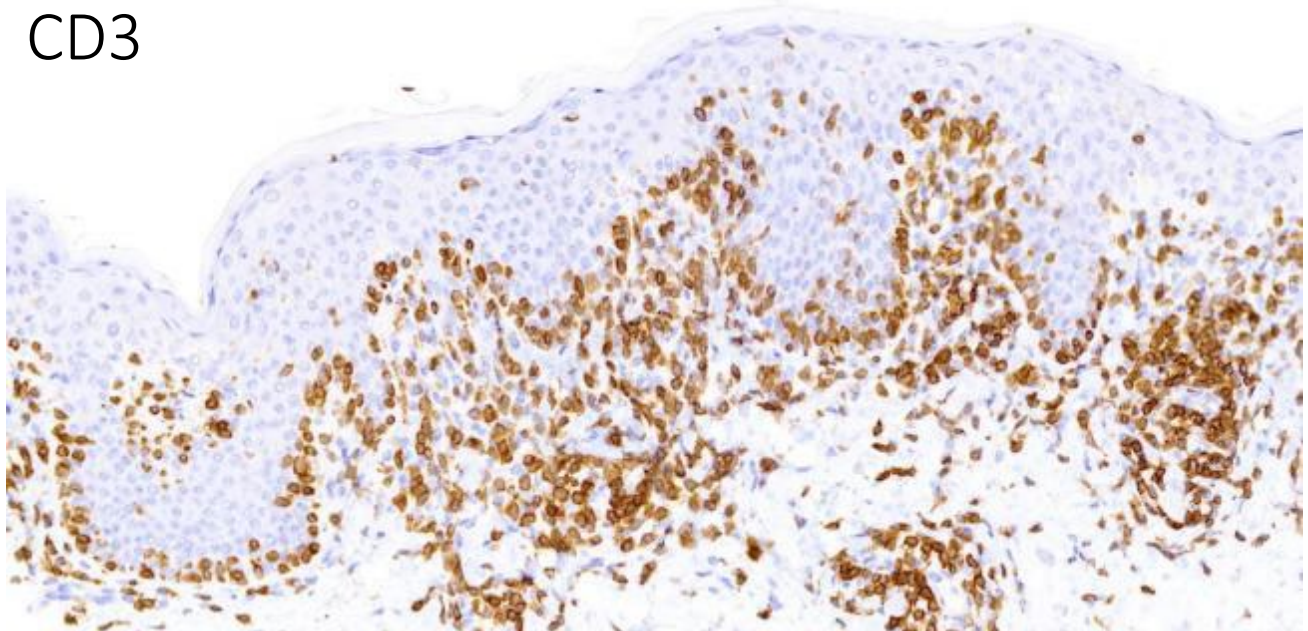
CD3



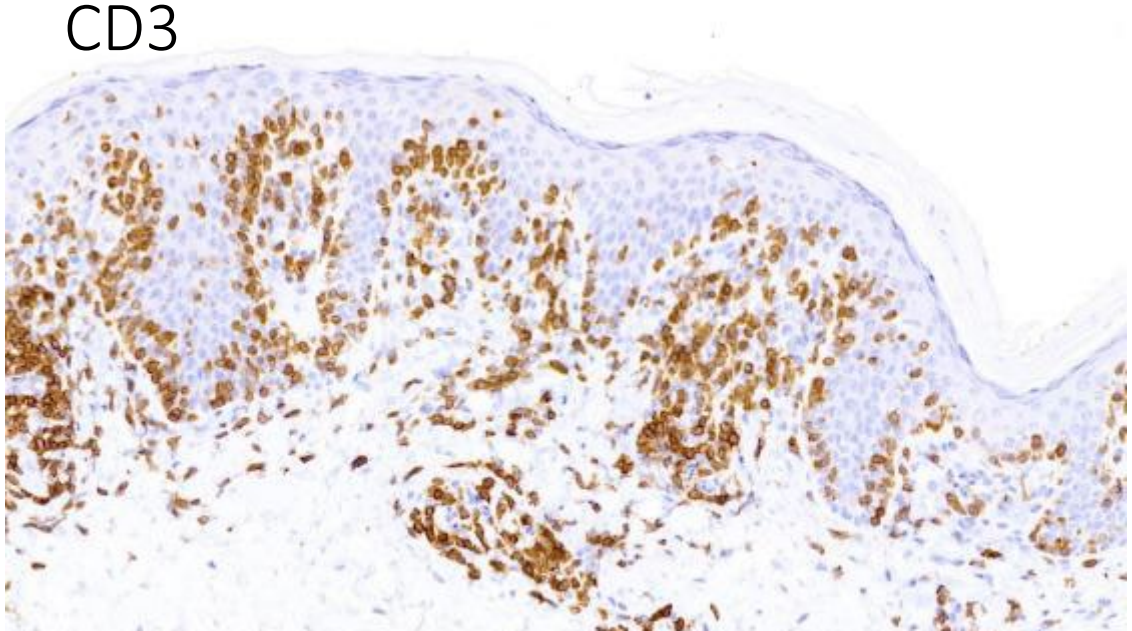
Ki67



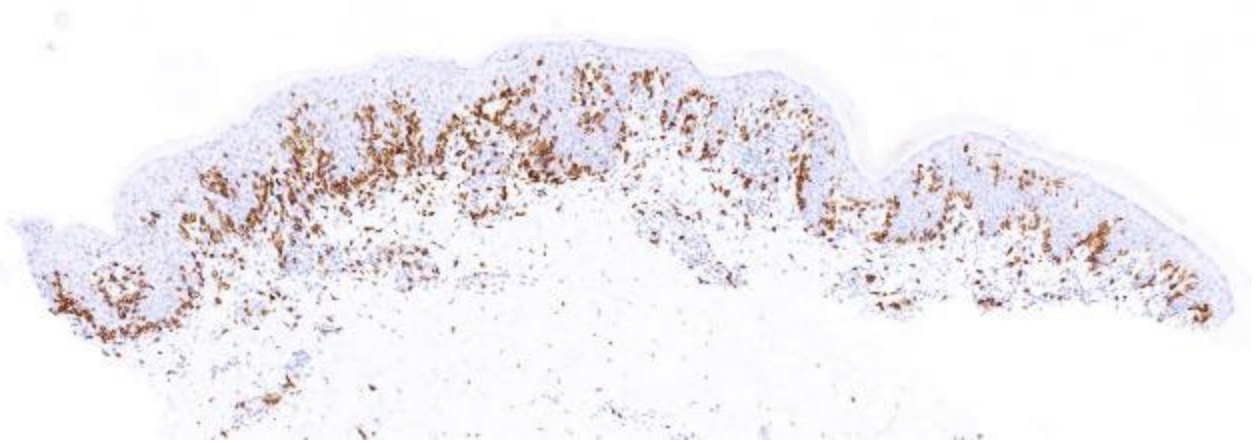
CD3

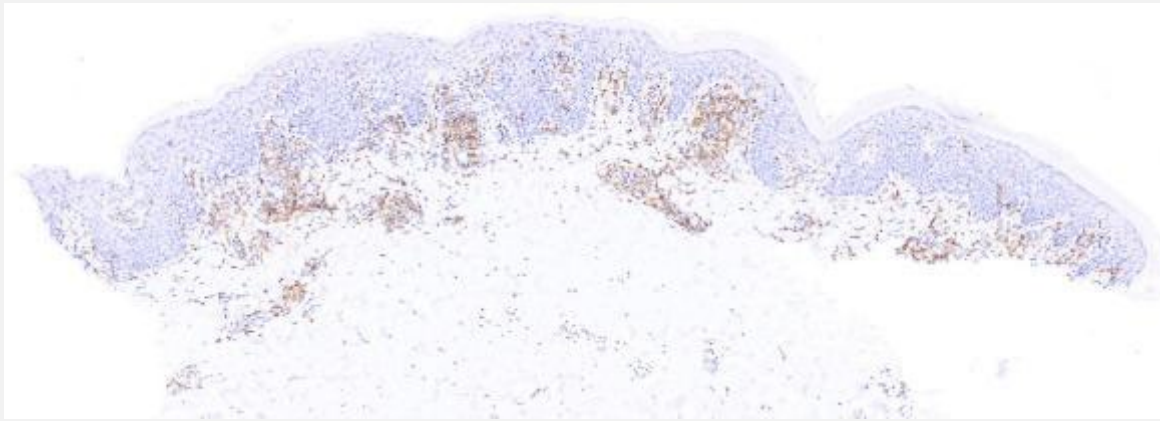


CD3

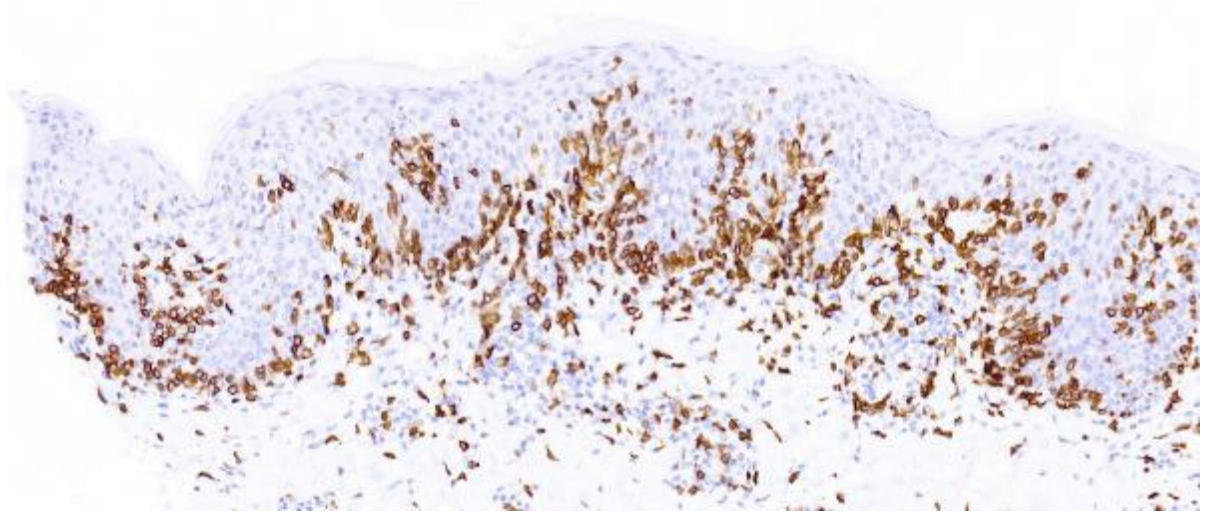


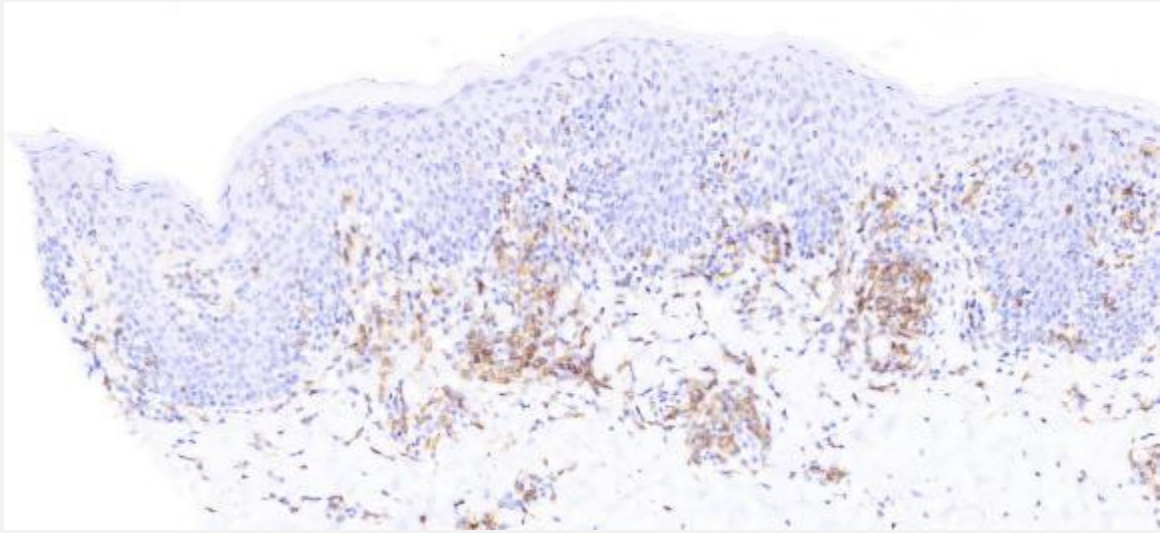
Immunohistochemistry:



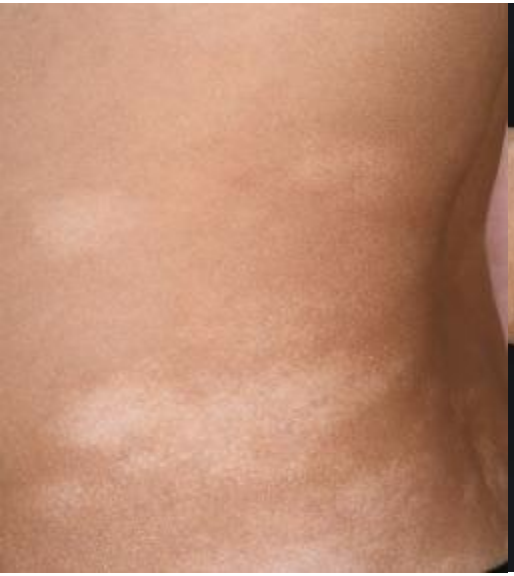


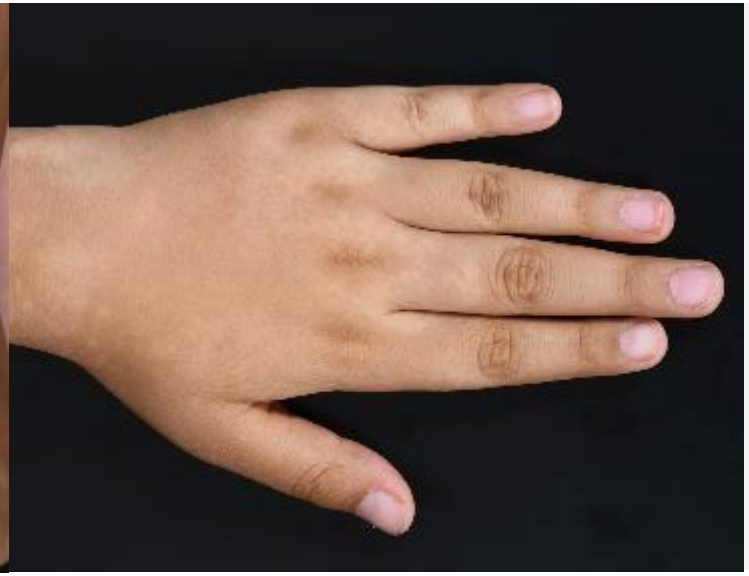
CD8 >> CD4





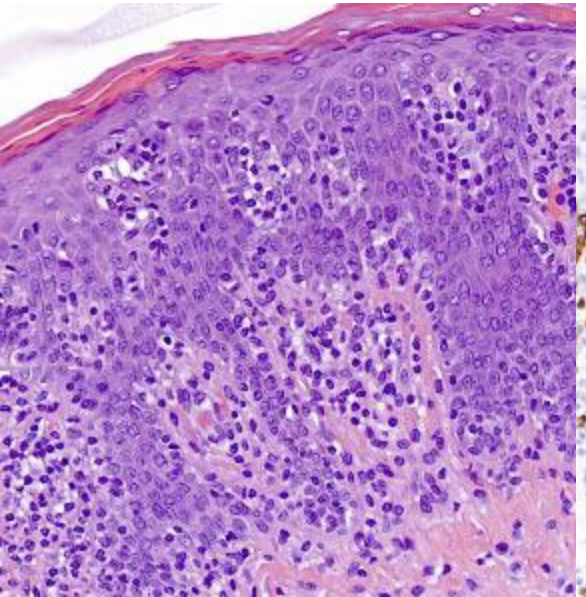
Summary of the findings:

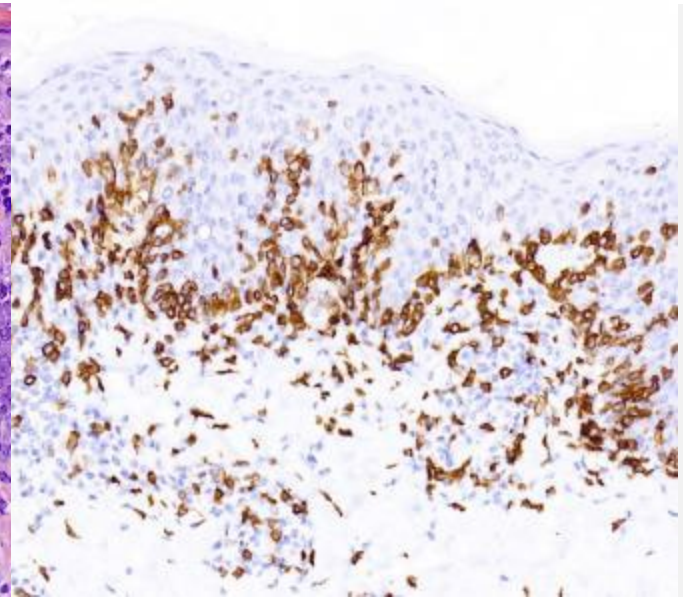




Clinical presentation:

- Sharply demarced hypopigmentations
- Child





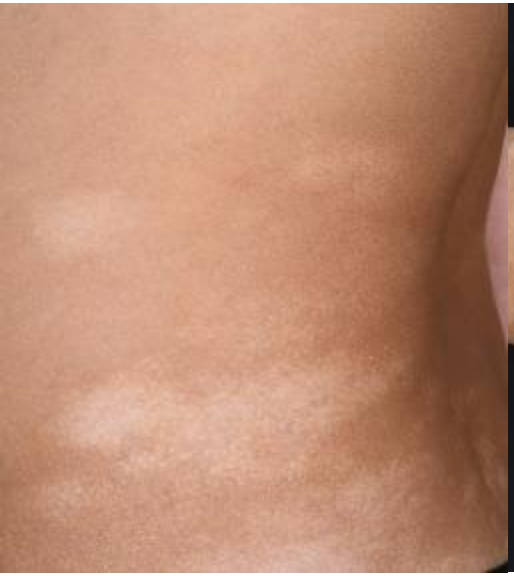
Histology:

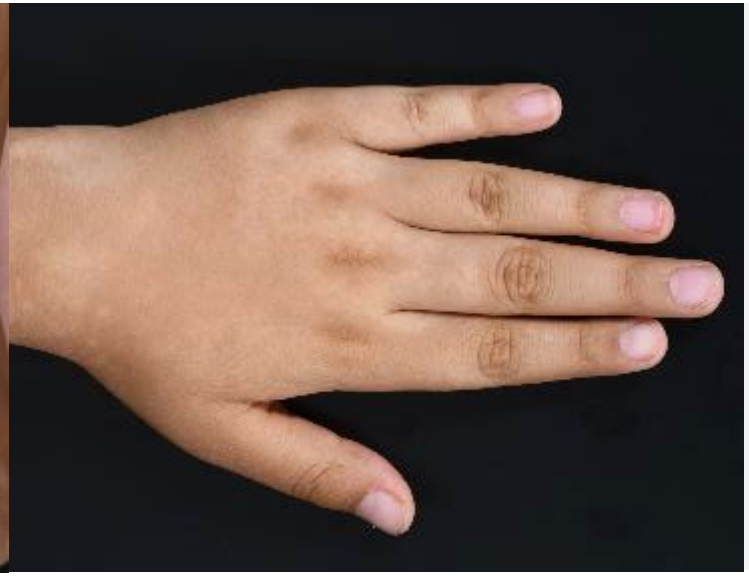
- Band-like lymphocytic infiltrate
- Prominent lining-up
- CD3+, CD8+, CD4-

diagnosis

?

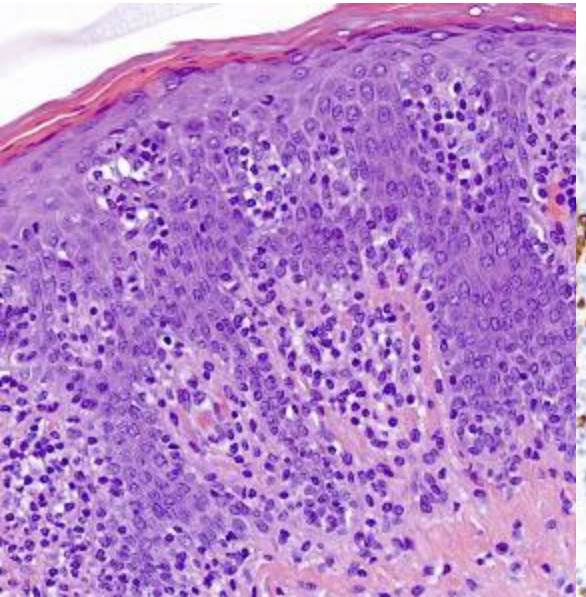
Summary of the findings:

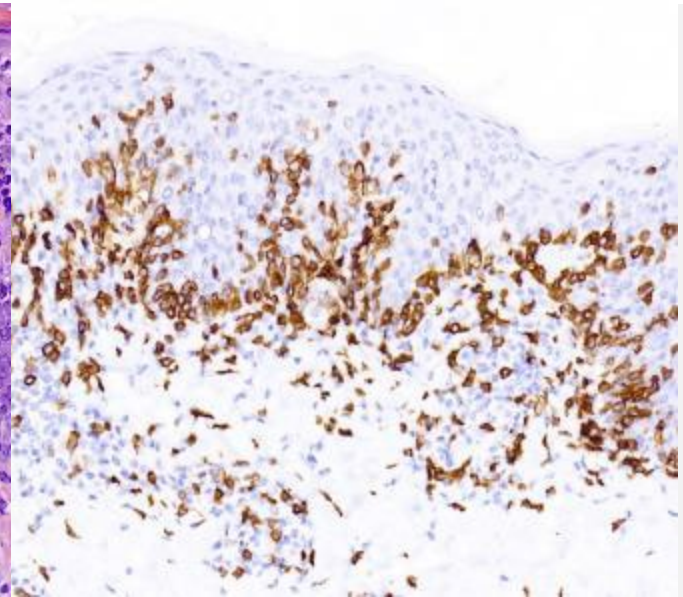




Clinical presentation:

- Sharply demarced hypopigmentations
- Child





Histology:

- Band-like lymphocytic infiltrate
- Prominent lining-up
- CD3+, CD8+, CD4-

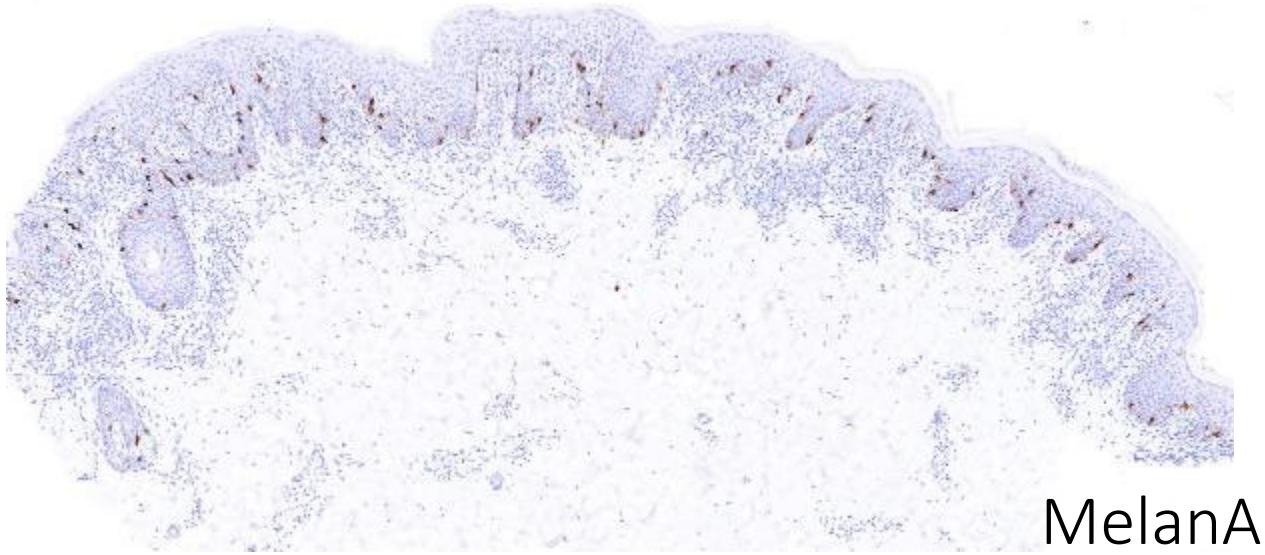
Vitiligo
or
hypopig-
mented
CD8+
Mycosis
fungoides?

Helpful tools to resolve the diagnostic dilemma?

Table 1: Histopathological distinctions between vitiligo and hypopigmented MF

	Vitiligo	Hypopigmented MF
Melanocytes	Total loss	Partial (focal) loss
Hydropic degeneration	Rare	More frequent
BM thickening	More frequent	Rare
Lymphocytes in papillary dermis	Less common	More frequent
Dermal infiltrate	Less common (lower density)	More frequent (higher density)
Dermal wiry fibrosis	Less common	More frequent

Adopted from El-Darouti *et al.*



MelanA

El-Darouti MA et al. Vitiligo vs. hypopigmented mycosis fungoides (histopathological and immunohistochemical study, univariate analysis). Eur J Dermatol 2006;16:17-22.

Loro et al. Inflammatory vitiligo versus hypopigmented mycosis fungoides in a 58-year-old Indian female. Indian Dermatol Online Journal 2013; 4

- Melanocytic density is not very helpful to differentiate between an **early** vitiligo and hypopigmented mycosis fungoides

Helpful tools to resolve the diagnostic dilemma?

Clinicopathologic correlation is essential:

Vitiligo:

Histology:
- CD8+ lymphocytic infiltrate
- Near-complete absence of melanocytes
- Basement membrane thickening
- Focal as opposed to diffuse epidermotropism
- Absence of T-cell clonal rearrangement

Clinical features:
- Very sharped boarders, often geographic
- “Chalk-white” (wood light)
- Face, hands, extensor sites, friction points, symmetric

Mycosis fungoides:

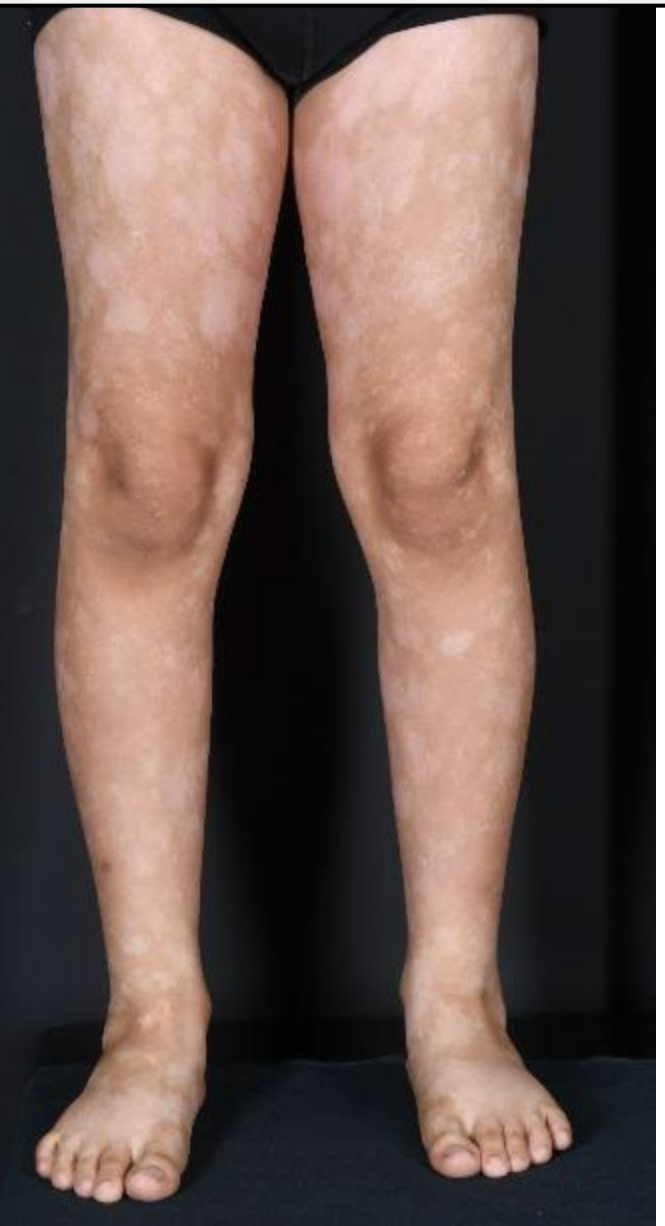
Histology:
- CD8+ lymphocytic infiltrate
- Relative decrease (<50%) of melanocytes
- Vacuolar degeneration
- Dermal wiry fibrosis
- Diffuse epidermotropism
- Clonal T-cell gene rearrangement

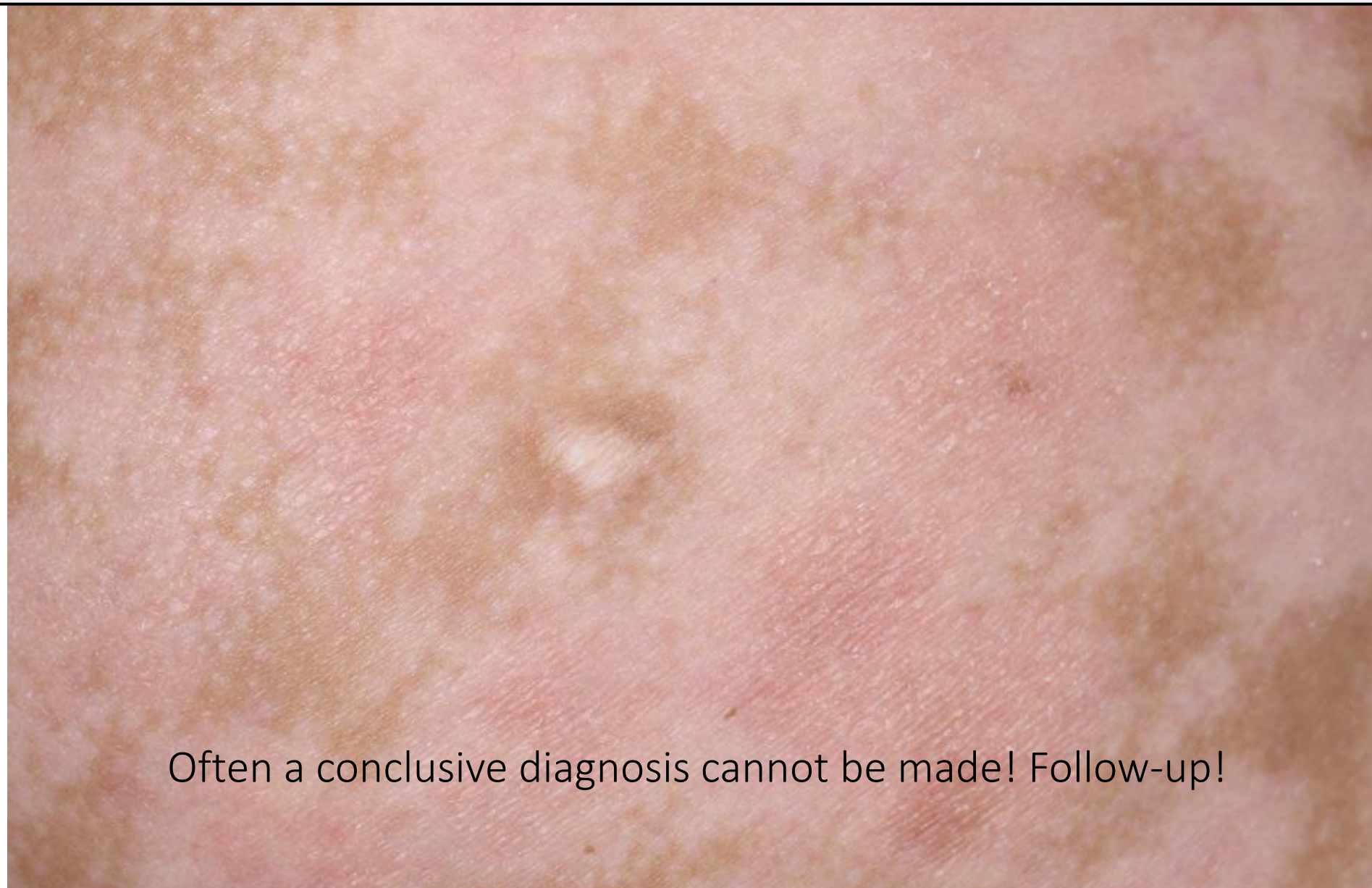
Clinical features:
- Erythematous and papular borders surrounding hypopigmented macules
- Often not completely depigmented
- Scaling, or poikiloderma
- Coexisting erythematous lesions
- Trunk, extremities, asymmetric

Abdel-Halim Ibrahim M. et al.
Thymocyte selection-associated high-mobility group box as a potential diagnostic marker differentiating hypopigmented mycosis fungoides from early vitiligo: A pilot study.
Ind J Dermatol Venereol Leprol. 2021: 87(6):819-825.

- 15 patients with hypopigmented mycosis fungoides (HMF) and 15 patients with early active vitiligo (V)
- Thymocyte selection-associated high-mobility group box (TOX)
- TOX+ :14/15 HMF cases (93.3%)
- TOX+: 1/15 vitiligo case (6.7%)

Clinical presentation:

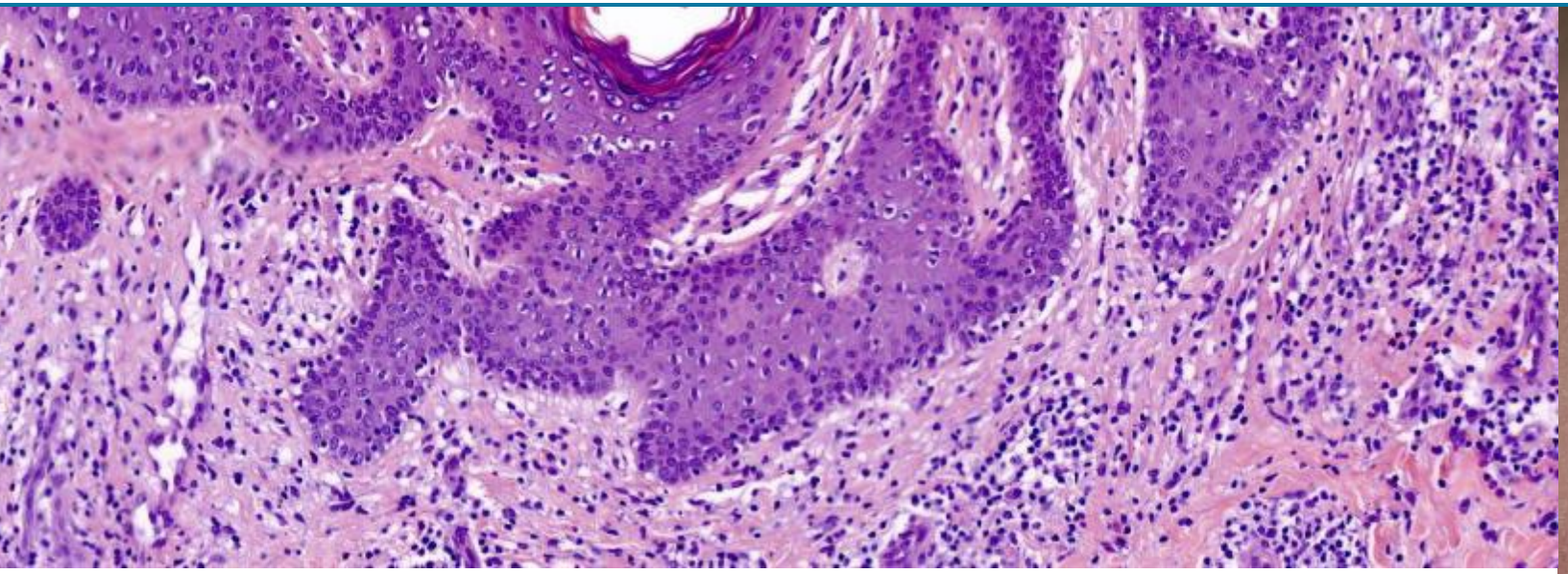


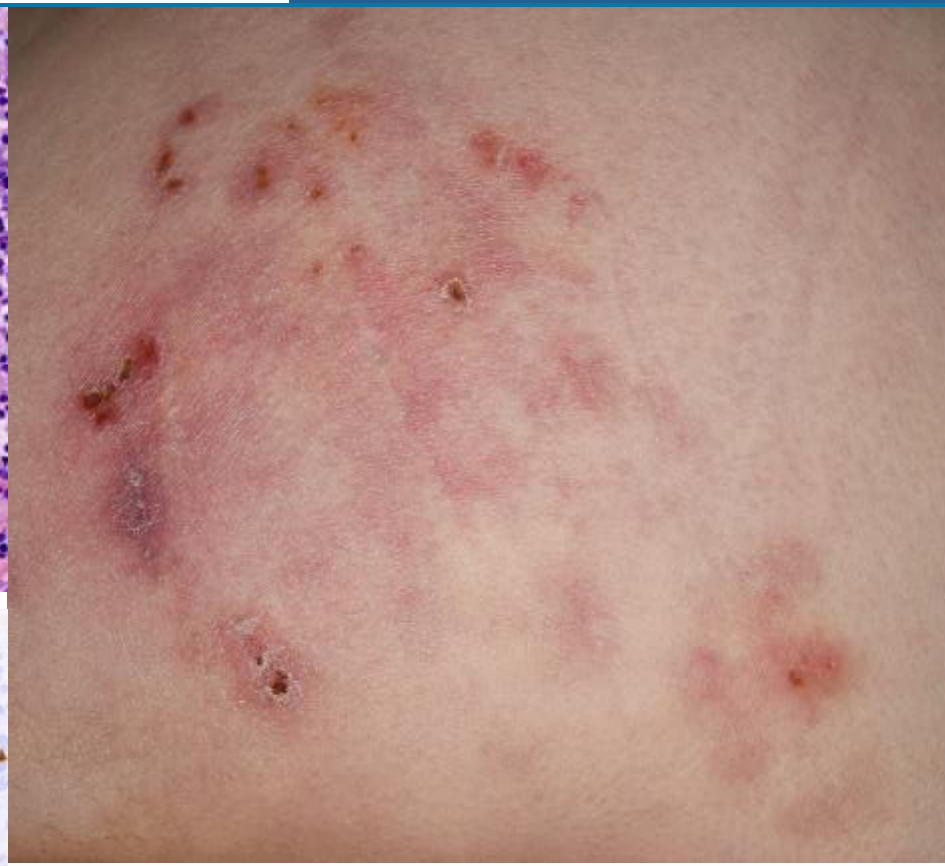


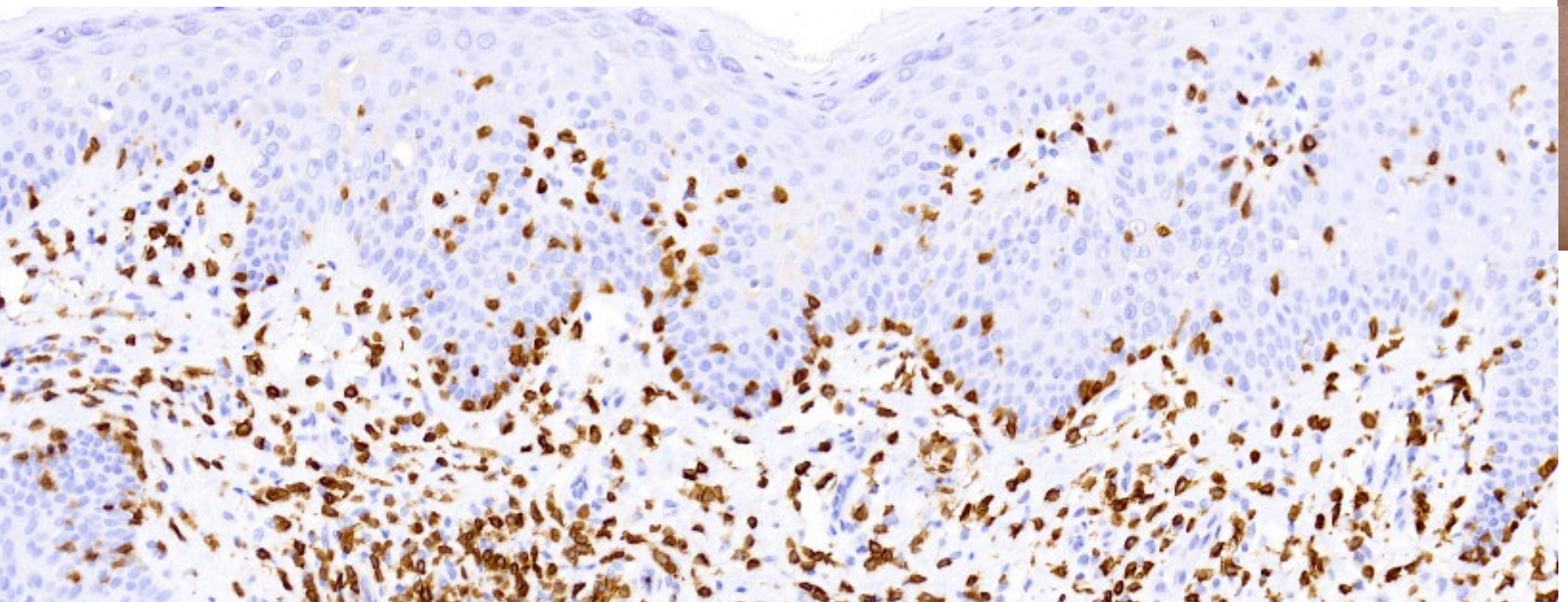
Often a conclusive diagnosis cannot be made! Follow-up!

DD of MF-like reactive T-cell lymphoproliferations

herpes



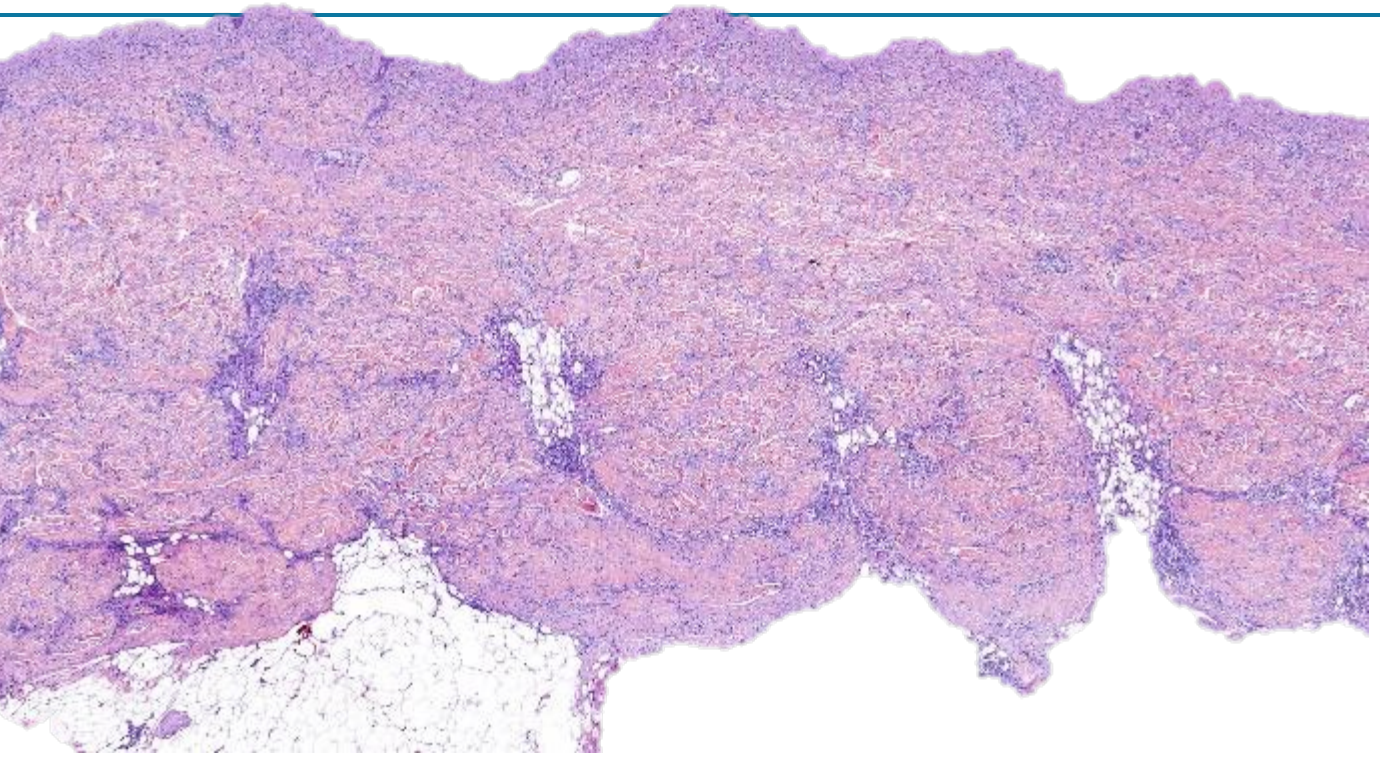


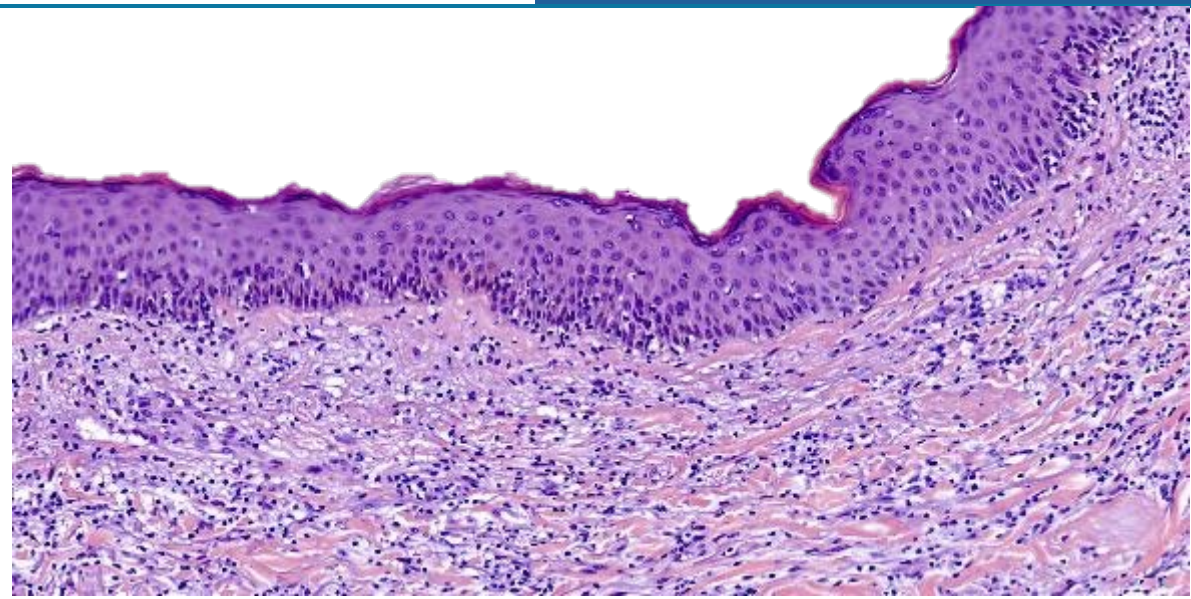


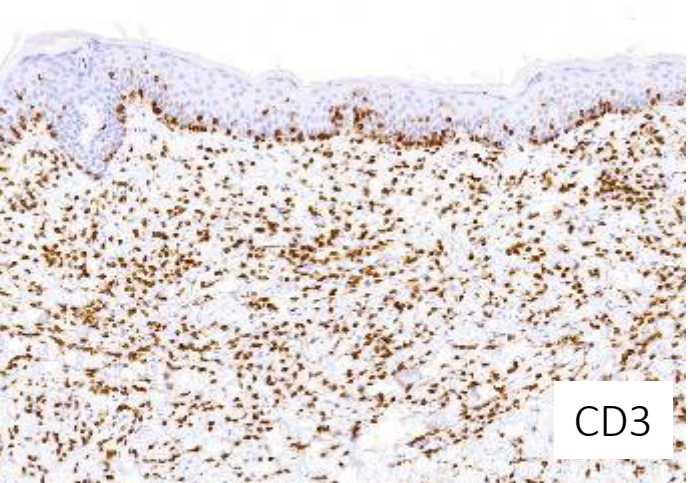
CD8

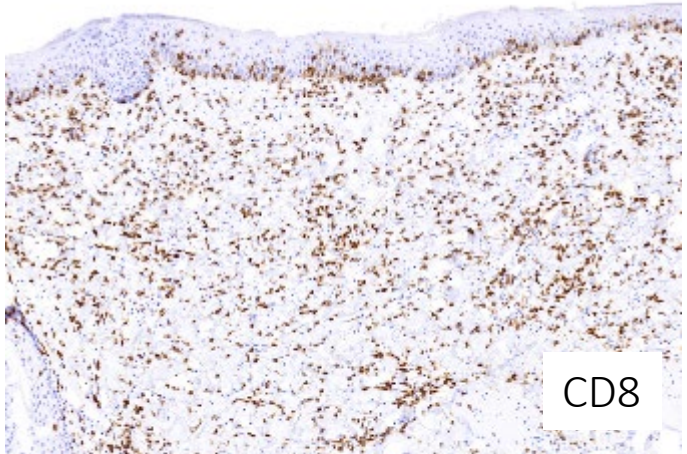
DD of MF-like reactive T-cell lymphoproliferations

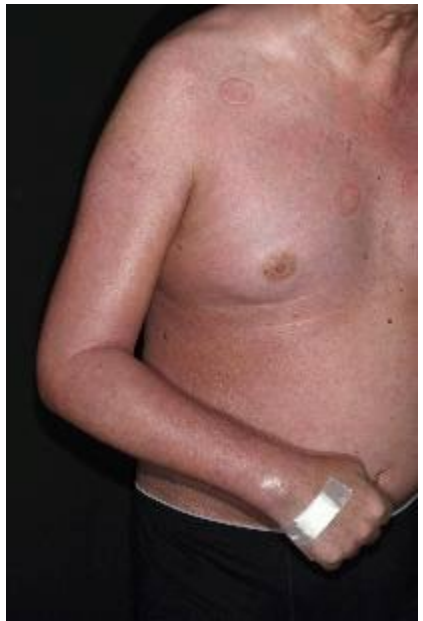
scleroderma

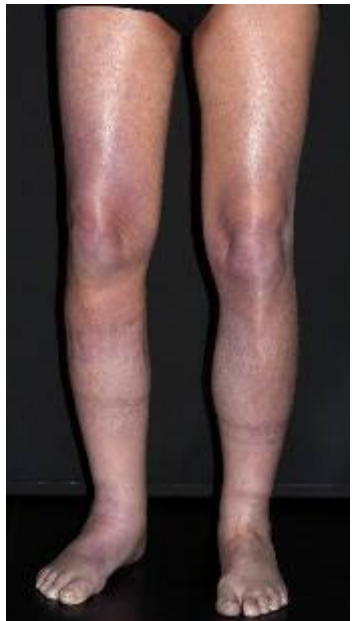


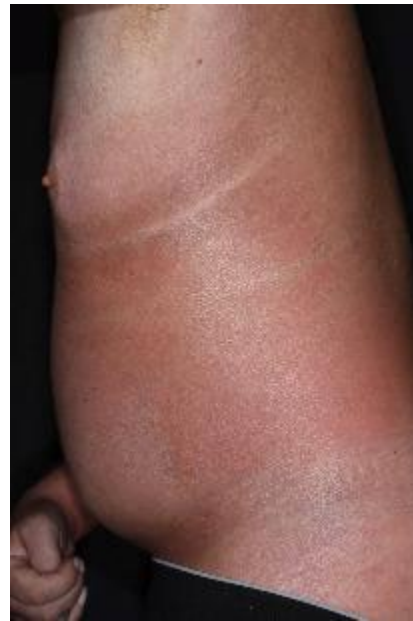






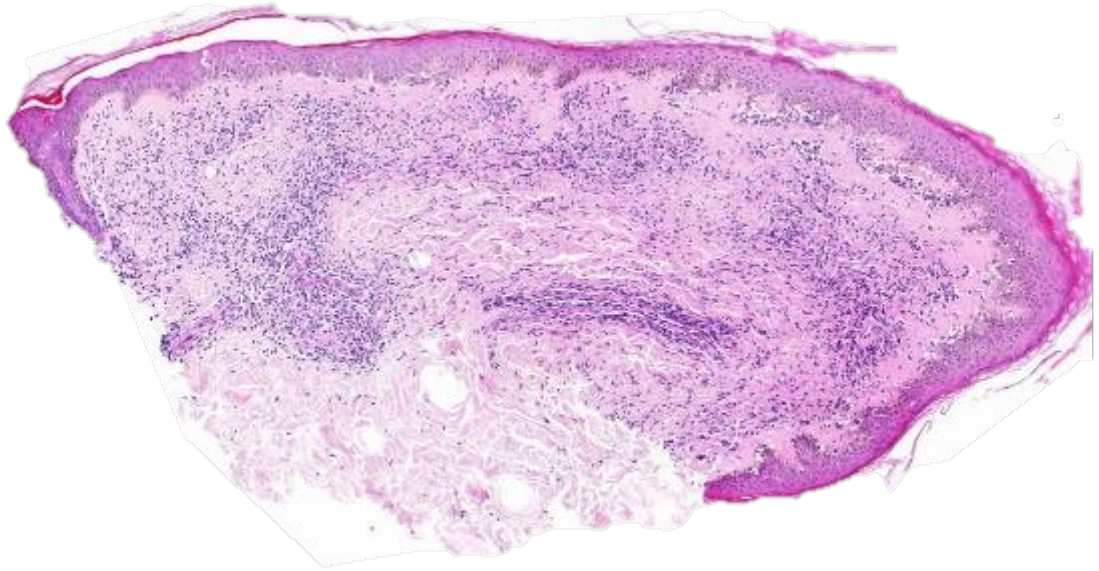


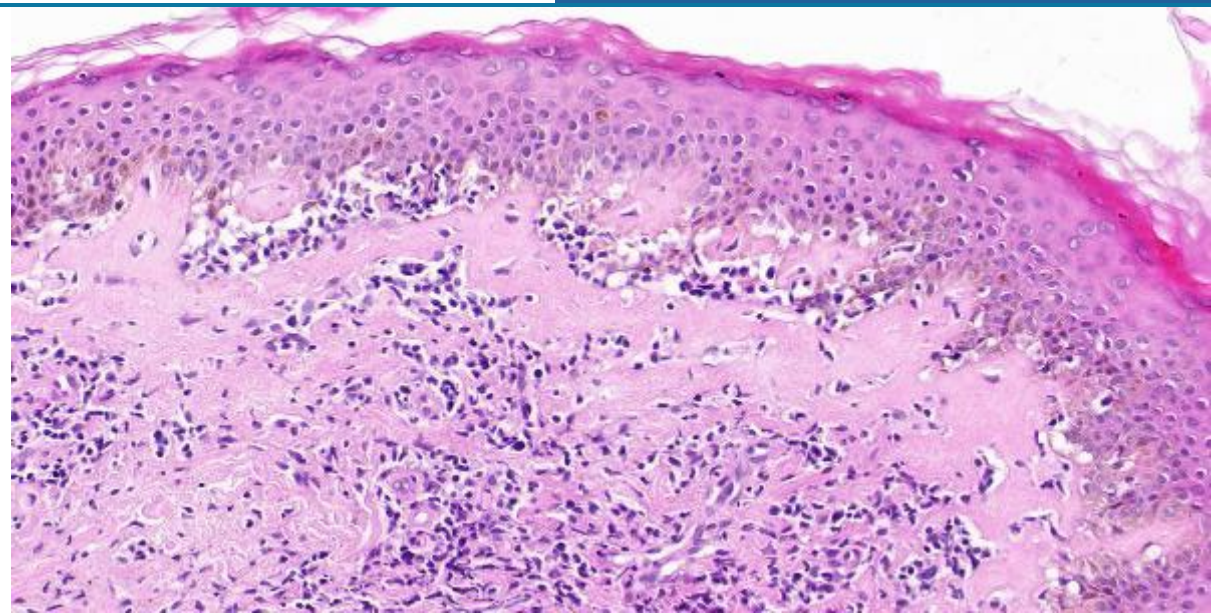


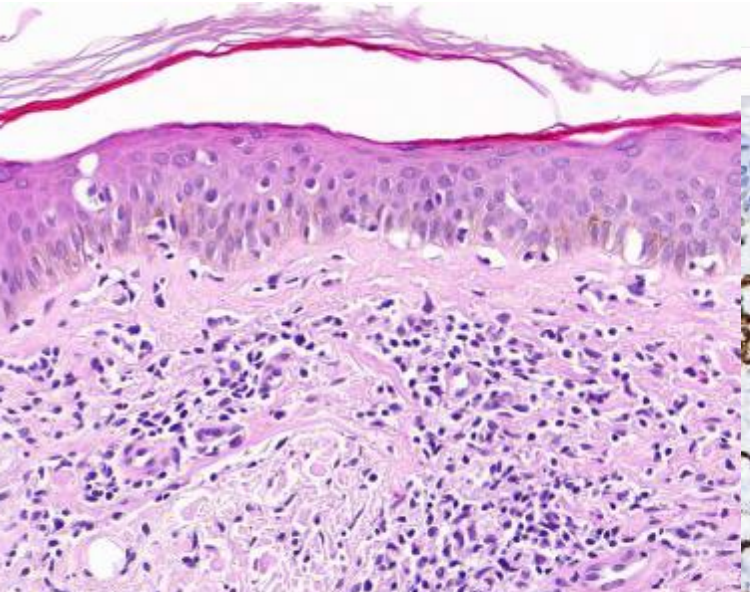


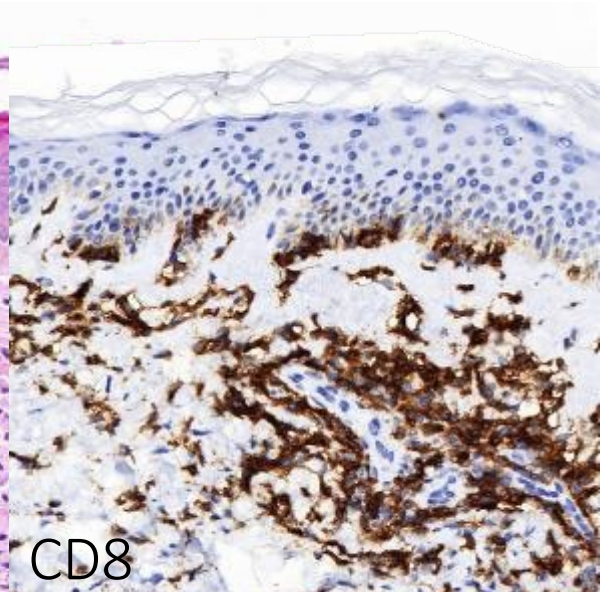
DD of MF-like reactive T-cell lymphoproliferations

LSA

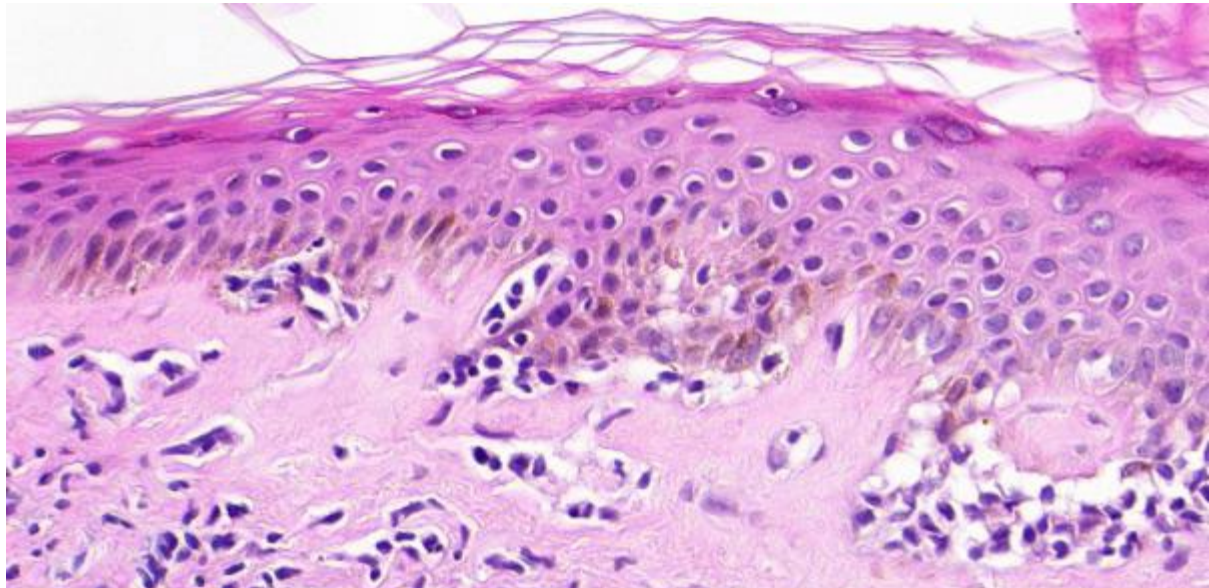








CD8



DD of MF-like reactive T-cell lymphoproliferations

LSA

ORIGINAL ARTICLE

Lichen Sclerosus et Atrophicus With Histopathologic Features Mimicking Mycosis Fungoides

A Large Series of Cases Comparing Genital With Extragenital Lichen Sclerosus

Eleonora Leoni, MD, Werner Kempf, MD,†‡ and Lorenzo Cerroni, MD§*

Abstract: Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory dermatosis of unknown etiology involving the genital and/or extragenital area, showing histopathologically a characteristic homogenization and sclerosis of the superficial collagen with variably dense lymphoid infiltrates. Intraepidermal lymphocytes may be observed, and in some cases may pose differential diagnostic problems with mycosis fungoides (MF). We studied the histopathologic features of 121 cases of LSA with dense lymphoid infiltrates (genital: 94; male:female: 93:1; age range: 2 to 87 y; median age: 11 y; extragenital: 27; male:female: 0.1:1; age range: 11 to 79 y; median age: 59 y), to better characterize the intraepidermal lymphoid infiltrate and to compare genital with extragenital cases. Epidermotropic lymphocytes mimicking the histopathologic features of MF were present in 93.6% of the genital specimens but none of the extragenital cases. Interestingly, typical features of LSA were missing in 39.4% of genital LSA, and in a further 25.5% were present only focally. Immunohistochemical analyses showed a predominance of $CD8^+$ T-lymphocytes within the epidermis. Molecular studies of the T-cell receptor genes revealed a monoclonal population of T-lymphocytes in nearly half of the cases. Our study shows that MF-like histopathologic features are extremely common in genital LSA but are never encountered in extragenital cases. A diagnosis of MF in the genital area should be made only upon compelling features, keeping in mind the frequent pseudolymphomatous aspects of LSA.

Key Words: lichen sclerosus, mycosis fungoides, cutaneous T-cell pseudolymphoma, cutaneous T-cell lymphoma, cutaneous pseudolymphoma, extragenital lichen sclerosus

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Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory dermatosis of unknown etiology involving the genital mucosa or the extragenital skin. The typical histopathologic findings are characterized by atrophic epidermis with orthohyperkeratosis overlying a papillary dermis showing homogenized, sclerotic collagen, and a variably dense lymphocytic infiltrate. In typical cases, the histopathologic diagnosis of LSA is not problematic; some cases, however, may lack typical features being characterized instead by a dense lymphocytic infiltrate with prominent exocytosis of lymphocytes within the epidermis, thus mimicking mycosis fungoides (MF).[1–3]

We studied the clinicopathologic features of LSA with particular emphasis on histopathologic features mimicking MF, and with comparison of genital with extragenital cases.

PATIENTS AND METHODS

One hundred twenty-one patients (male:female = 3.8:1; age range: 2 to 87 y; mean age: 30.6 y; median age: 15 y) presenting with LSA have been included in our study. Ninety-four cases were from the anogenital area (male:female = 93:1; age range: 2 to 87 y; median age: 11 y), and 27 from the extragenital area (male:female = 0.1:1; age range: 11 to 79 y; median age: 59 y). The cases were collected at the Department of Dermatology of the Medical University of Graz and at the Kempf und Pfaltz Histologische Diagnostik, Zürich, Switzerland. Variably dense lymphoid infiltrates were present in all cases (cases devoid of lymphoid infiltrates were excluded). Partial data on 9 cases had been published previously.[1] The study has been approved by the ethical committee of the Medical University of Graz.

A total of 123 formalin-fixed, paraffin-embedded biopsy specimens were available for histopathologic analysis. Following histopathologic features were evaluated: presence of typical aspects of LSA; presence of epidermotropic lymphocytes (pseudo-MF aspects); presence of pronounced hemorrhage (pseudovascular aspects); presence of associated morphea; presence of granulomatous phlebitis; presence of germinal centers; presence of perineural inflammatory infiltrates.

In 69 cases (27 from the extragenital, 42 from the genital area), immunohistochemical analyses were performed with a standard immunoperoxidase technique using

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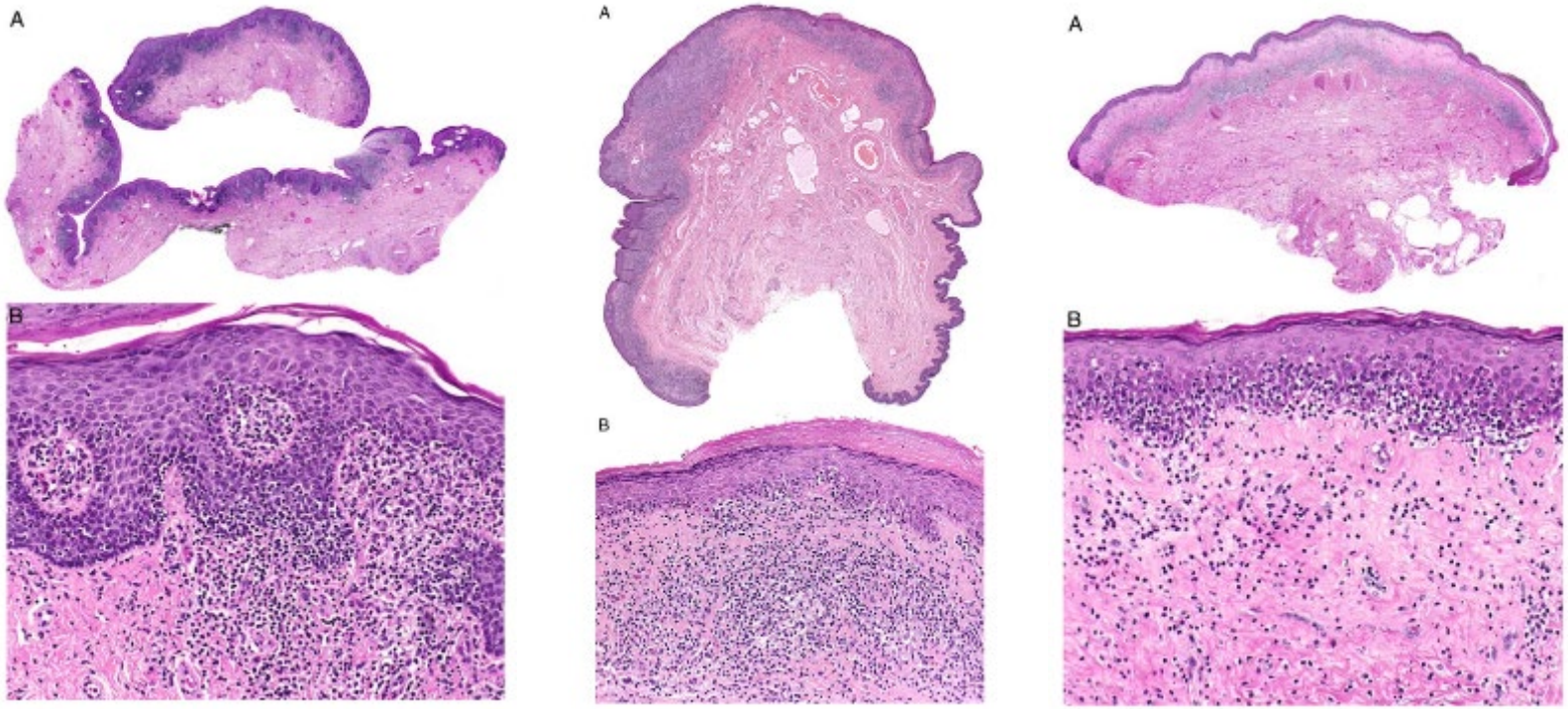


TABLE 1. Summary of Histopathologic Features of Cases Included in Our Study

	n/N (%)	
	Extragenital	**Genital**
Typical histopathologic features of LSA	29/29 (100)	57/94 (60.6) (prominent 35.1%, focal 25.5%)
Pseudo-MF histopathologic features	0	88/94 (93.6)
Granulomatous phlebitis	0	6/94 (6.4)
Associated morphea	1/29 (3.4)	0
Prominent hemorrhage ("pseudovascular" features)	6/29 (20.7)	4/94 (4.3)
Perineural infiltrate	5/29 (17.2)	1/94 (1.1)
Germinal centers	0	6/94 (6.4)
B. burgdorferi DNA by PCR	1/20 (5)	1/10 (10)

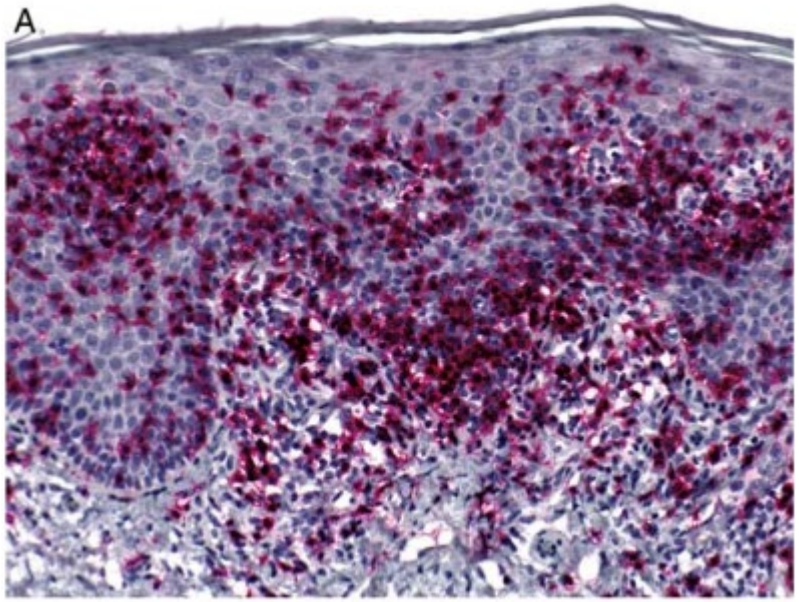
DD of MF-like reactive T-cell lymphoproliferations

LSA

TABLE 2. Assessment of the Phenotype and Clonality in Cases of LSA in the Literature (Including This Study)

References	No. Cases	CD8 > CD4 (Cases/Total No. of Cases)	CD4 > CD8 (Cases/Total No. of Cases)	TCR Clonality Assessment (Monoclonal/Total No. of Cases) (%)
Regauer et al[7]	50	50/50	0/50	19/38 (50.0)
Regauer and Beham-Schmid[8]	33	ND	ND	18/33 (54.5)*
Lukowsky et al[9]	39	Relative ratio not presented	Relative ratio not presented	19/39 (48.7)
Citarella et al[1]	9	ND	ND	1/9 (11.1)
This study	94 (genital)	29/42 (epidermis) 12/42 (dermis)	13/42 (epidermis) 30/42 (dermis)	16/33 (48.5)
	27† (extragenital)	12/17 (epidermis)‡ 1/27 (dermis)	5/17 (epidermis)‡ 26/27 (dermis)	

*18/33 cases with standard PCR analysis; subsequent fragment analysis showed a monoclonal population of T-lymphocytes in 8/18 positive specimens (44.4%).
†Two cases had 2 biopsies each.
‡The intraepidermal lymphocytes did not show an MF-like pattern in cases of extragenital LSA (see text); several cases of extragenital LSA did not show intraepidermal lymphocytes.
ND indicates not determined.



- monoclonal TCR in about 50%
- intraepidermal lymphocytic population had mainly a cytotoxic phenotype (CD8+, CD4–)

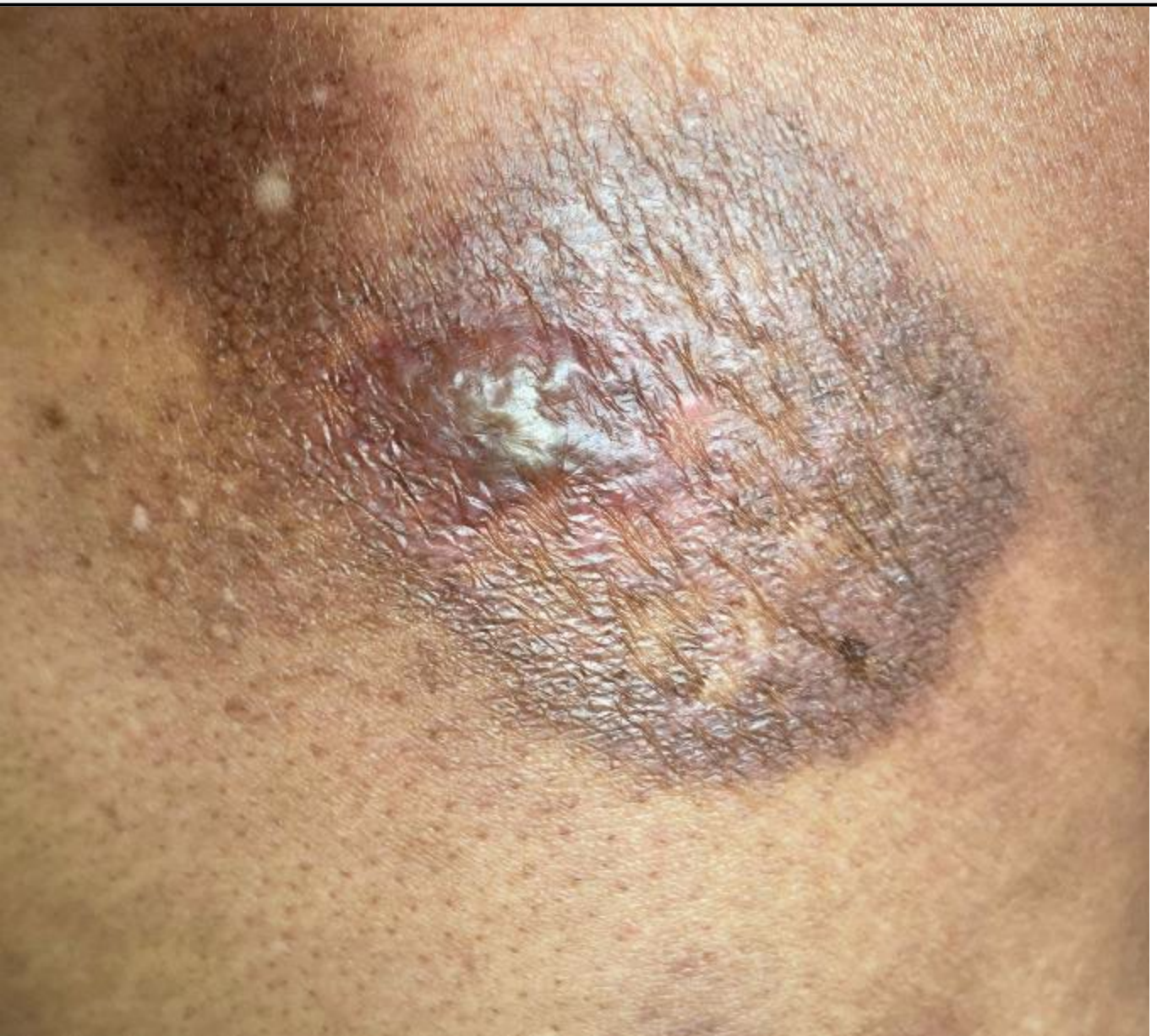
Lymphoma prototype and simulator

Mycosis fungoides: lining-up

- Morphea / scleroderma
- Lichen sclerosus et atrophicus (LSA)
- Vitiligo
- Herpes
- Borreliosis
- Syphilis
- Lymphomatoid drug eruption
- Lymphomatoid contact dermatitis
- Mogamulizumab-associated rash (MAR)

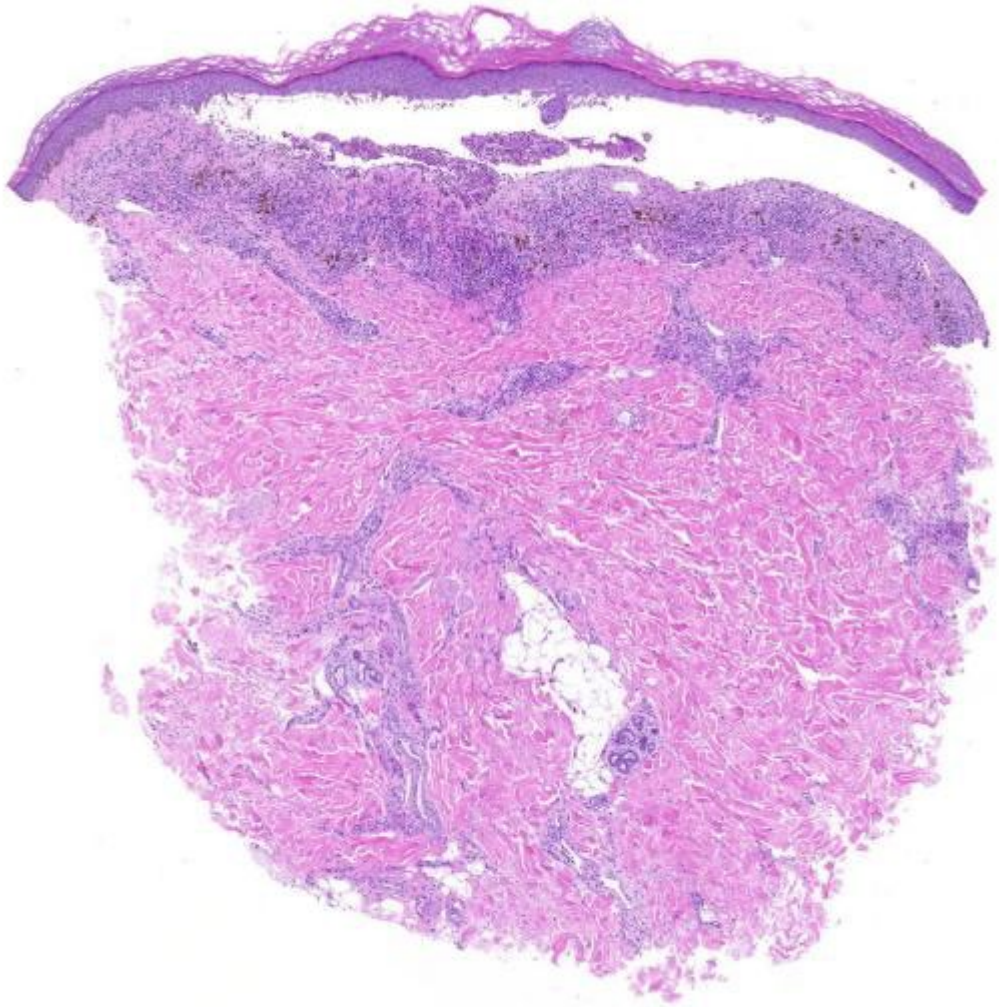
Case 4

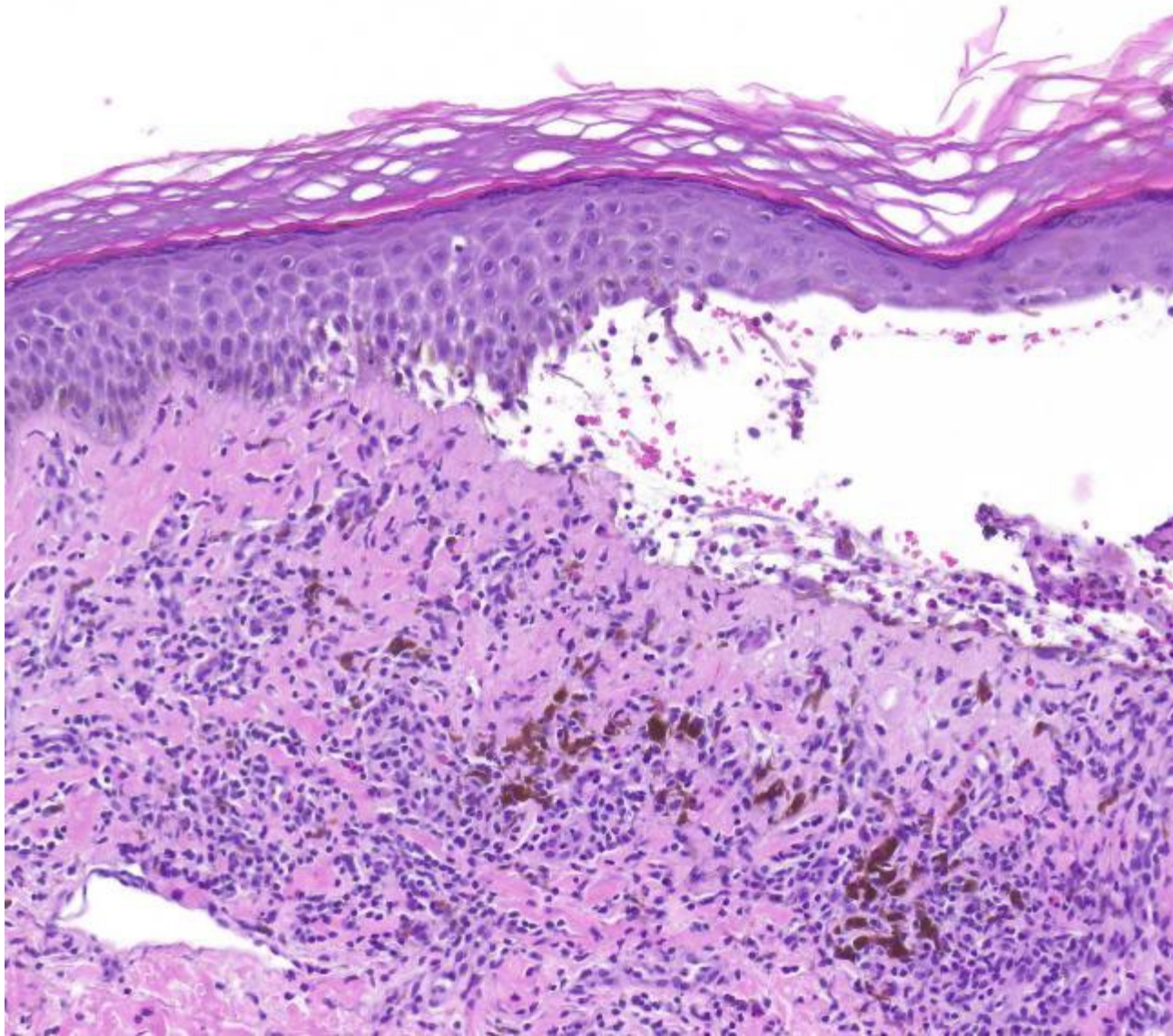
Clinical presentation:

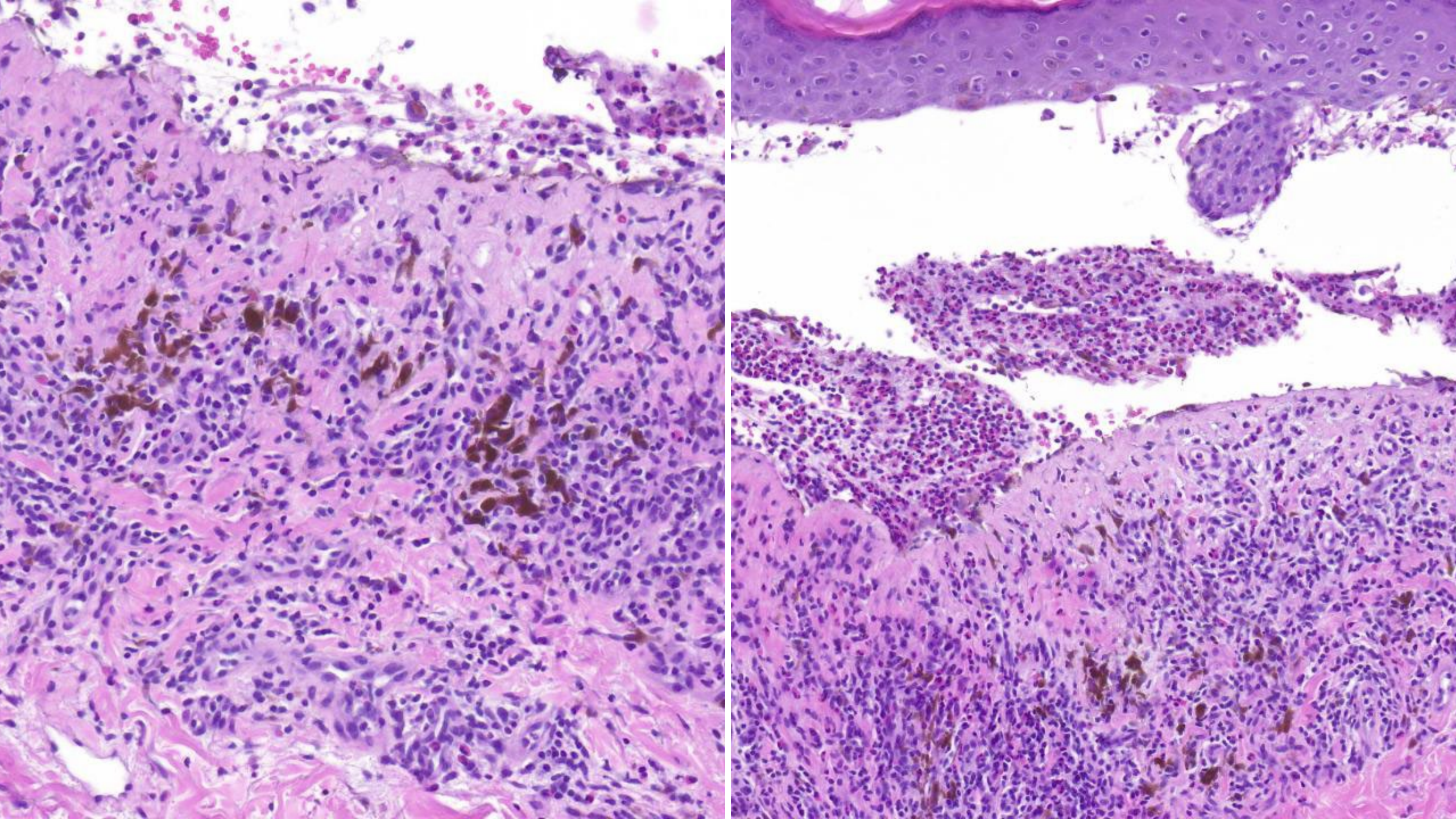


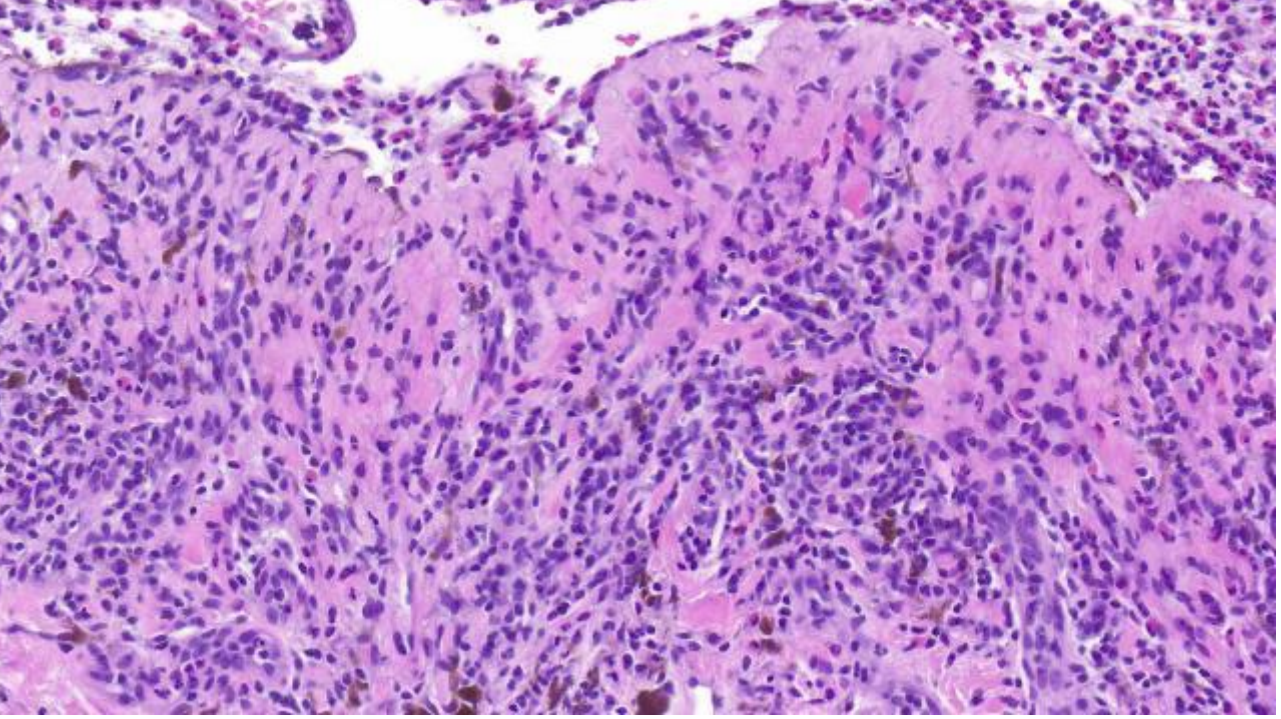
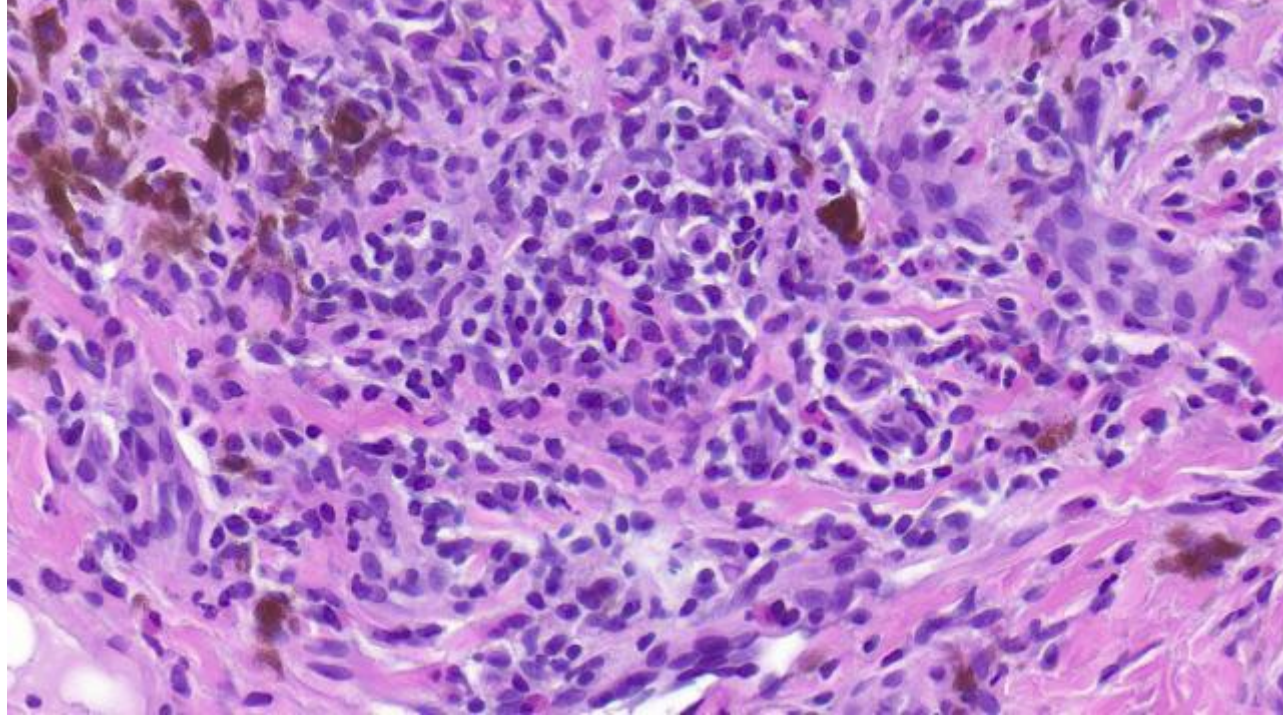
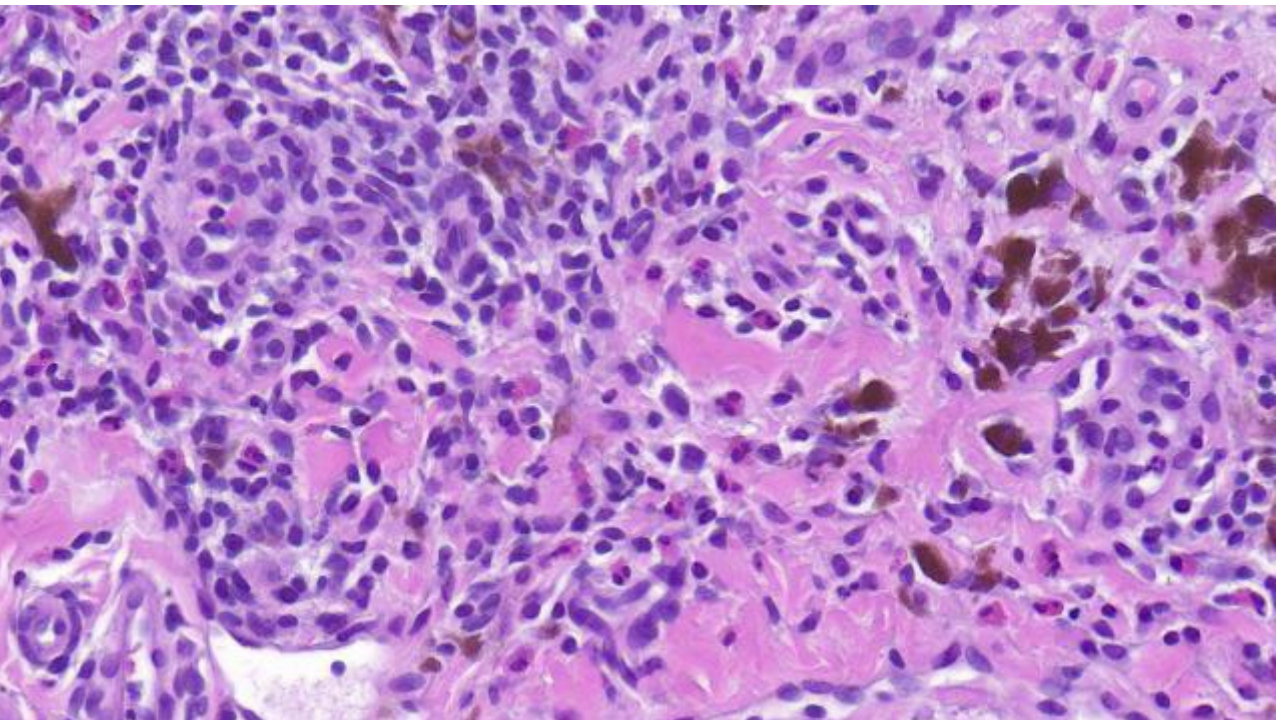
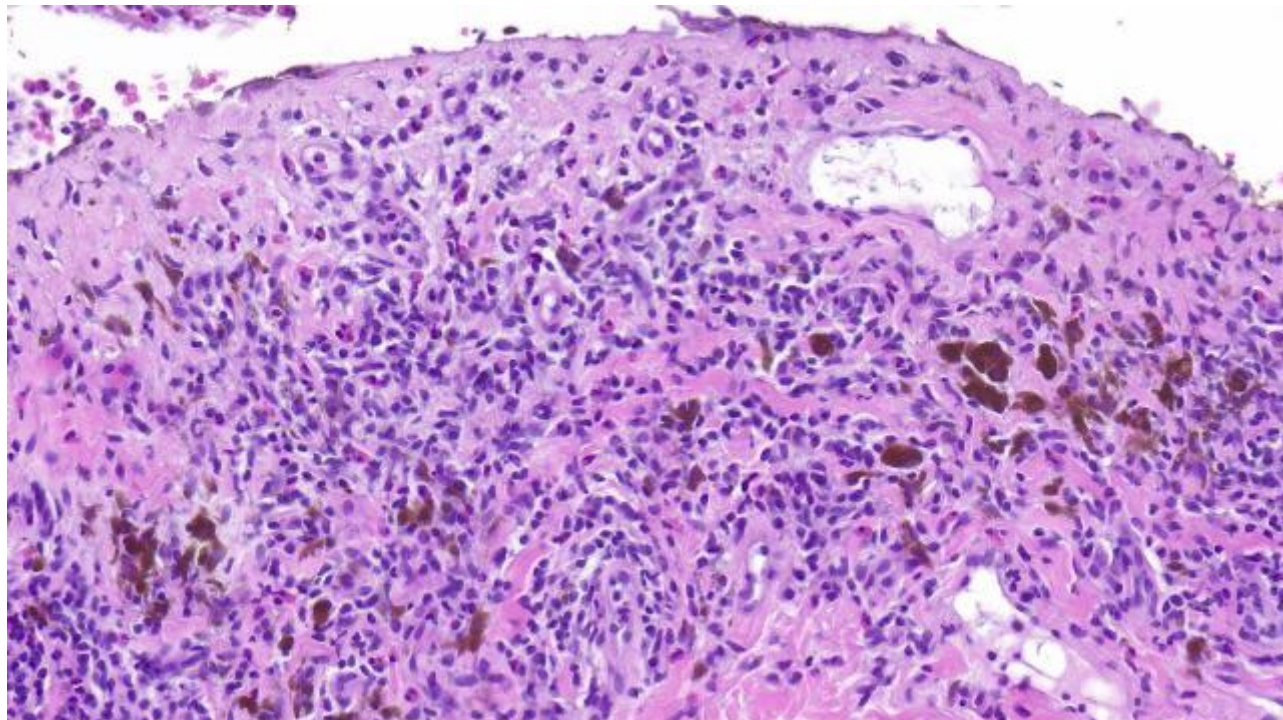
- A 19-year-old patient
- Fitzpatrick skin type V
- Skin lesions since early childhood.
- The condition was characterized by hyperpigmented plaques on the trunk
- Recurrent episodes of blister formation.
- Hand and face not effected
- No other abnormalities, no family history
- Initially, a diagnosis of atopic dermatitis had been considered.
- The patient reported mild pruritus but no other associated symptoms.
- Treatment with topical corticosteroids had been ineffective.

Histology:

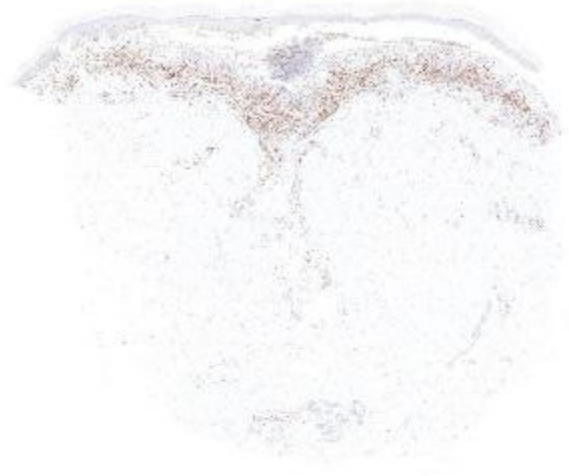






CD3



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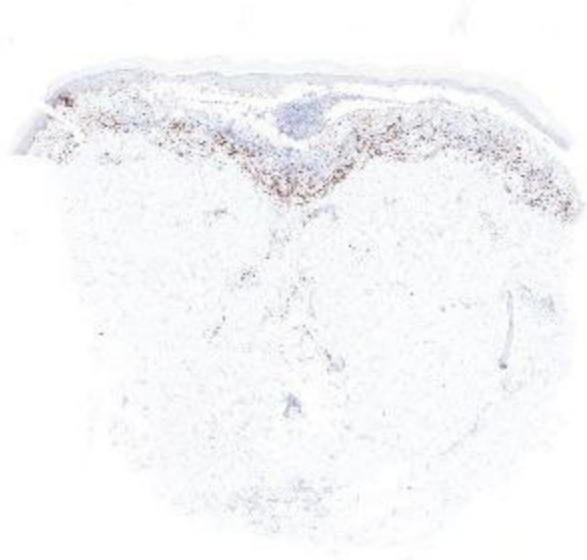
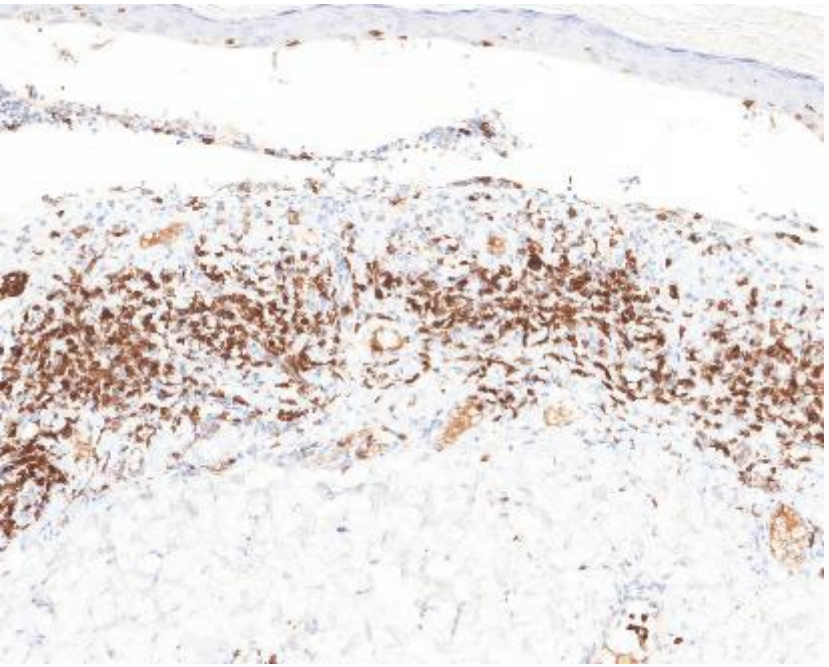
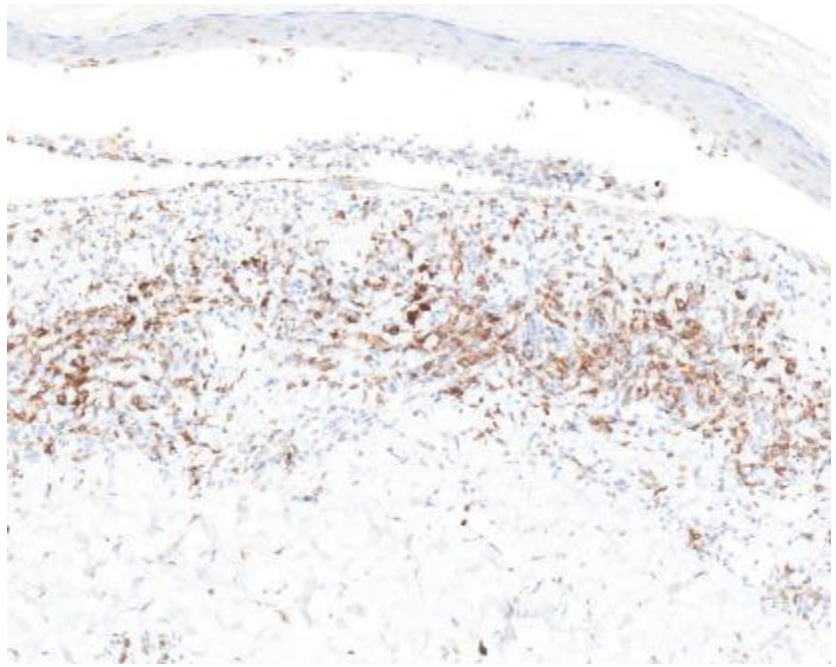
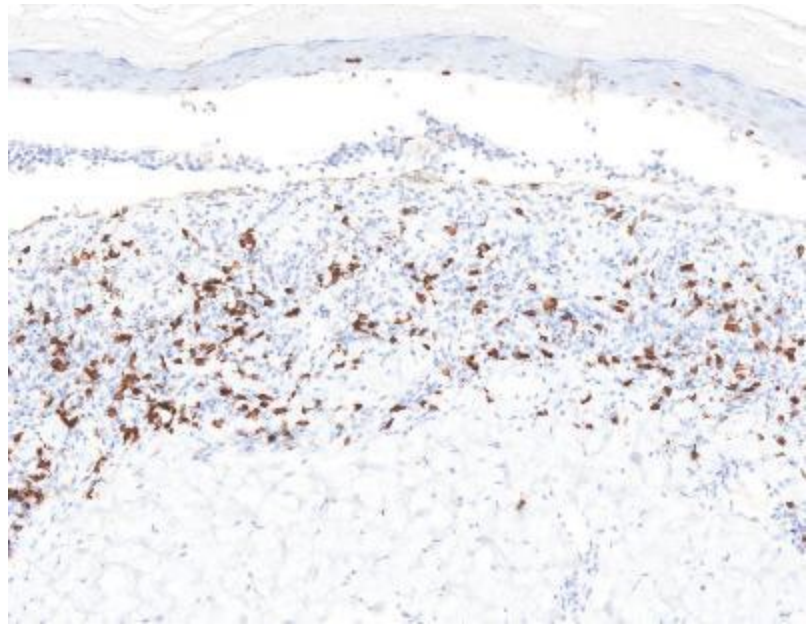
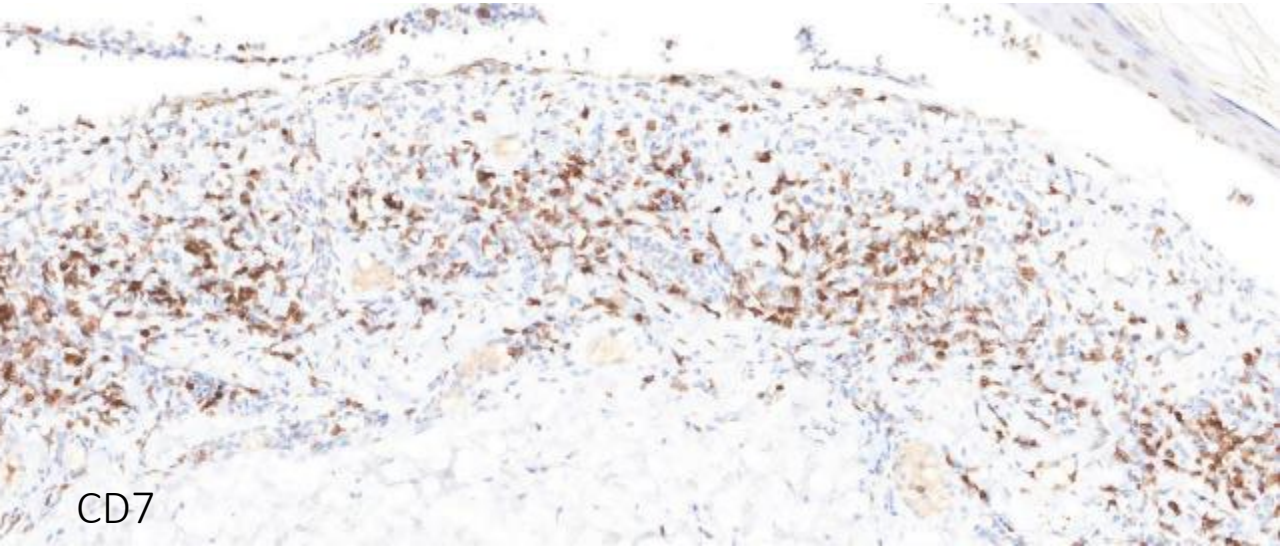


CD8





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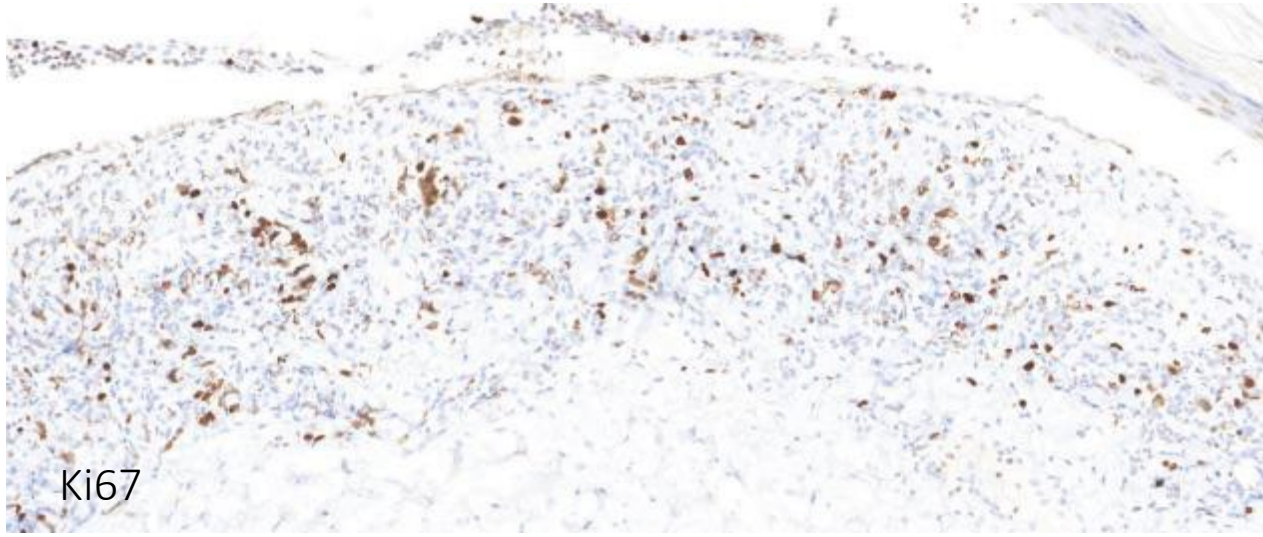
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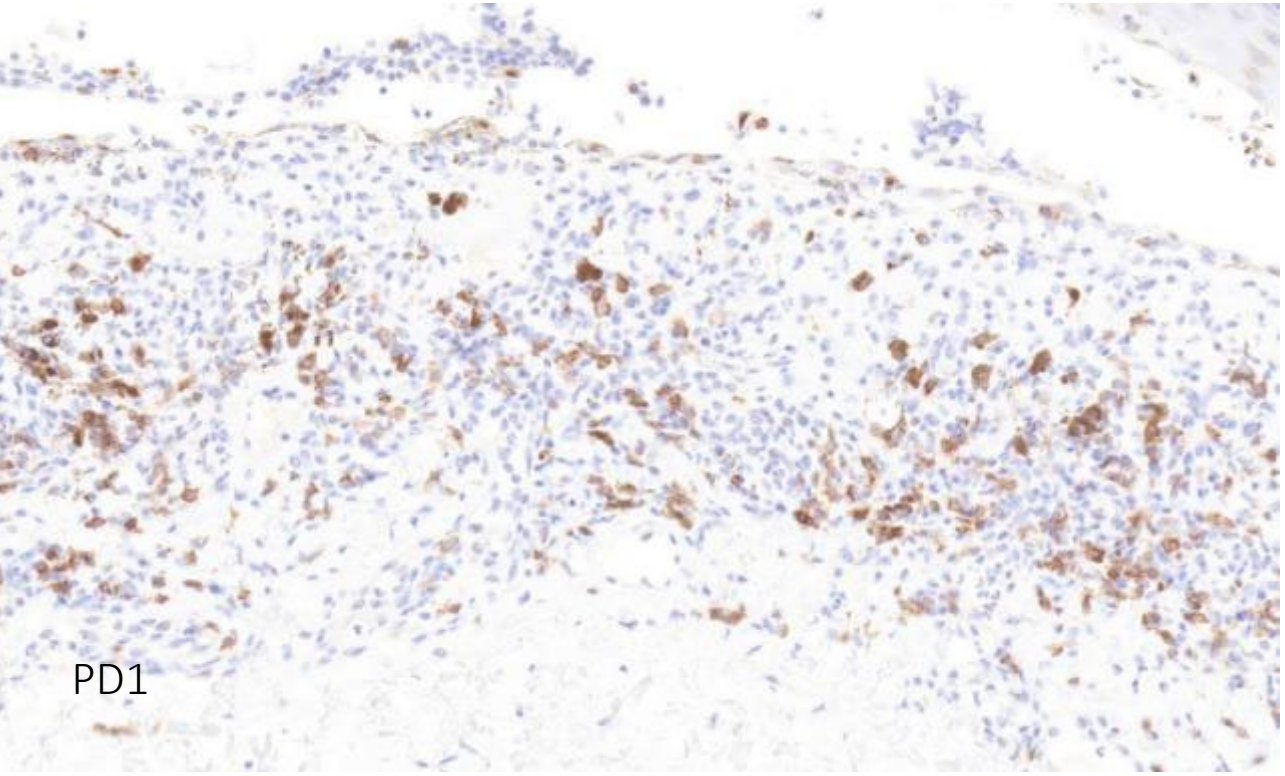
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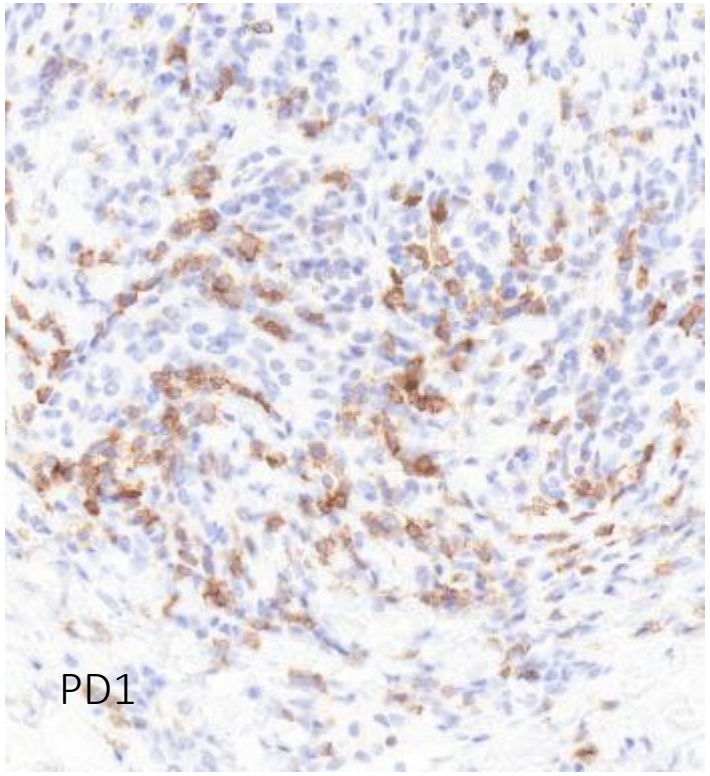
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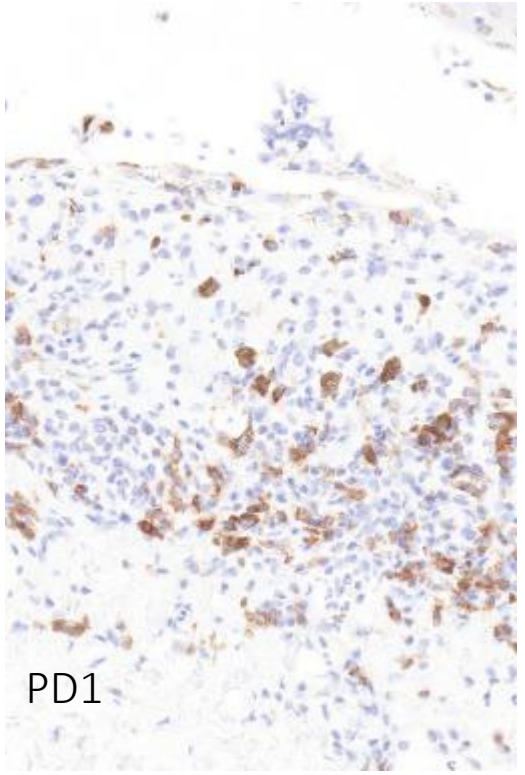
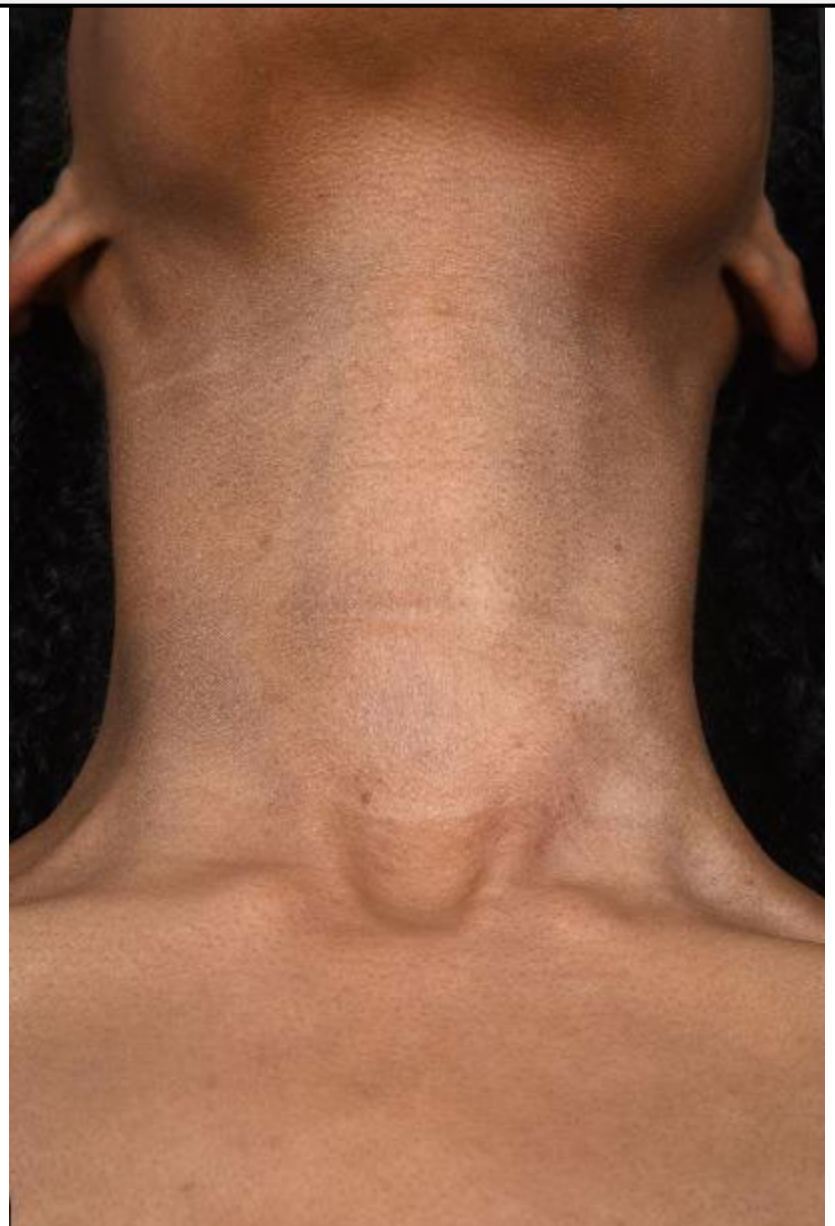
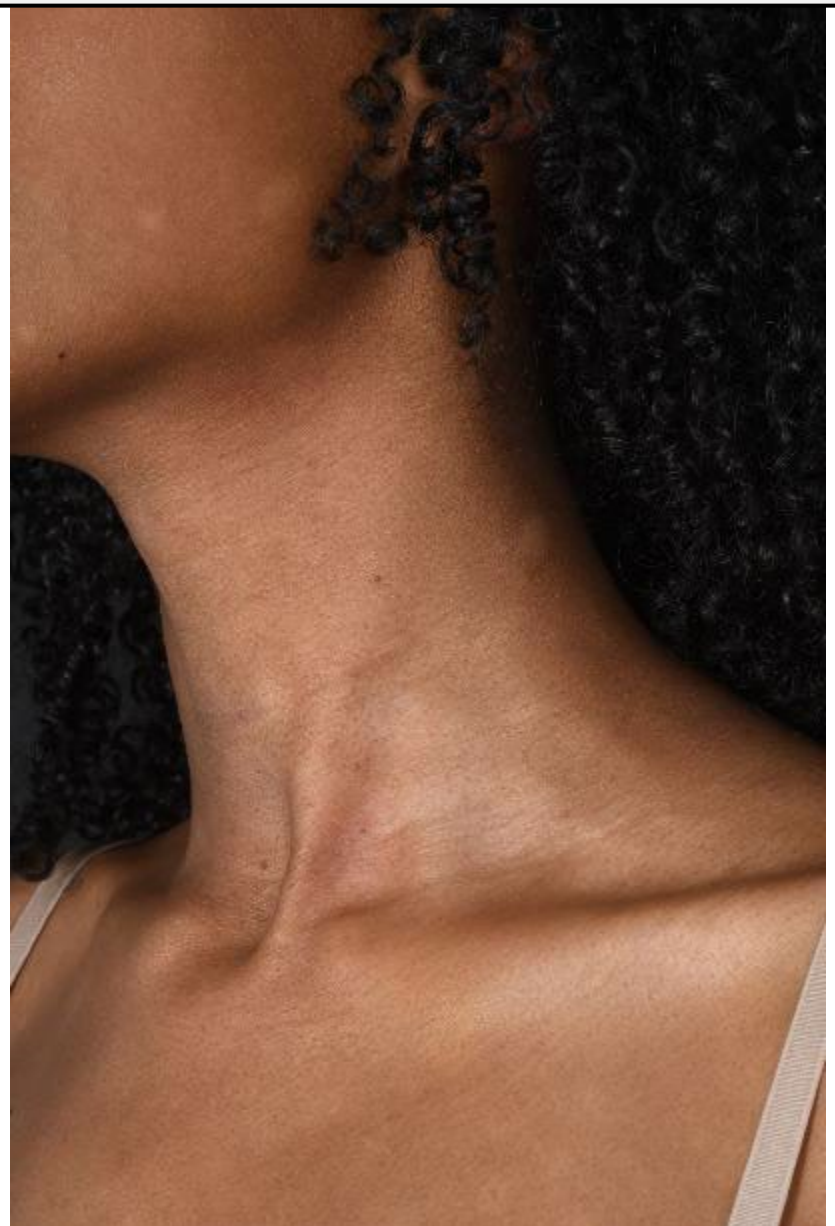
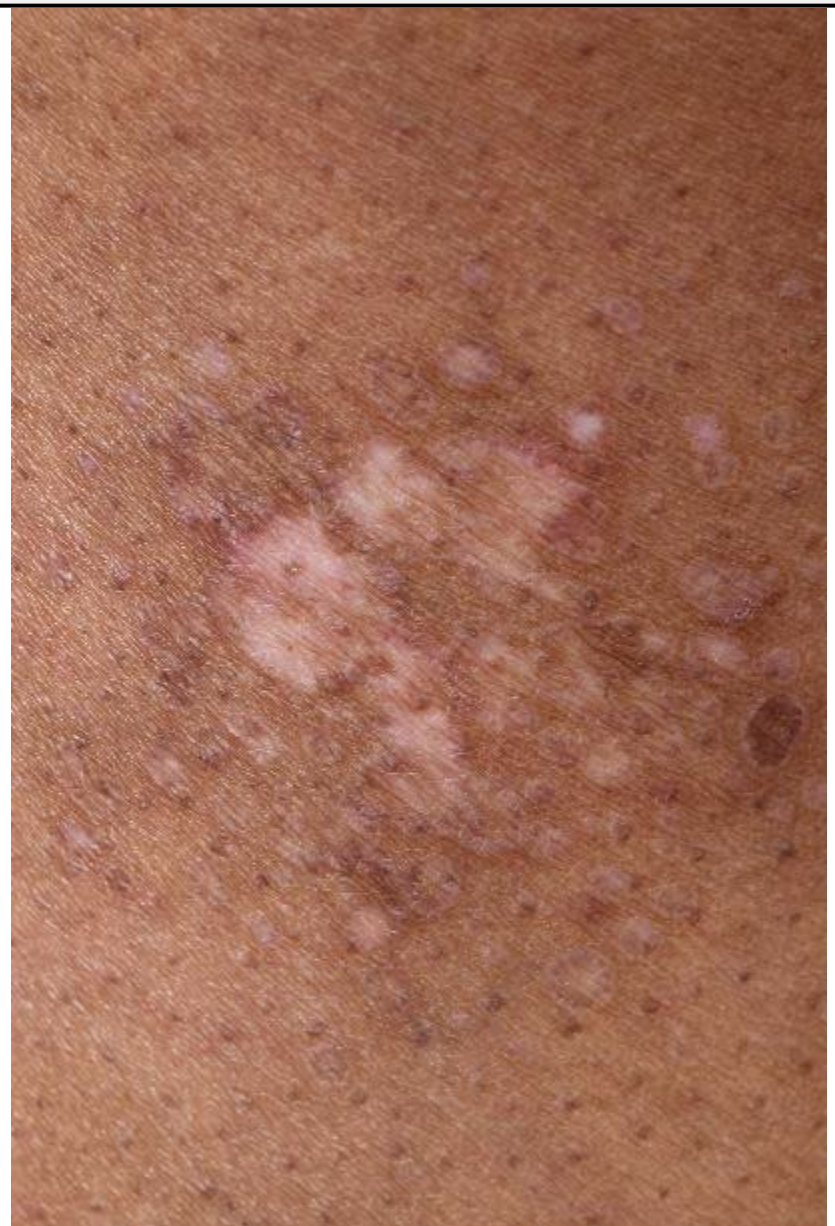

PD1

Clinical presentation:







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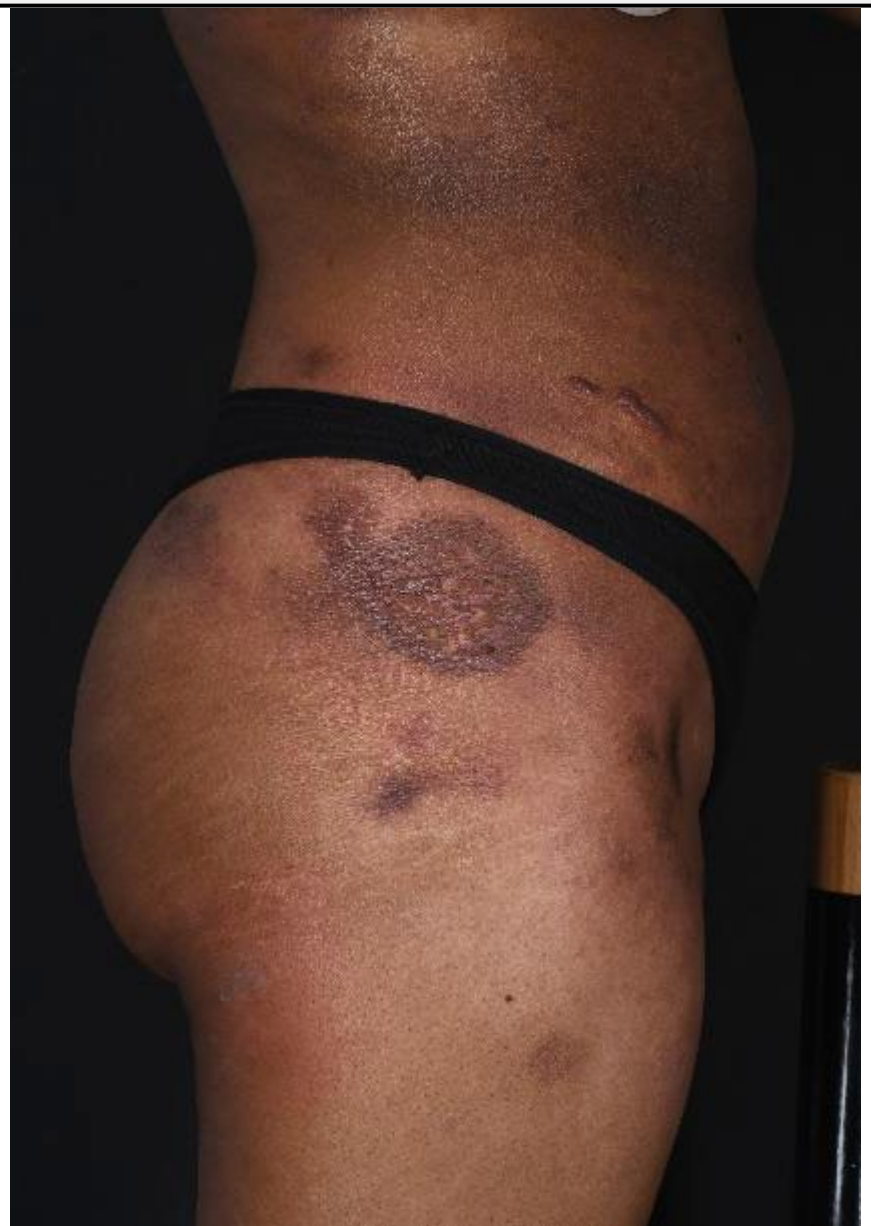

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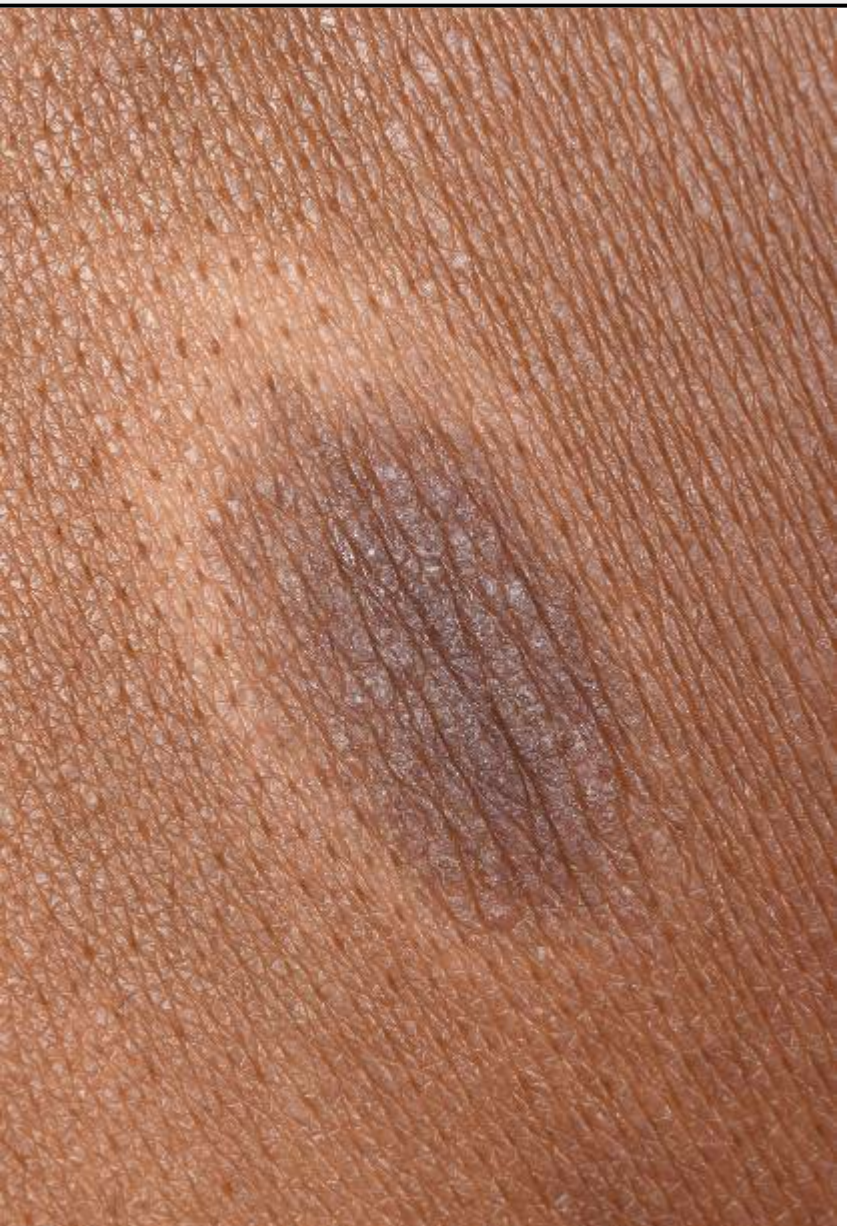

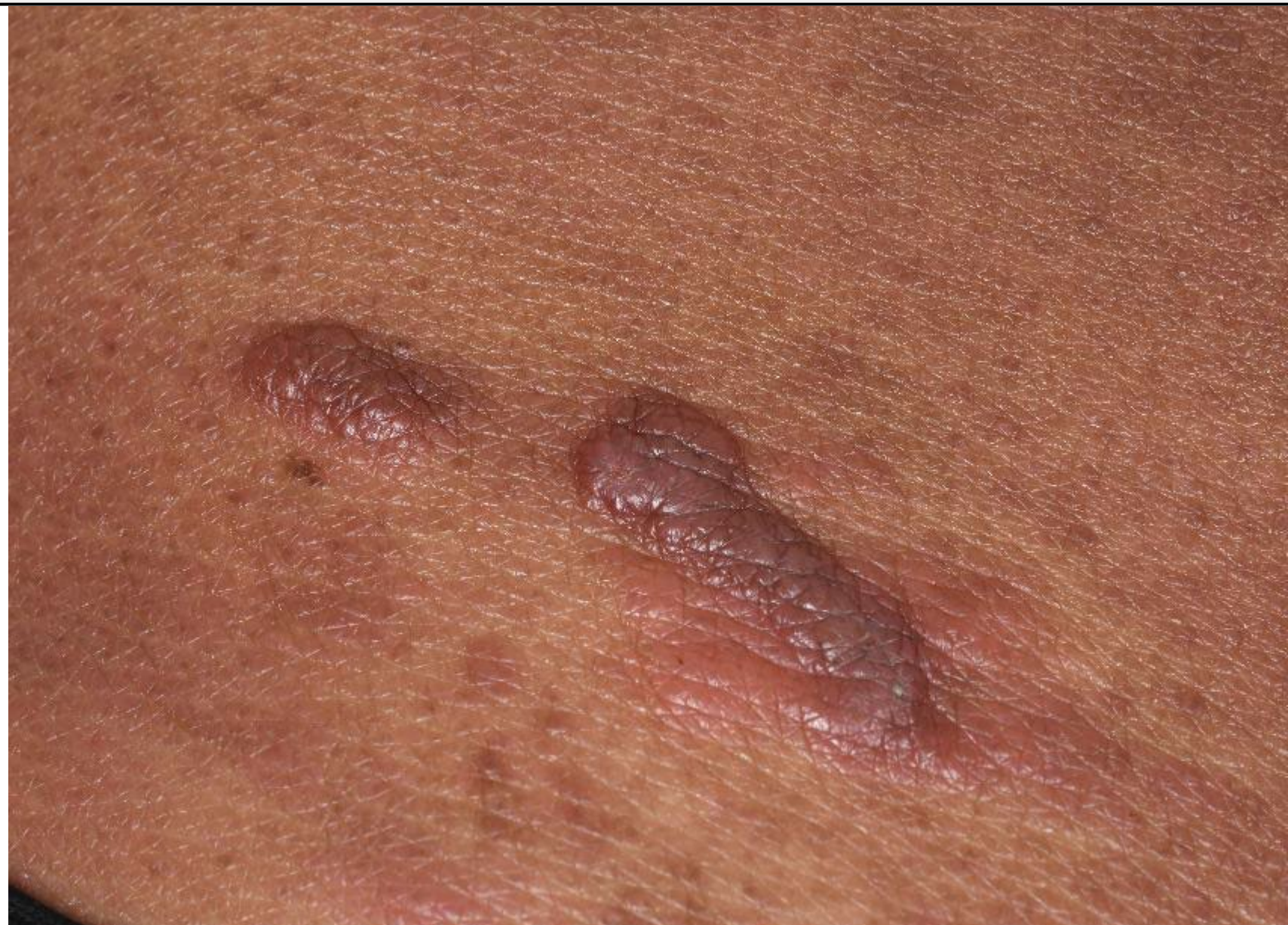

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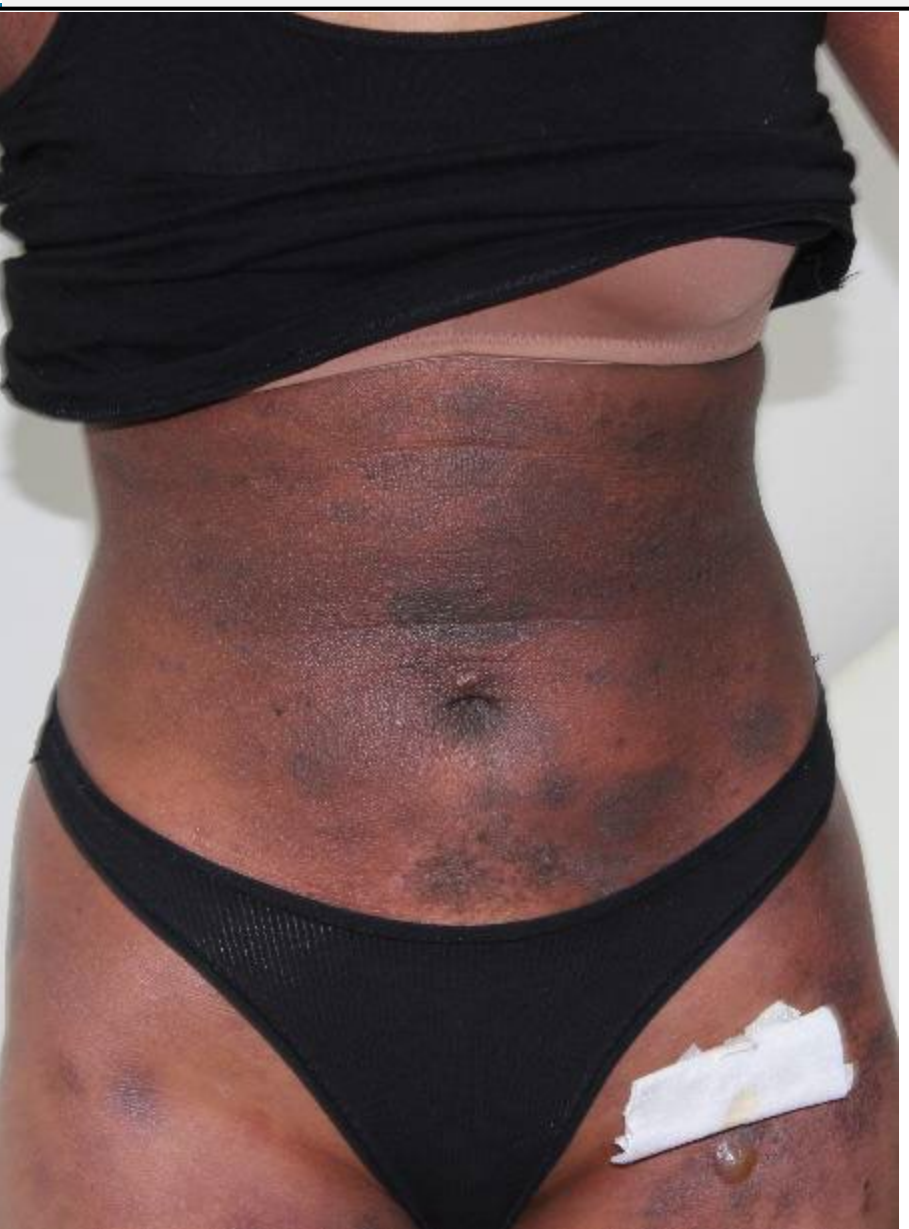
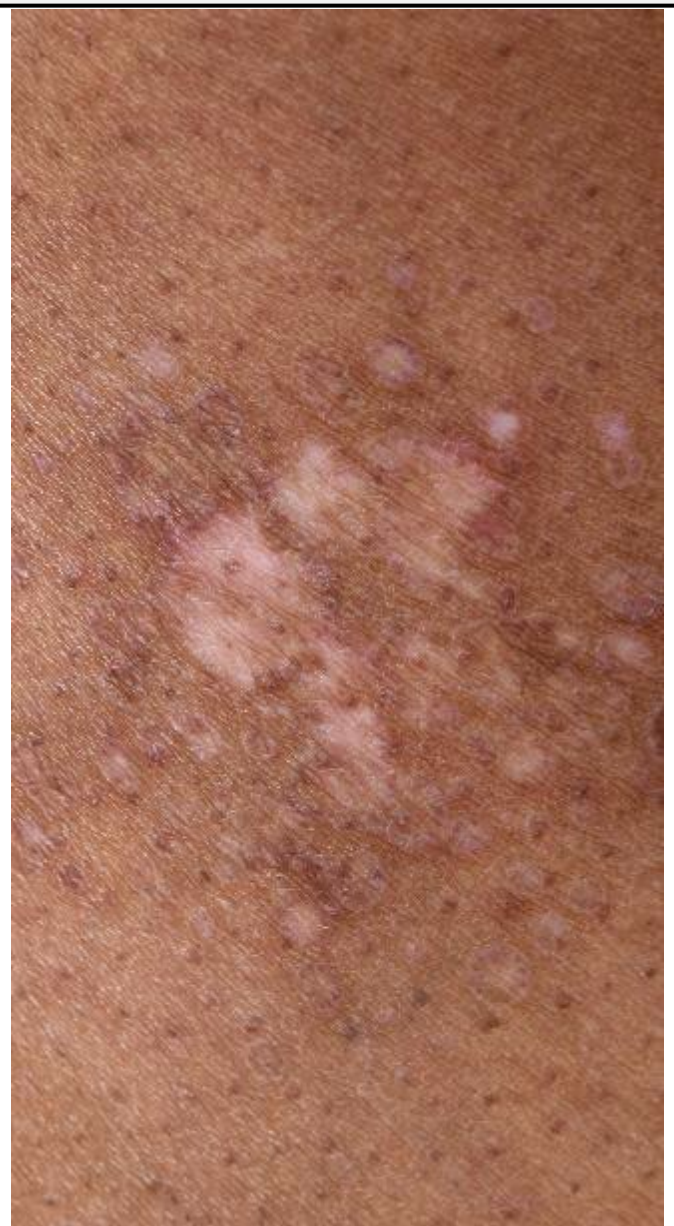
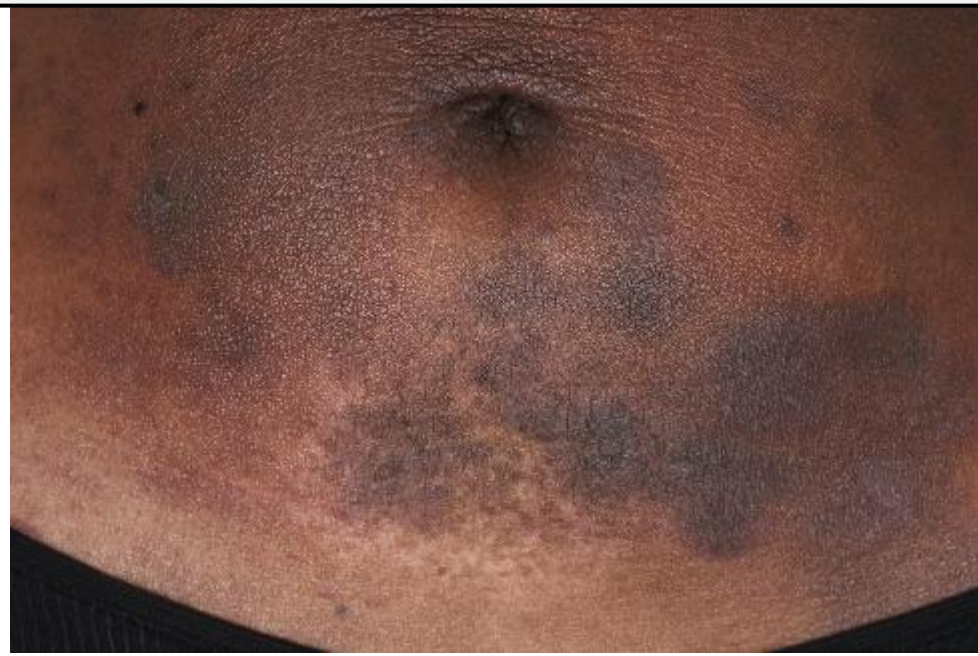
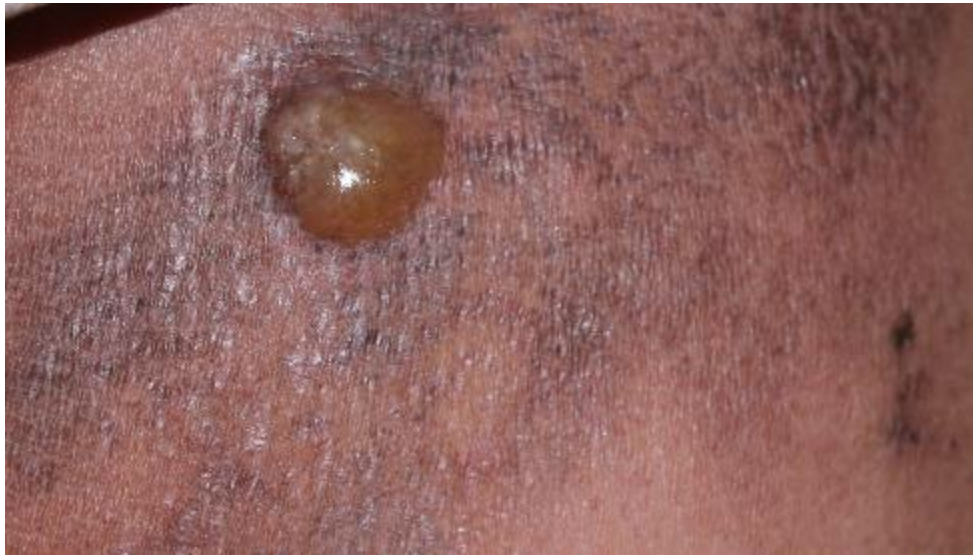
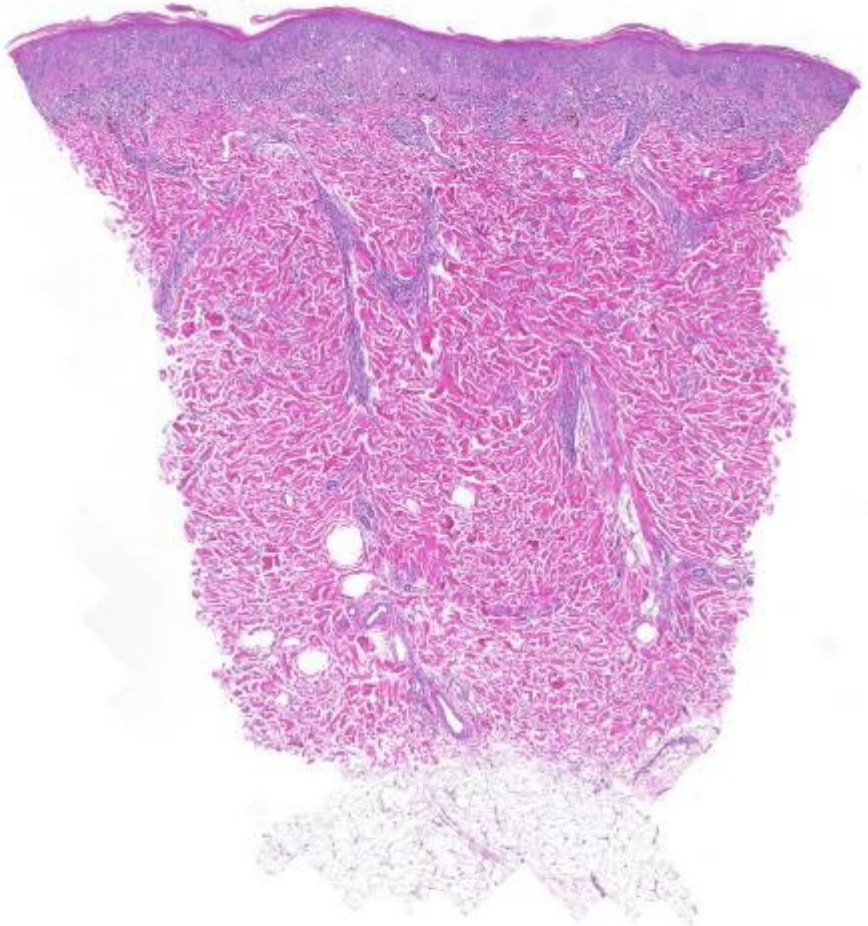


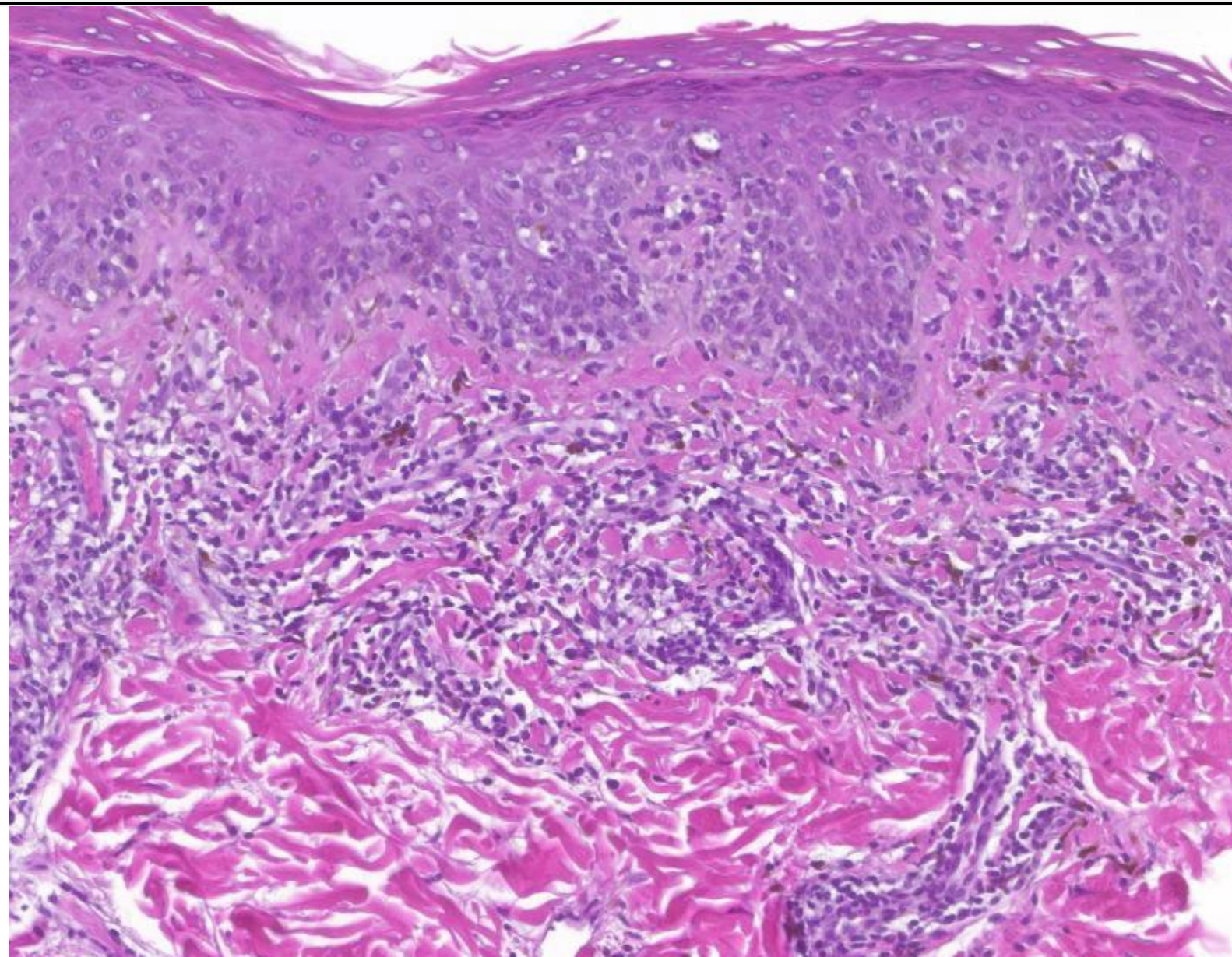


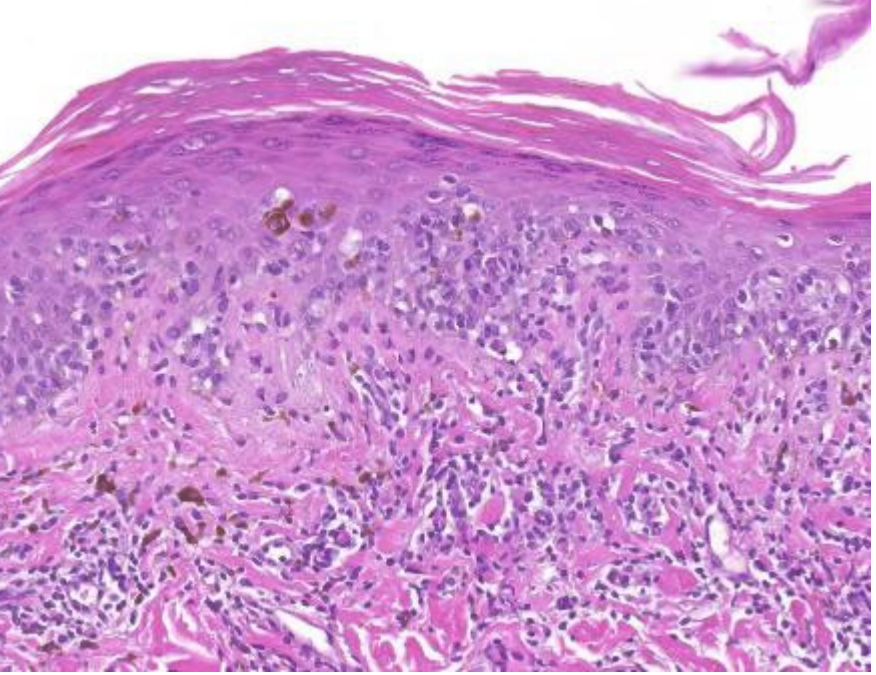
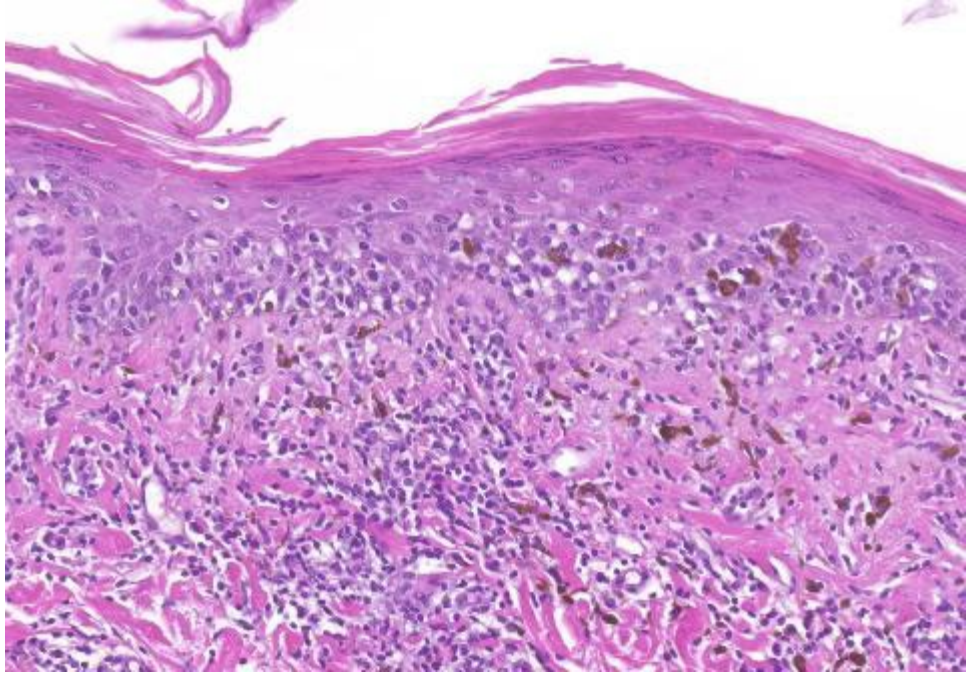
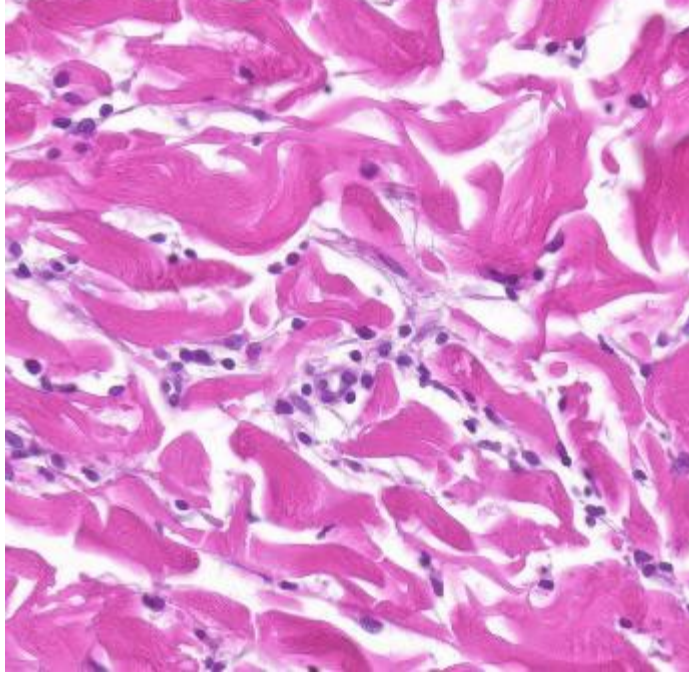
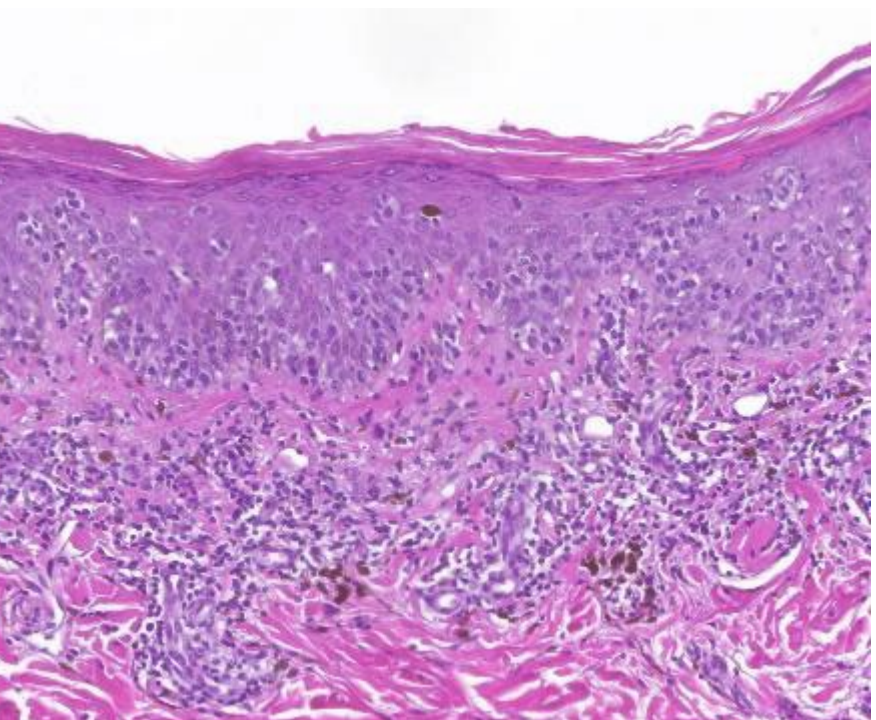
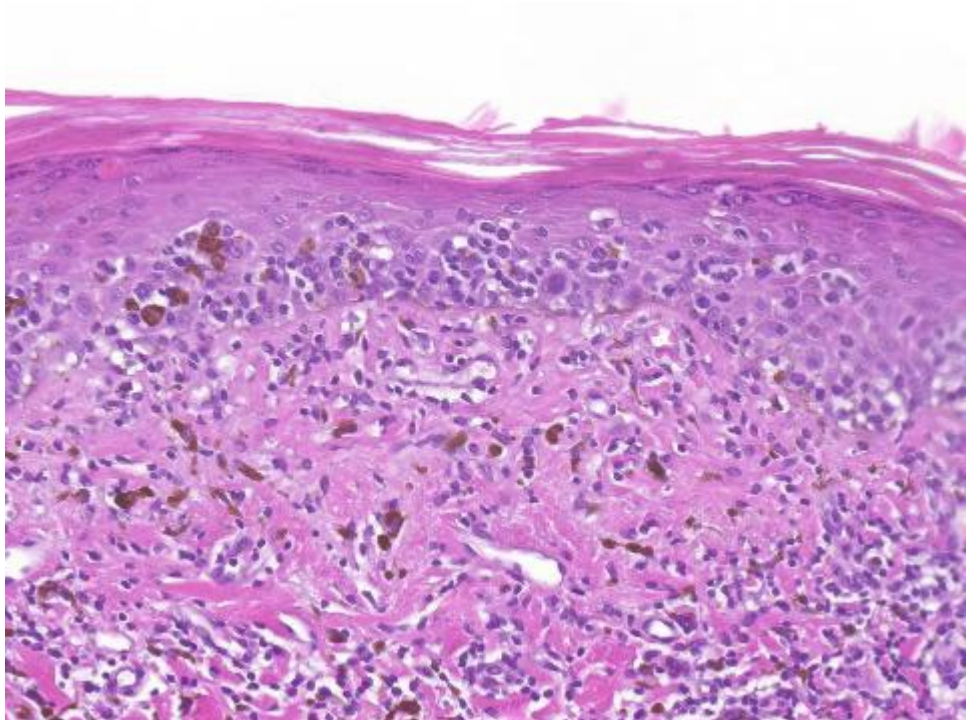
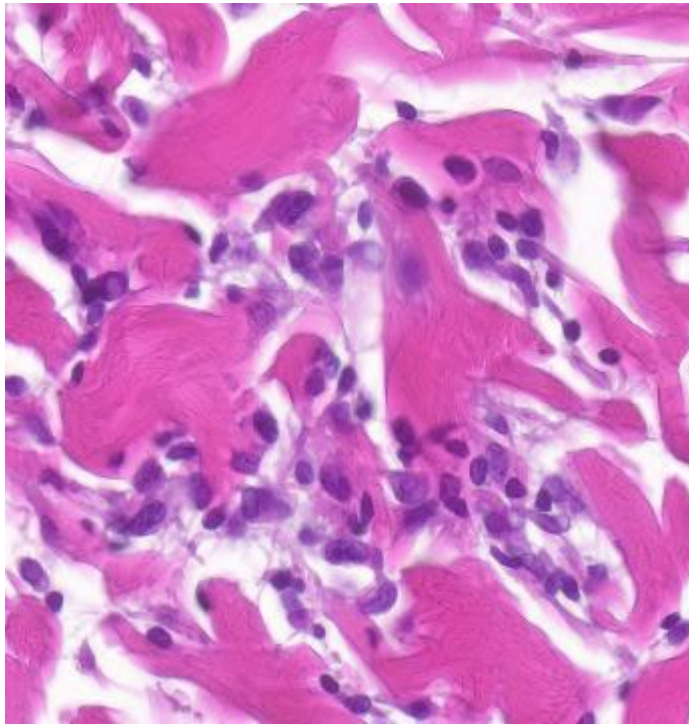




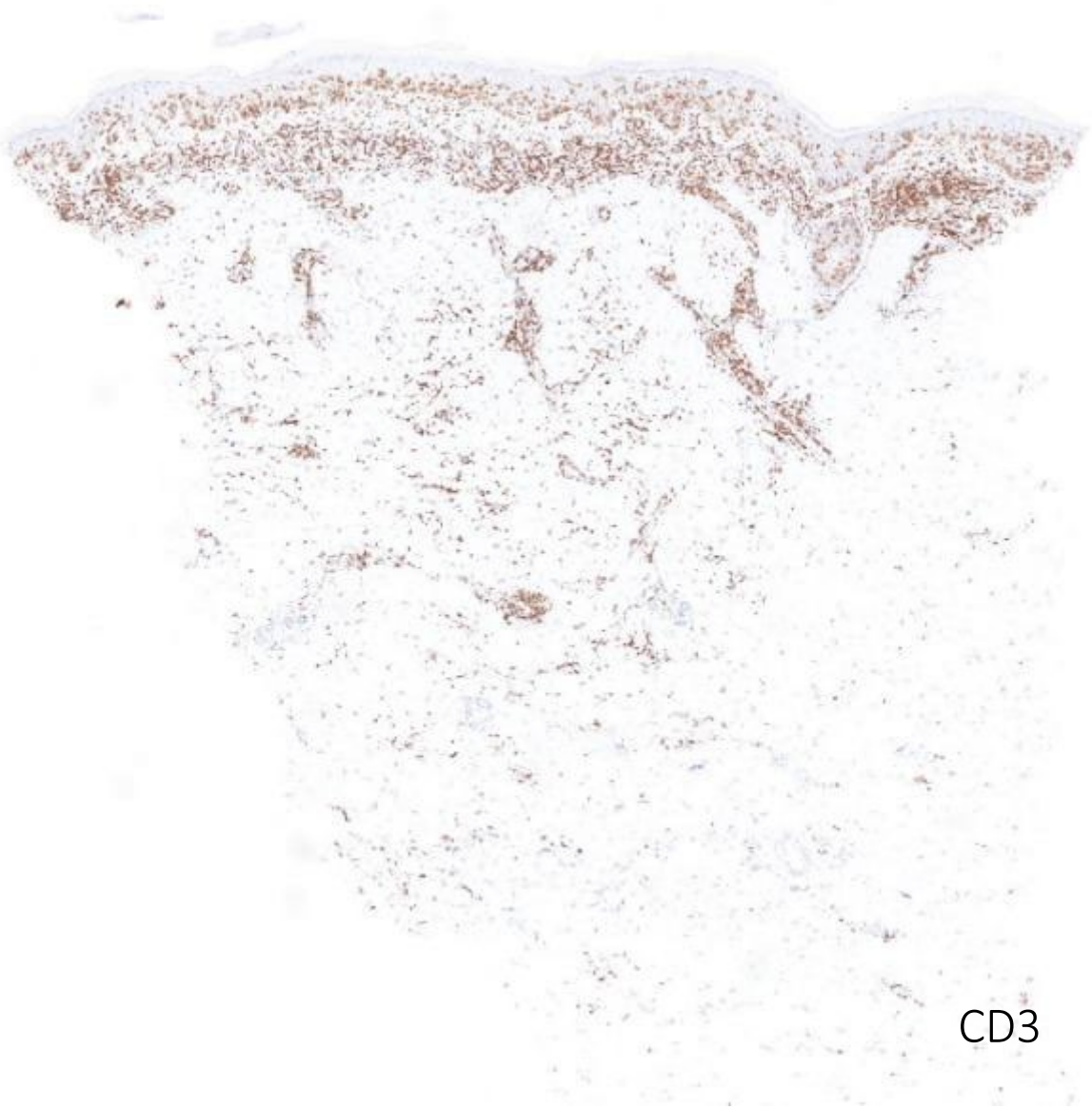
Histology:



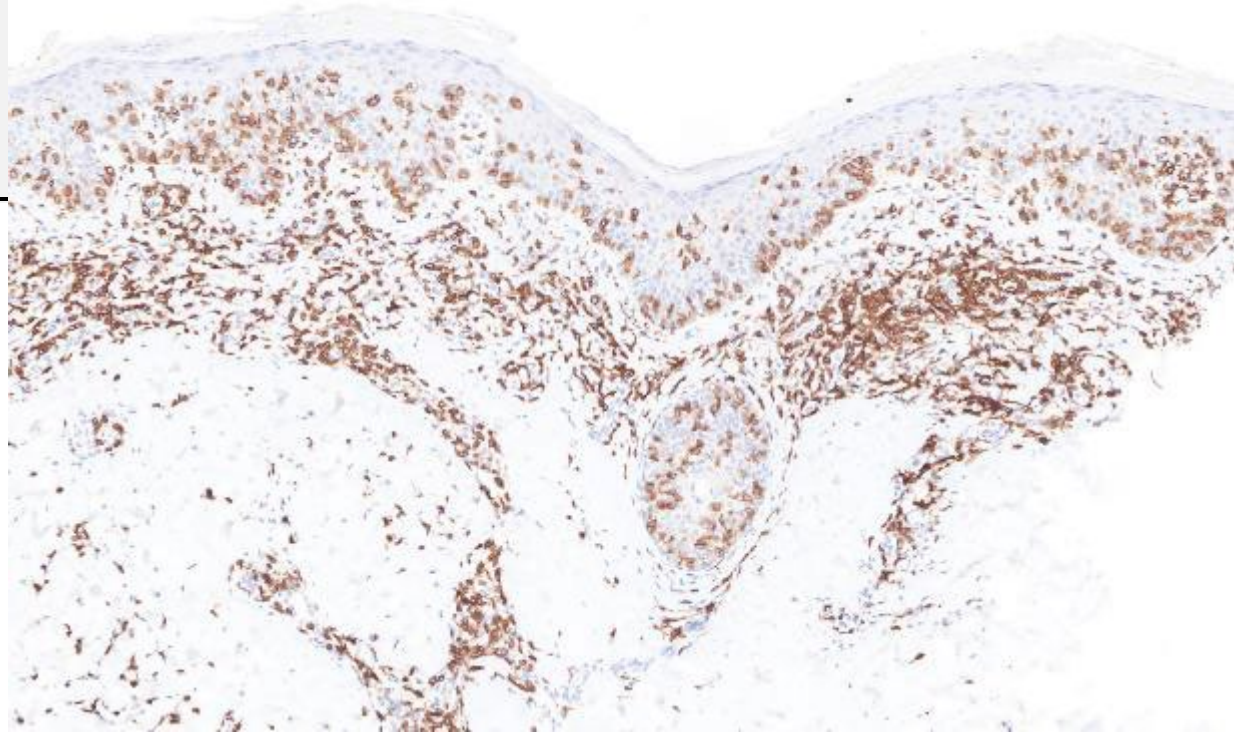


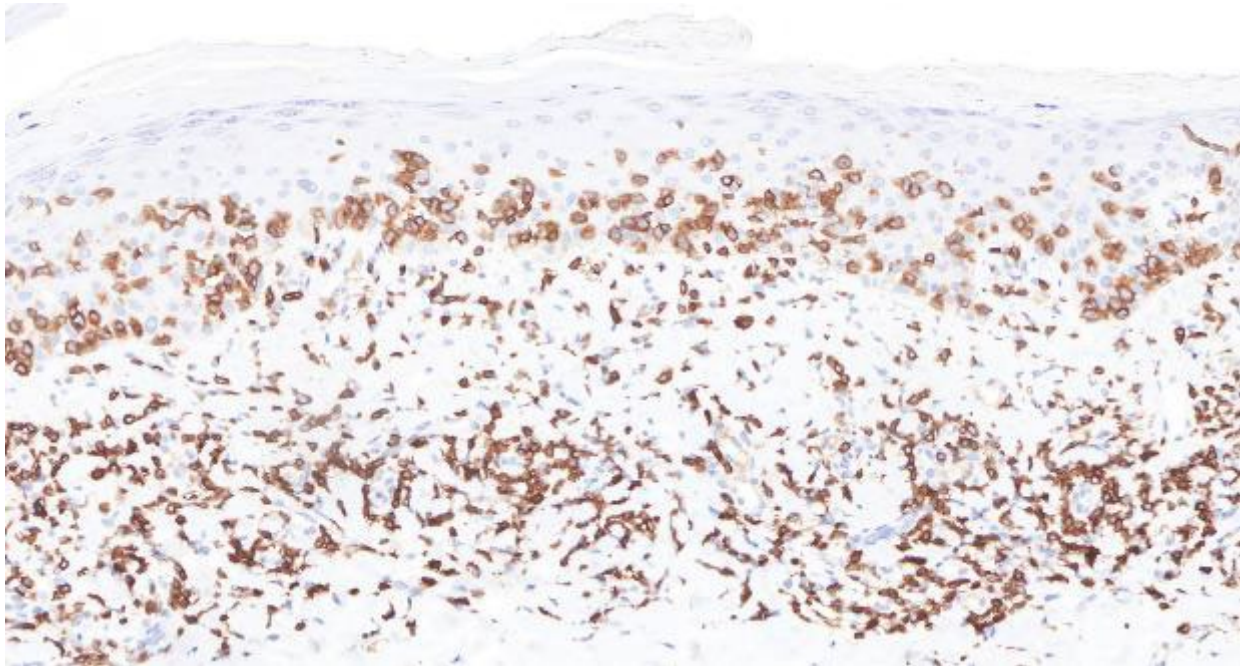







Immunohistochemistry:



CD3





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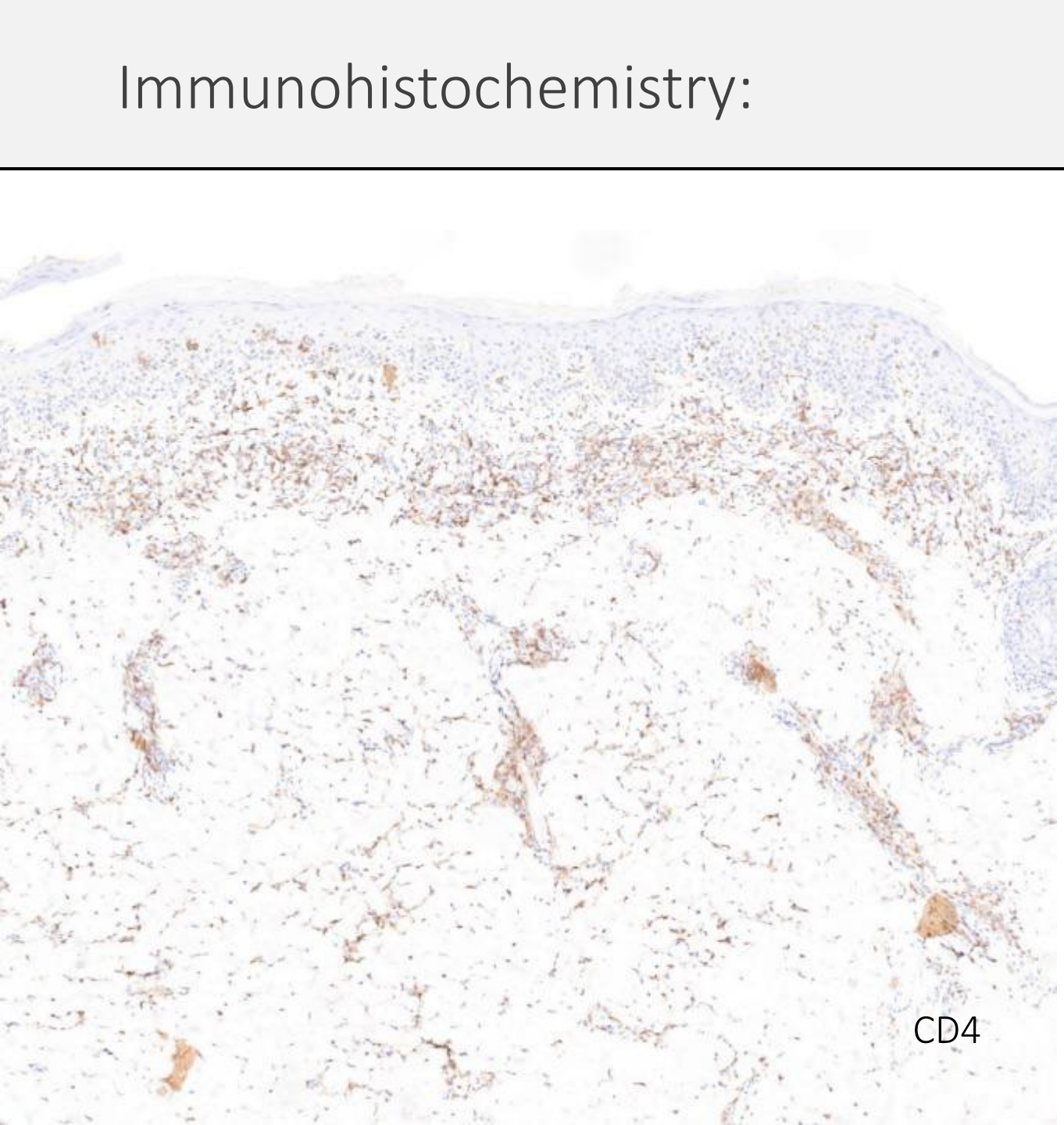

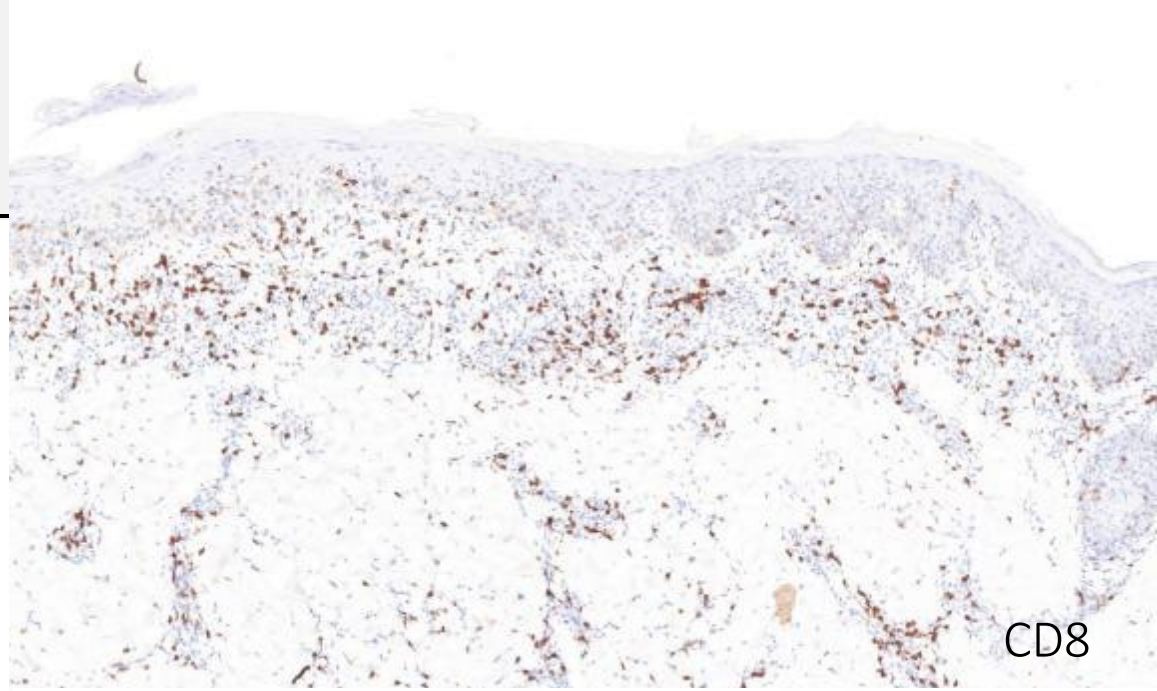

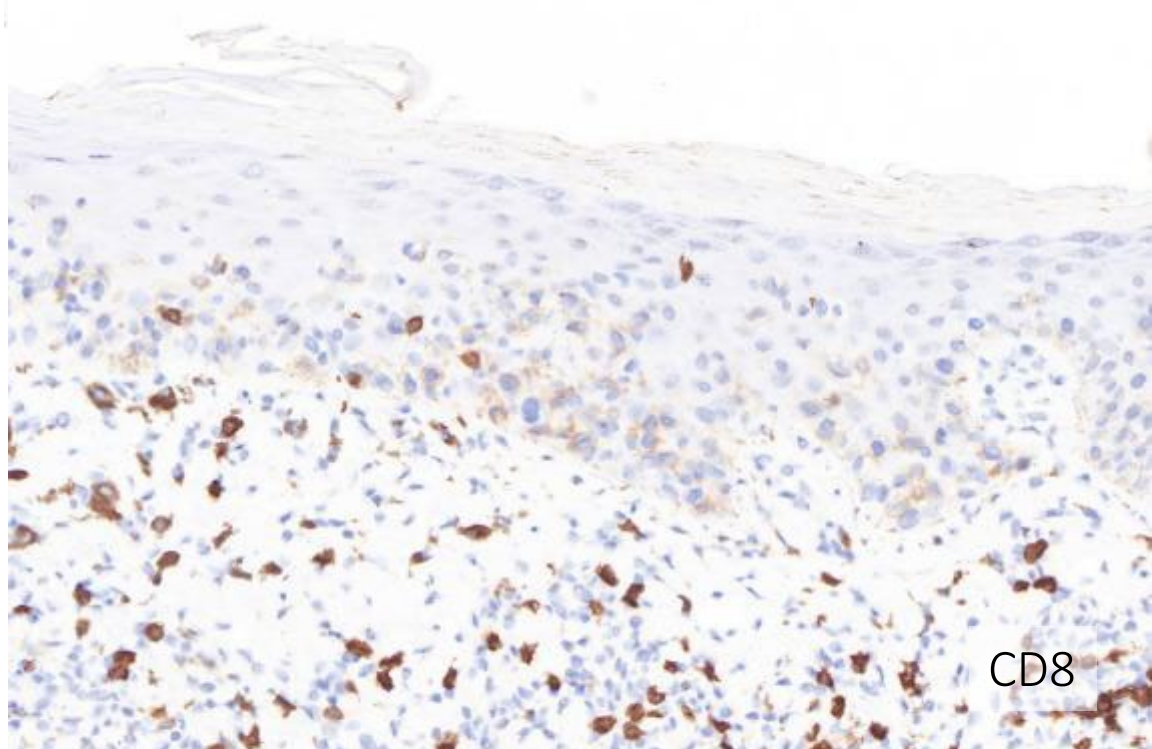

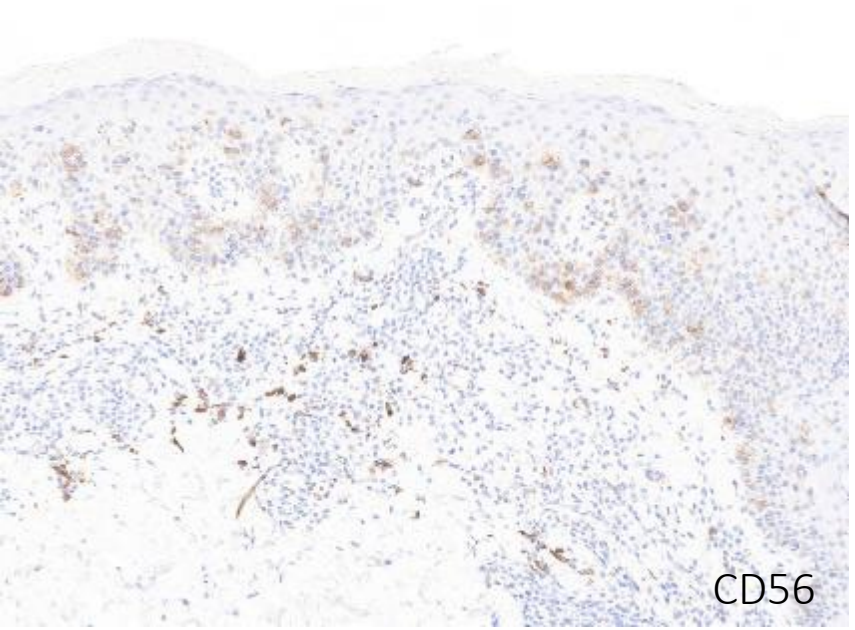
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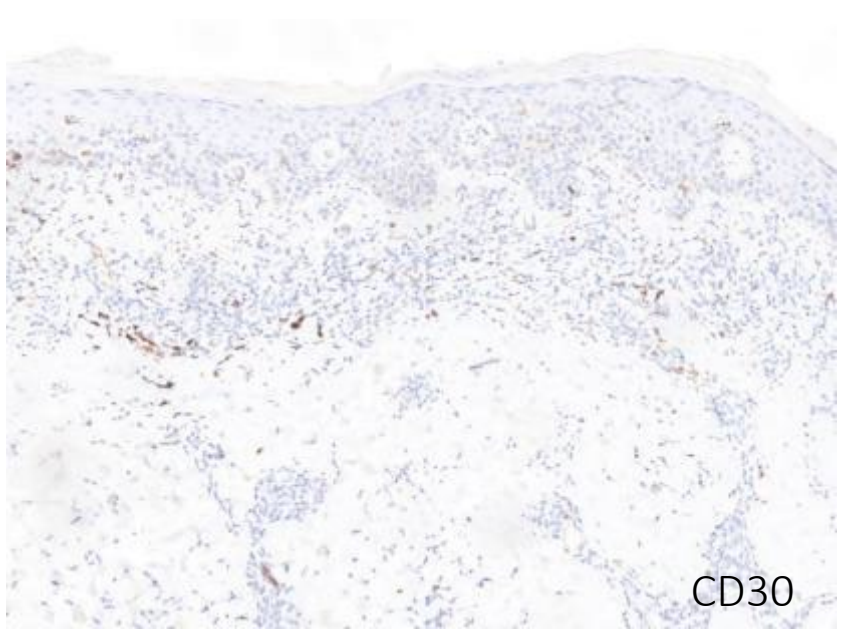
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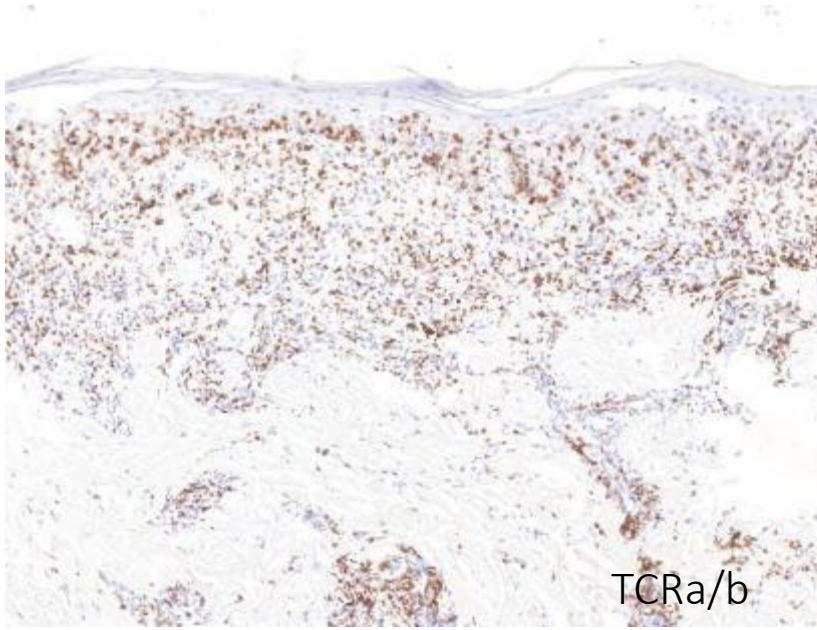
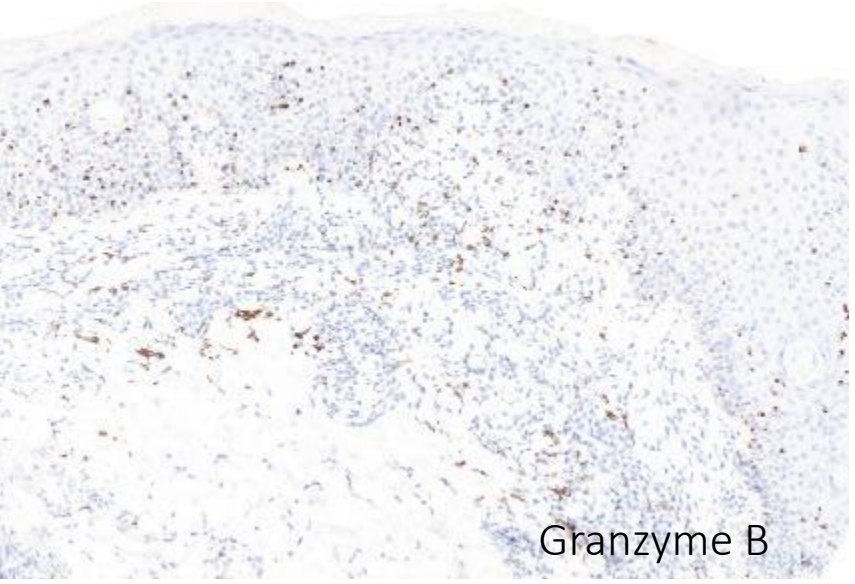
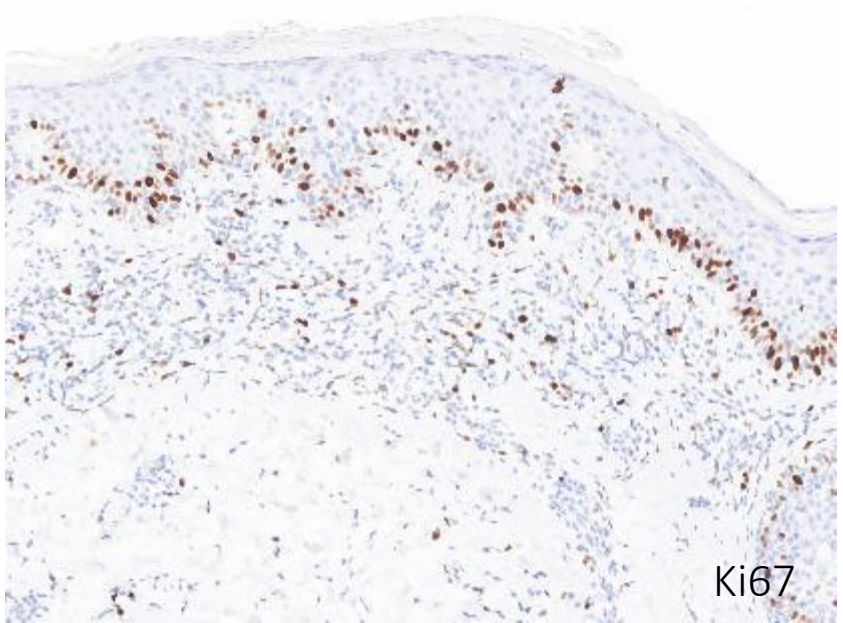
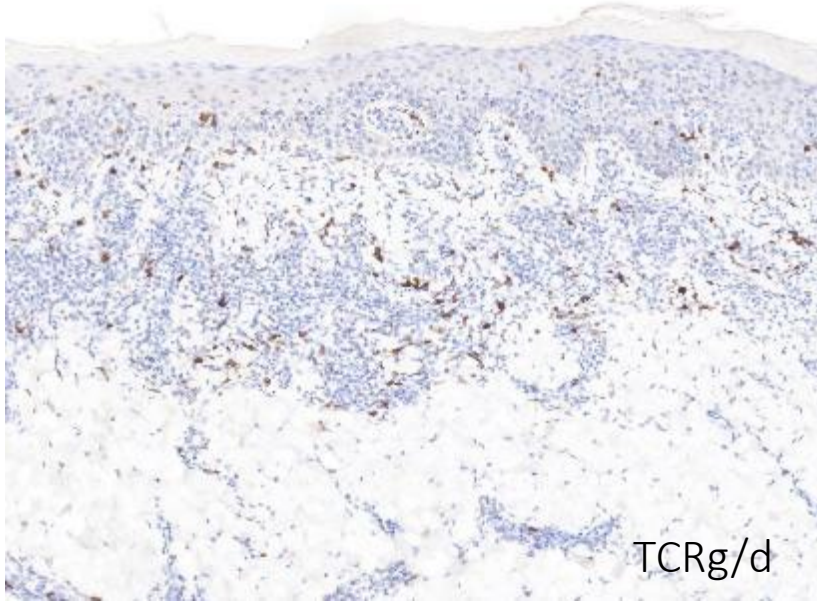
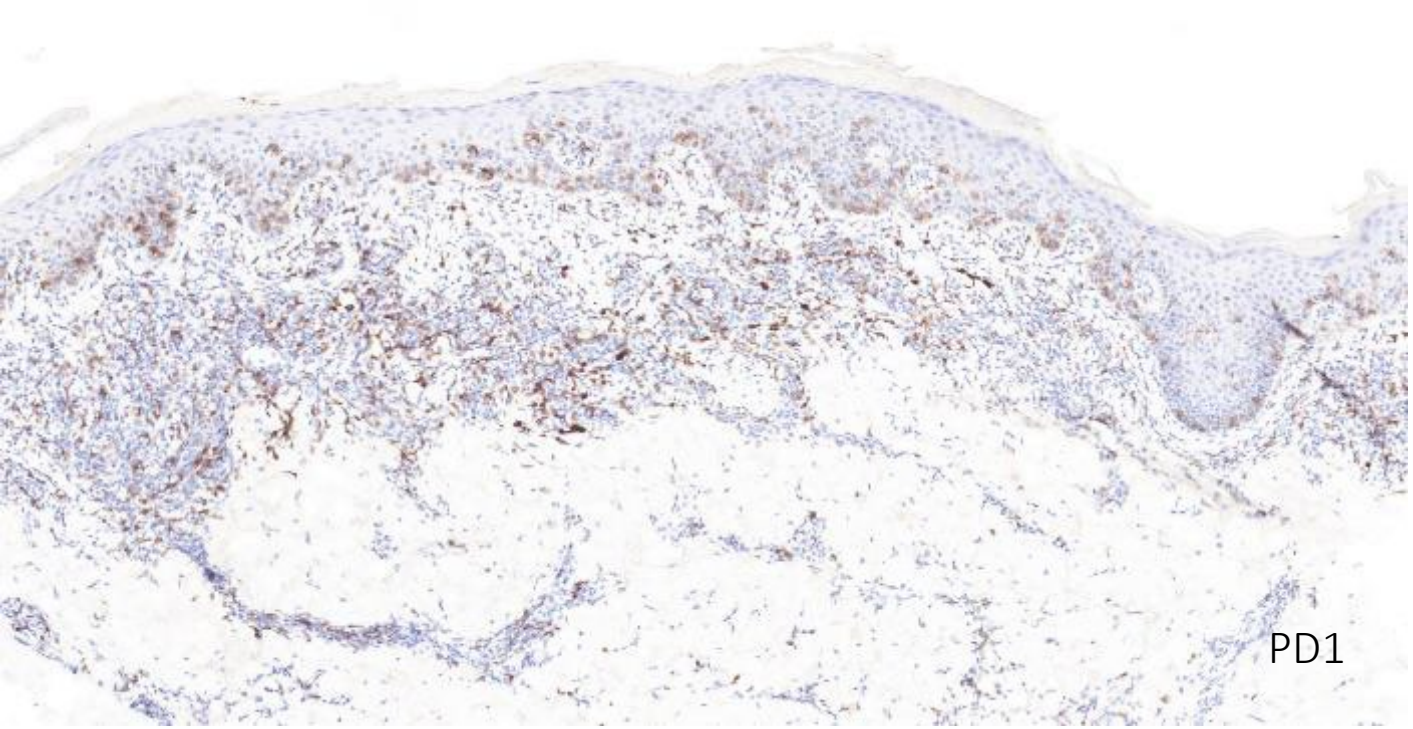

TCRa/b


Granzyme B


Ki67


TCRg/d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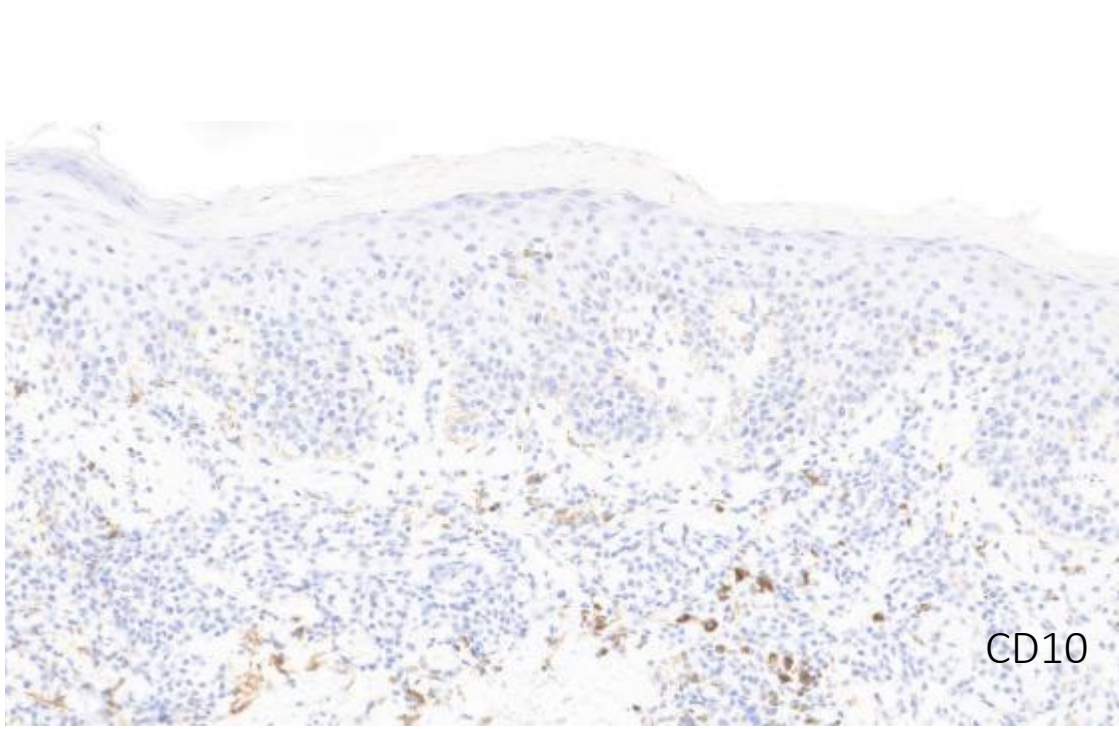
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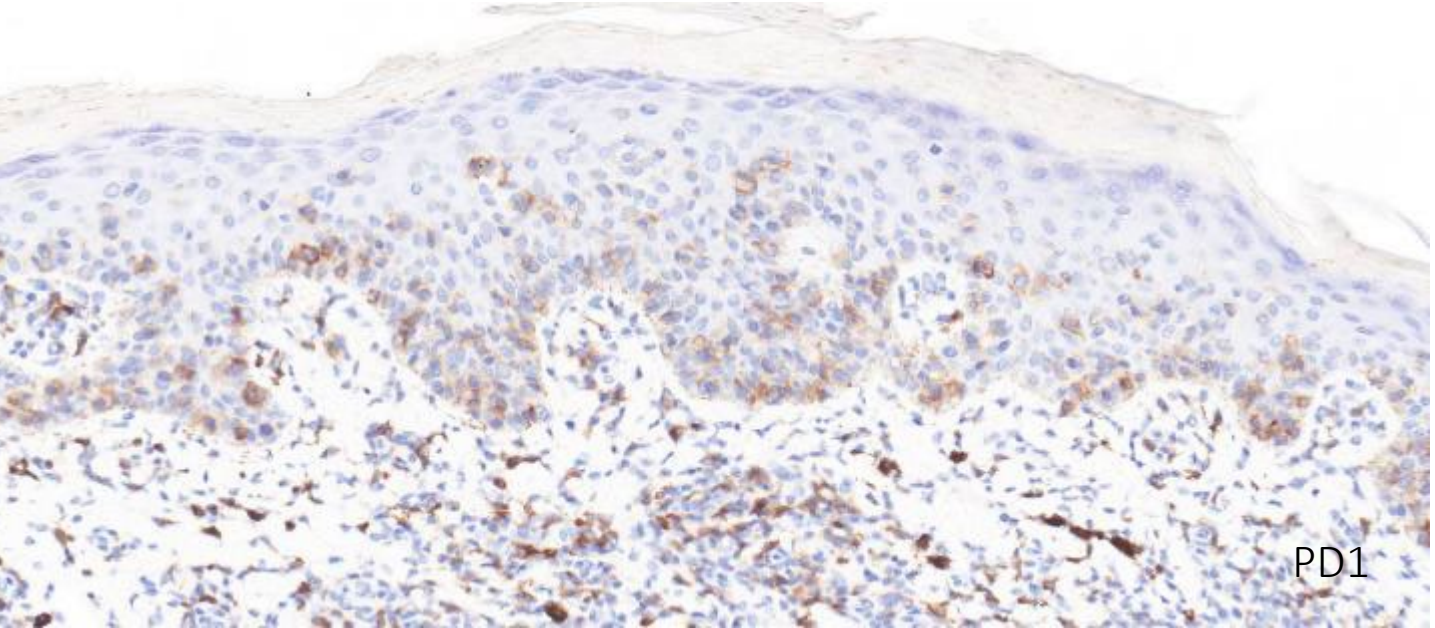
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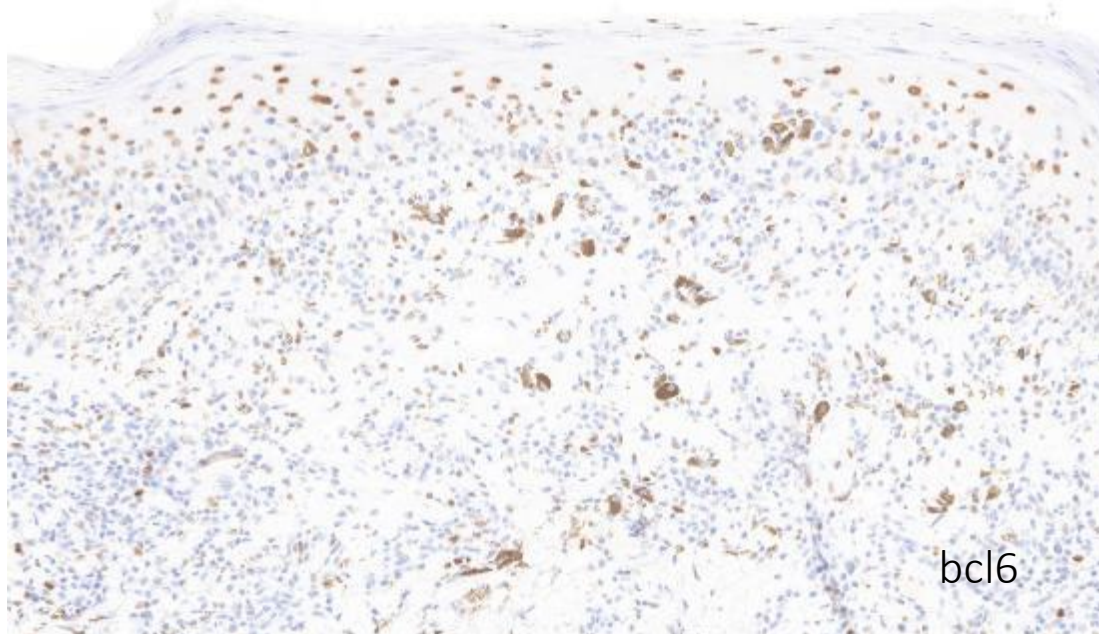
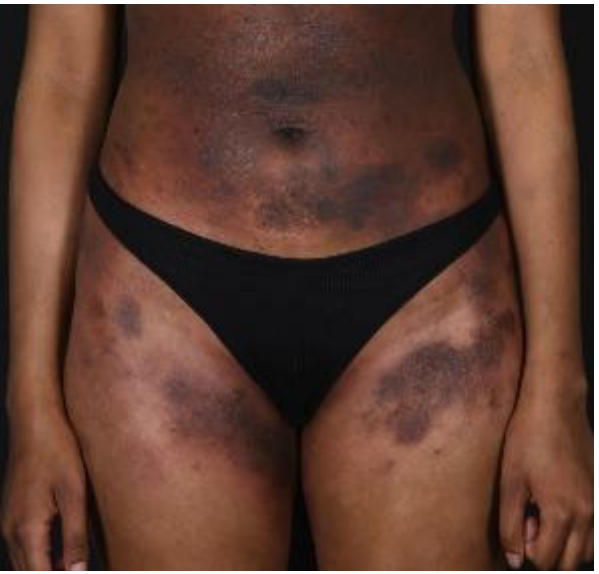
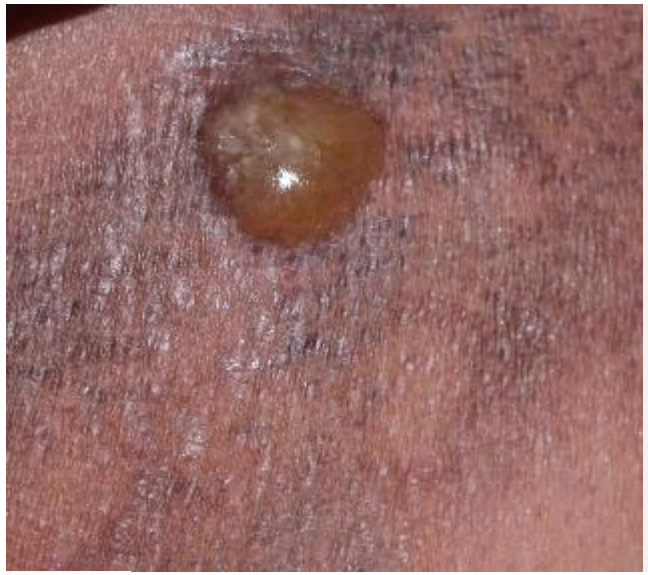

bcl6

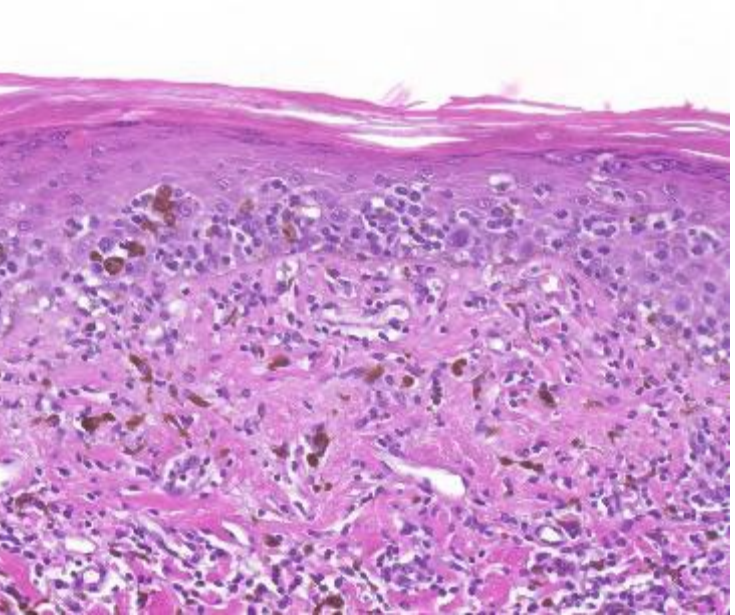
Summary of the findings:

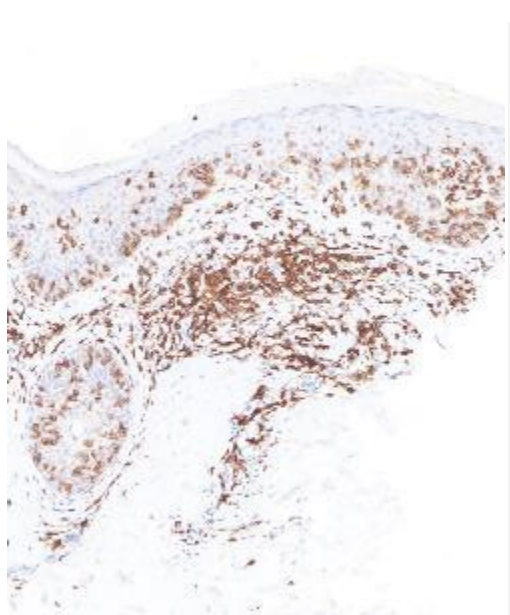




Clinical presentation:

- Disseminated hyper- und hypopigmented plaques
- Blitsers
- First skin eruption in early childhood





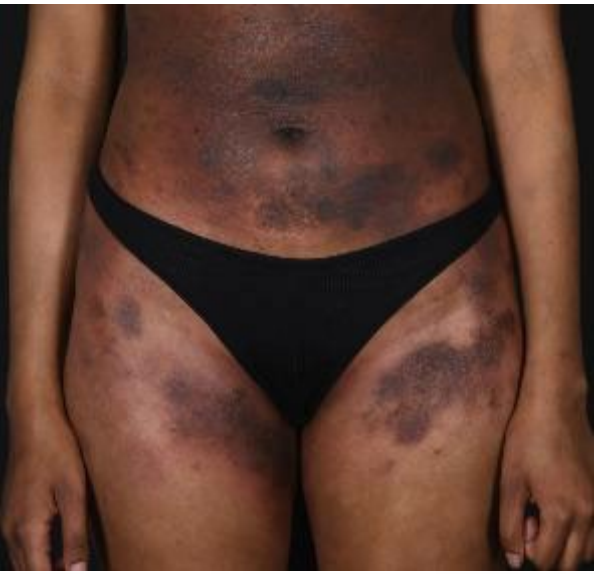
Histology:

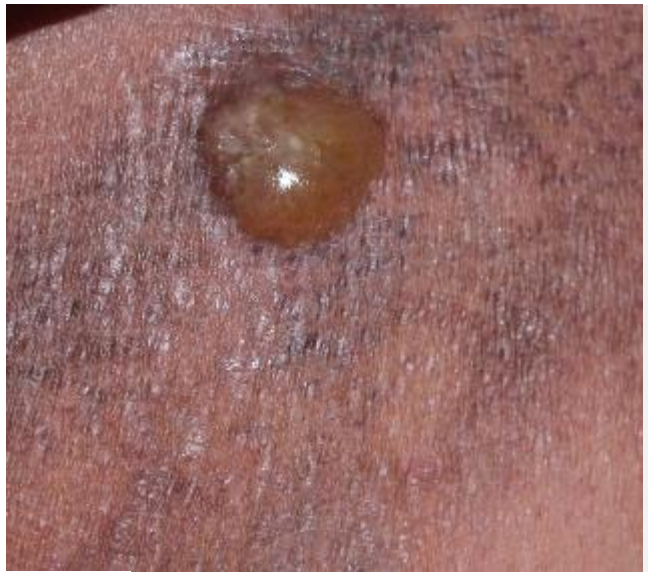
- Dense band-like infiltrate
- CD3+, CD4-, CD8+ (dim)
- CD56+ (dim)
- PD1+ (dim)
- TCRbeta+
- TCR gamma/delta-
- TCR clonal

diagnosis

?

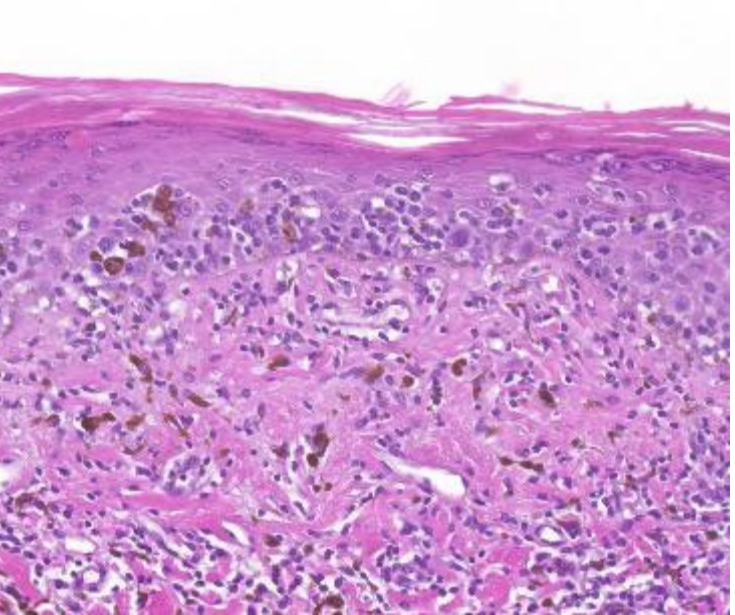
Summary of the findings:

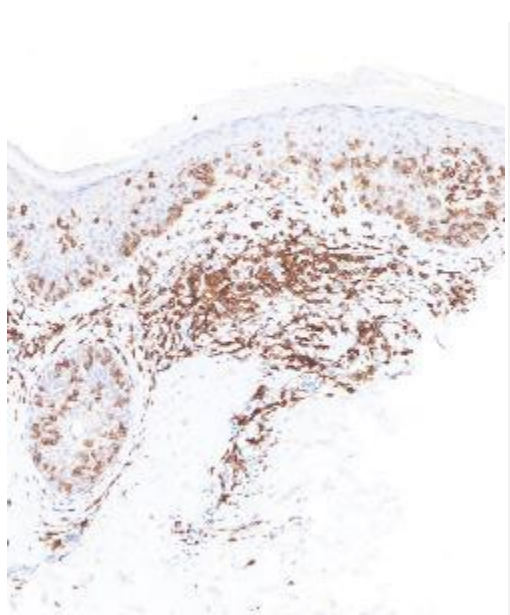




Clinical presentation:

- Disseminated hyper- und hypopigmented plaques
- Blitsers
- First skin eruption in early childhood





Histology:

- Dense band-like infiltrate
- CD3+, CD4-, CD8+ (dim)
- CD56+ (dim)
- PD1+ (dim)
- TCRbeta+
- TCR gamma/delta-
- TCR clonal

Bullous Mycosis fungoides
CD8+ (dim)
CD56+ (dim)
PD1+ (dim)

Bullous mycosis fungoides

- Very rare: approximately 20 cases have been reported
- Mycosis fungoides bullosa is largely restricted to older patients with an more aggressive clinical course
- Vesicles and blisters usually arise in typical plaques and tumours but also in normal-appearing skin.
- The pathological mechanism is unclear.

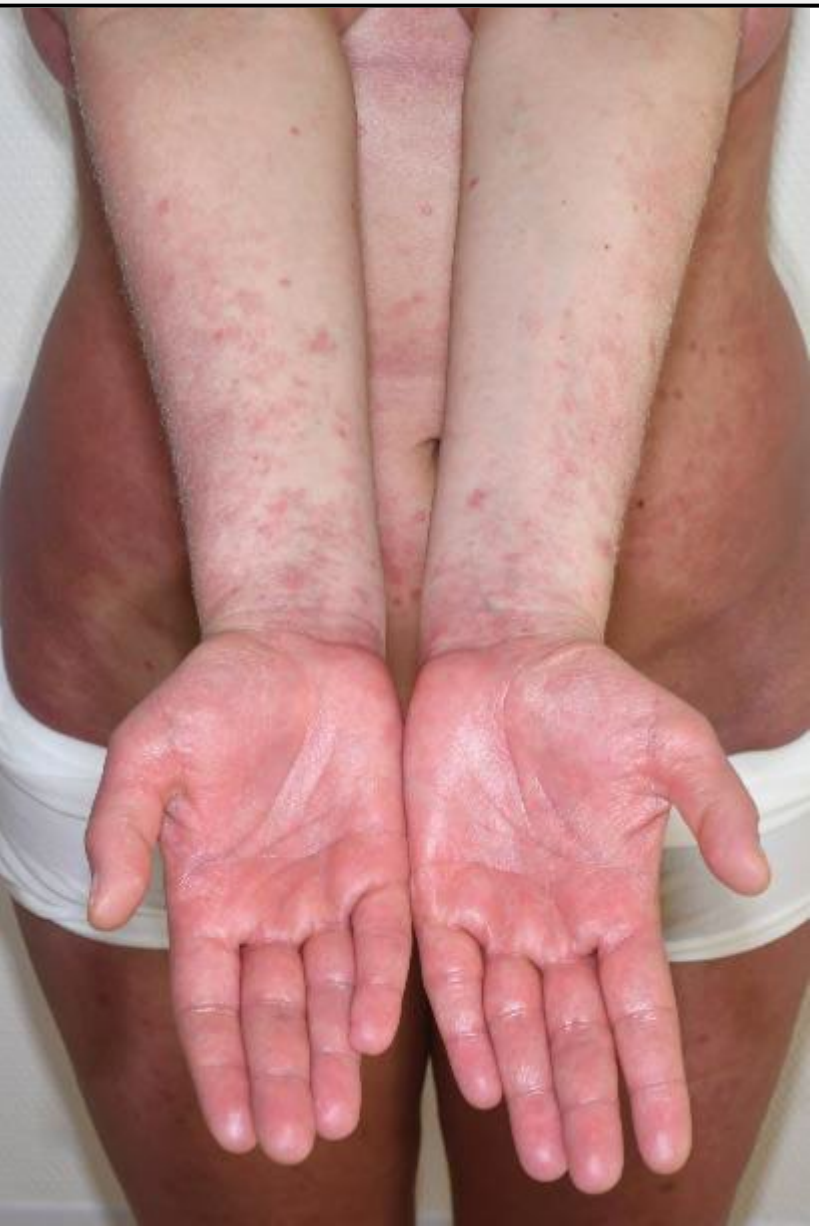
Recommended diagnostic criteria:

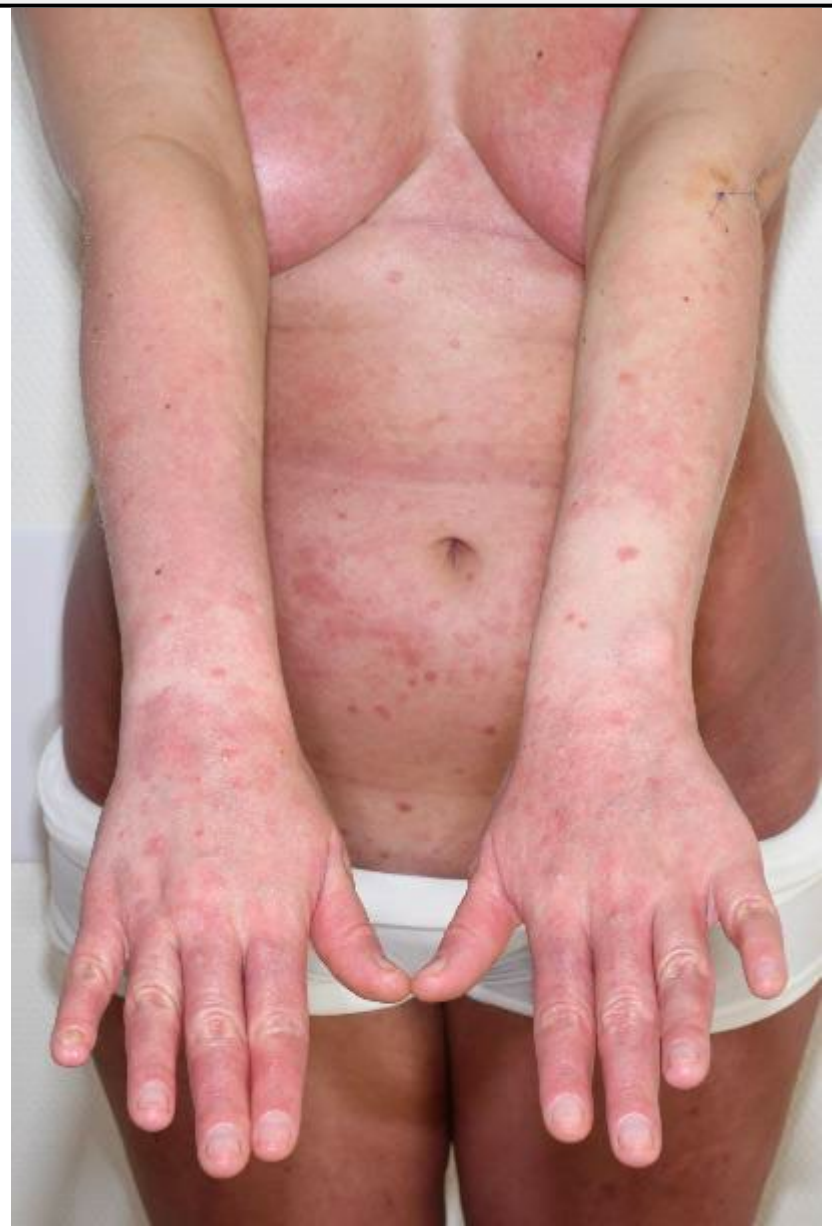
(1) Clinically apparent vesiculobullous lesions, with or without typical mycosis fungoides lesions (patches, plaques, tumors)
(2) Typical histologic features of mycosis fungoides (atypical lyphoid cells, epidermotropism, Pautrier's microabscesses) with intraepidermal or subepidermal blisters;
(3) Negative immunofluorescence (both direct and indirect, if possible) to rule out concomitant autoimmune bullous diseases;
(4) Negative evaluation for other possible causes of vesiculobullous lesions (eg, medications, bacterial or viral infection, porphyria, phototherapy).

Bowman PH, Hogan DJ, Sanusi ID. Mycosis fungoides bullosa: report of a case and review of the literature. Am Acad Dermatol. 2001;45(6):934-9.
Kneitz H, Bröcker E-V, Becker J. Mycosis fungoides bullosa: a case report and review of the literature. J Med Case Rep. 2010 Mar 3;4:78.

Case 5

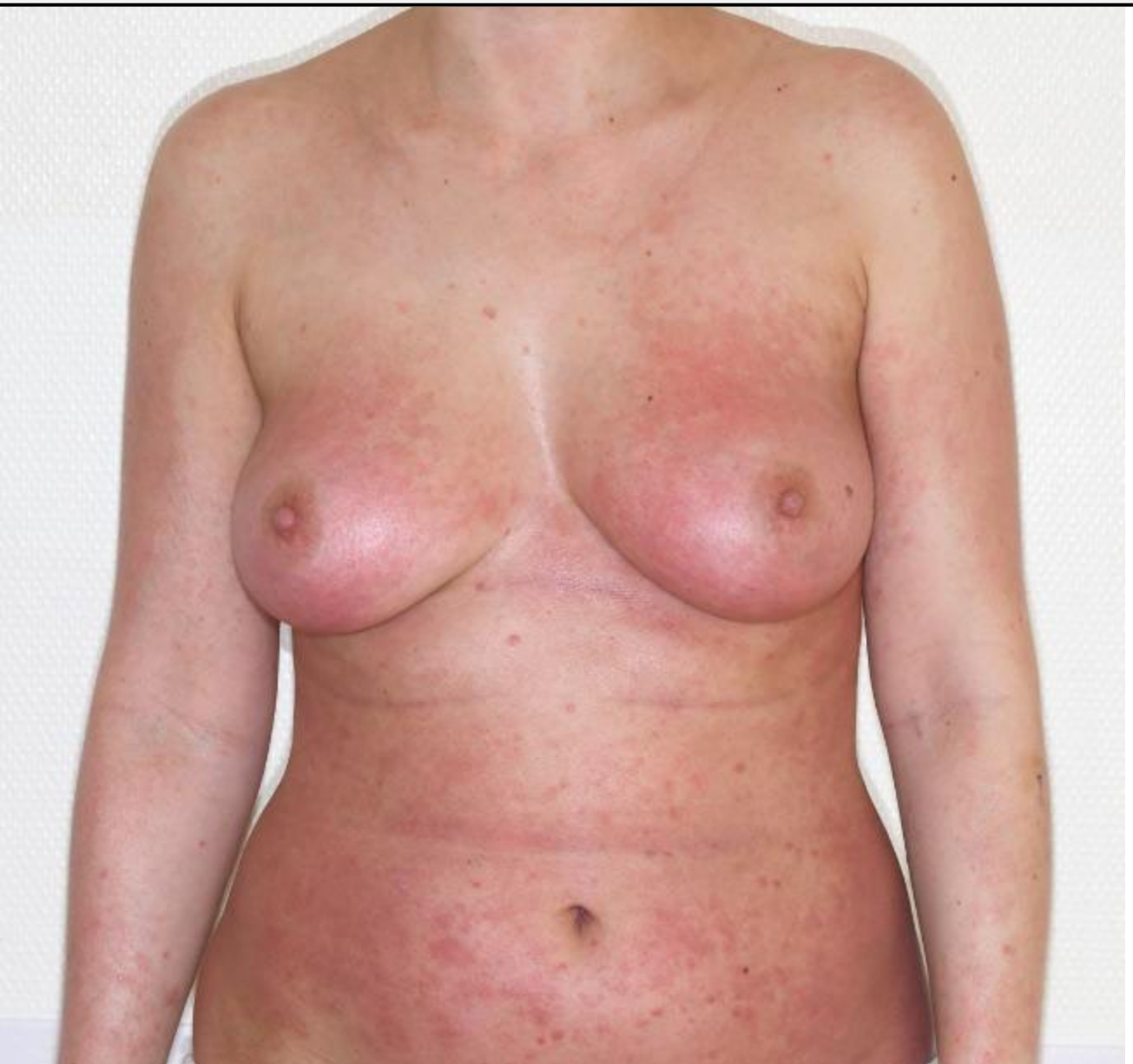
Clinical presentation:

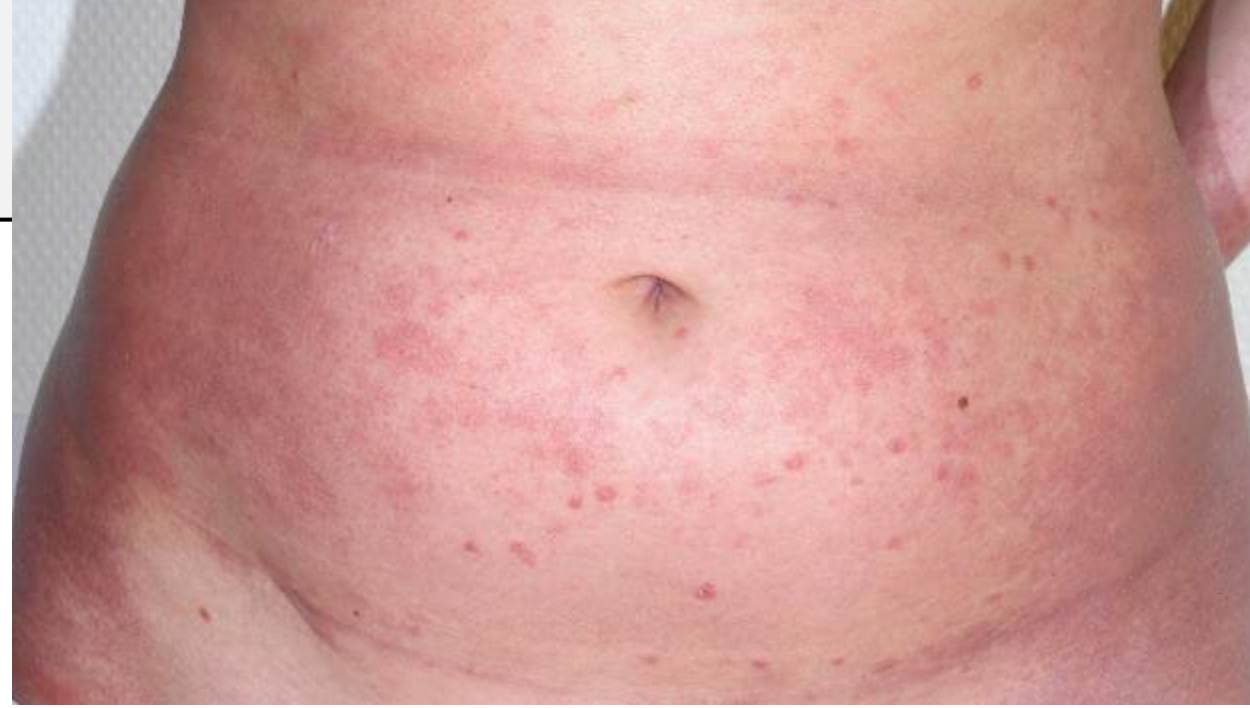


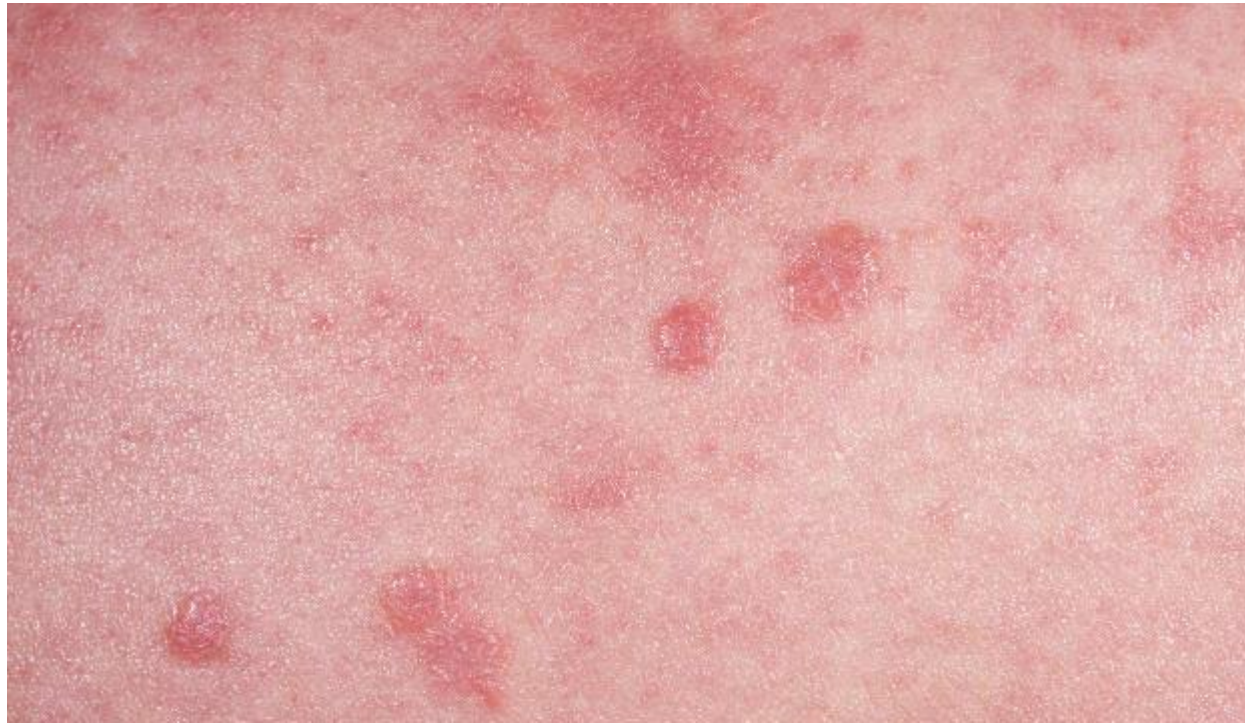


- 31-year-old female patient from Ukraine
- Skin eruption for 12 months
- No history of atopy
- Itching erythematous maculae and flat annular plaques
- Erythema on the hands and feed
- Suspected clinical diagnosis: eczema, rule out CTCL

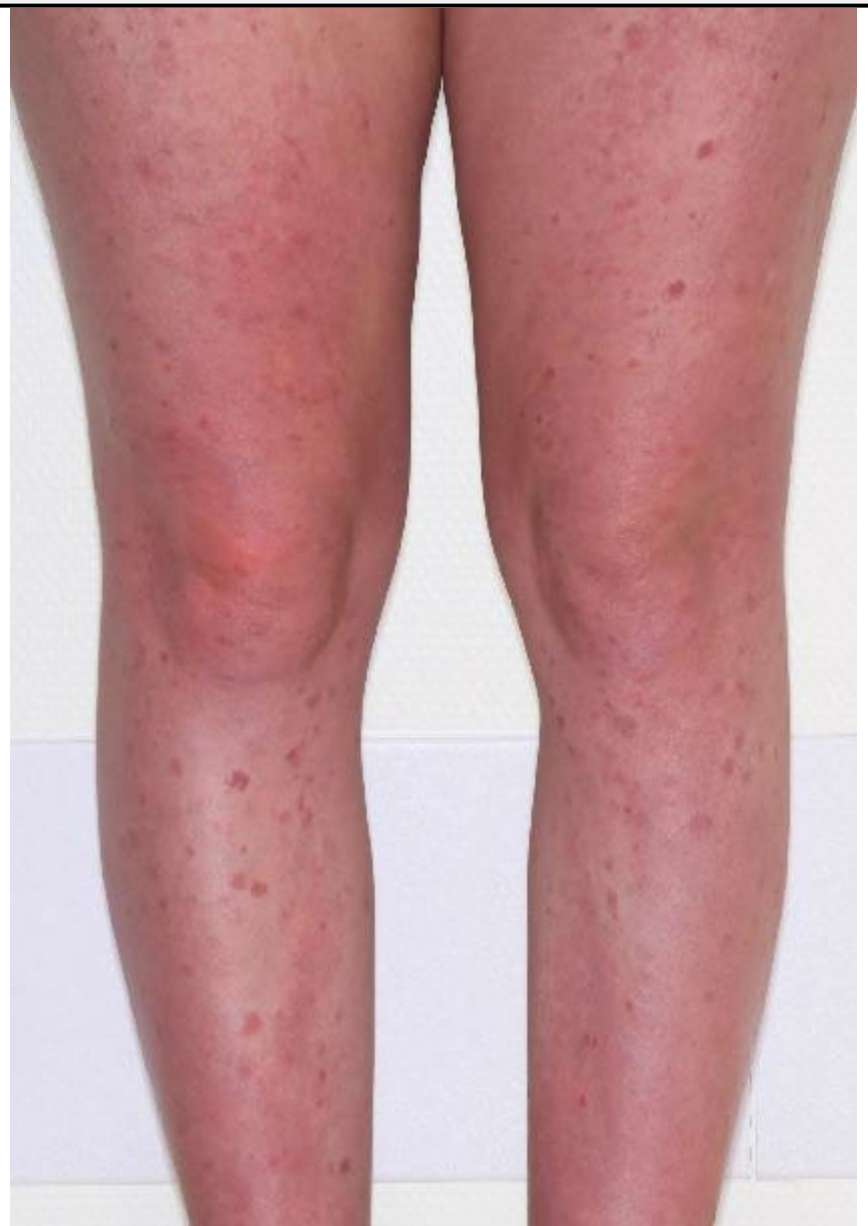
Clinical presentation:

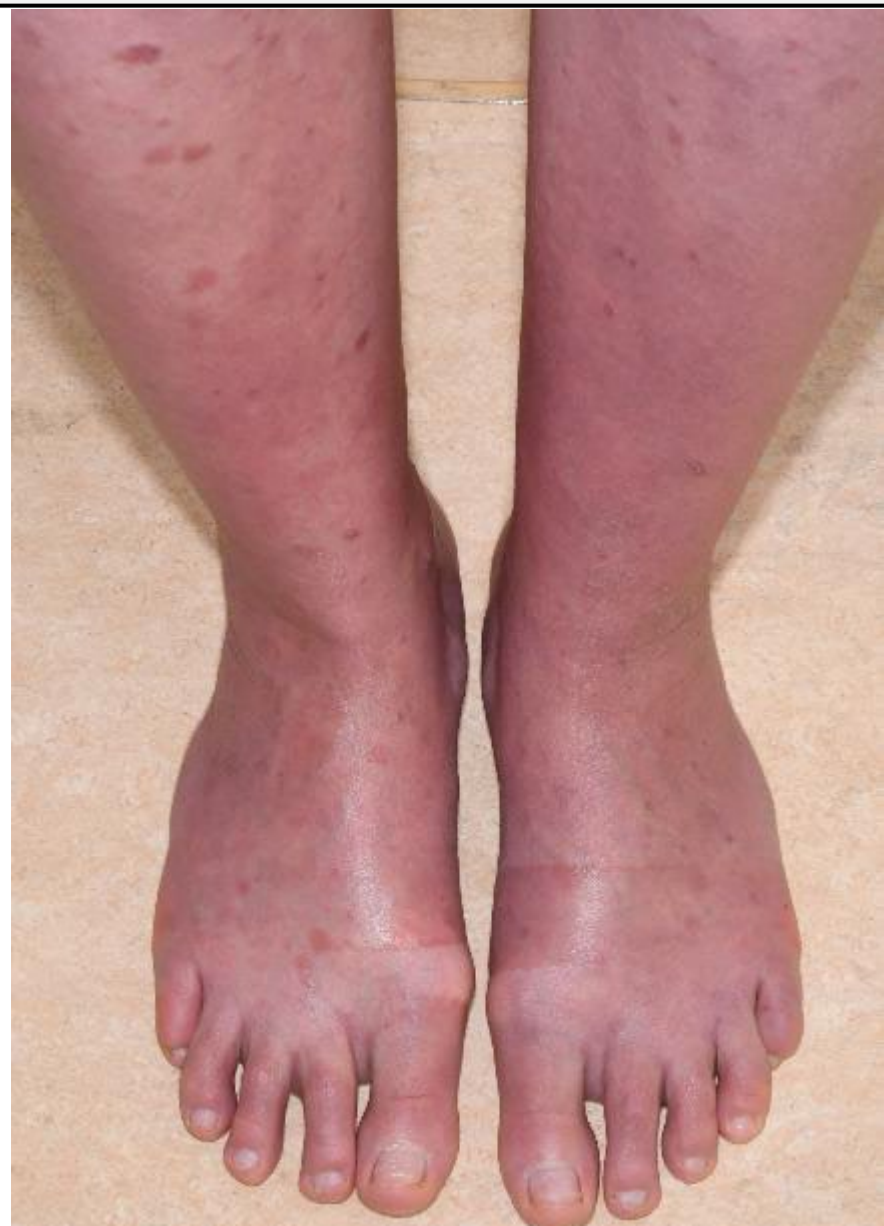




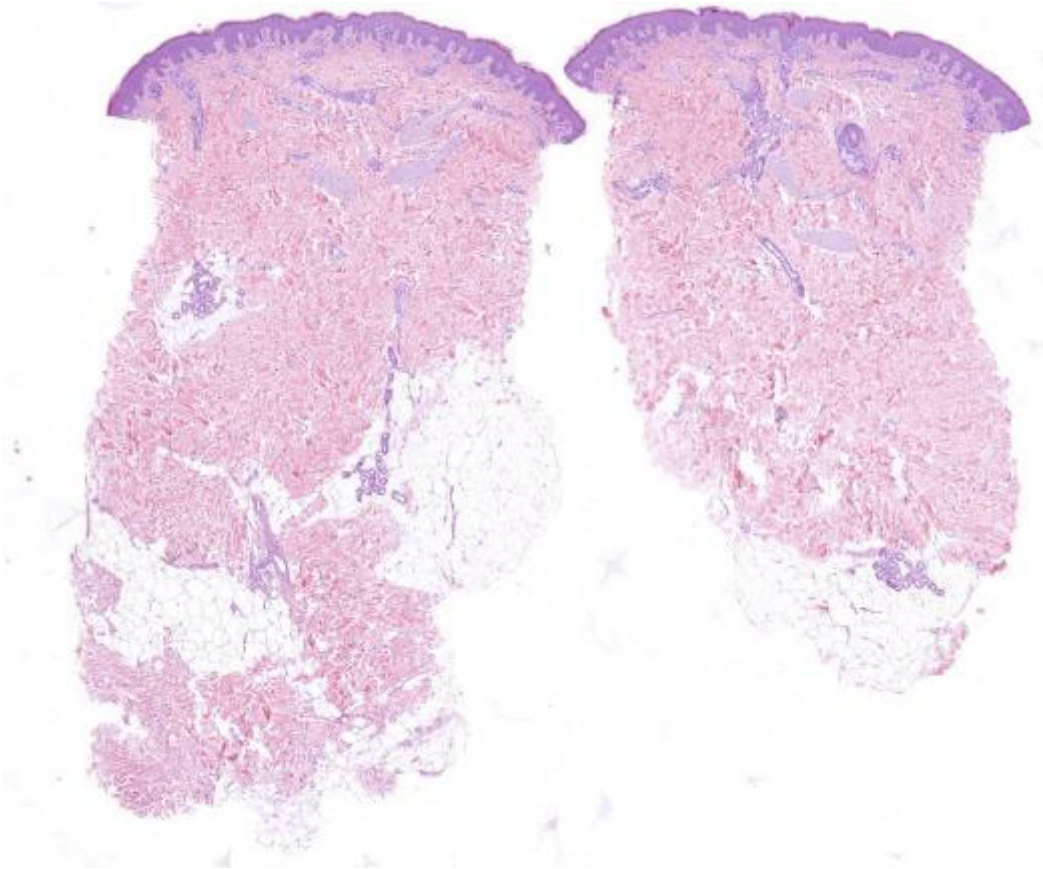


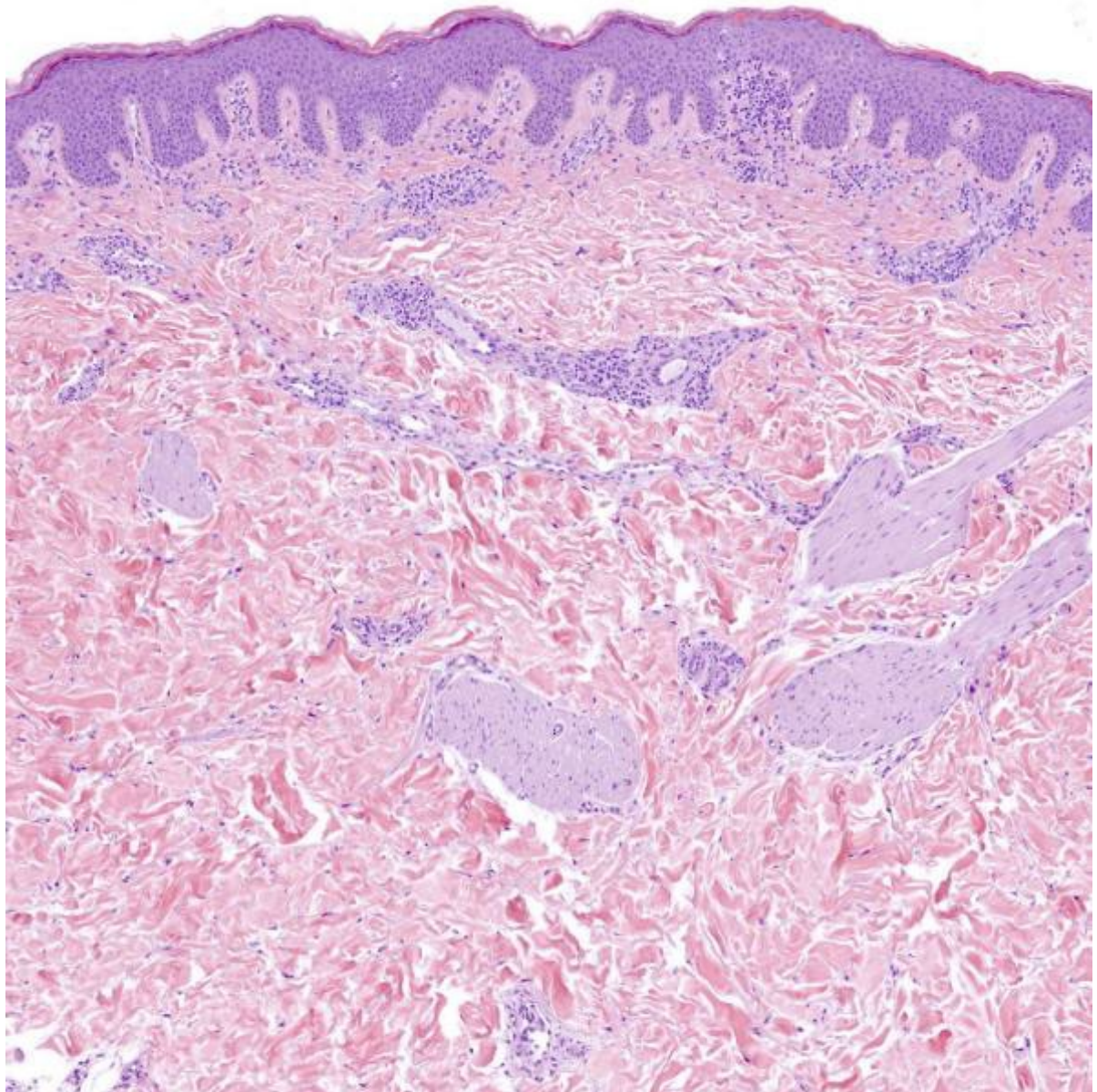
Clinical presentation:

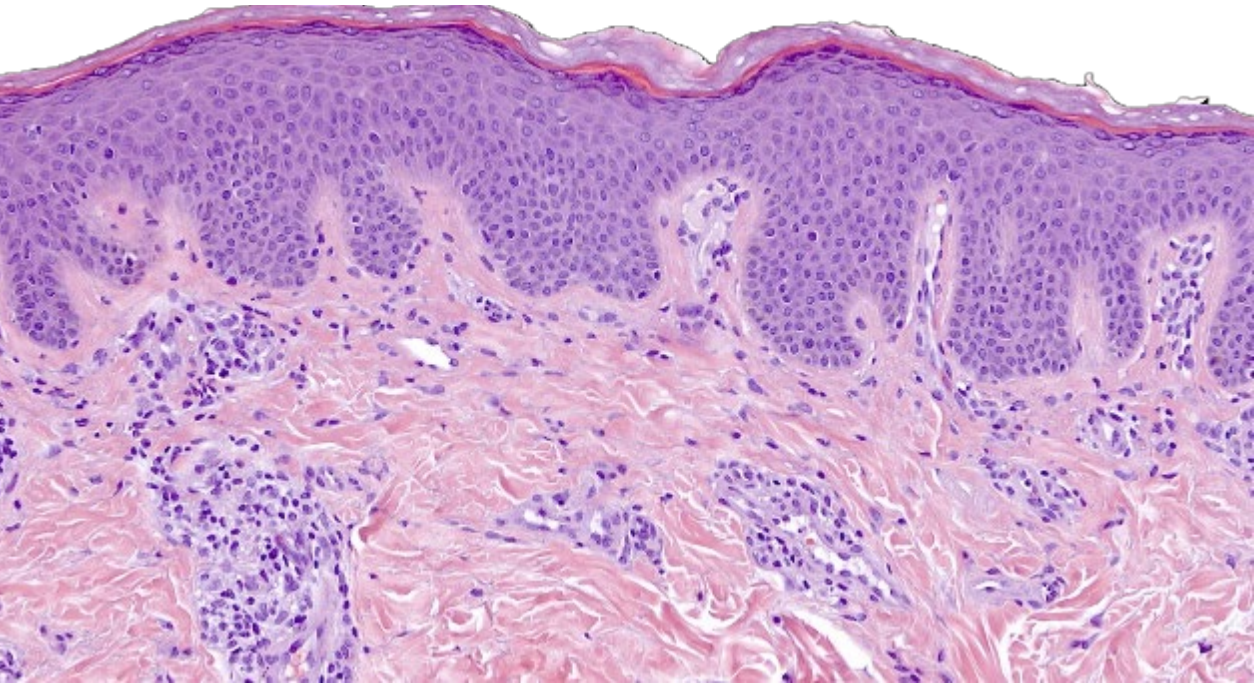


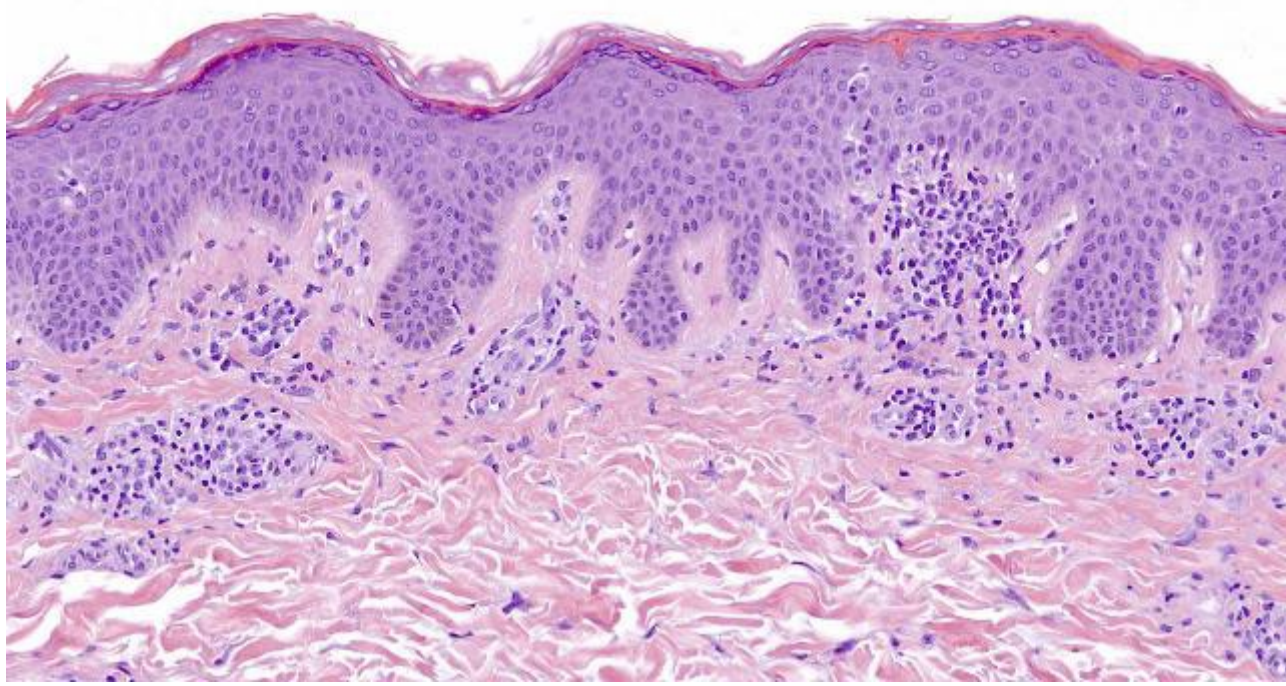


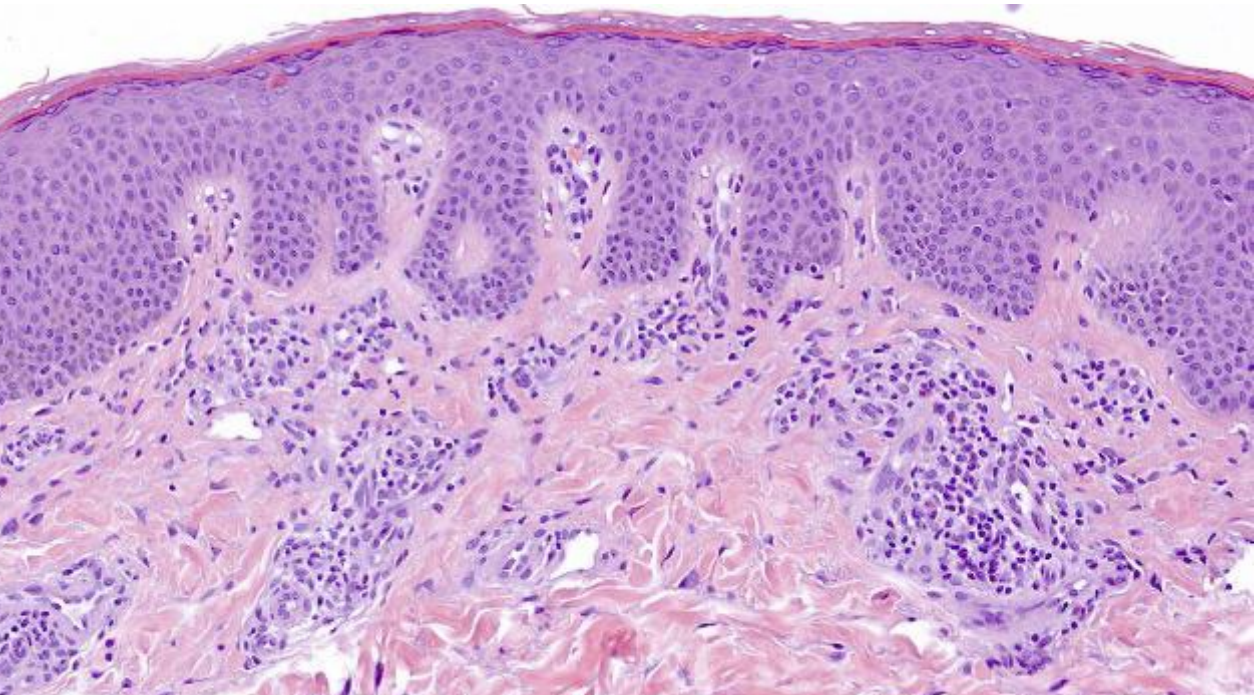
Histology:

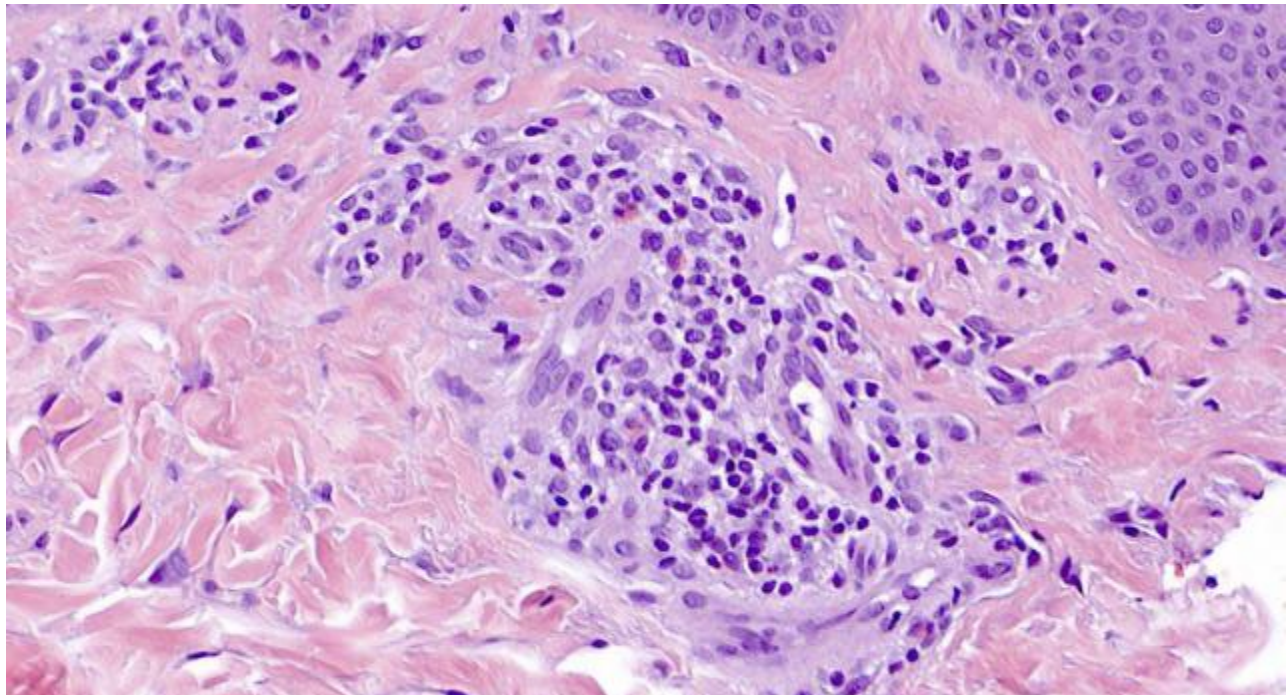






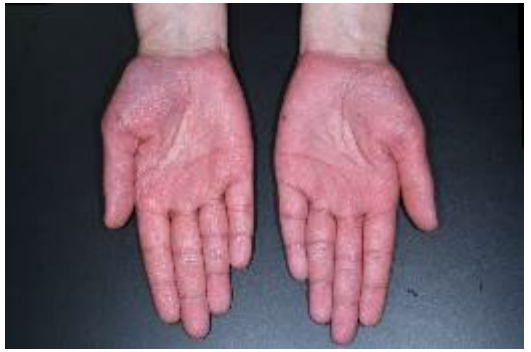


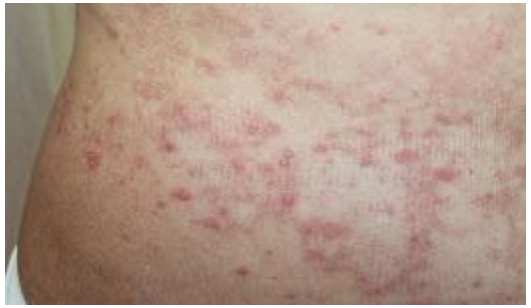




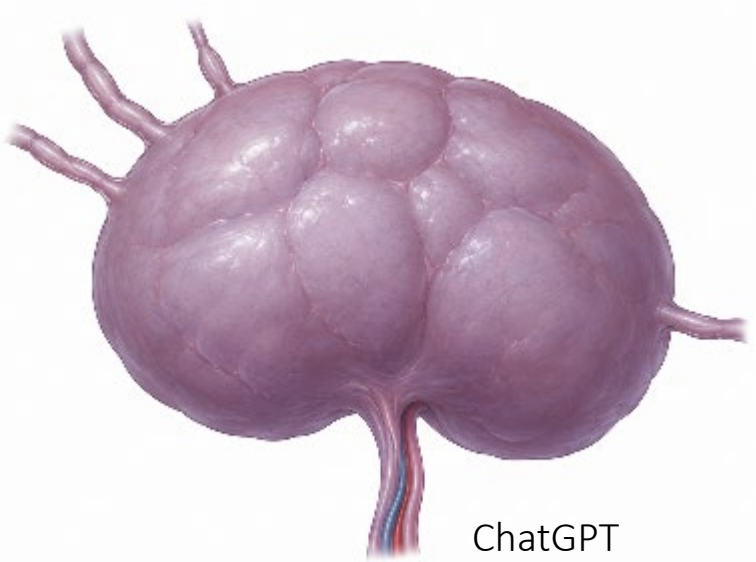
Feedback from the clinician:

Call from the clinician:
Please recheck the histology





Progression of skin lesion:
Sharply demarked palmar erythema
Enlarged lymph nodes cervical



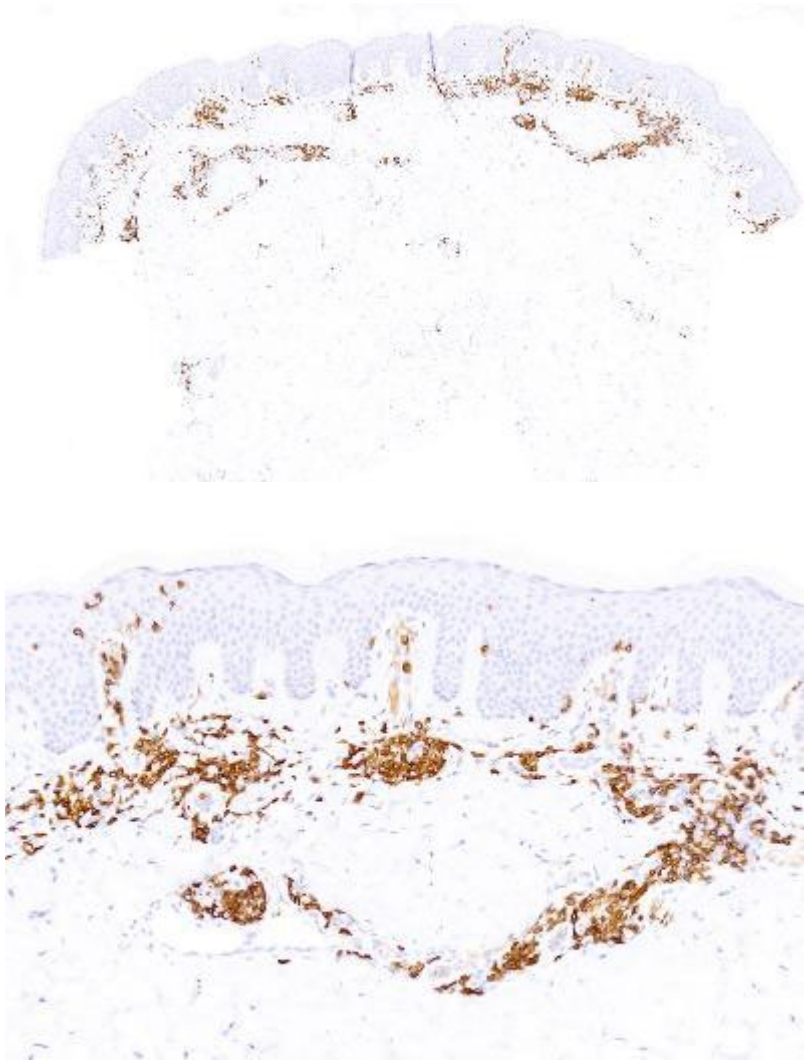
ChatGPT

Ultrasound:
Cervical LN: lymphoma suspected
CT scan: only peripheral LN enlarged

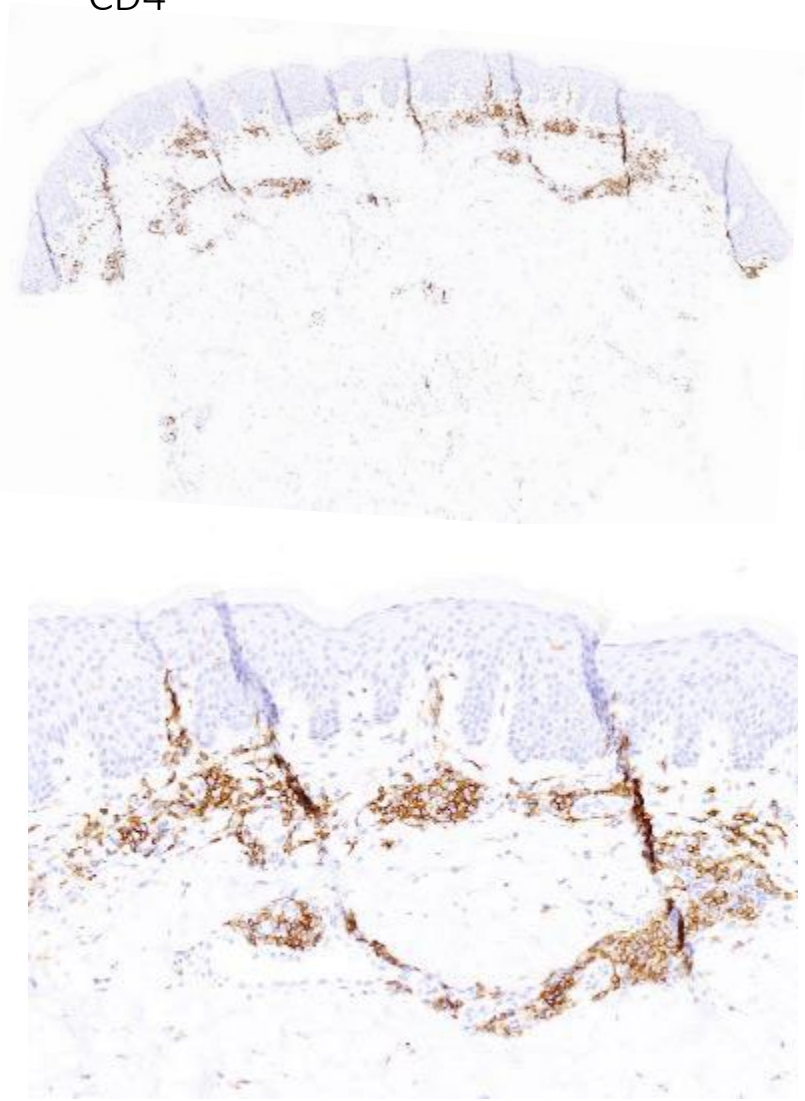
FACS:
no neoplastic CD4+CD7- T-cell-population

Immunohistochemistry:

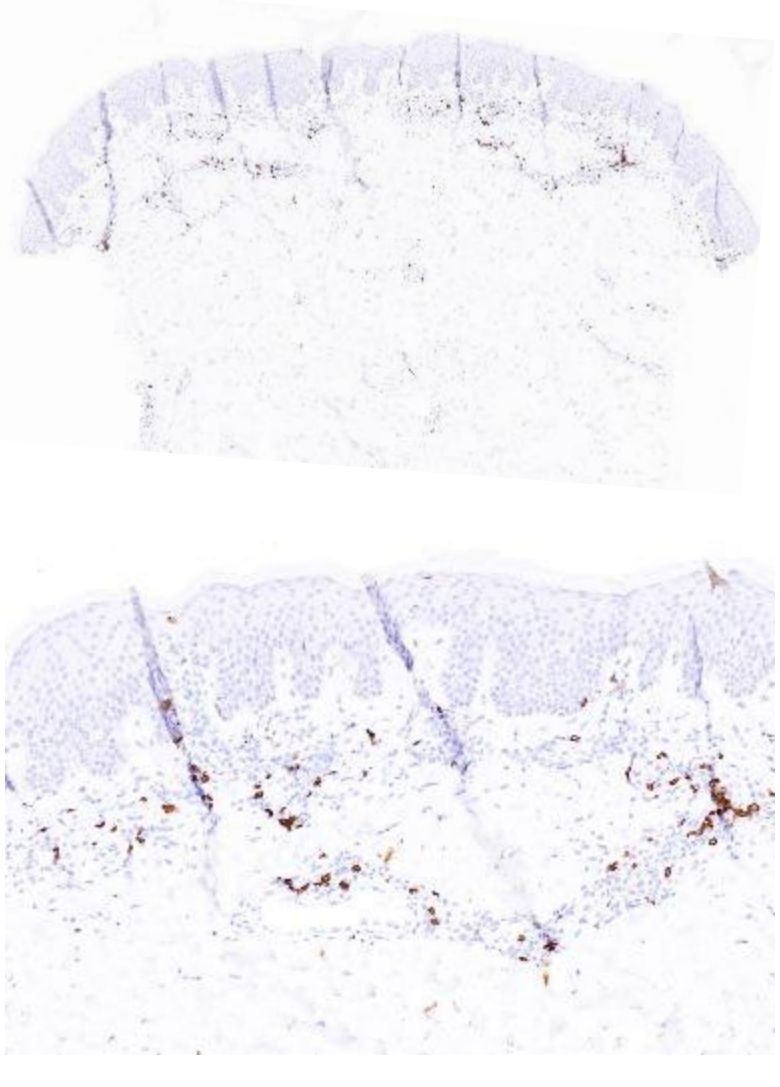
CD3



CD4

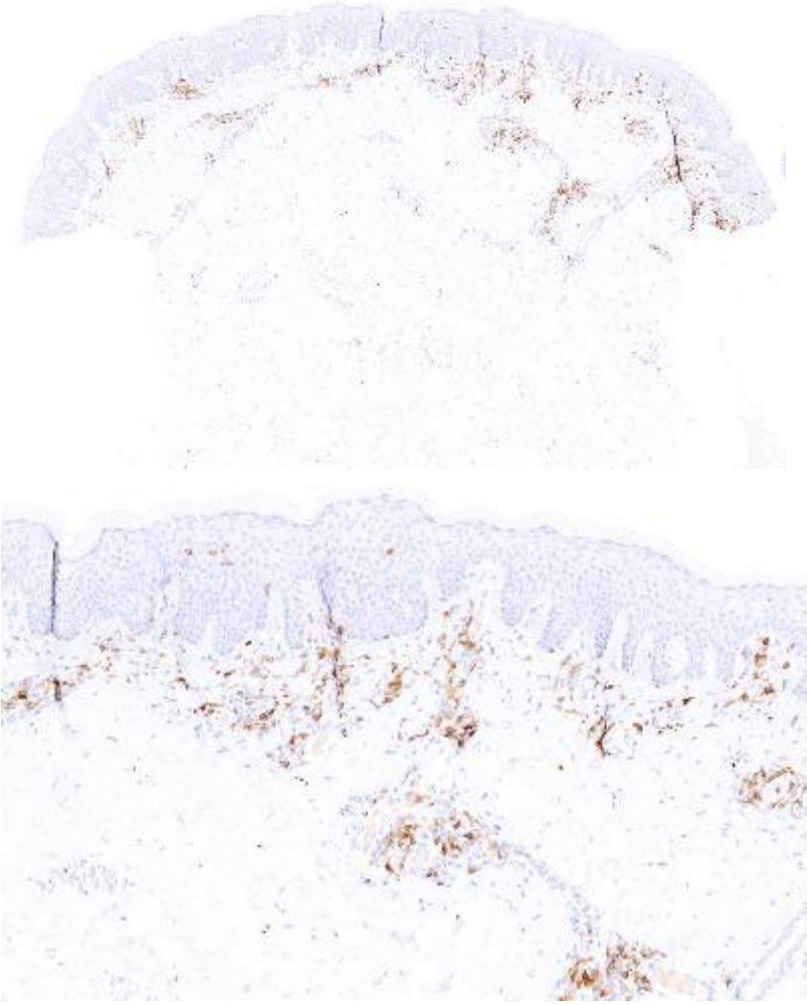


CD8

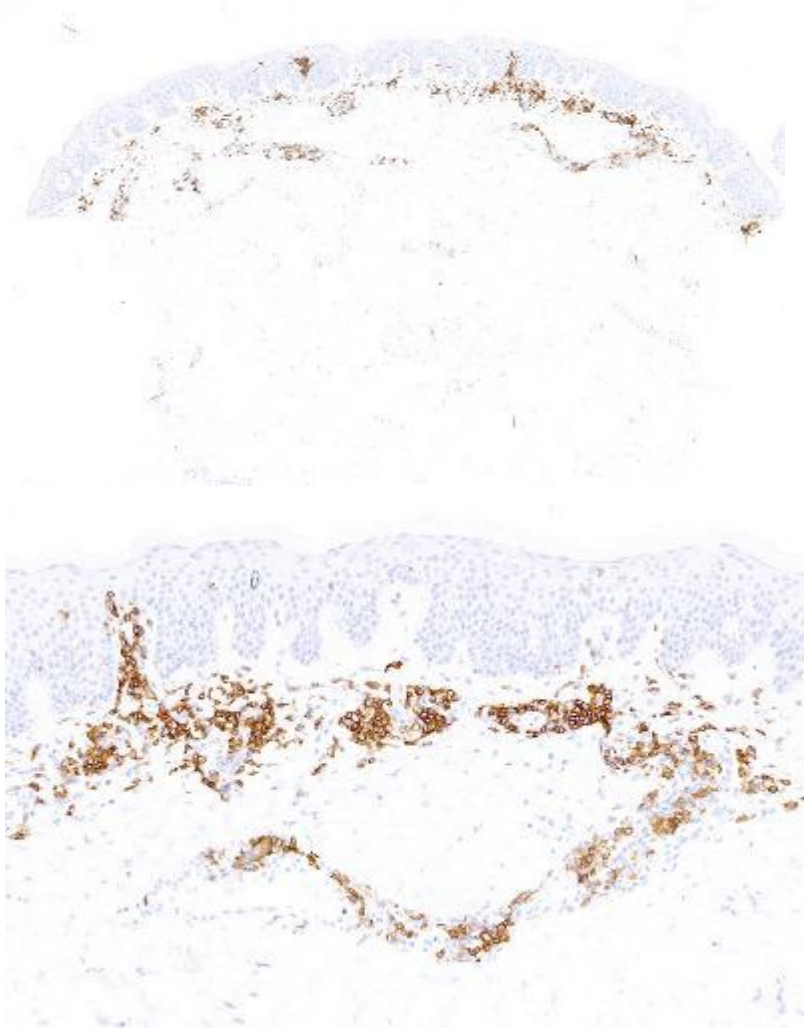


Immunohistochemistry:

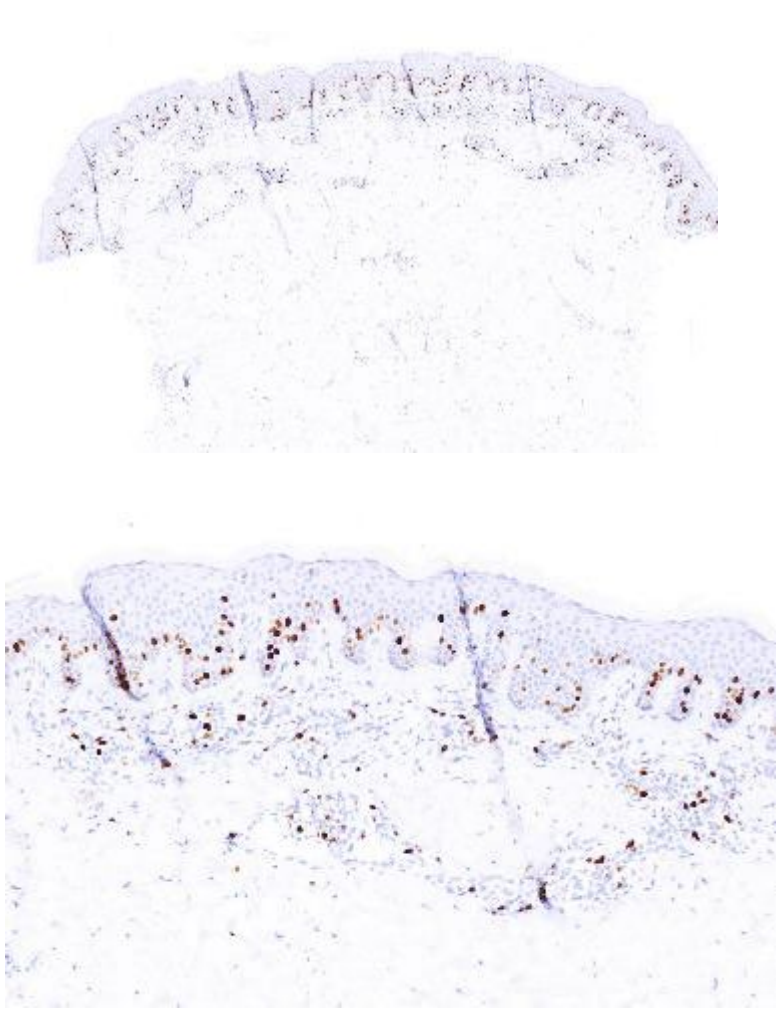
CD7



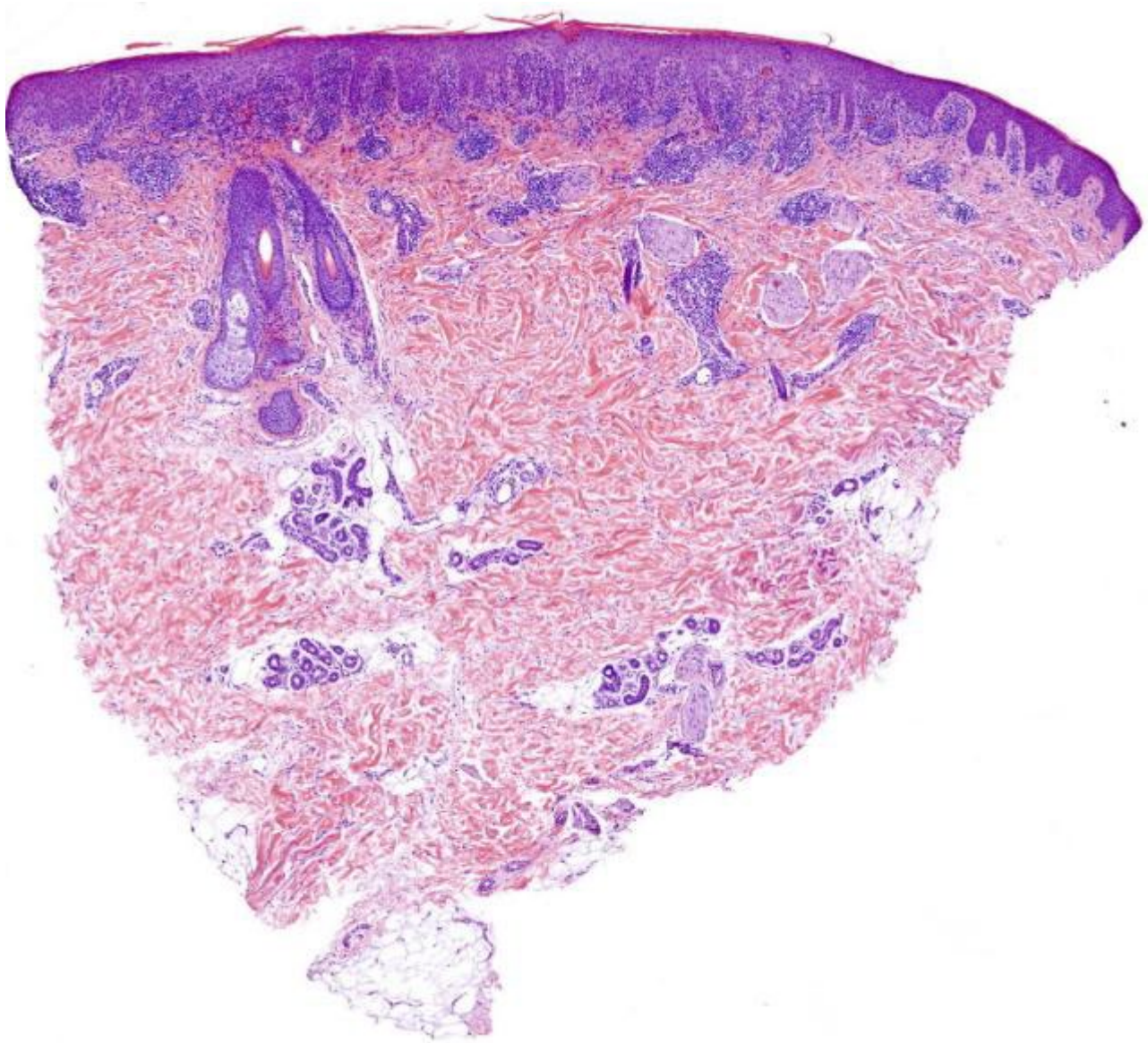
PD1

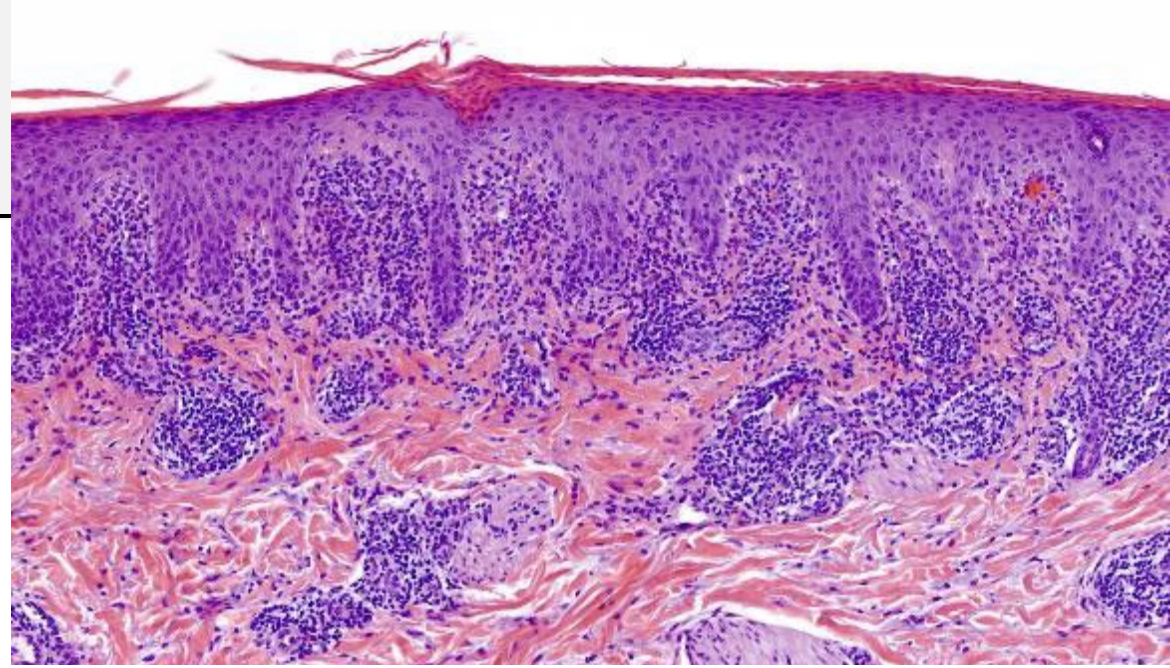


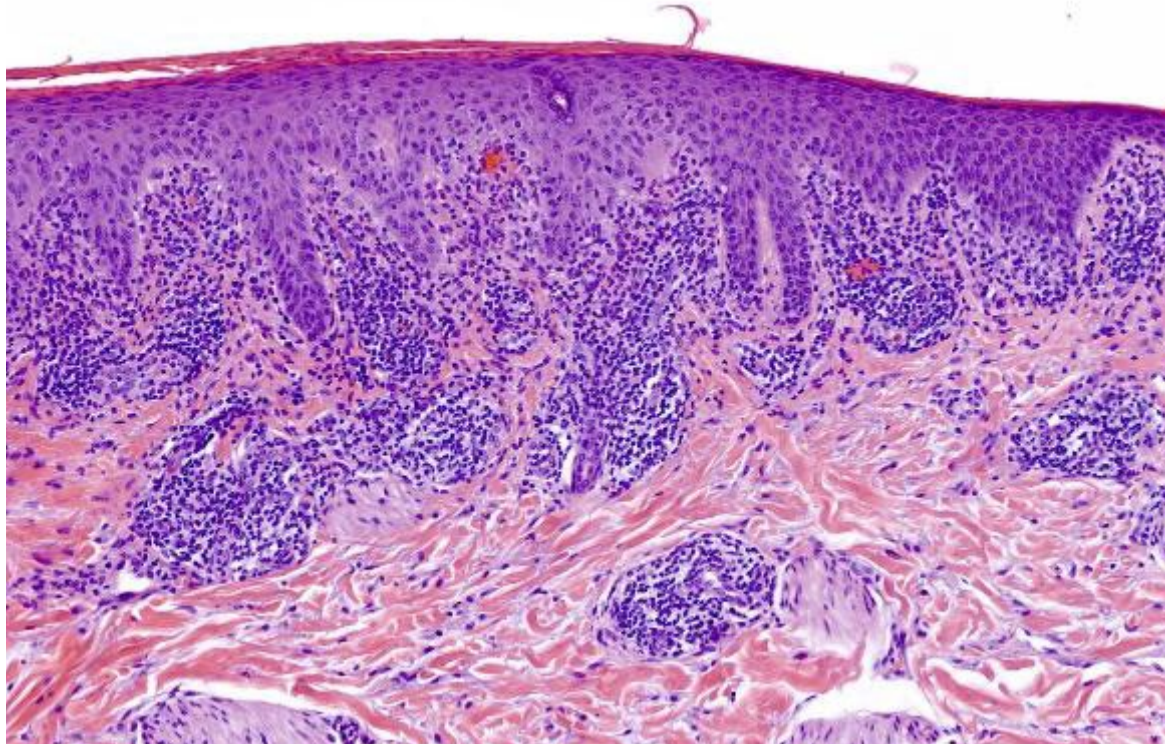
Ki67

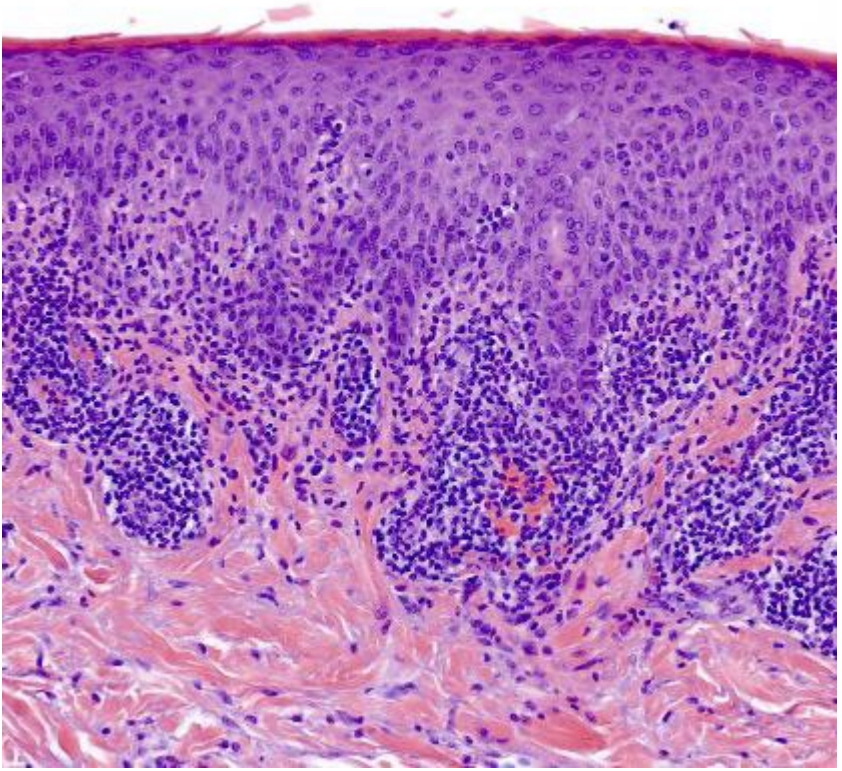


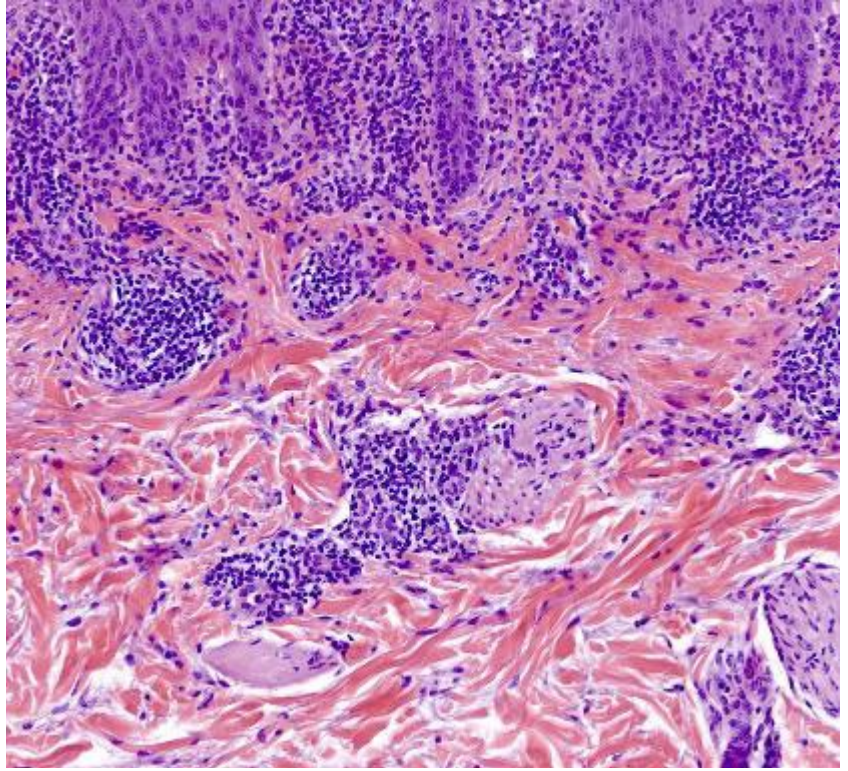
Histology II:



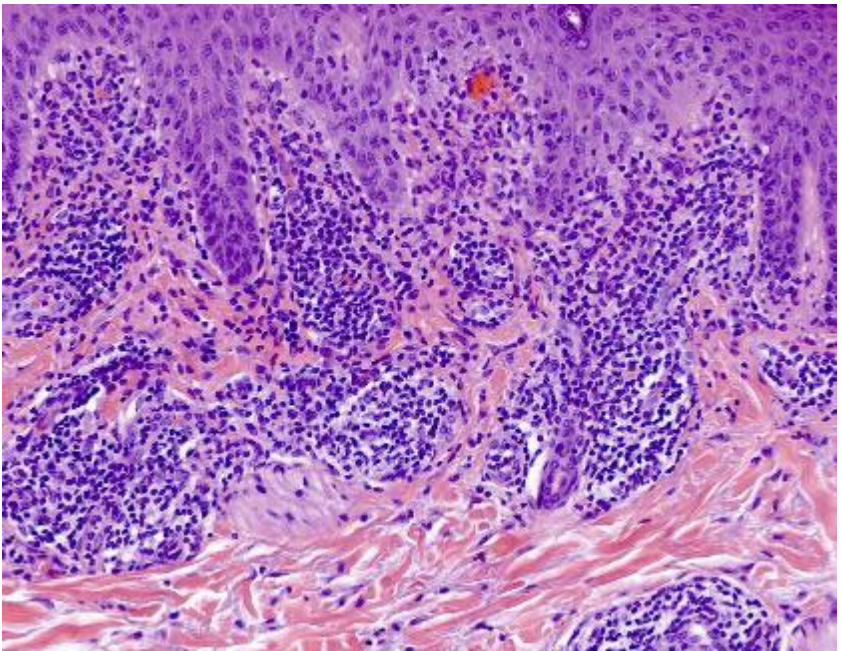


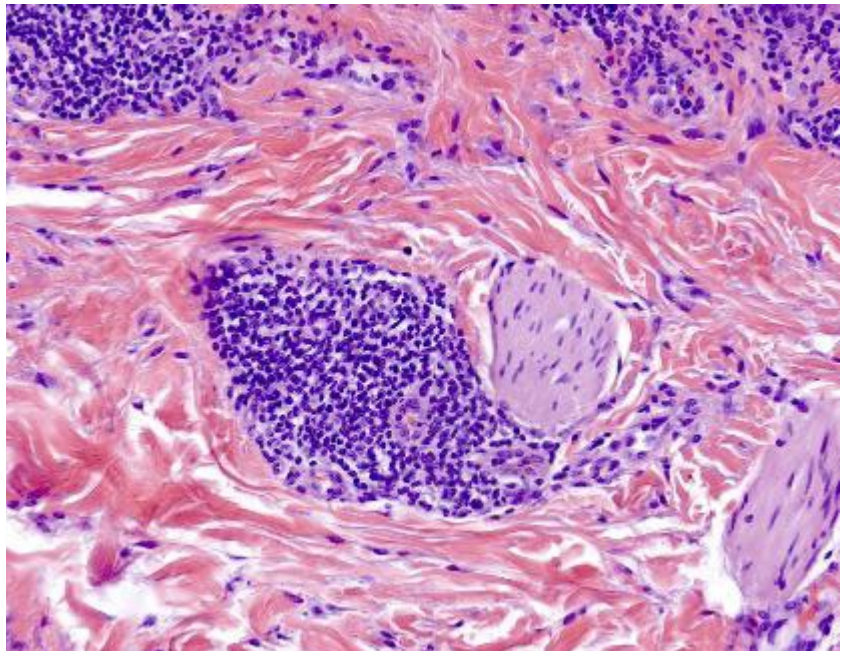


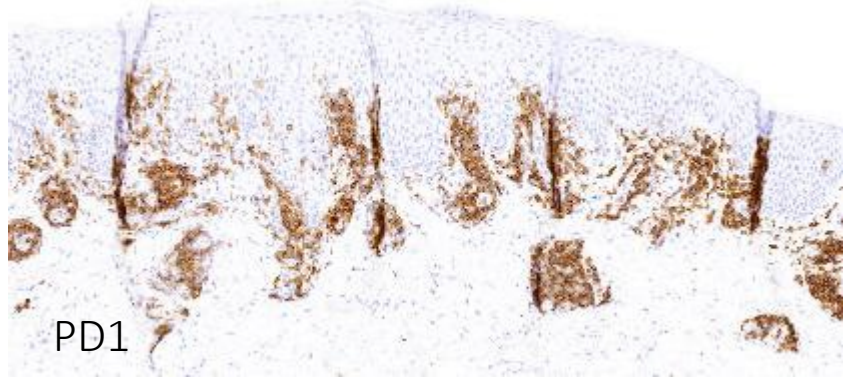


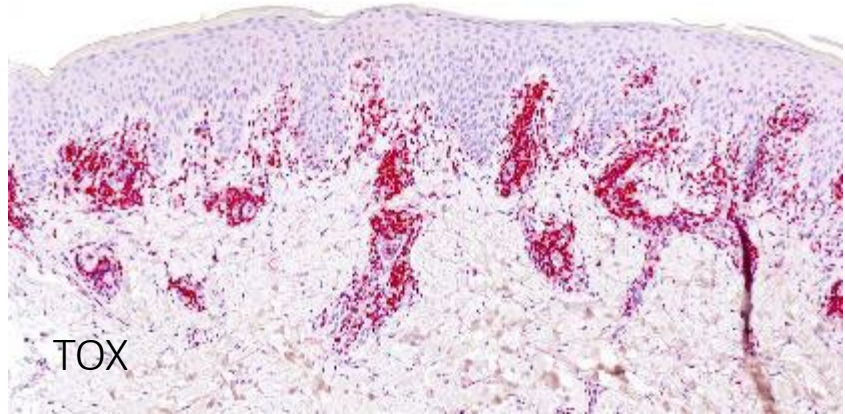



CD7

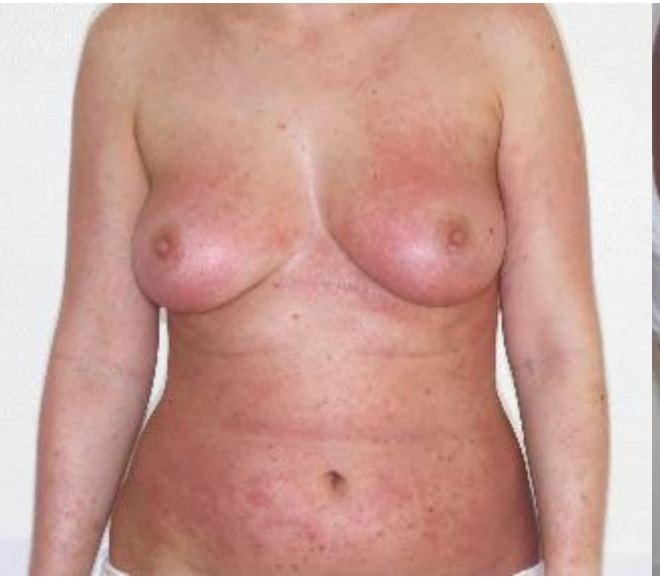


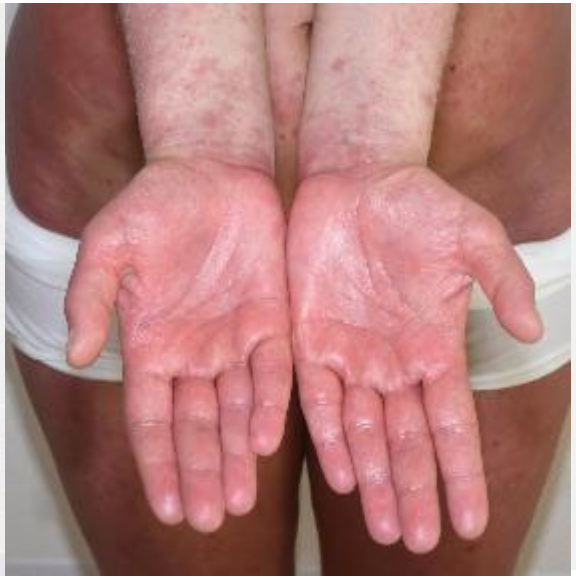



PD1


TOX

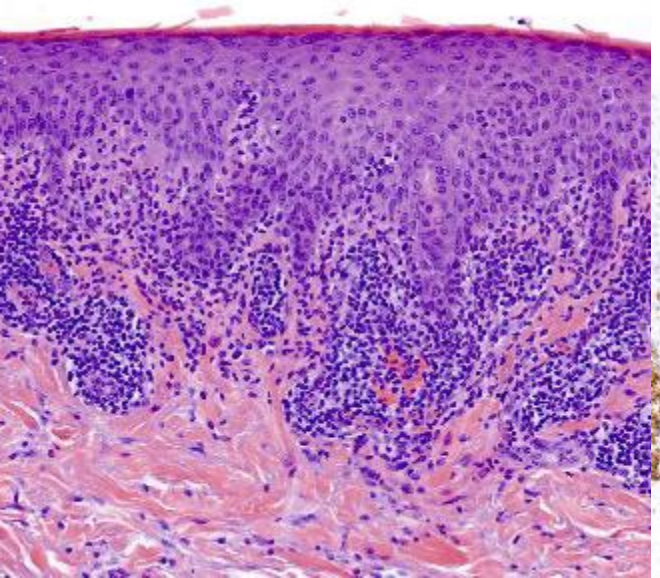
Summary of the findings:

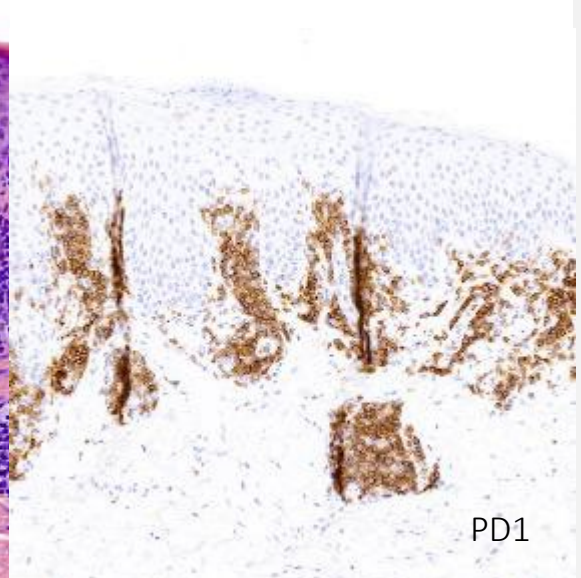




Clinical presentation:

- Confluent scaly maculae
- Palmar erythema
- Mild pruritus





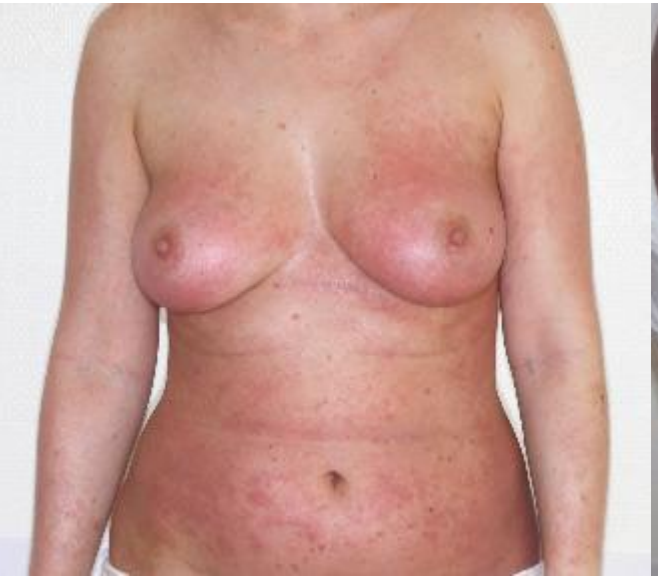
Histology:

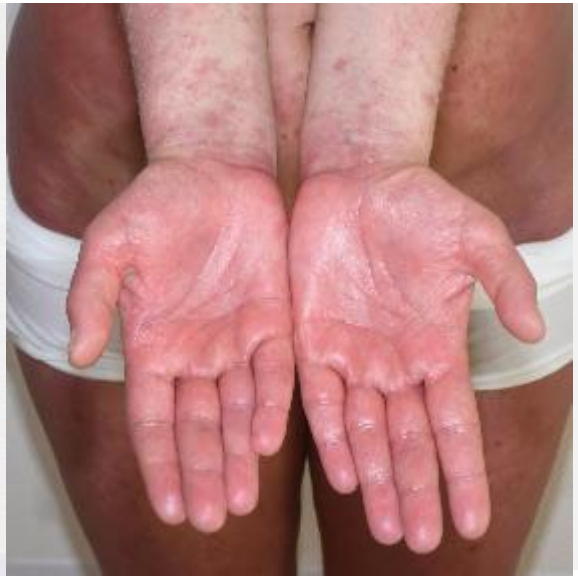
- Superficial perivascular/ band like lymphocytic infiltrate, few eosinophils
- Sparse epidermotropism
- No significant atypia
- CD3+, CD4+, CD8-, CD7 +/-, PD1+, Ki67 low

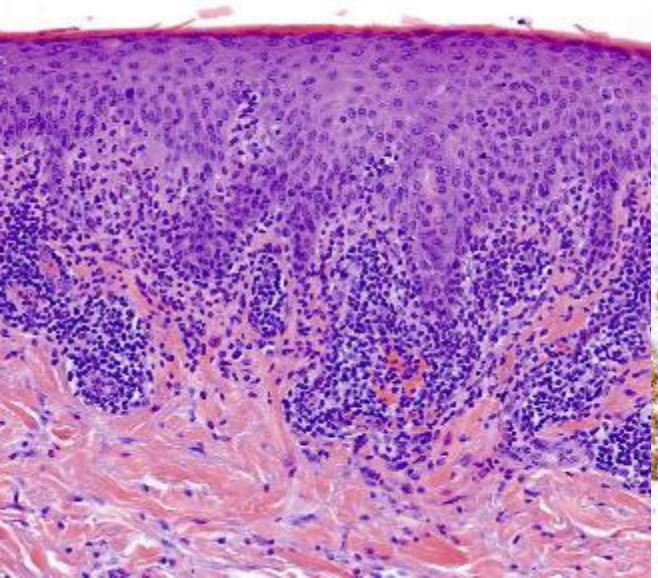
diagnosis

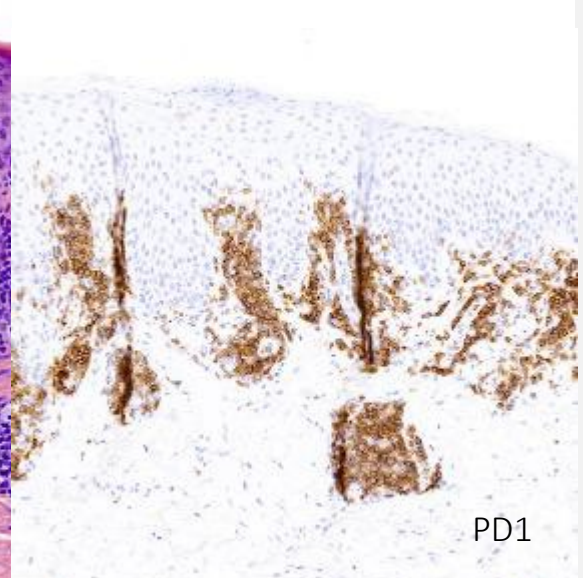
?

Summary of the findings:









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Sézary syndrome

without erythroderma at initial presentation

Essential diagnostic criteria WHO 5th ed.:

1) Erythroderma > 80 % of body surface

2) Detection of blood involvement by a neoplastic T-cell population (TCR clonal)

3) Absolute Sézary count ≥ 1000/µL or

expanding $CD4^+$ T-cell population with a CD4:CD8-ratio > 10 or

expanding $CD4^+$ T-cell population with an abnormal phenotype of

$CD4^+/CD7^-$ T-cell ≥ 40 % or $CD4^+/CD26^-$ T-cells ≥ 30 %

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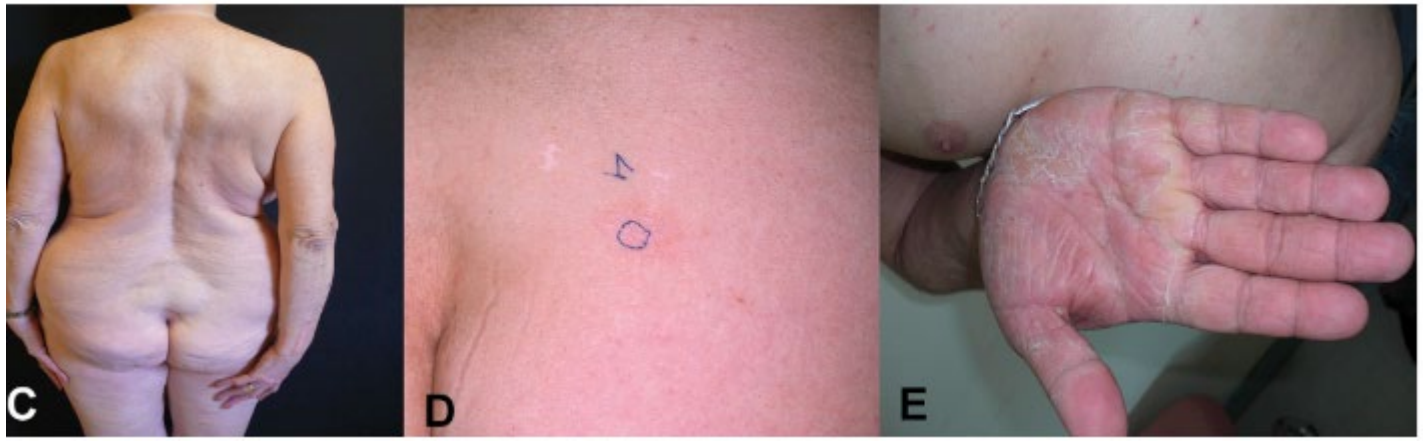
Sézary syndrome without erythroderma?



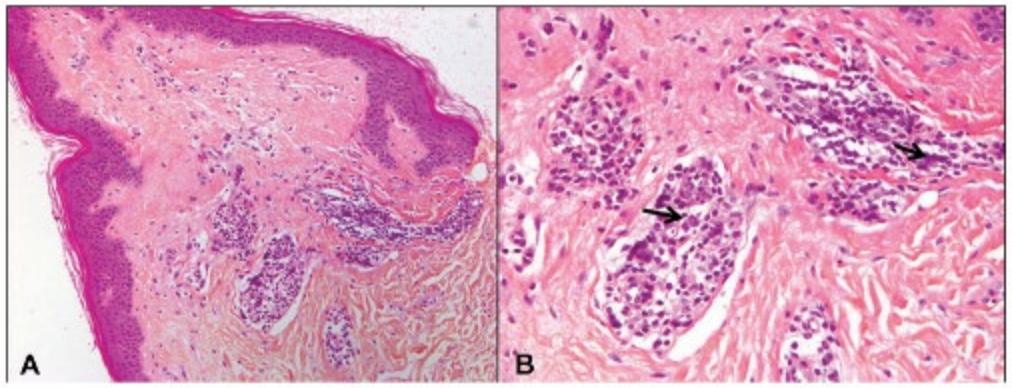


Fig 2. Histopathologic characteristics of Sézary syndrome without erythroderma (patient 3). **A**, Intradermal perivascular lymphoid infiltrate without epidermotropism. **B**, Perivascular atypical cells (*arrows*). (**A** and **B**, Hematoxylin-eosin stain; original magnifications: **A**, ×100; **B**, ×200.)

Table I. Clinical features of Sézary syndrome without erythroderma at diagnosis

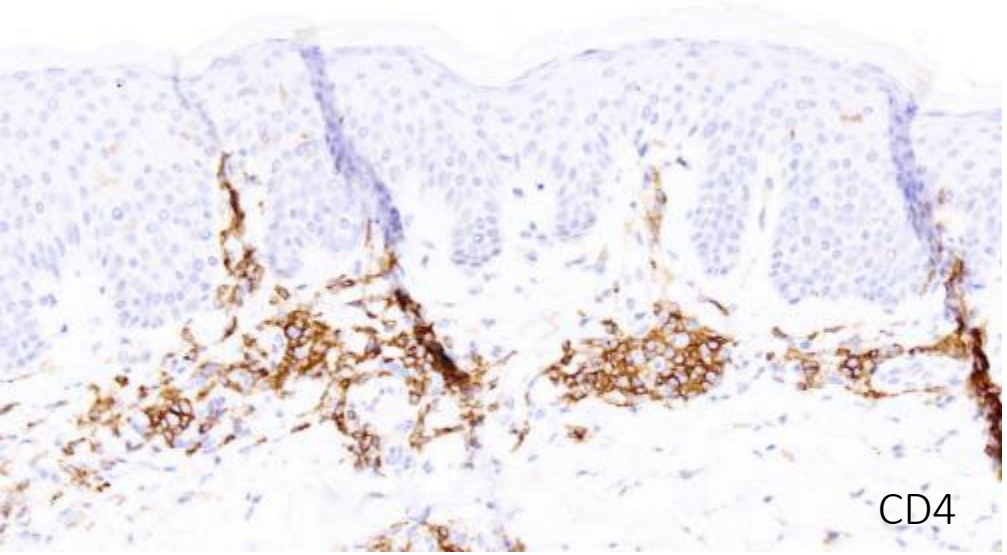
Patient	Gender	Age at diagnosis, y	Pruritus	Pruritus duration before diagnosis, y	Specific CTCL lesions	Peripheral/central lymph nodes
1	F	82	Yes	6	None	–/–
2	F	73	Yes	8	None	–/–
3	F	93	Yes	0.5	None	–/–
4	F	76	Yes	4	None	–/–
5	F	67	Yes	2	Patch (T1*)	–/–
6	M	47	Yes	1	Infiltrated skin, PPK	+†/–

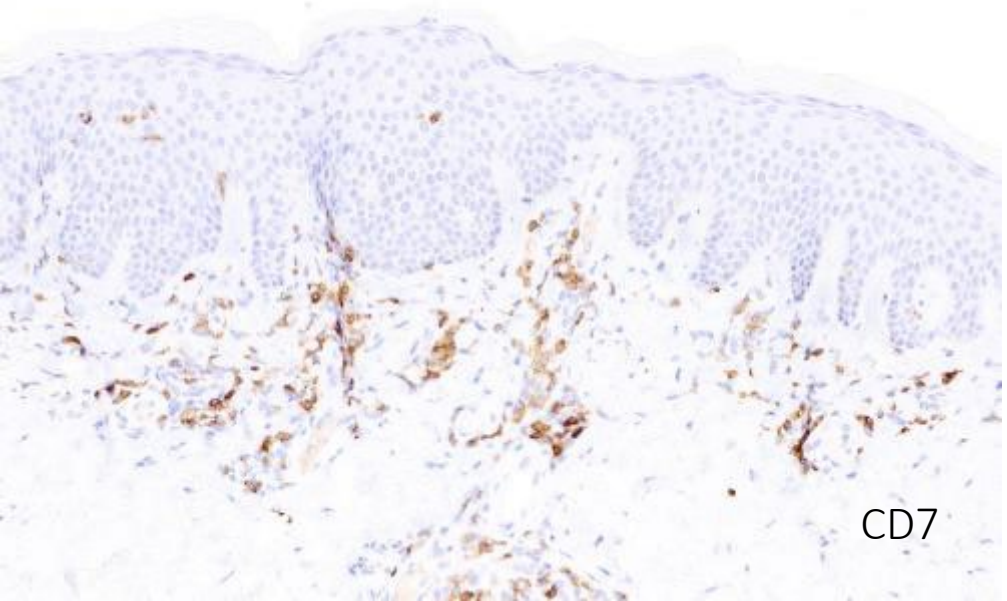
CTCL, Cutaneous T-cell lymphoma; *F*, female; *M*, male; *PPK*, palmoplantar keratoderma.
*Patches involving <10% of total body surface area.
†Axillary: 1.5 cm.

Biopsies from non-erythrodermic skin showed a superficial dermal infiltrate, mild atypia, typical immunophenotype (PD1+)

Henn A. et al. Sézary syndrome without erythroderma. J Am Acad 2015; 72:1003-9.

Sézary syndrome: CD7 loss > 50%





CLINICAL AND LABORATORY INVESTIGATIONS

BJD
British Journal of Dermatology

Histopathological and immunophenotypical criteria for the diagnosis of Sézary syndrome in differentiation from other erythrodermic skin diseases: a European Organisation for Research and Treatment of Cancer (EORTC) Cutaneous Lymphoma Task Force Study of 97 cases

C.D. Klemke,[1] N. Booken,[2] C. Weiss,[2] J.P. Nicolay,[1] S. Goerdt,[1] M. Felcht,[1] C. Géraud,[1] W. Kempf,[3] C. Assaf,[4] N. Ortonne,[5] M. Battistella,[6] M. Bagot,[7] R. Knobler,[8] P. Quaglino,[9] B. Arheiliger,[10] M. Santucci,[11] P. Jansen,[12] M.H. Vermeer[12] and R. Willemze[12]

- 65% of Sézary syndrome samples showed a loss in contrast to only 6% in erythrodermic inflammatory dermatoses ($p < 0.001$)
- Strongly supports the diagnosis of SS
- Look for the neoplastic population (e.g. CD4+)

Sézary syndrome: Ki67 high

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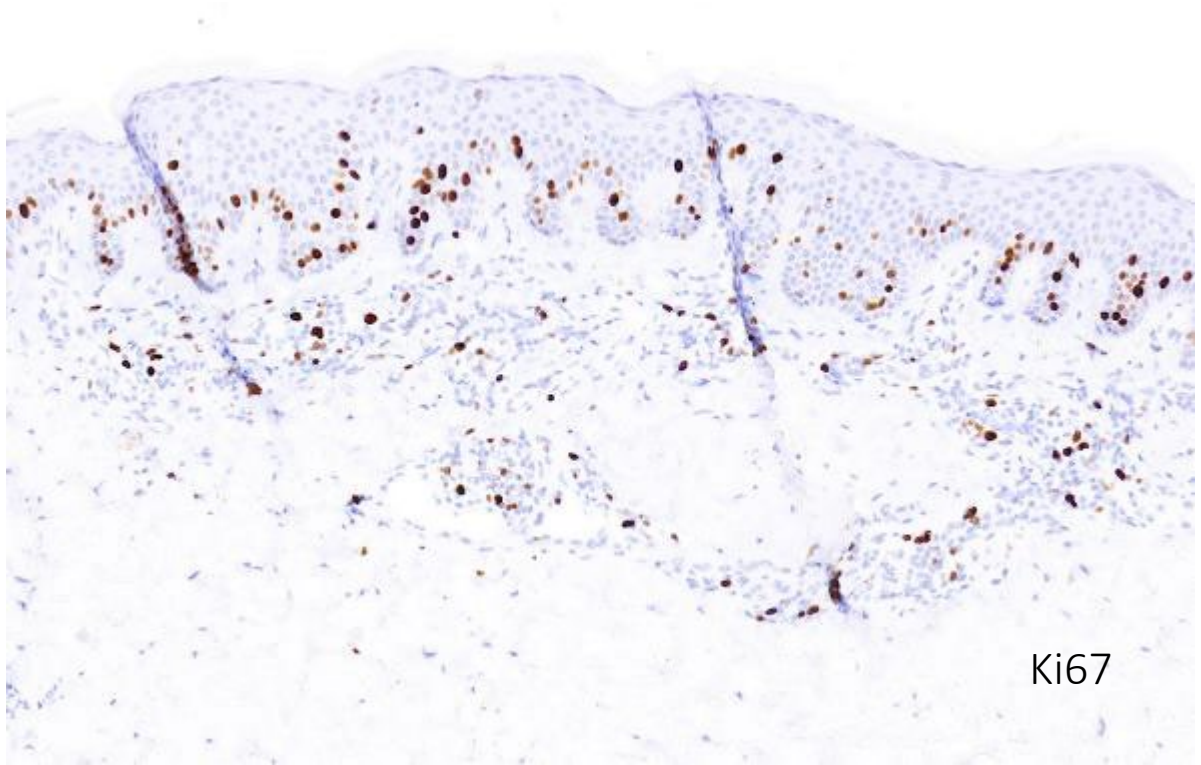

Ki67

	SS	EID	Test	P-value
Number of patients	57	40		
Ki-67			Trend	< 0·001
Negative	–	1		
< 5%	6	13		
5–10%	17	11		
11–25%	9	6		
26–50%	4	–		
> 50%	4	–		
Not determined	17	9		

- Higher Ki67 supports the diagnosis of Sézary syndrome ($p < 0.001$)
- Especially helpful if the proliferative rate is high

Sézary syndrome: PD1 expression > 50%

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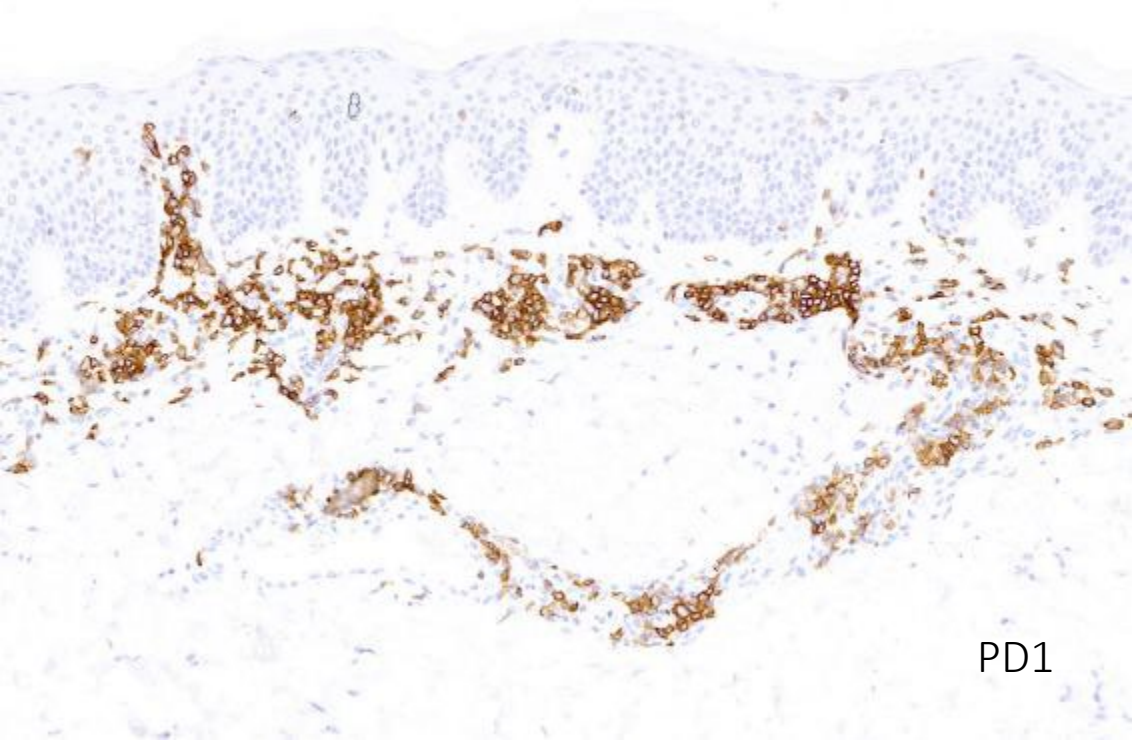
	SS	EID	Test	P-value
Number of patients	57	40		
PD-1 expression > 50%	41/55 (75)	17/37 (46)	χ^2	0·0053

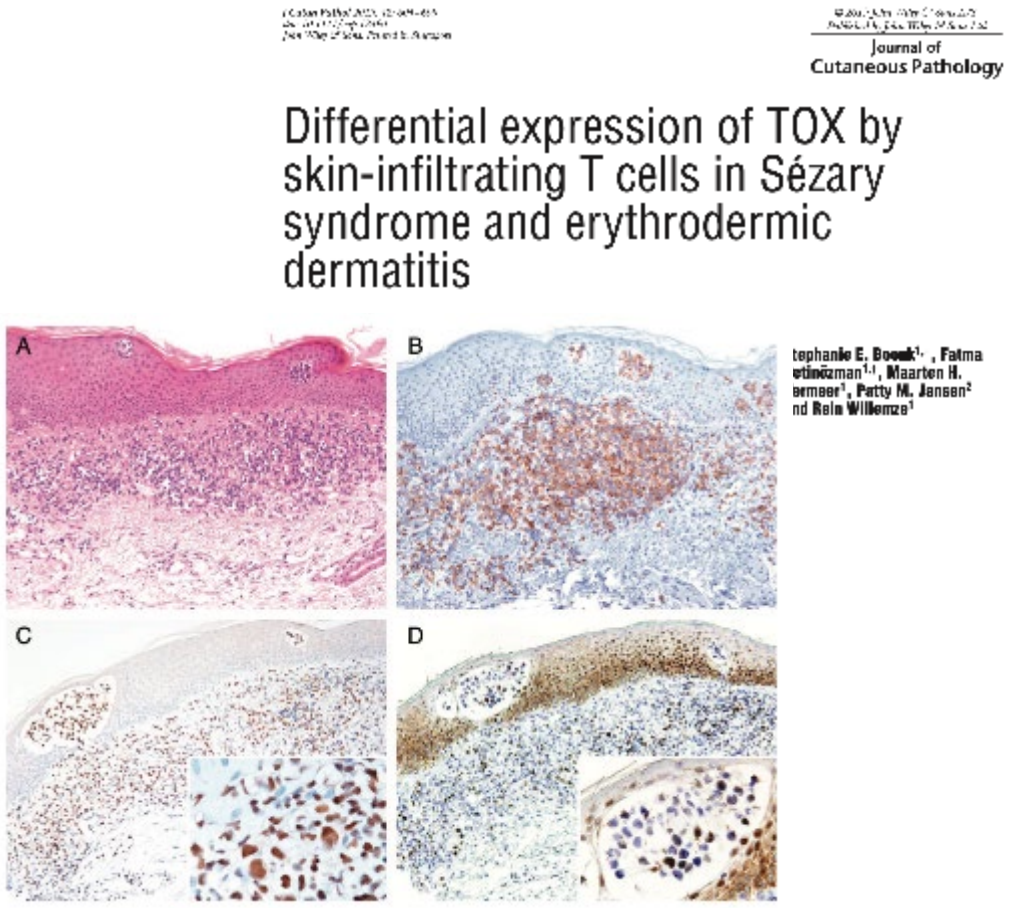


PD1

- 75% of Sézary syndrome samples showed a PD1 expression, but also 46% of the erythrodermic inflammatory dermatosis were PD1+ ($p < 0.0053$)
- PD1 expression supports the diagnosis of SS

Why TOX?

= Thymocyte selection-associated high mobility group box factor



Journal of Cutaneous Pathology

Differential expression of TOX by skin-infiltrating T cells in Sézary syndrome and erythrodermic dermatitis

tephanie E. Boonk[1], Fatma etinözman[1,†], Maarten H. ermeer[1], Patty M. Jansen[2] nd Rein Willemze[1]

Table 1. Summary of the immunohistochemical stainings

	Diagnosis	TOX (%)	C-MYC (%)	PD-1 (%)	Antigen loss
1	SS	++++	+	++++	–
2	SS	++++	+	++++	CD7, CD2
3	SS	++++	+	++++	CD7
4	SS	+++	–	++++	CD7
5	SS	++++	+	+++	CD7
6	SS*	++ (w)	–	+++	–
7	SS	++++	++	+++	CD7
8	SS	+++	–	+++	–
9	SS	++++	++	++++	CD7
10	SS	++++	+	++++	CD7
11	SS	++++	+	+	–
12	SS	++++	+	+++	CD7, CD2
13	SS	+++	–	++++	CD7
14	SS	++++	+	++++	CD7
15	SS*	++ (w)	–	+++	CD7
16	AE†	+ (w)	–	+	–
17	AE	+ (w)	–	++	–
18	AE†	+ (w)	–	+++‡	–
19	AE†	++ (w)	–	+++‡	CD7
20	PSOR	+ (w)	+	++	–
21	PSOR	+ (w)	–	+	–
22	PSOR	+ (w)	–	–	–
23	IE	+ (w)	+	–	–
24	IE	++ (w)	–	–	–
25	IE	++ (w)	+	–	–
26	IE	+ (w)	+	–	–
27	IE	++ (w)	–	–	–
28	IE	+ (w)	+	+	–
29	IE	+ (w)	+	+	CD7
30	IE	+ (w)	+	+	–
31	IE	+ (w)	–	+	–
32	PARA	+ (w)	+	+	–

AE, atopic erythroderma; IE, idiopathic erythroderma; PARA, paraneoplastic erythroderma; PD-1, programmed death-1; PSOR, psoriatic erythroderma; SS, Sézary syndrome; TOX, C-MYC and PD-1: –, <10%; +, 11–25%; ++, 26–50%; +++, 51–75%; ++++, >75%. Antigen loss: loss of CD2, CD3, CD4, CD5 or CD7 expression by more than 50% of the (neoplastic) T cells; –: no loss.

*SS cases showing no or minimal cellular atypia.

†Cases showing considerable numbers of small to medium-sized atypical T cells.

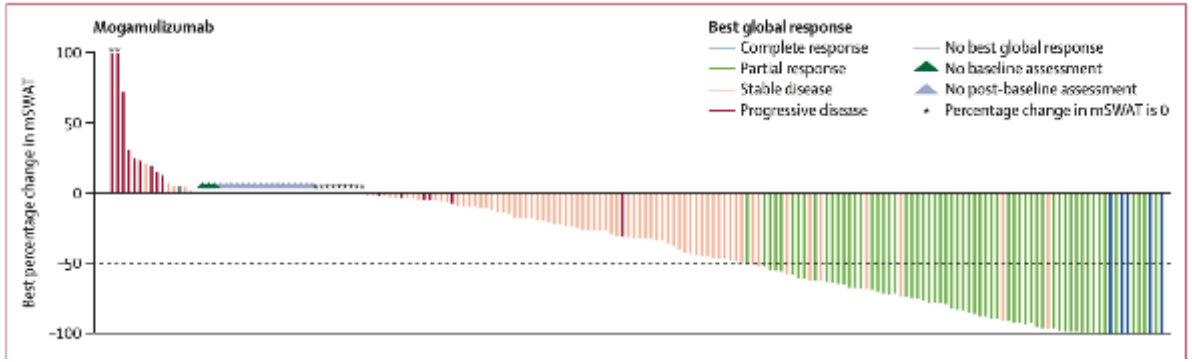
‡CD8$^-$ T cells; (w): weak staining.

- Strong TOX staining >50% of the T-cells in 87% of Sézary syndrome
- In erythrodermic dermatitis only weak staining in 11-50% of the T-cells was observed

Mogamulizumab = POTELIGEO®

PFS: Mogamulizumab 7,7 vs. Vorinostat 3,1 months

Approved for second-line treatment in adult patients



Planned procedure in our patient:

1. ECP and MTX 15mg /week, if no response switch to Mogamulizumab

Pre-treatment assessment:

1. Beta HCG+
2. Ultrasound confirmed twin pregnancy
3. no MTX, no Moga
4. Embryotox center and ethic board discussion

	Mogamulizumab (n=186)	Vorinostat (n=186)
Proportion of patients with an overall response by global assessment*†	52/186 (28%)	9/186 (5%)
Overall responses in patient subgroups		
Mycosis fungoides	22/105 (21%)	7/99 (7%)
Sézary syndrome	30/81 (37%)	2/87 (2%)
Stage IB or IIA	7/36 (19%)	5/49 (10%)
Stage IIB	5/32 (16%)	1/23 (4%)
Stage III	5/22 (23%)	0/16 (0)
Stage IV	35/96 (36%)	3/98 (3%)
Duration of response, months	14·1 (8·4–19·2)	9·1 (5·6–NE)
Mycosis fungoides	13·1 (4·7–18·0)	9·1 (5·6–NE)
Sézary syndrome	17·3 (9·4–19·9)	6·9 (6·9–6·9)
Compartment response*‡		
Skin	78/186 (42%)	29/186 (16%)
Blood	83/122 (68%)	23/123 (19%)
Lymph nodes	21/124 (17%)	5/122 (4%)
Viscera	0/3 (0%)	0/3 (0%)

Data are n/N (%) or median (IQR). The proportion of patients achieving an overall response is based on the Global Composite Response score. NE=not estimable. *Proportion of patients with an overall response or compartmental response is the percentage of patients with confirmed complete response or confirmed partial response. †p<0·0001. ‡Denominator includes patients with measurable compartmental disease at baseline.

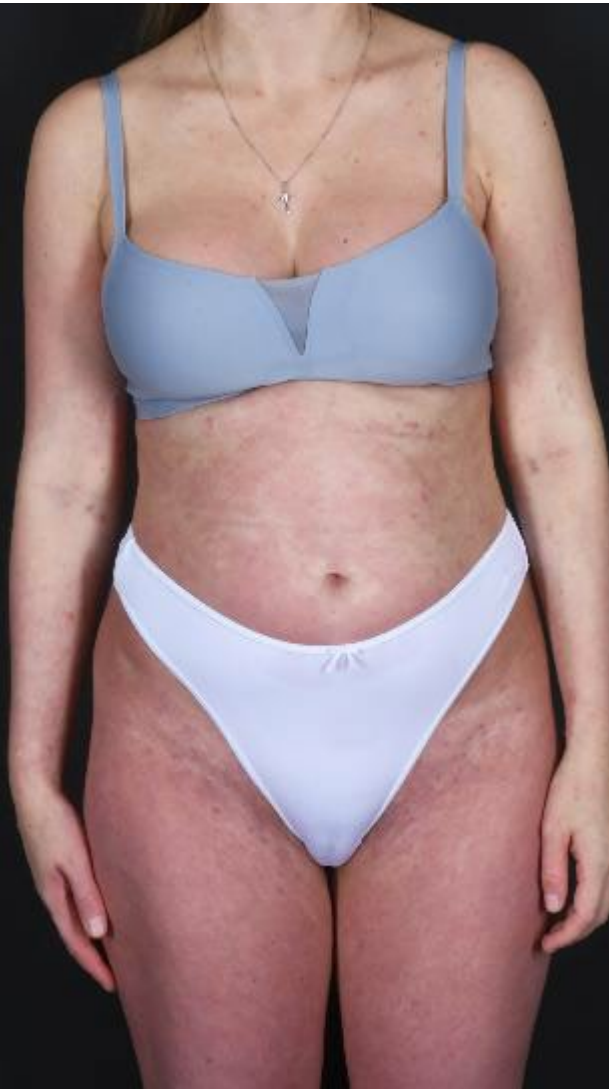
***Table 2:* Measures of response by investigator assessment**

Kim YH et al. Mogamulizumab versus vorinostat in previously treated cutaneous T-cell lymphoma (MAVORIC): an international, open-label, randomised, controlled phase 3 trial. Lancet Oncology 2018

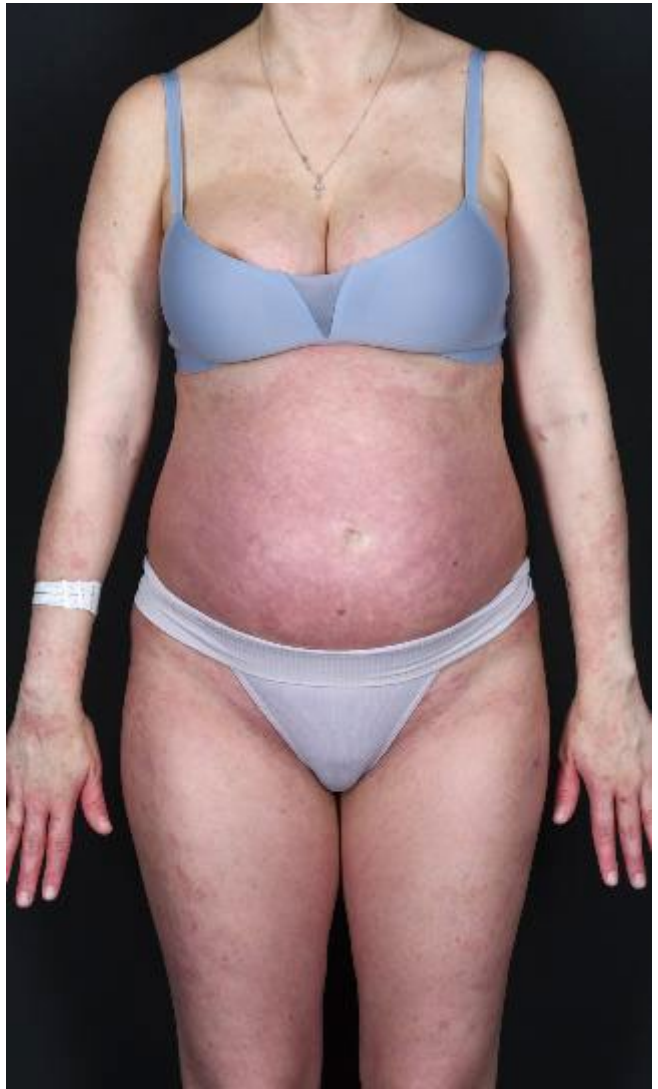
15.08.2025

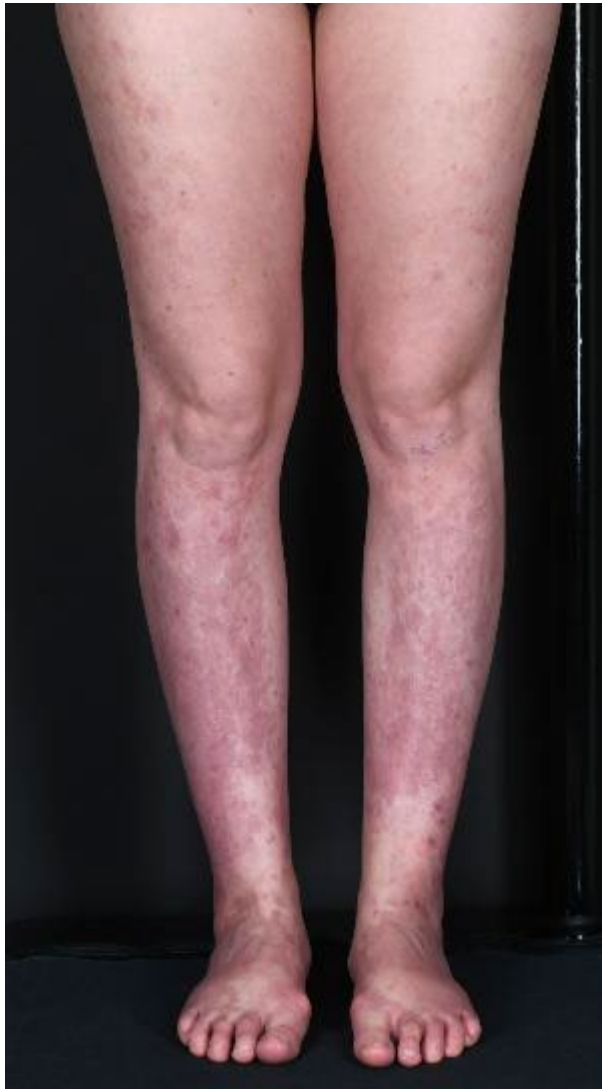
19.09.2025

B1: ECP weekly



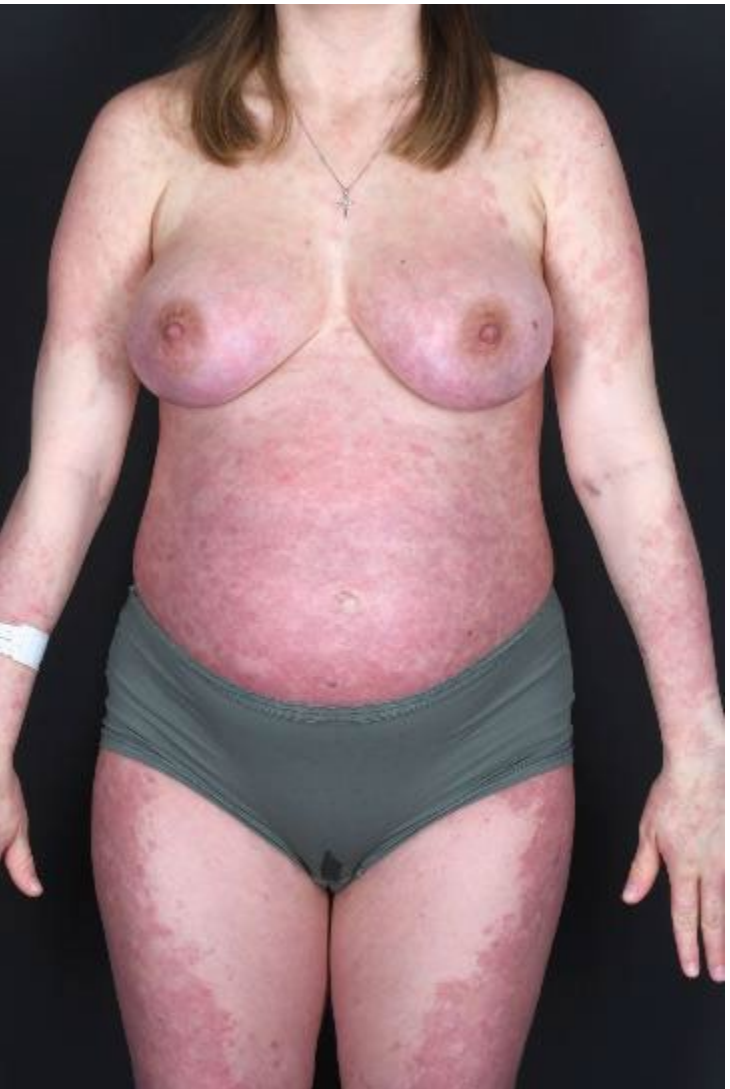
B1: ECP weekly

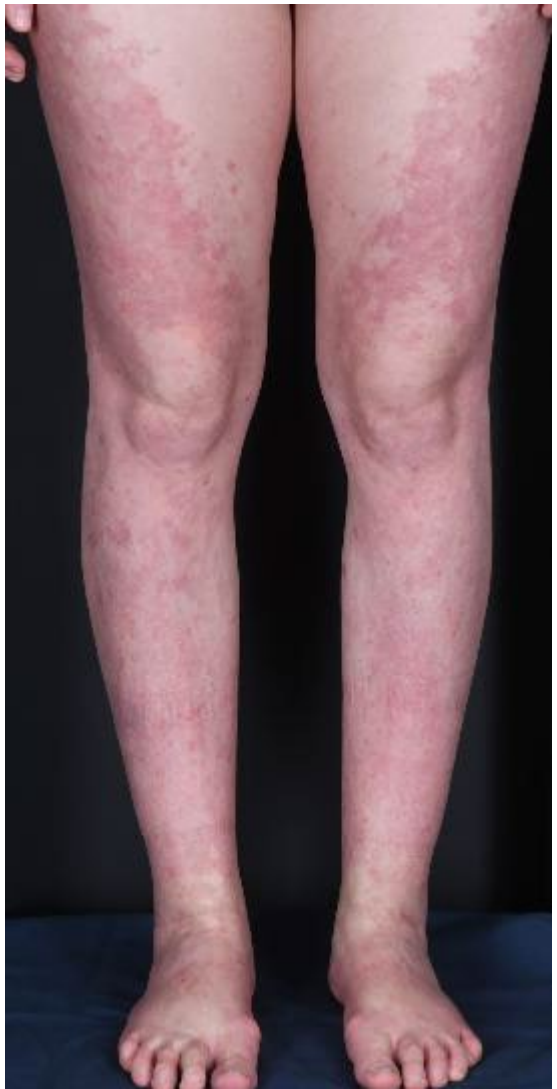




17.10.2025

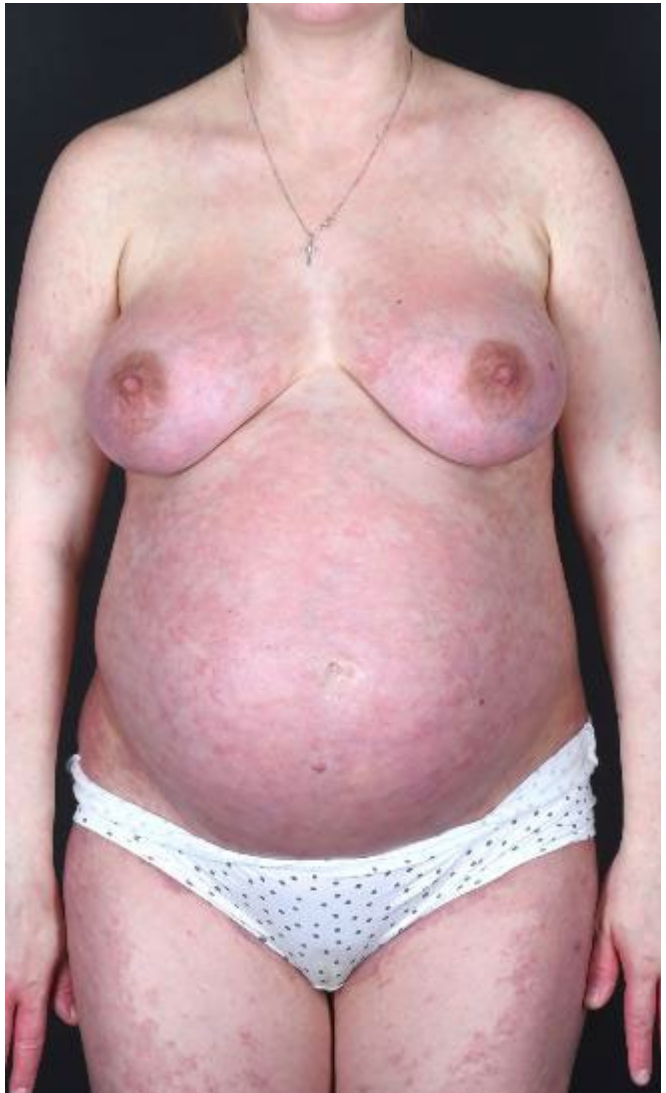
B1: ECP weekly and Peg INF 180 µg

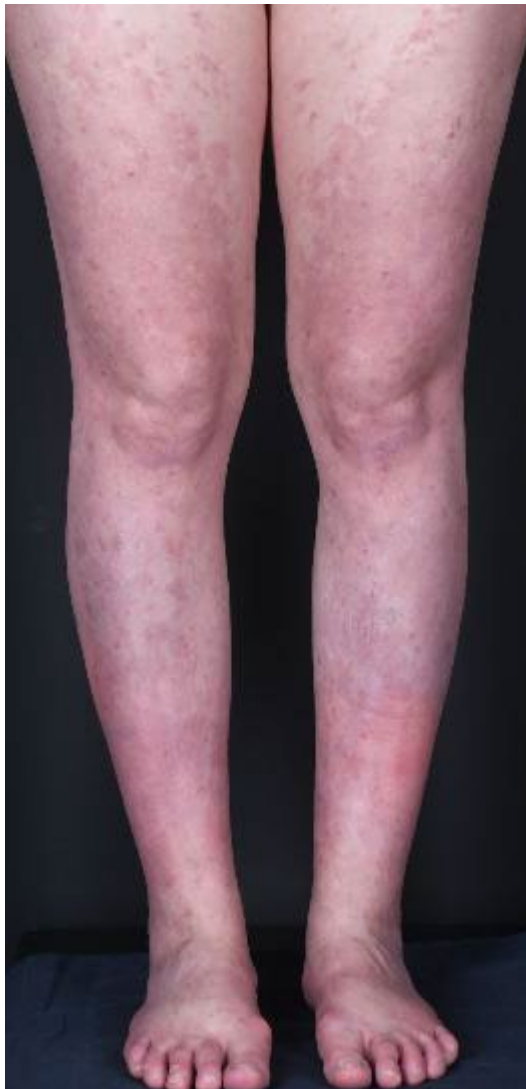




13.11.2025

B1: ECP weekly and Peg INF 180 µg



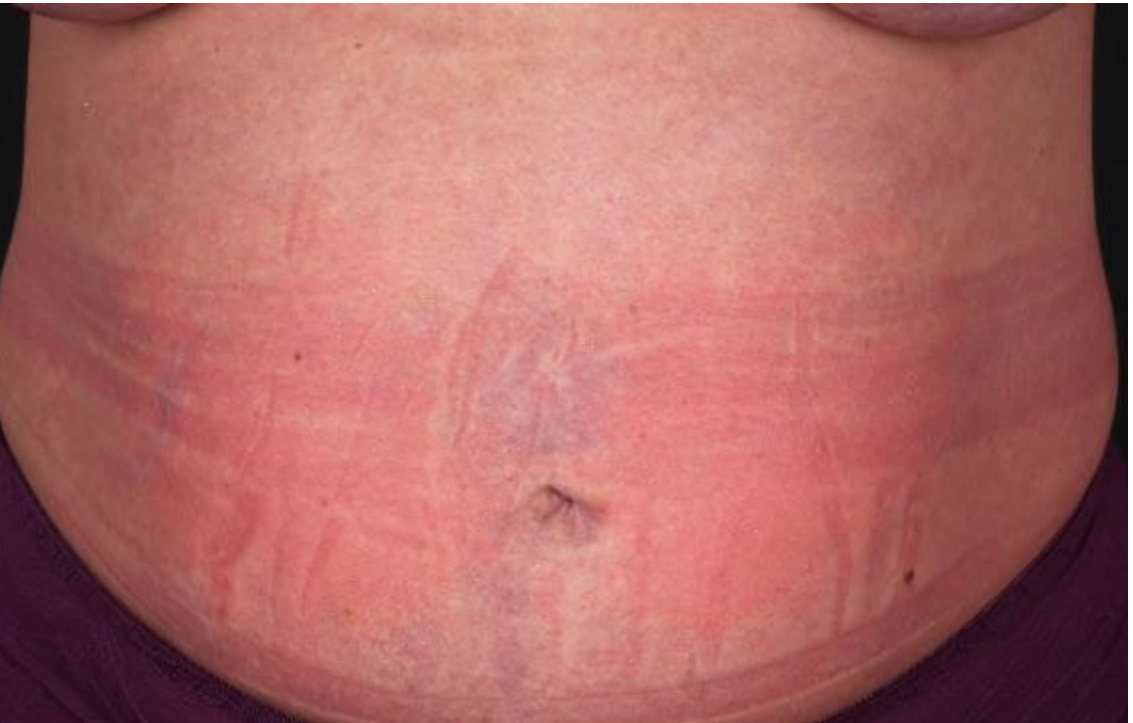


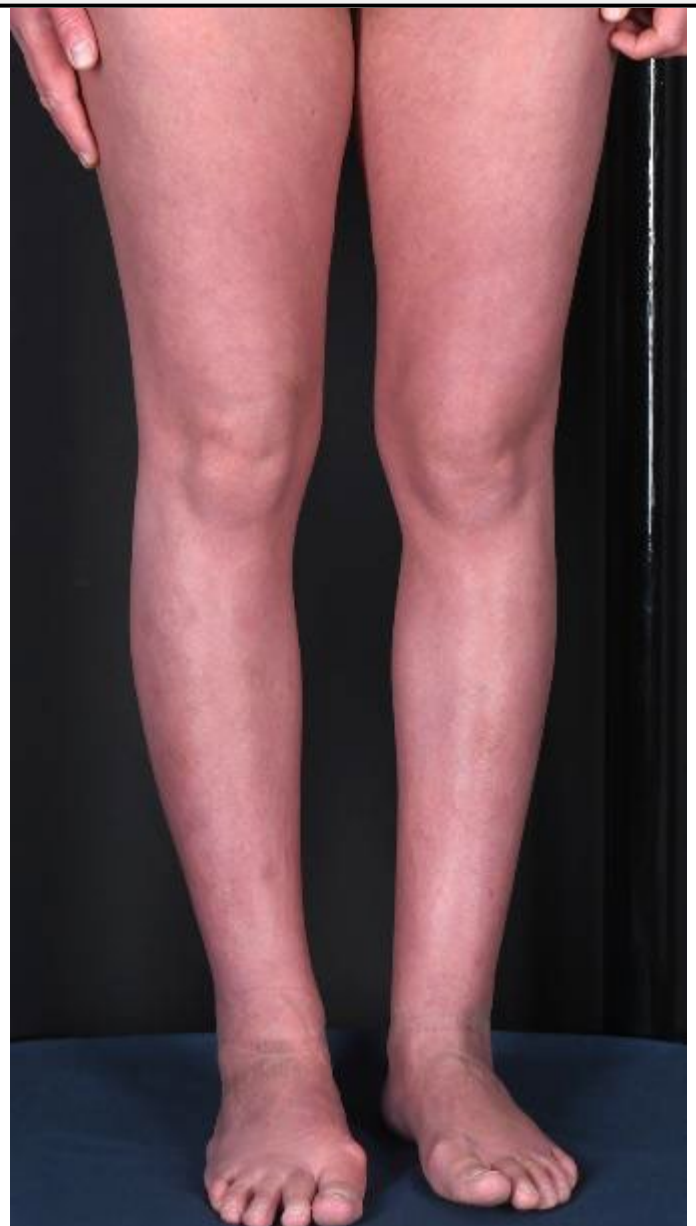
25.03.2026

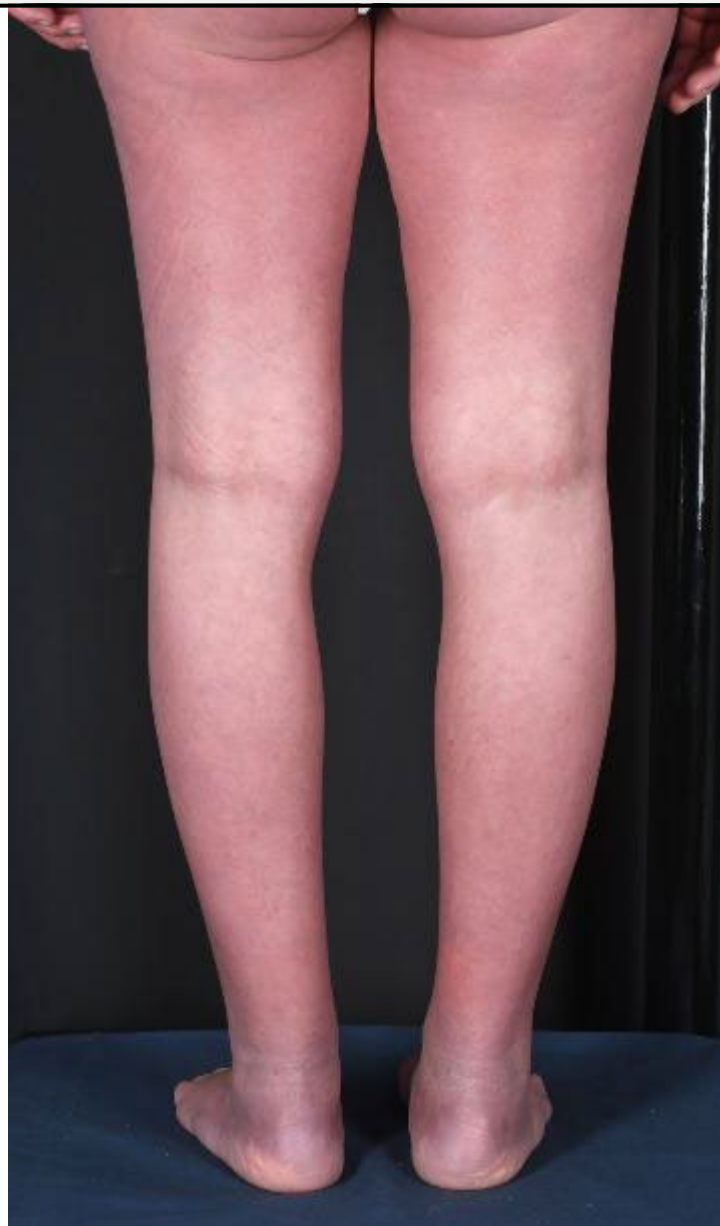
- Birth of two healthy twins
- Postpartal:
  - necrotizing pancreatitis
  - cardiomyopathy: LVEF 40%
  - amemia: red blood cell transfusion (4x)
  - intensive care unit

No specific Sézary therapy possible: progression erythroderma, pruritus, B2

CMV reactivation and Quantiferon +

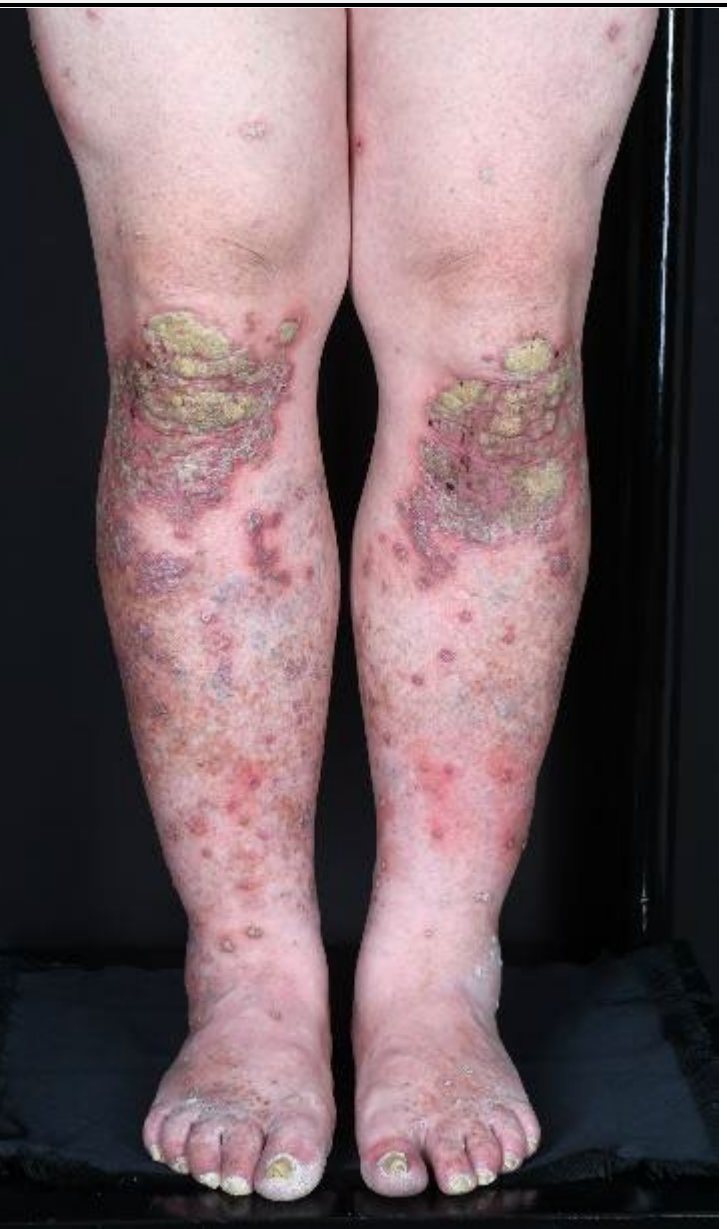


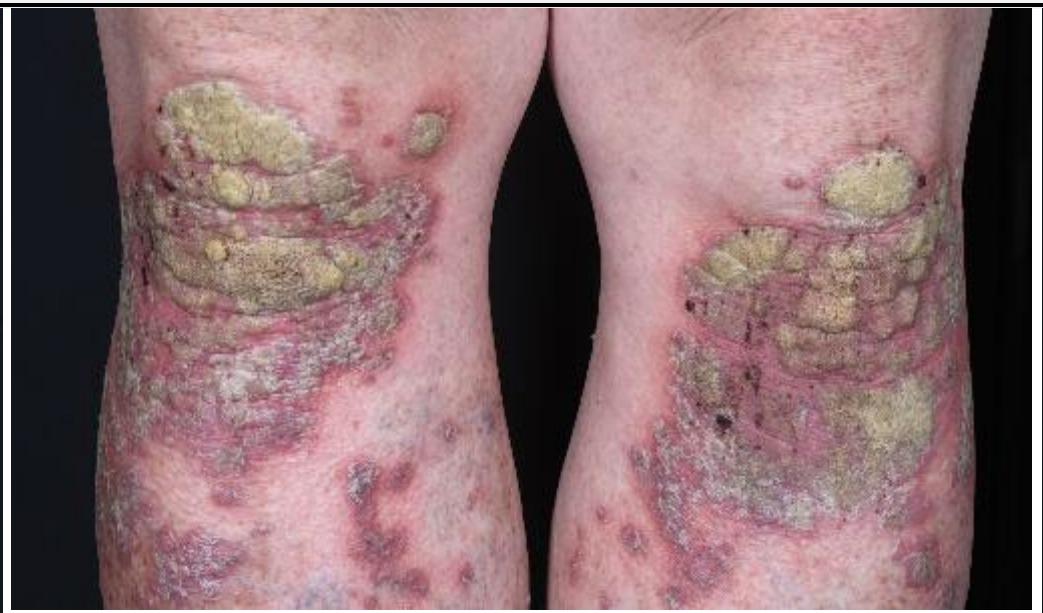


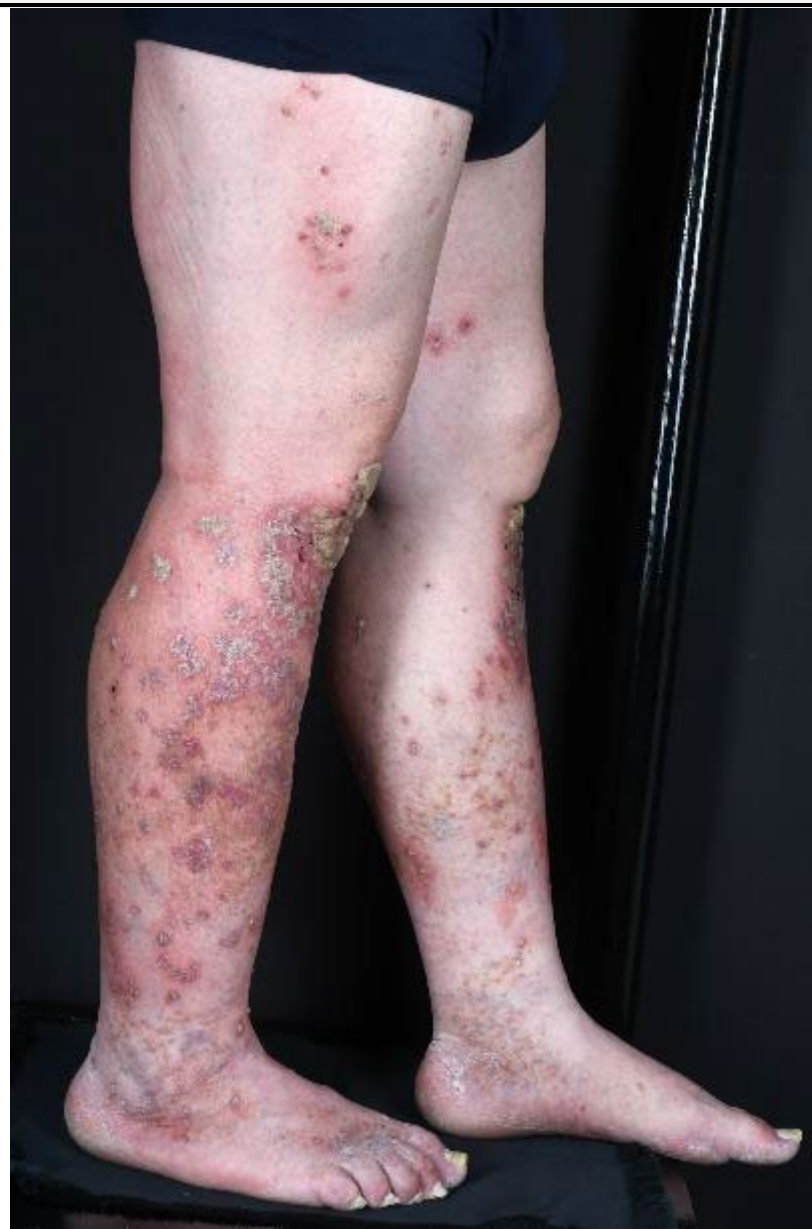


Case 6

Clinical presentation – intitial presentation

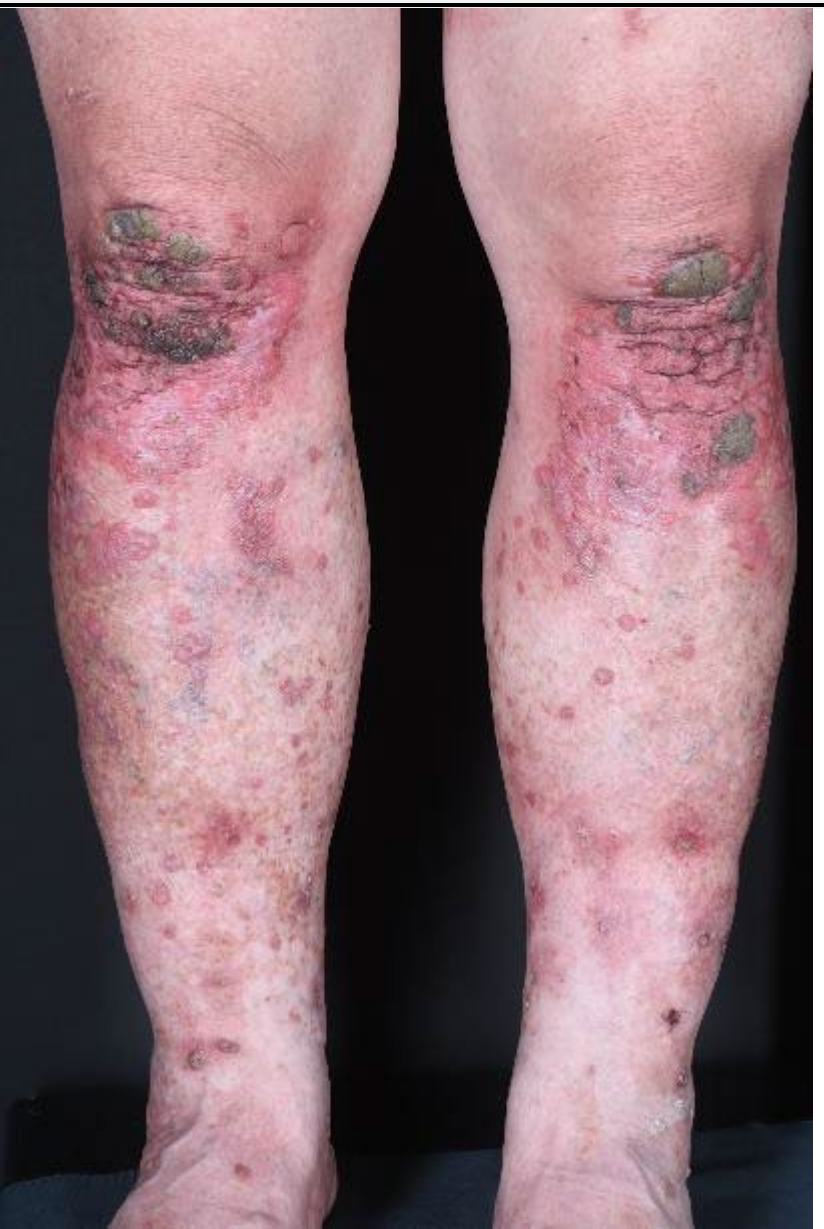


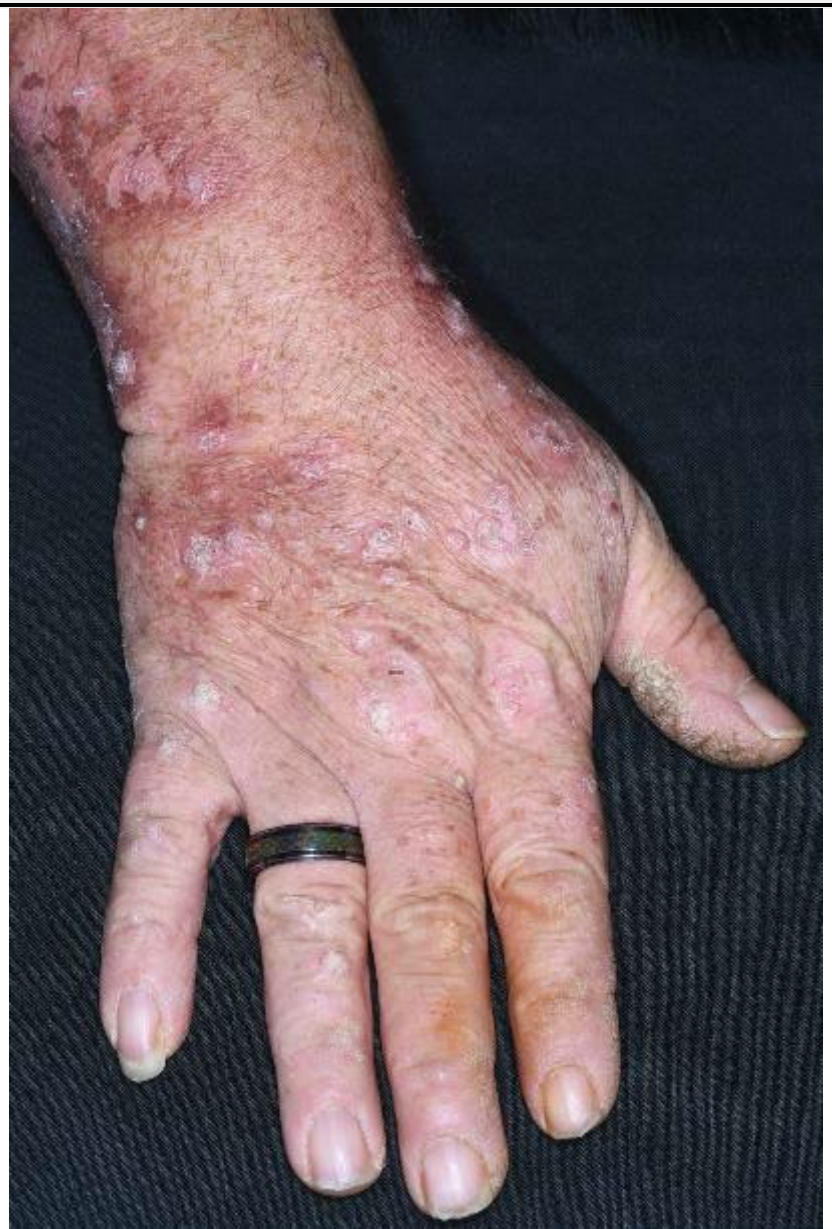


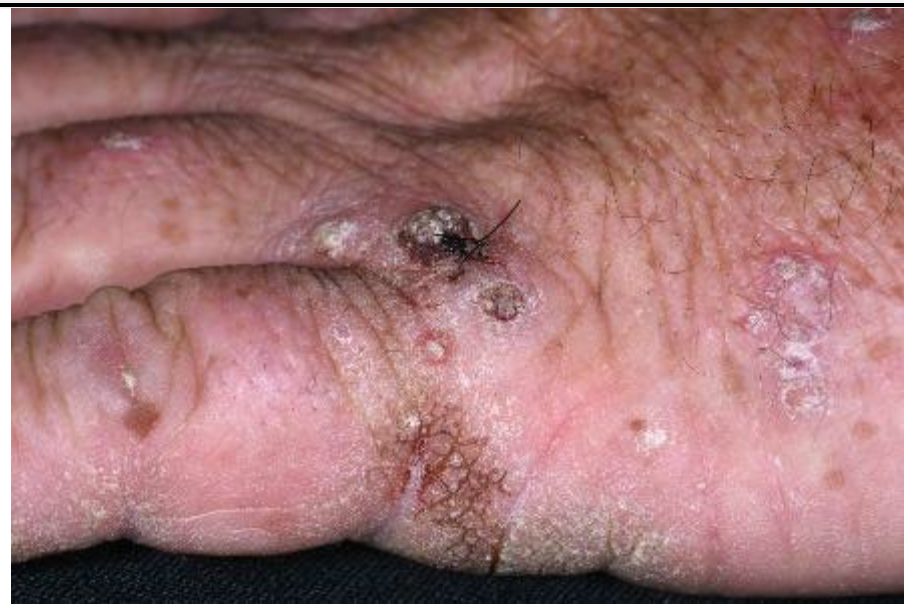


- 54-year-old male patient
- Psoriasis since childhood without arthritis
- UV-light therapy in the past
- Previous psoriasis specific treatments without any effects:
  - Fumaracid 18 months
  - Secukinumab 18 months
  - Risankizumab 4 months
  - Ixekizumab 80 mg 4 months

Clinical presentation – after treatment



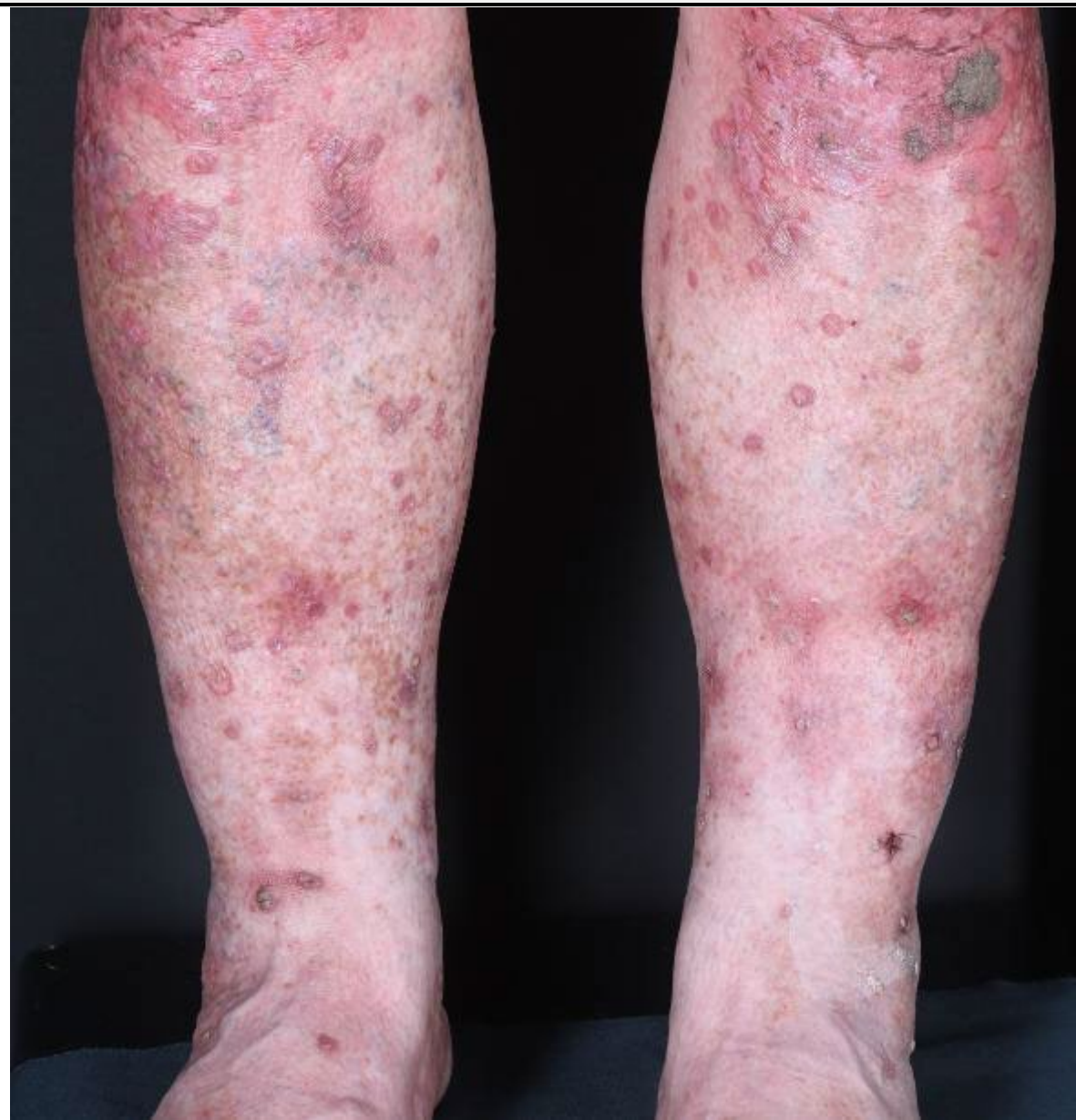




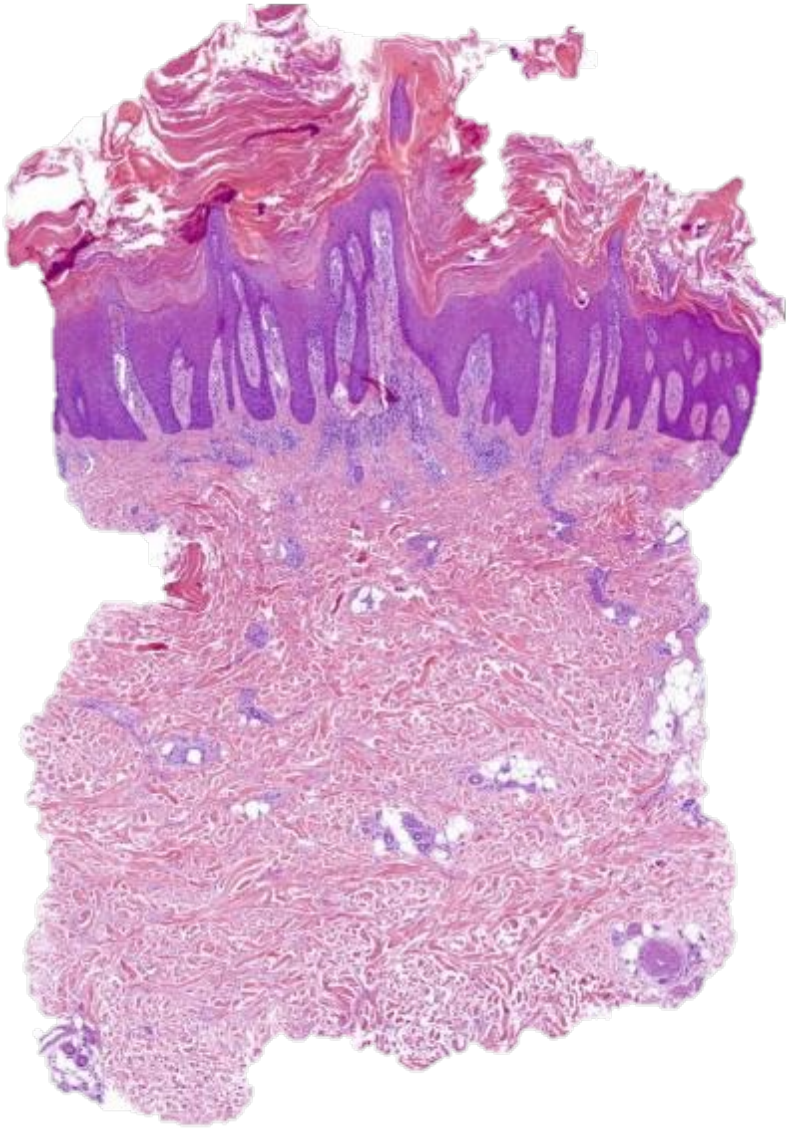
- Topical treatment with salicylacid in Vaseline 2% and cignolin up to 1%: reduction of hyperkeratosis, but no additional significant effects
- Clinician suspected additional verrucae vulgaris on the back of the hands

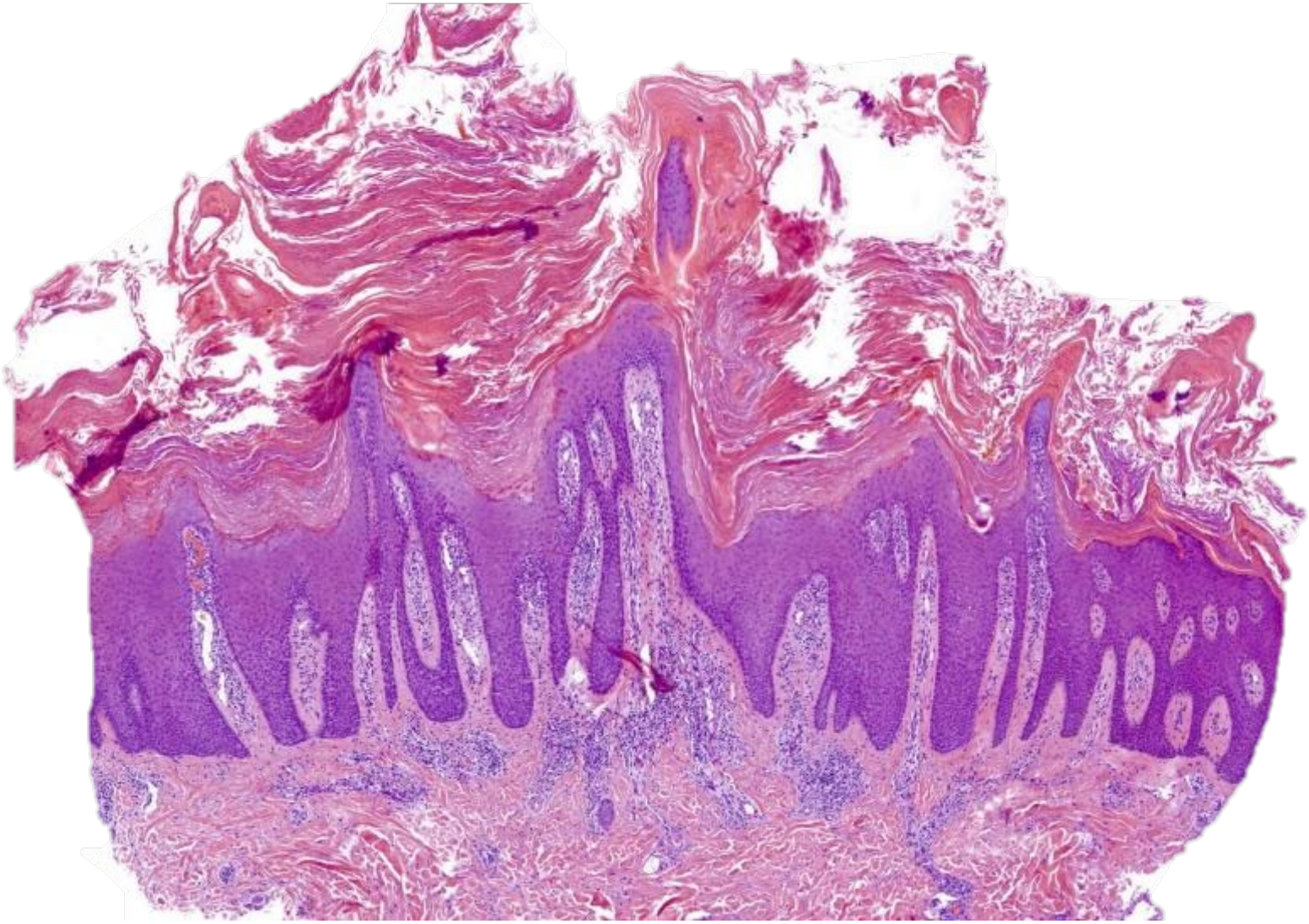
What is your clinical diagnosis?

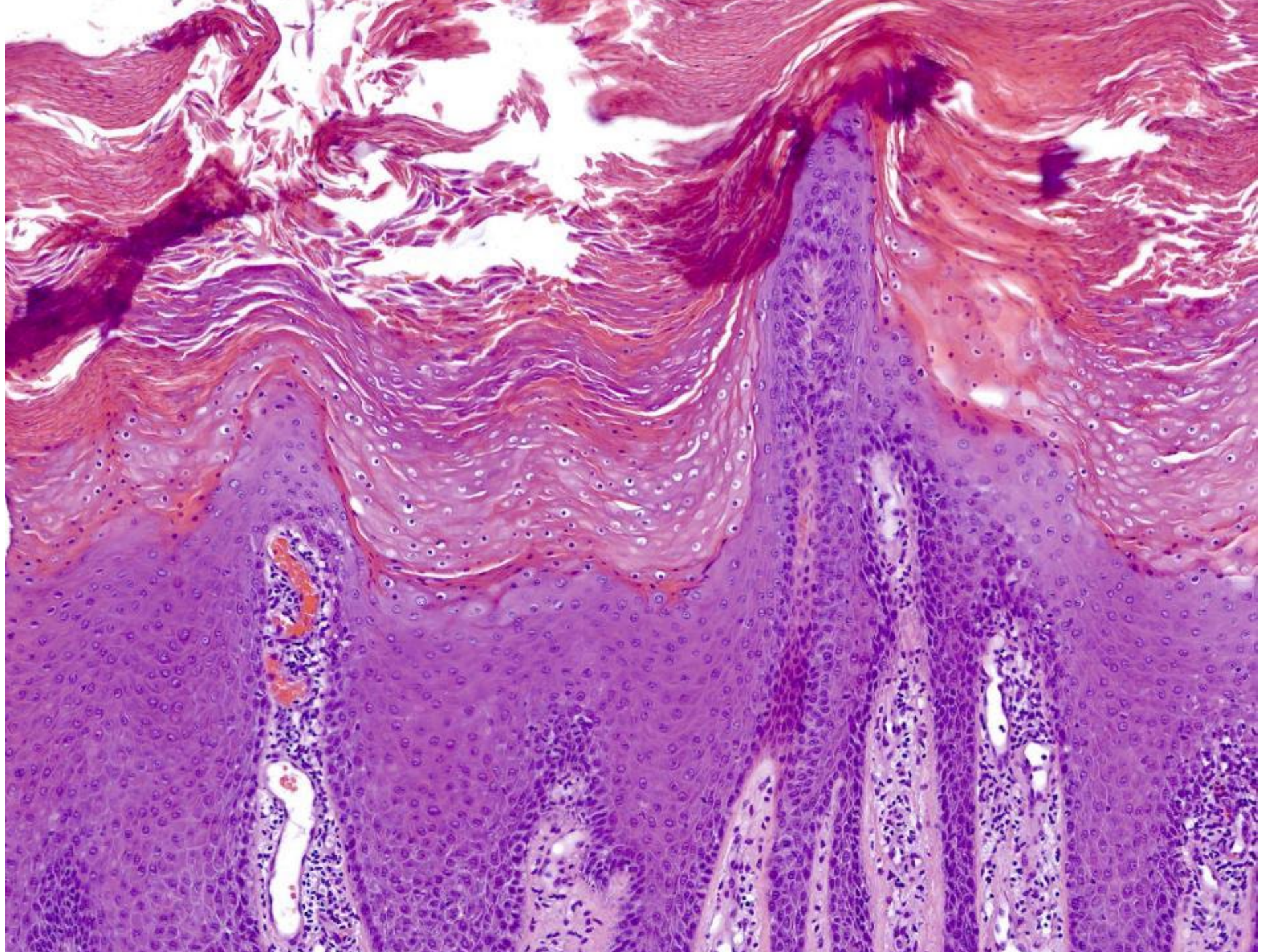
- Psoriasis vulgaris? But why no response to treatment?
- Verrucae vulgaris? Typical clinical presentation?
- Collision with a secondary diagnosis?
- Different diagnosis simulating psoriasis?

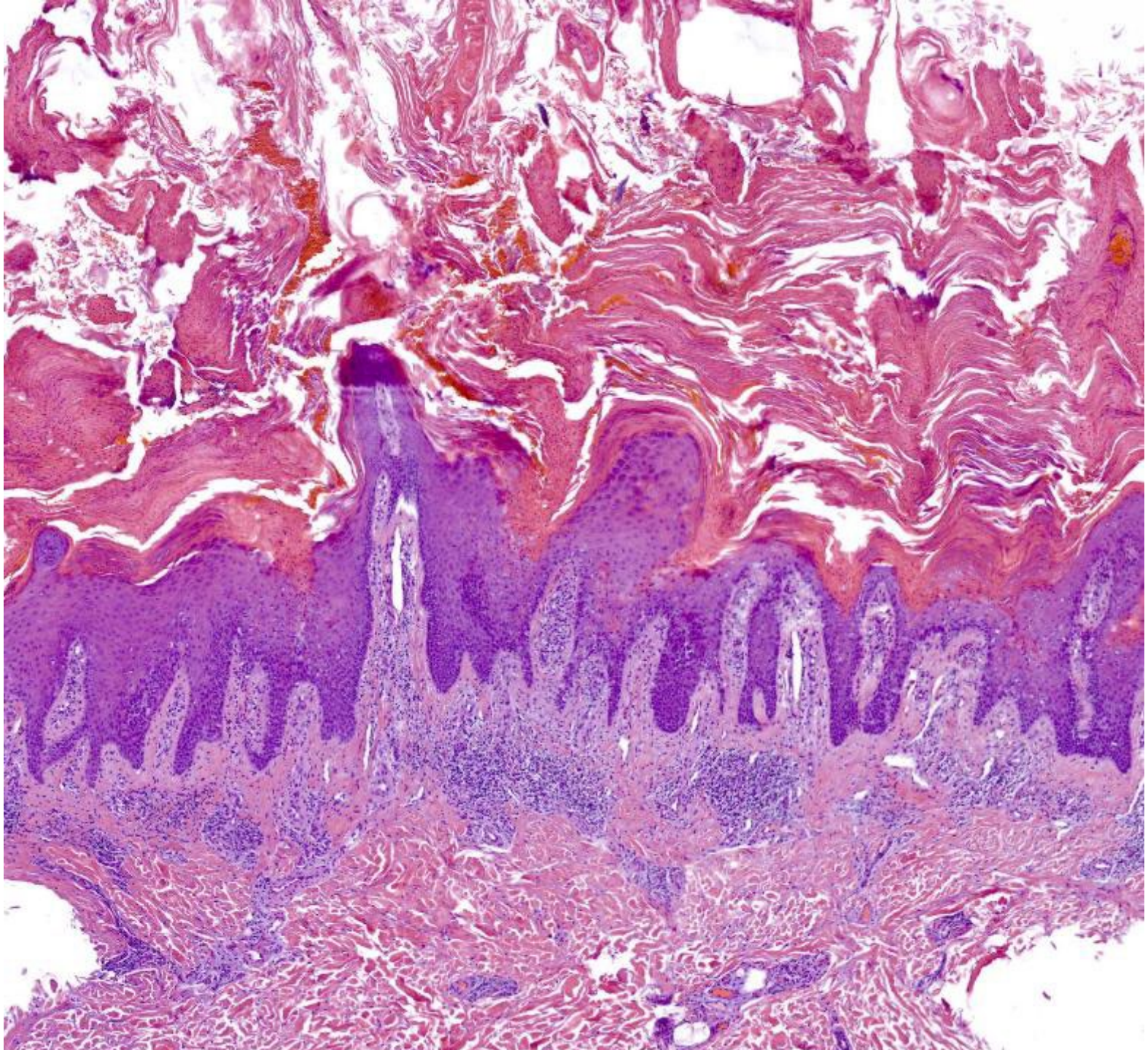


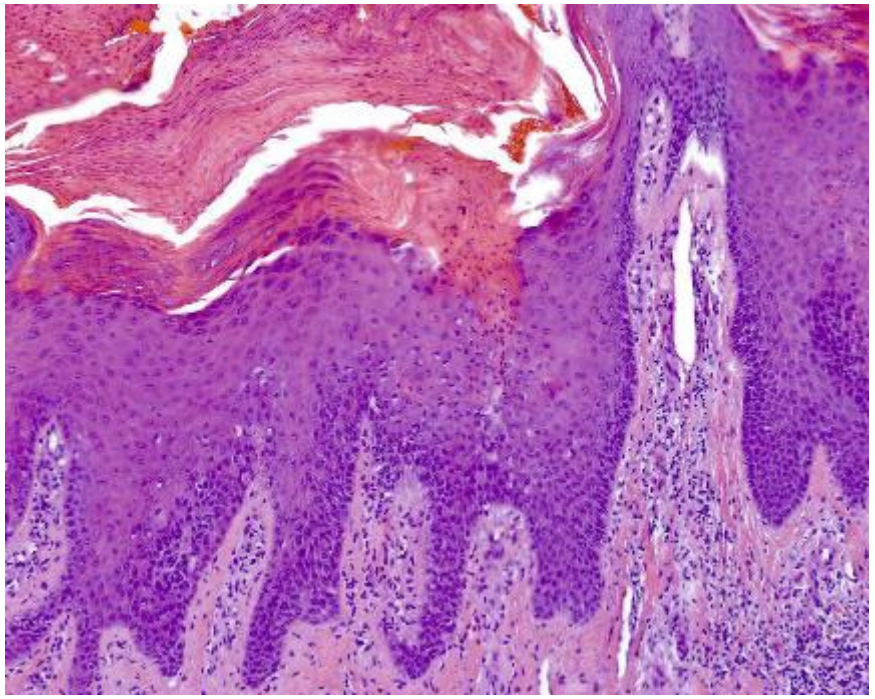
Histology

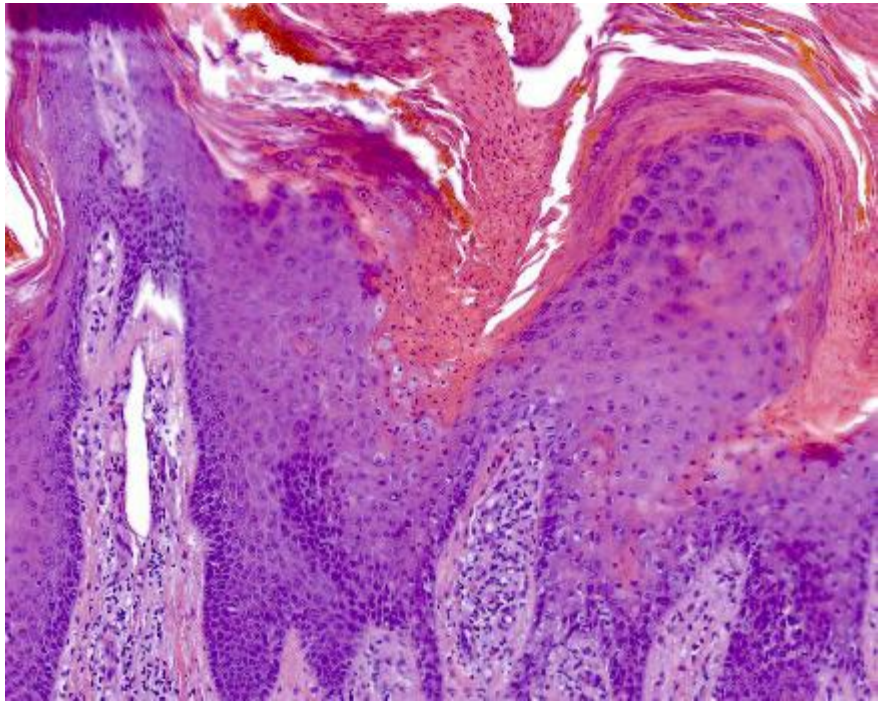




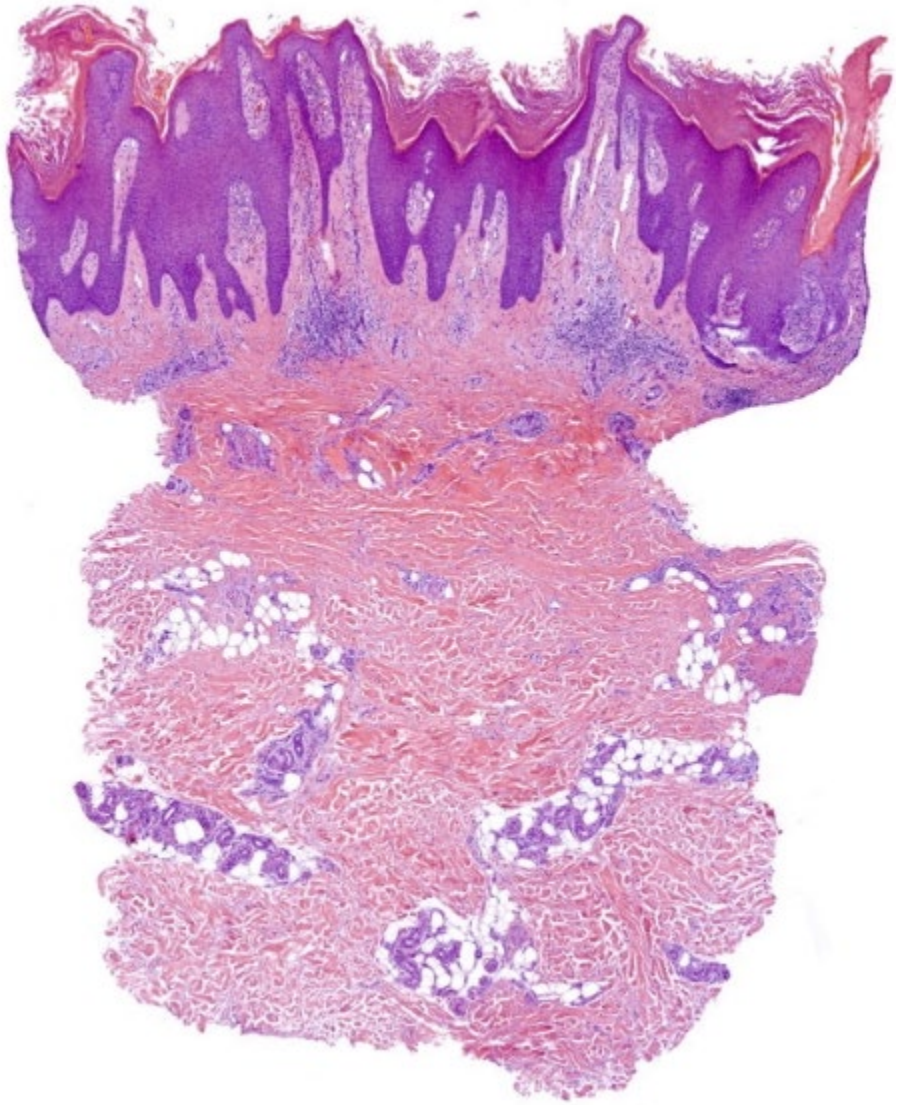


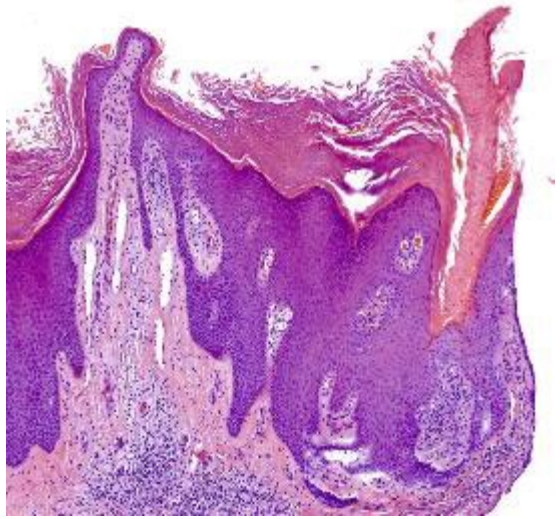


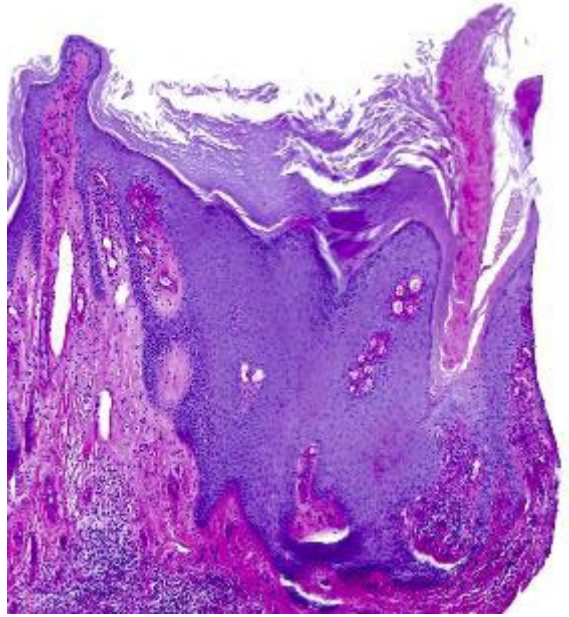


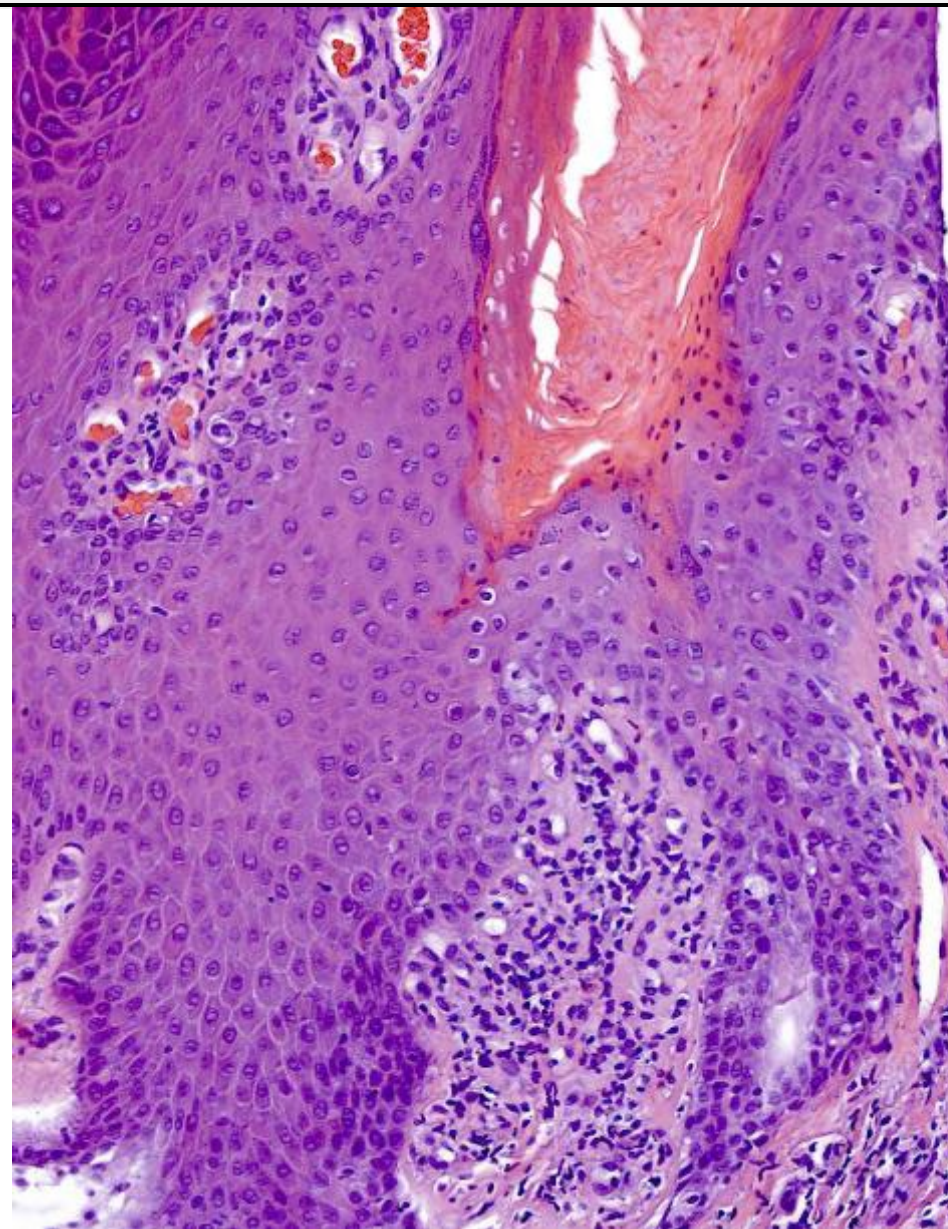


Histology

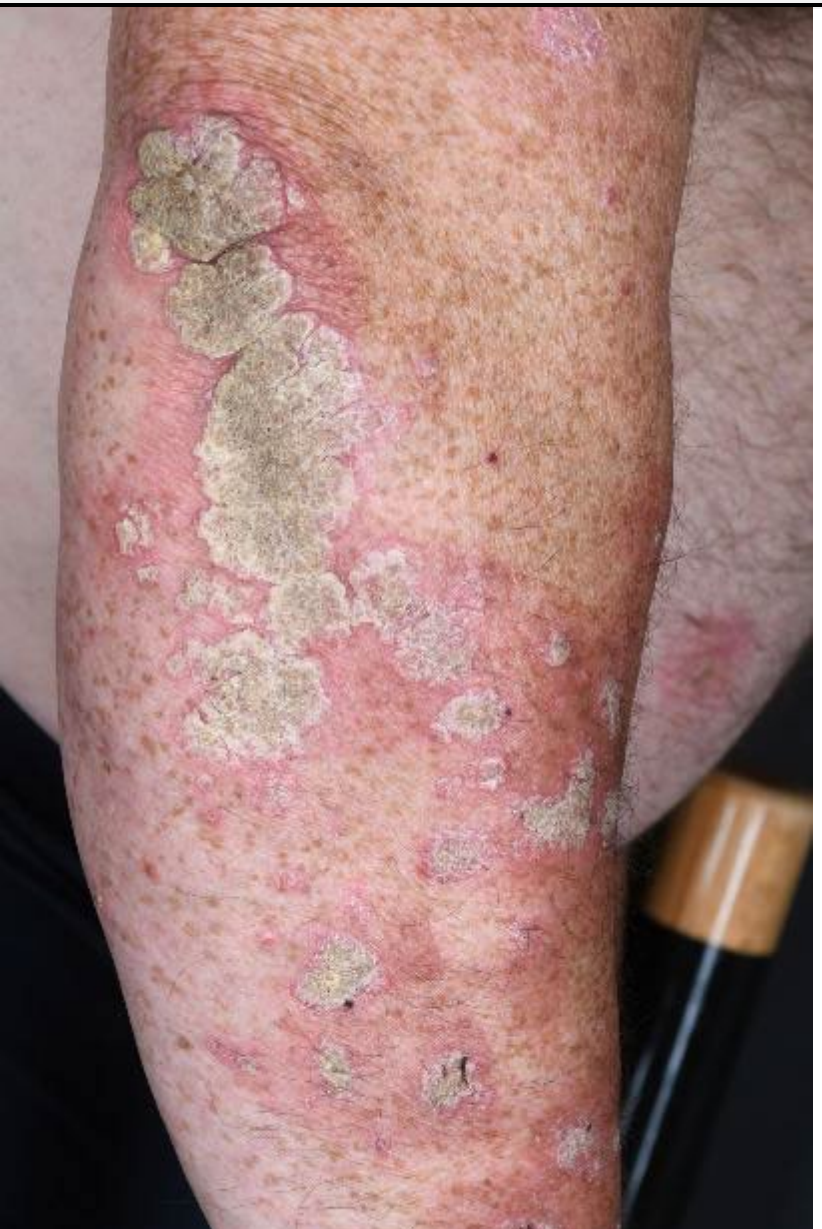


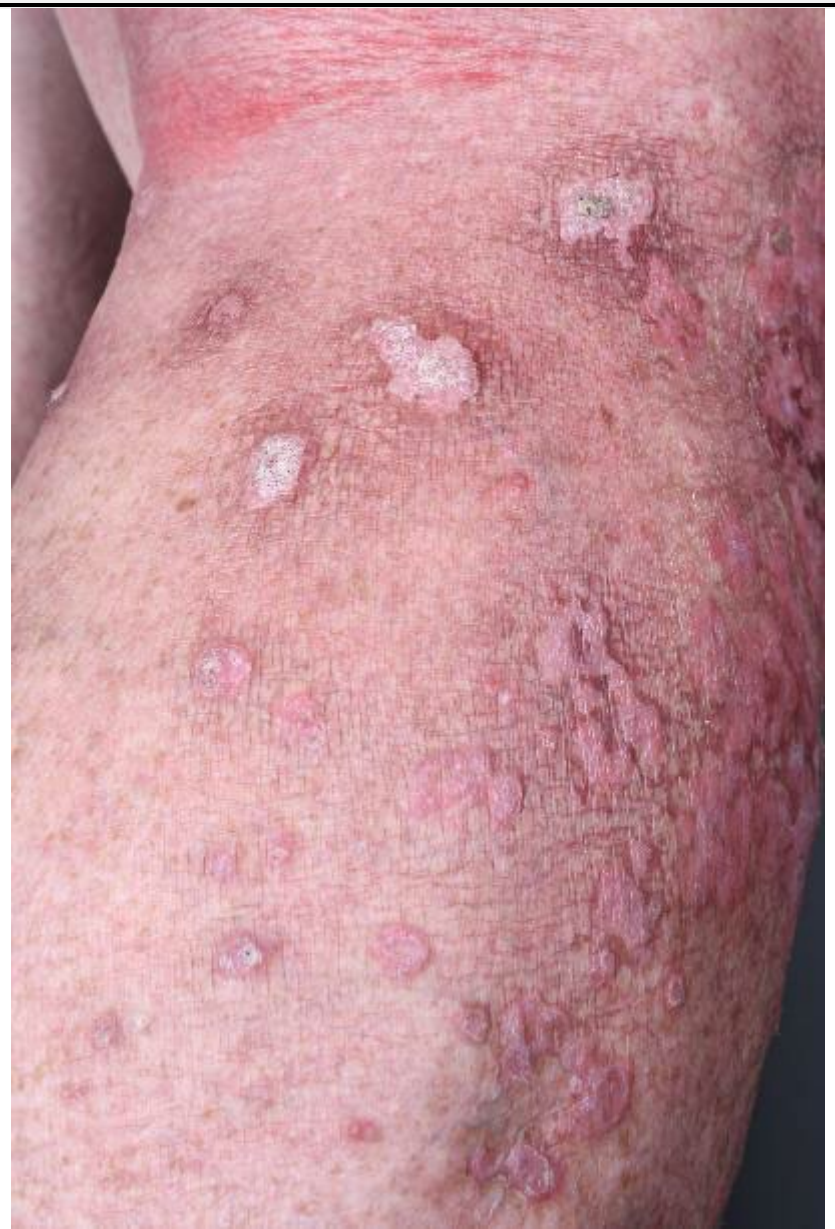


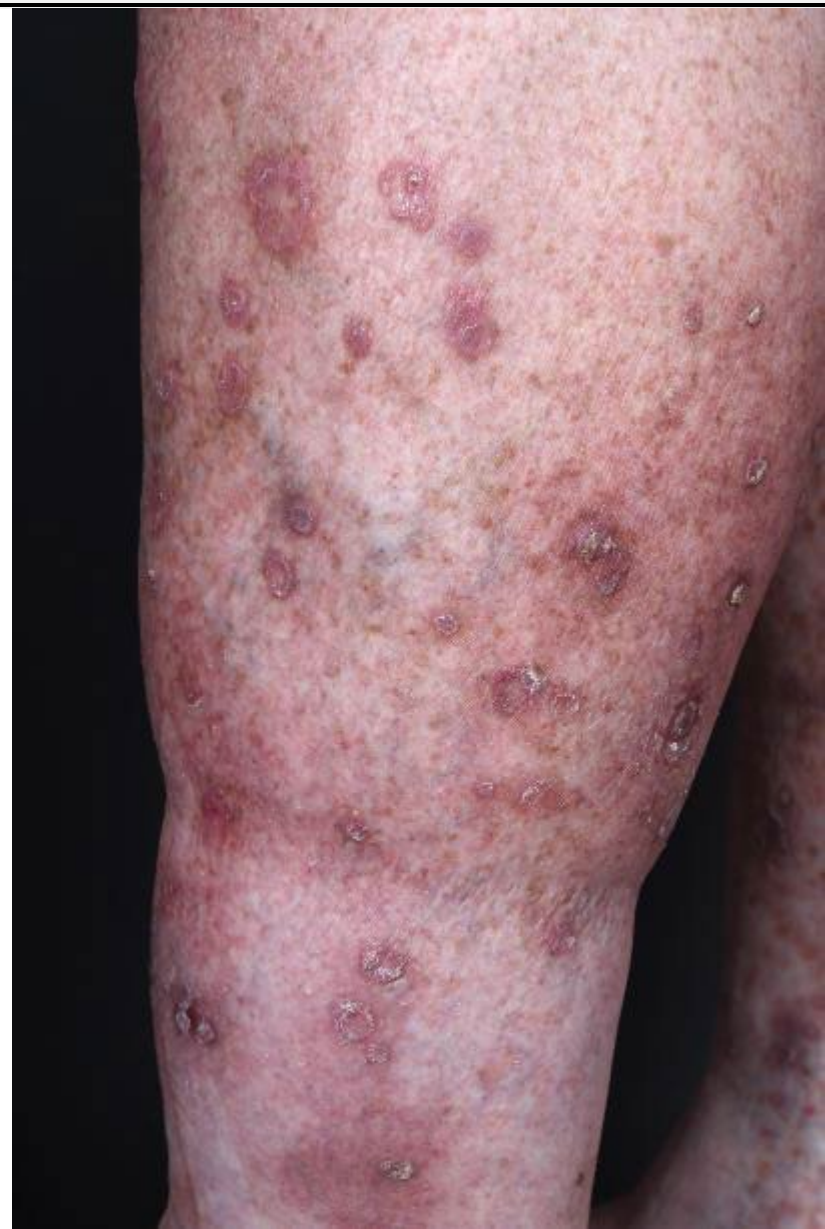




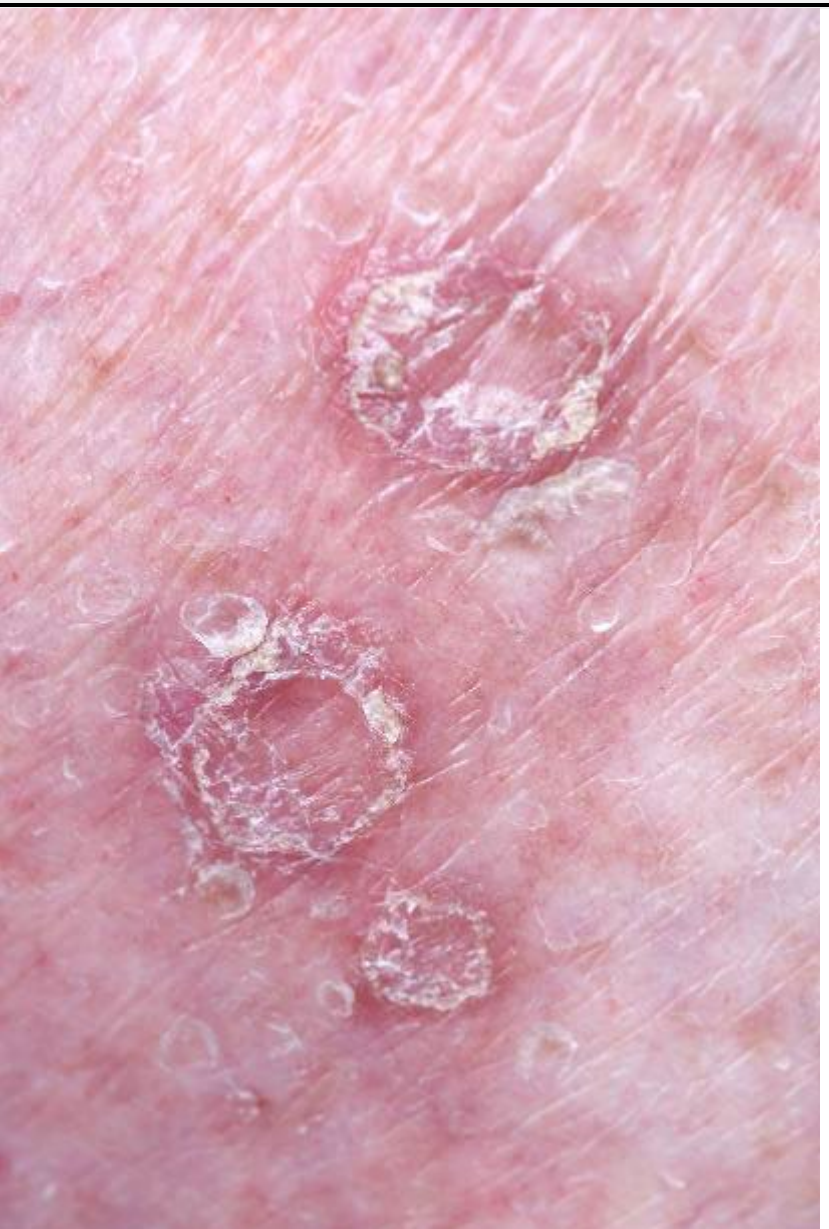
Clinical correlation

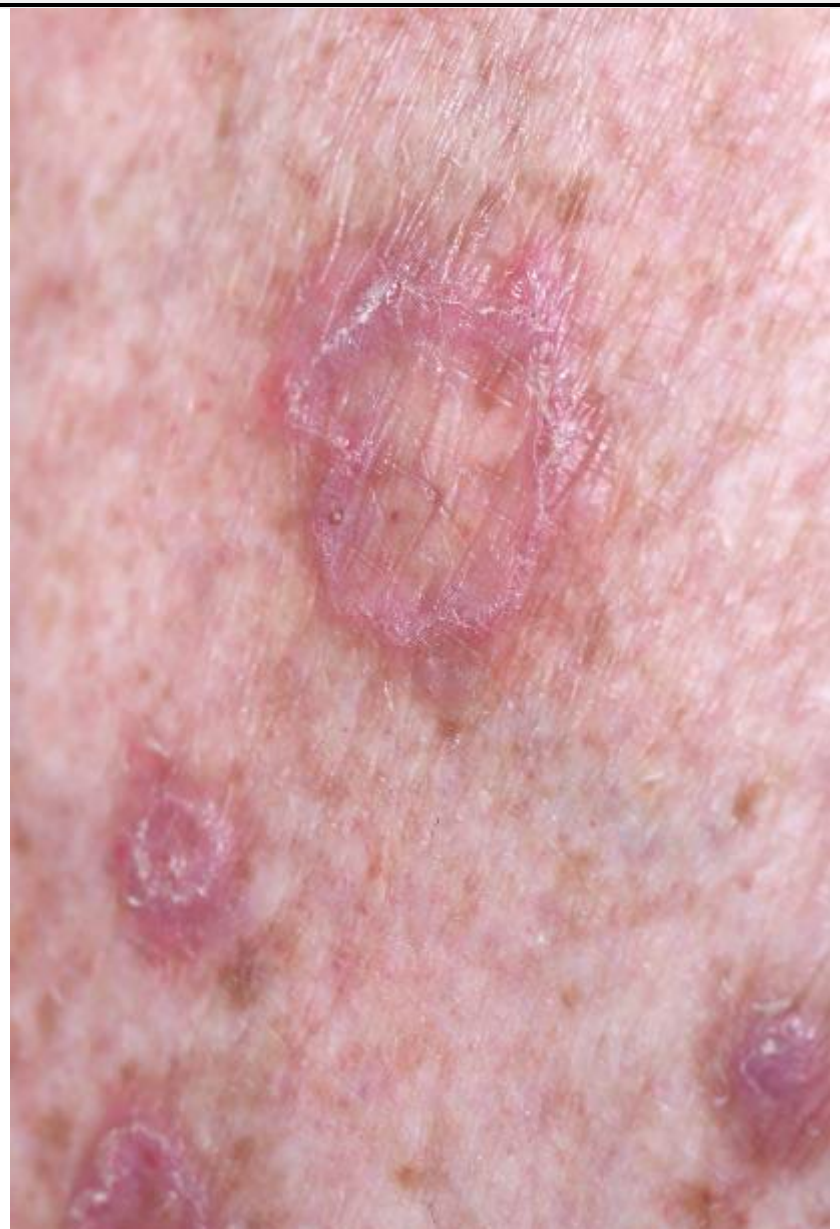


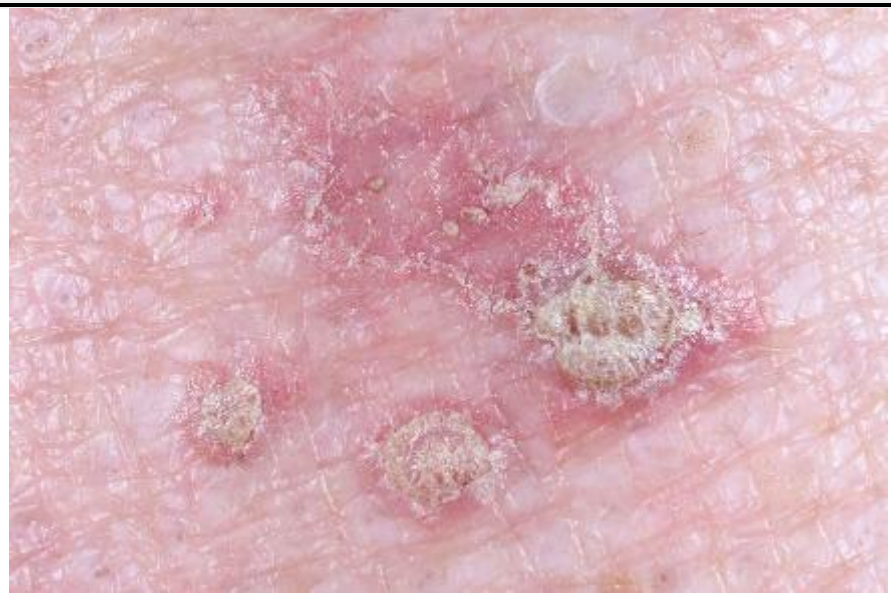


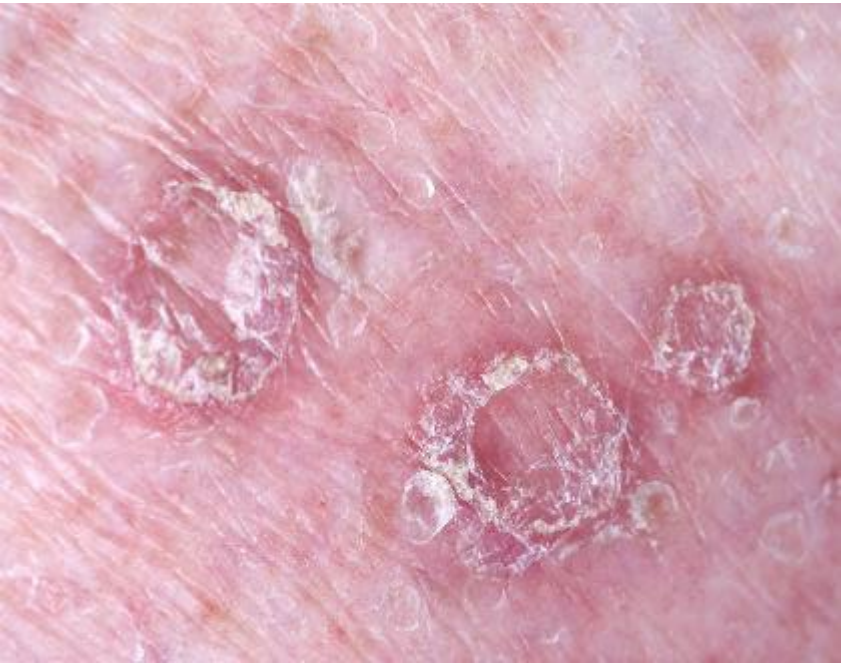
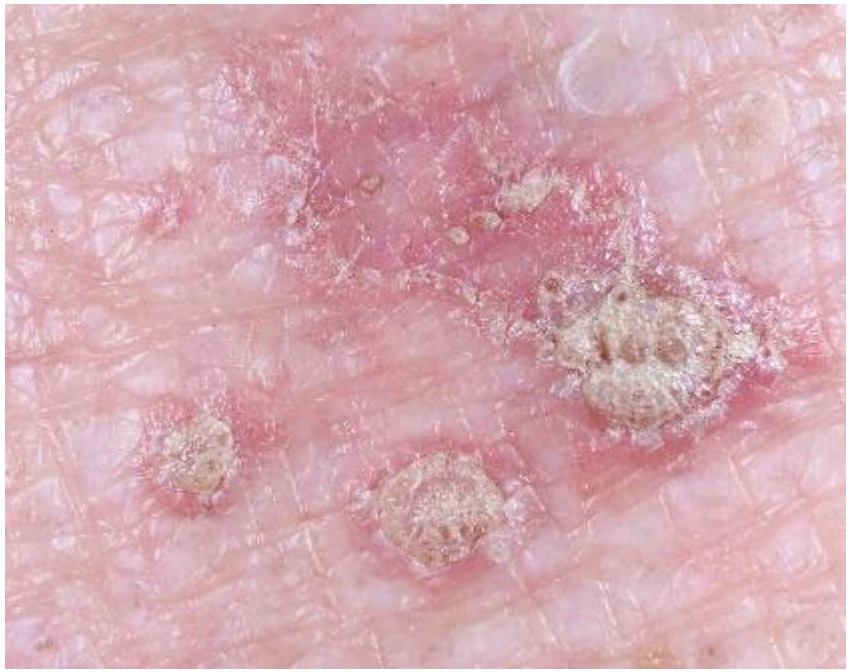
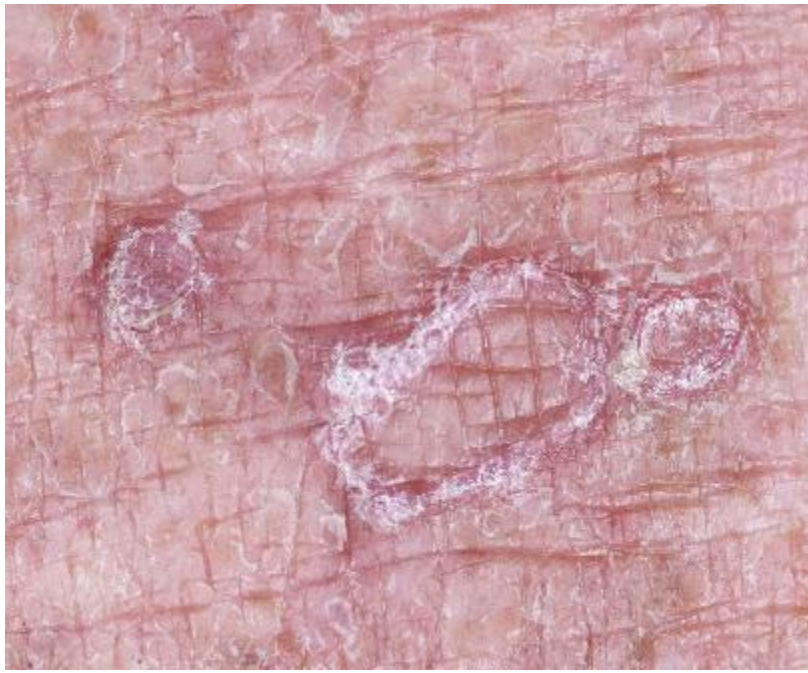
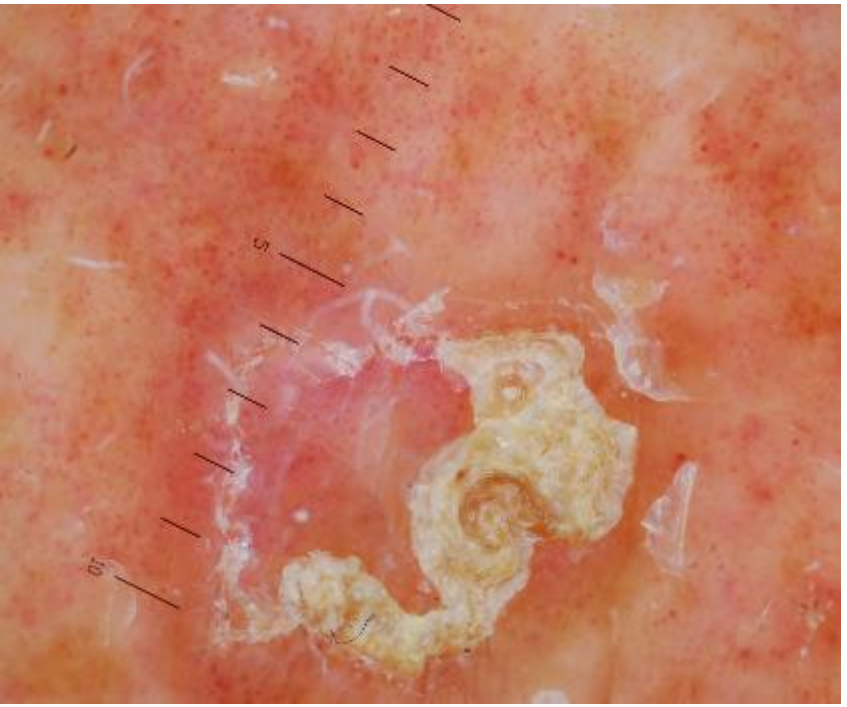
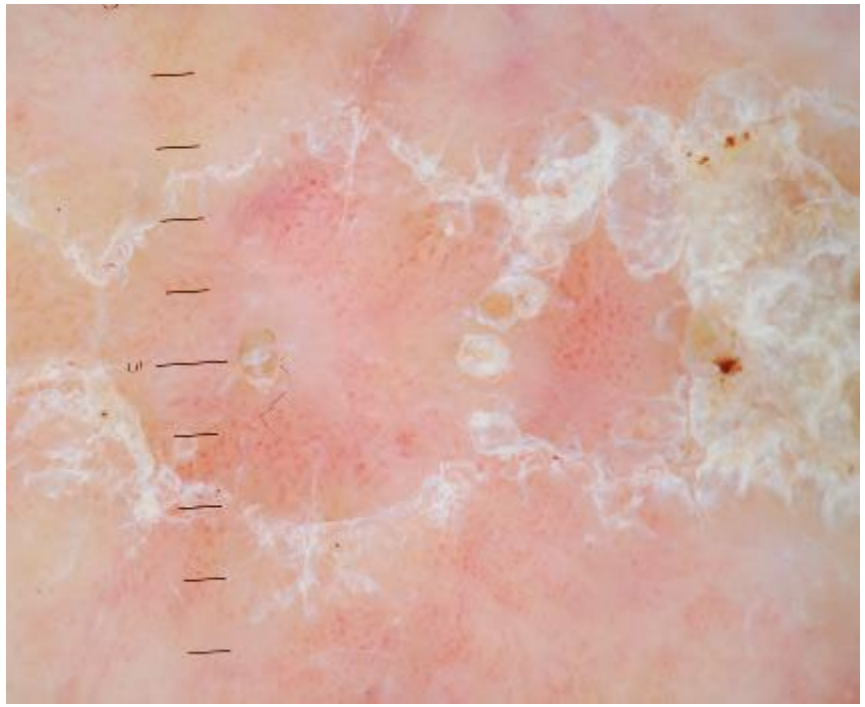
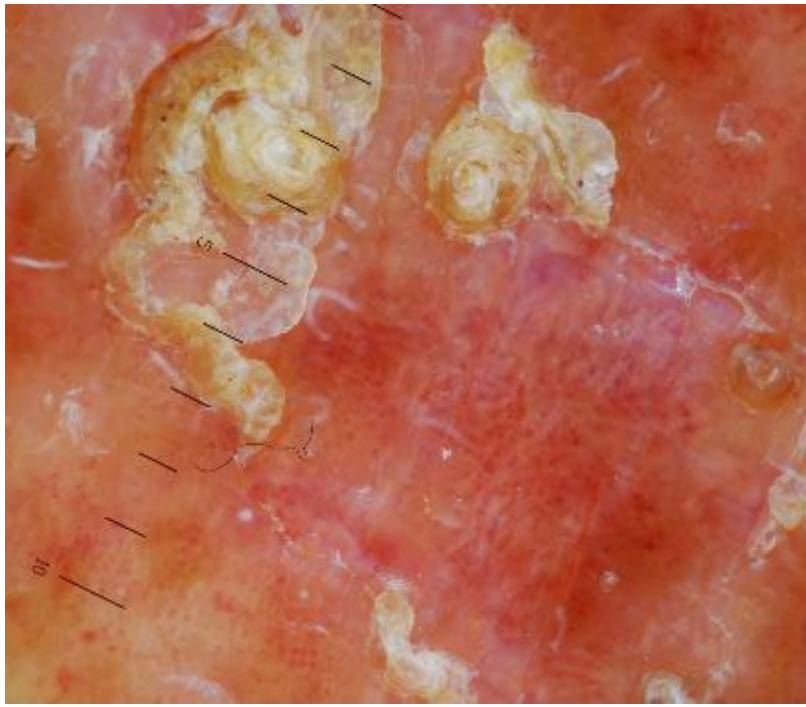


Clinical correlation

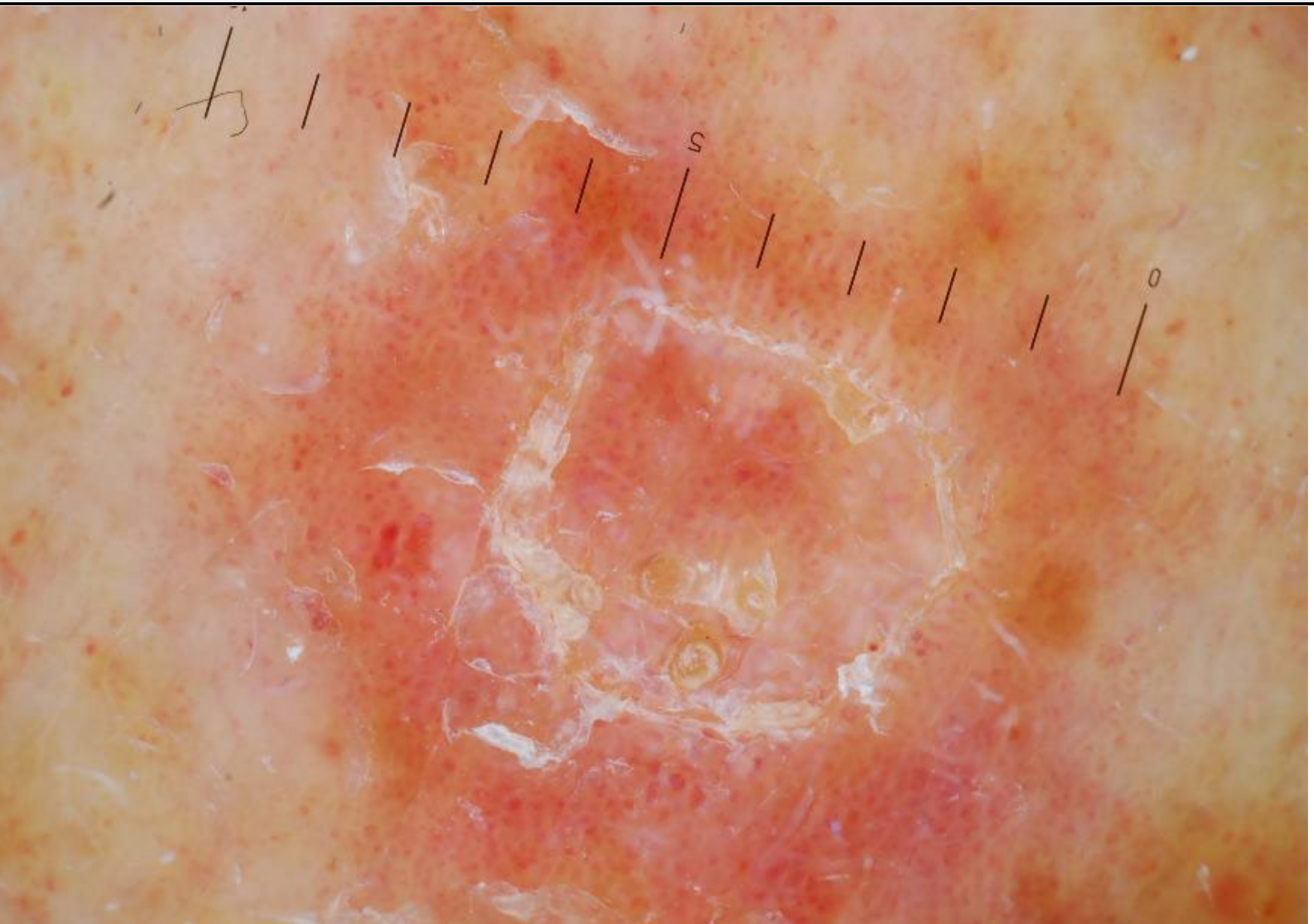






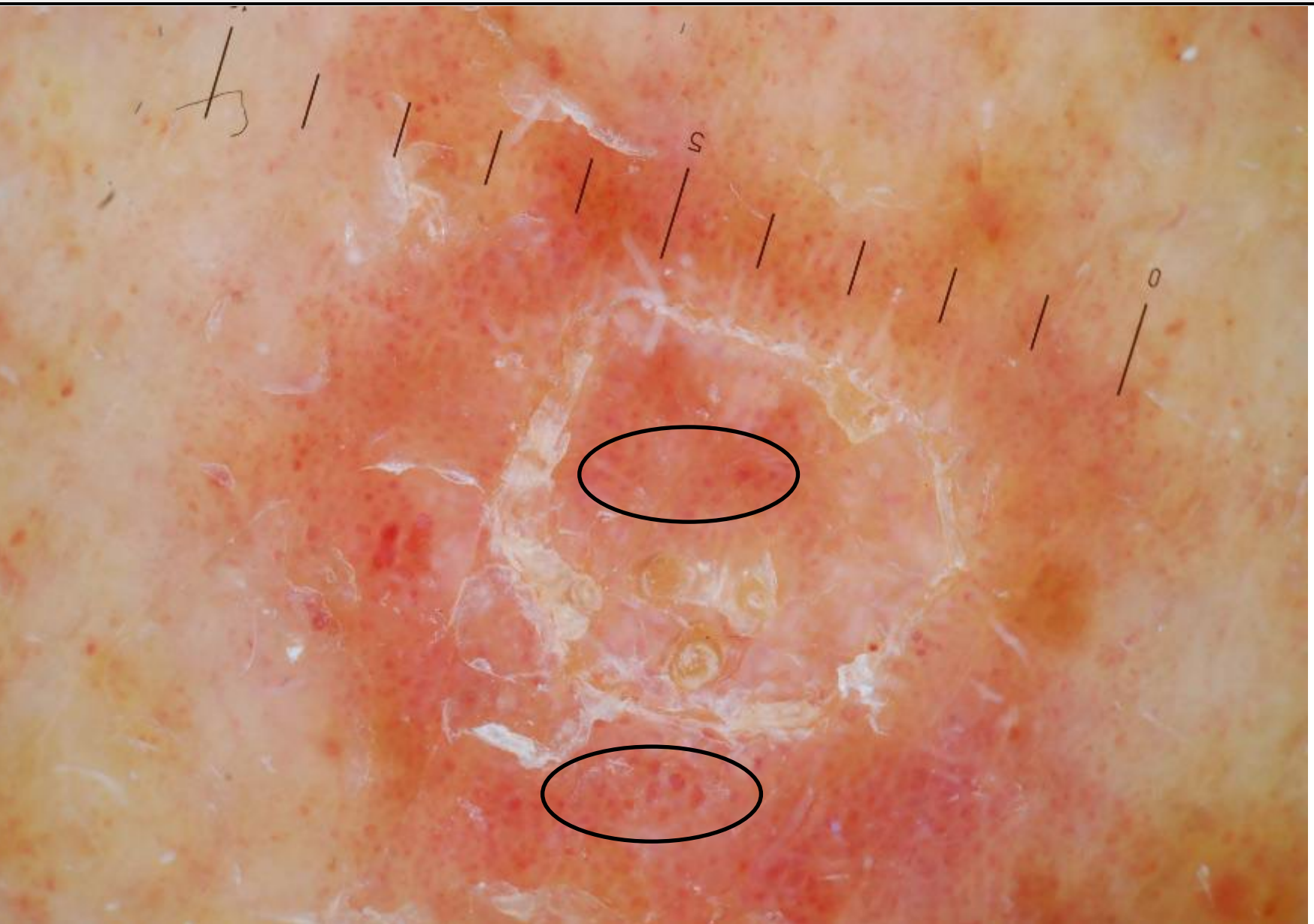







Clinical correlation



- Dotted and coiled vessels
  = elongated vessels
- Continuous rim of scale
  = cornoid lamella
- Follicular plaques
  = cornoid lamella

Clinical correlation



- Dotted and coiled vessels
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- Follicular plugs
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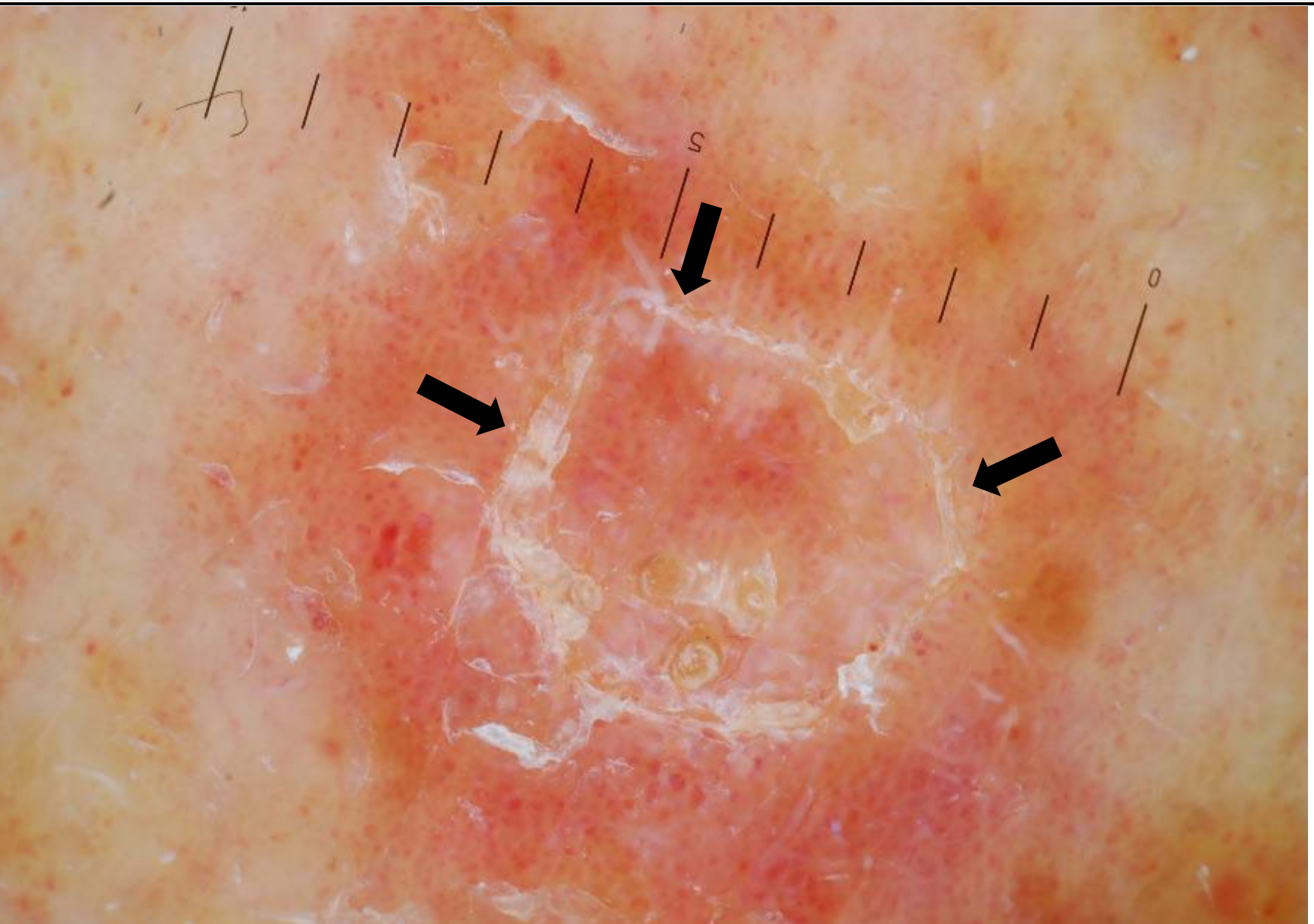
Clinical correlation



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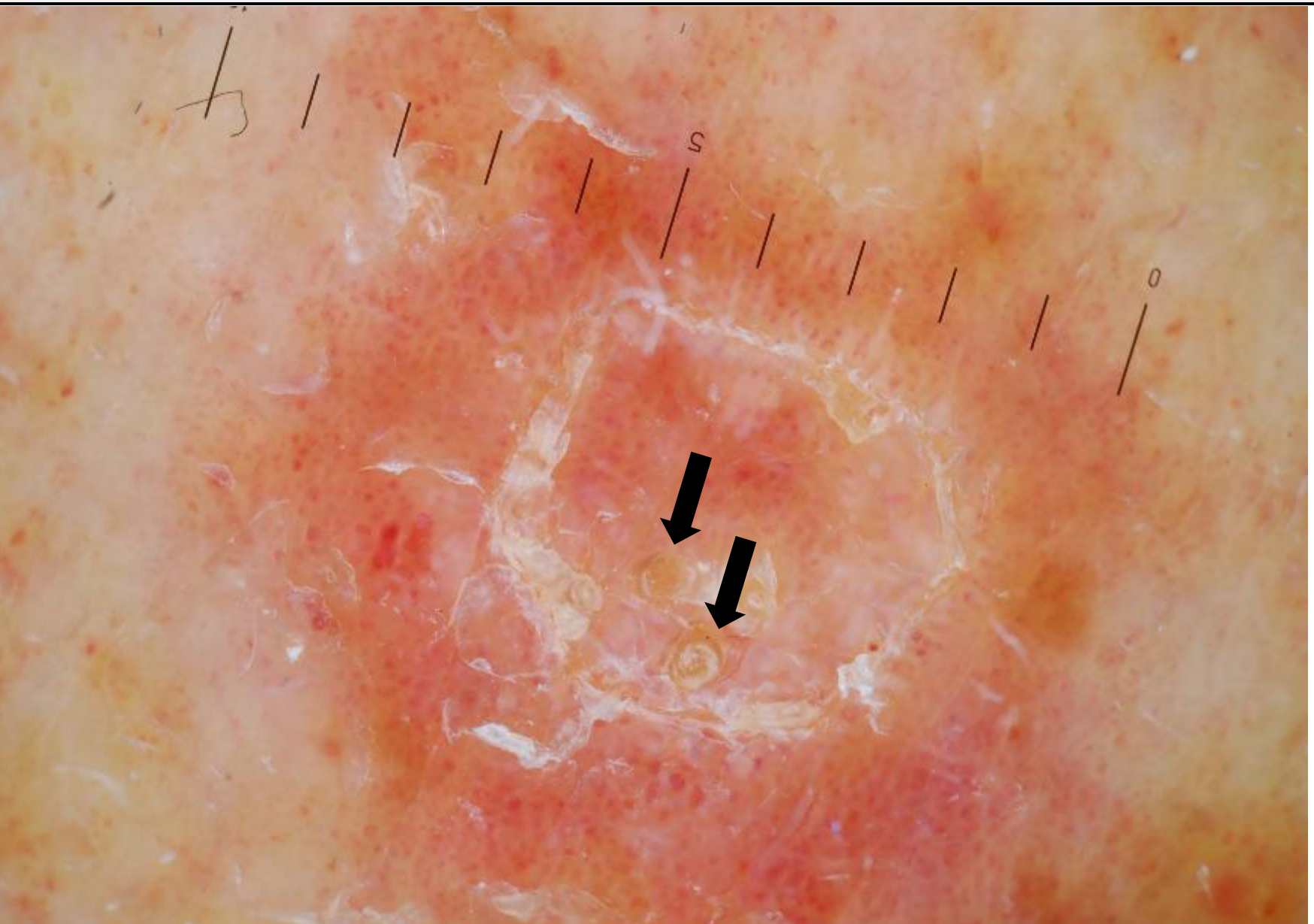
Clinical correlation



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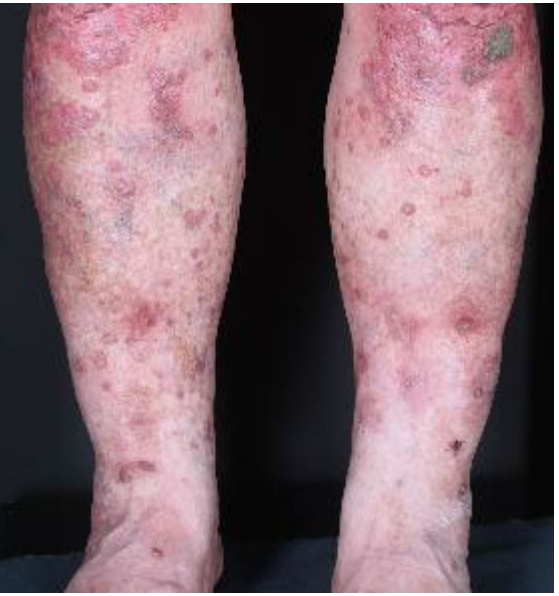
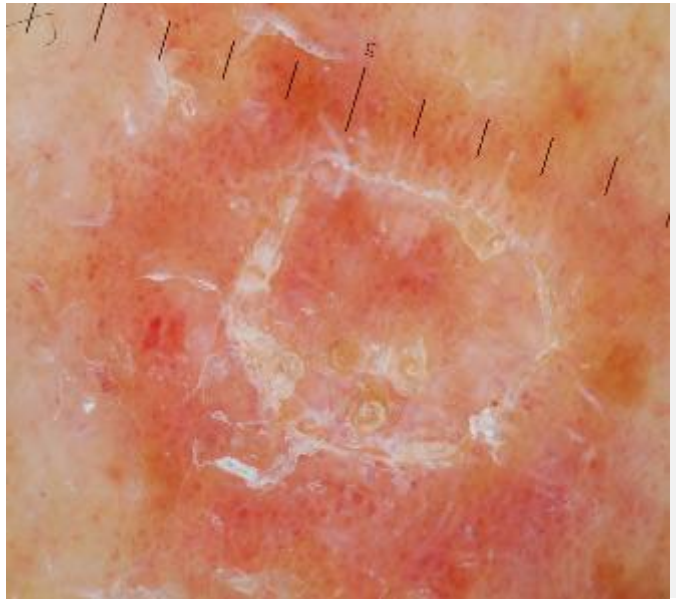
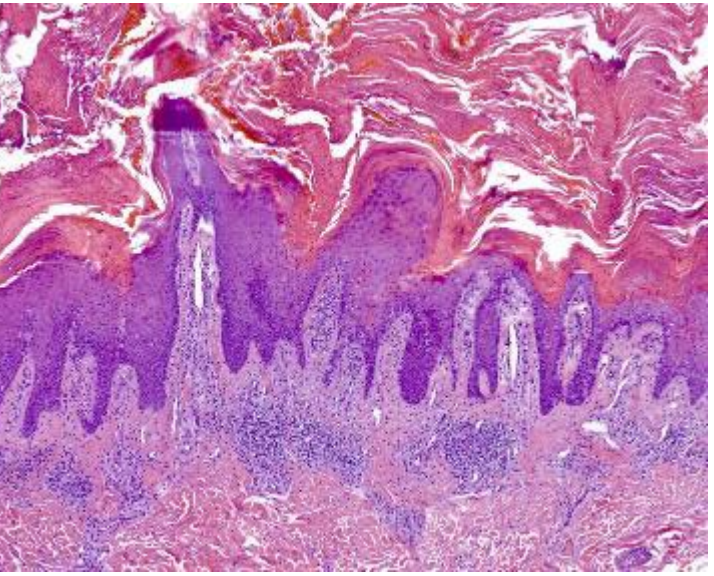
Summary of the findings:

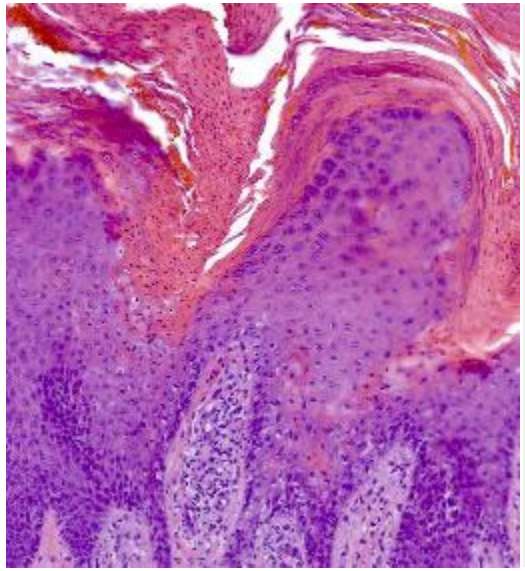




Clinical presentation:

- Verrucous psoriasiform plaques
- Ring and dot like cornification
- Predilection site: extensor surfaces extremities





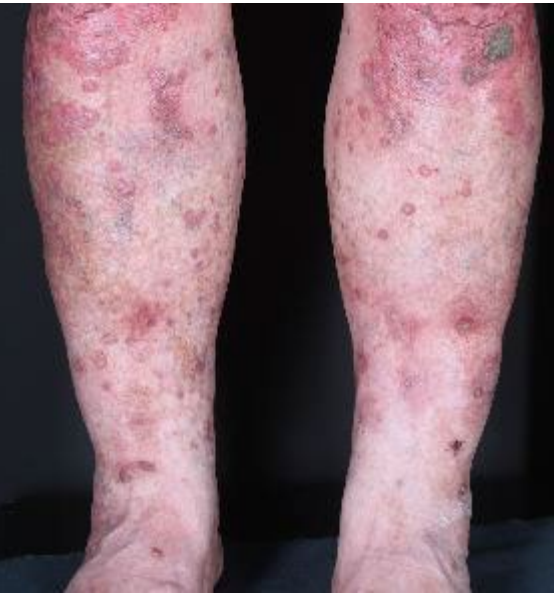
Histology:

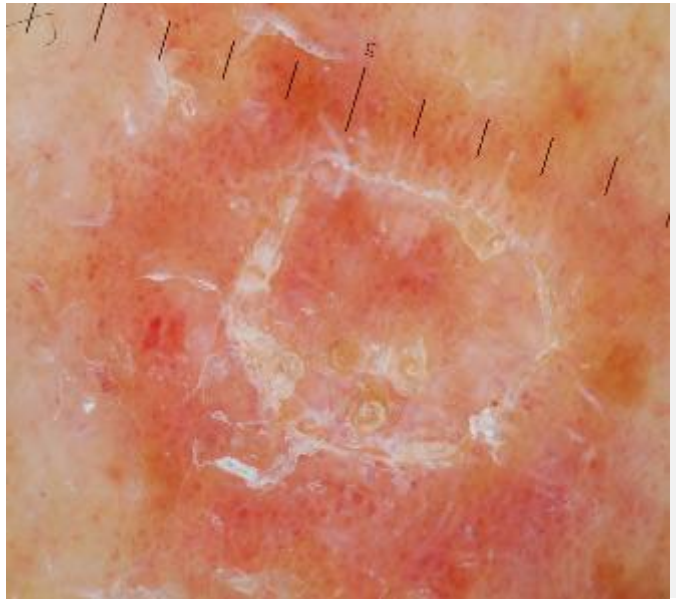
- Acanthosis and papillomatosis
- hyperkeratosis
- multiple cornoid lamellae

diagnosis

?

Summary of the findings:





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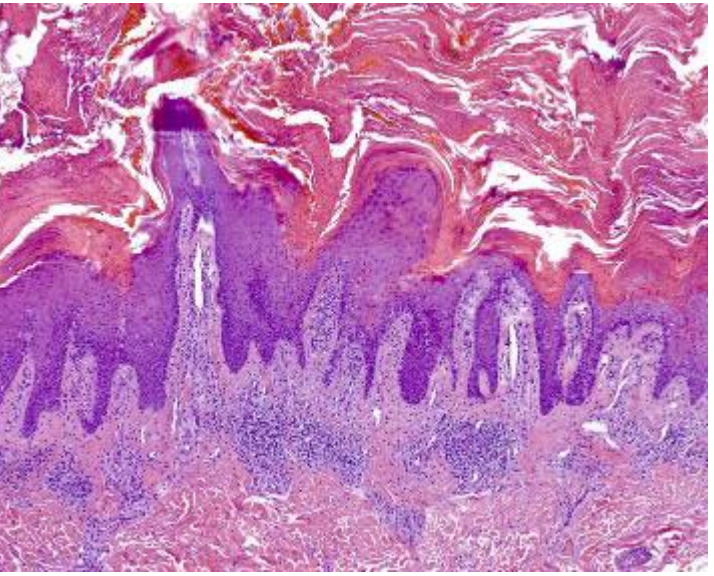
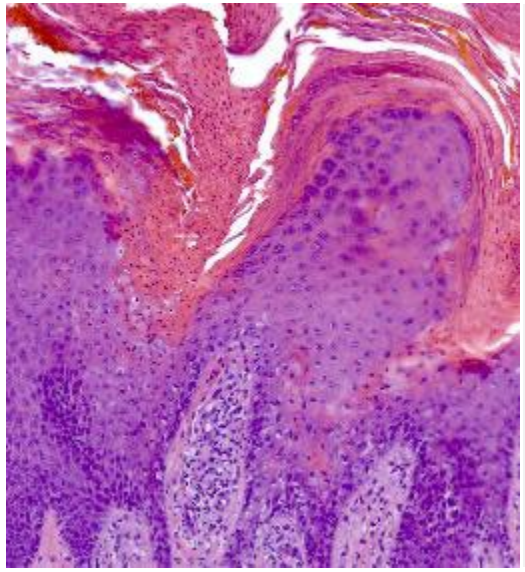




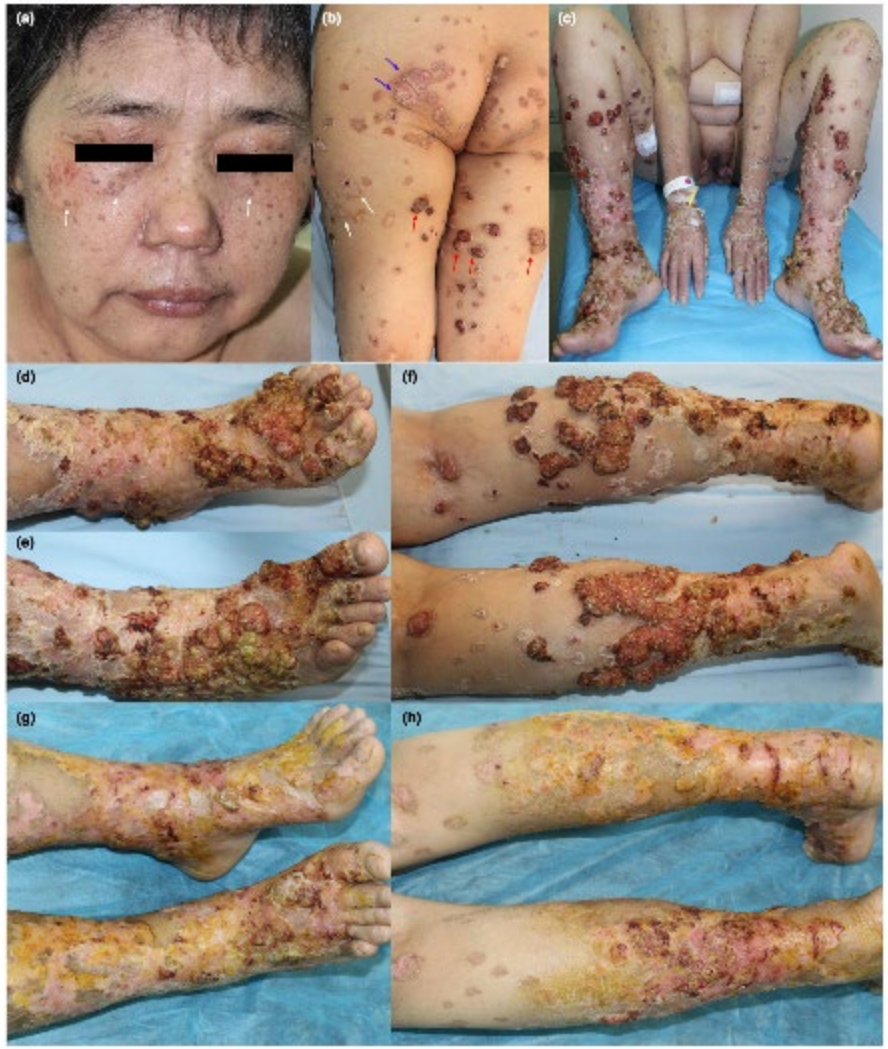
Histology:

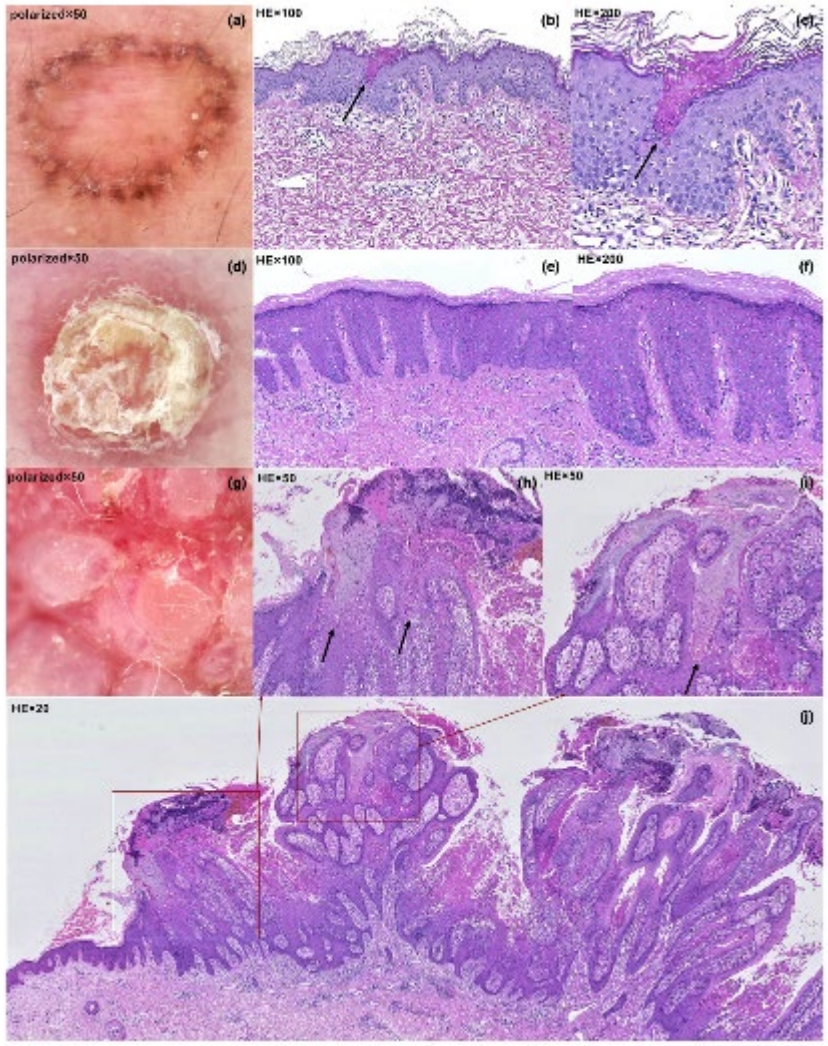
- Acanthosis and papillomatosis
- hyperkeratosis
- multiple cornoid lamellae

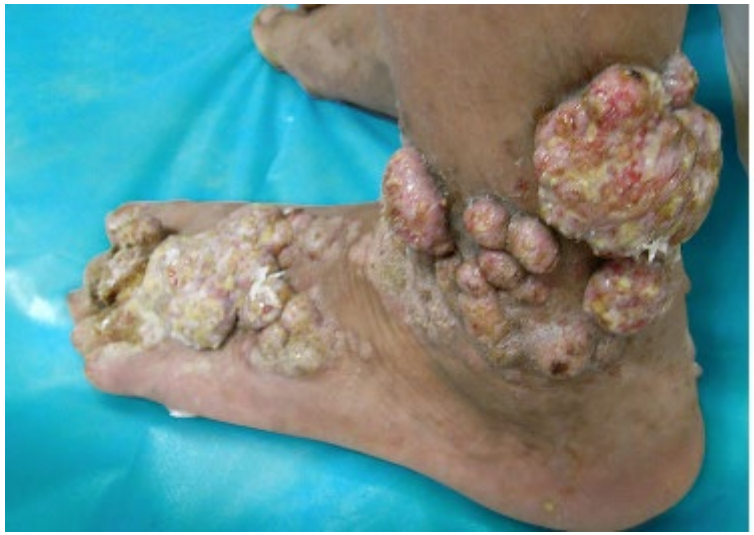
multiple porokeratomas

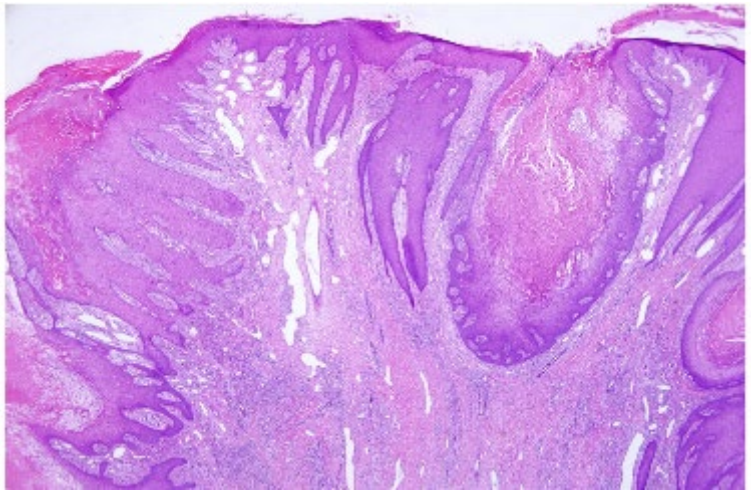
coexisting with disseminated superficial porokeratosis in a patient with psoriasis?

Literature







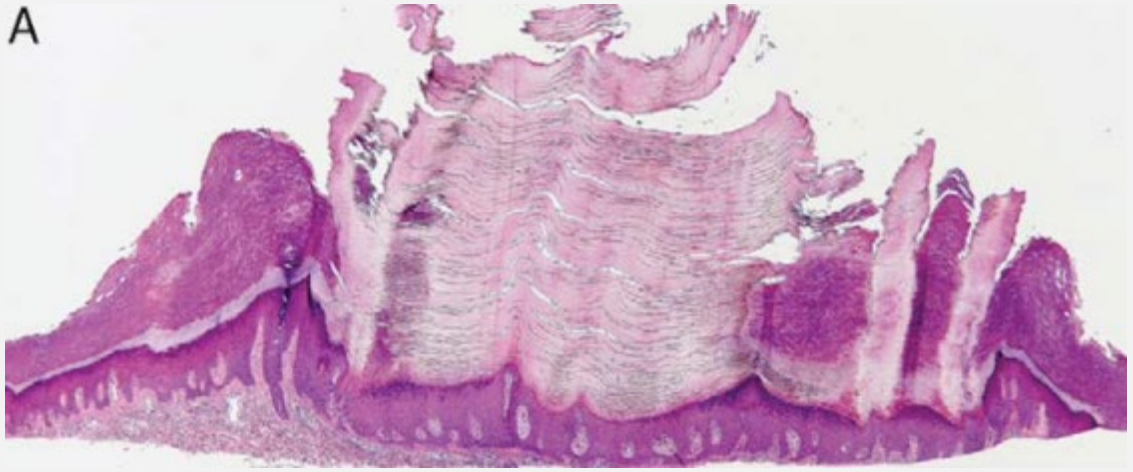


Xu X. et al. Multiple porokeratomas (porokeratotic acanthoma) coexisting with disseminated superficial porokeratosis: Clinical, dermoscopic and pathological observations, and review of the published work. J Dermatol 2020:47:787-791. **Zhang et al.** Multiple Verrucous Nodules and Masses in an Adolescent Boy With Porokeratoma: A Case Report and Literature Review. Pediatiric Dermatology Pediatric Dermatology, 2026; 43:132–136

Porokeratoma

First described by Walsh et al. in 2007:

- Solitary tumor like lesions with multiple columns of parakeratosis
- Mean age 57.2 (31-77) years
- More common in man (4.5:1)



Zhang et al. in 2026:

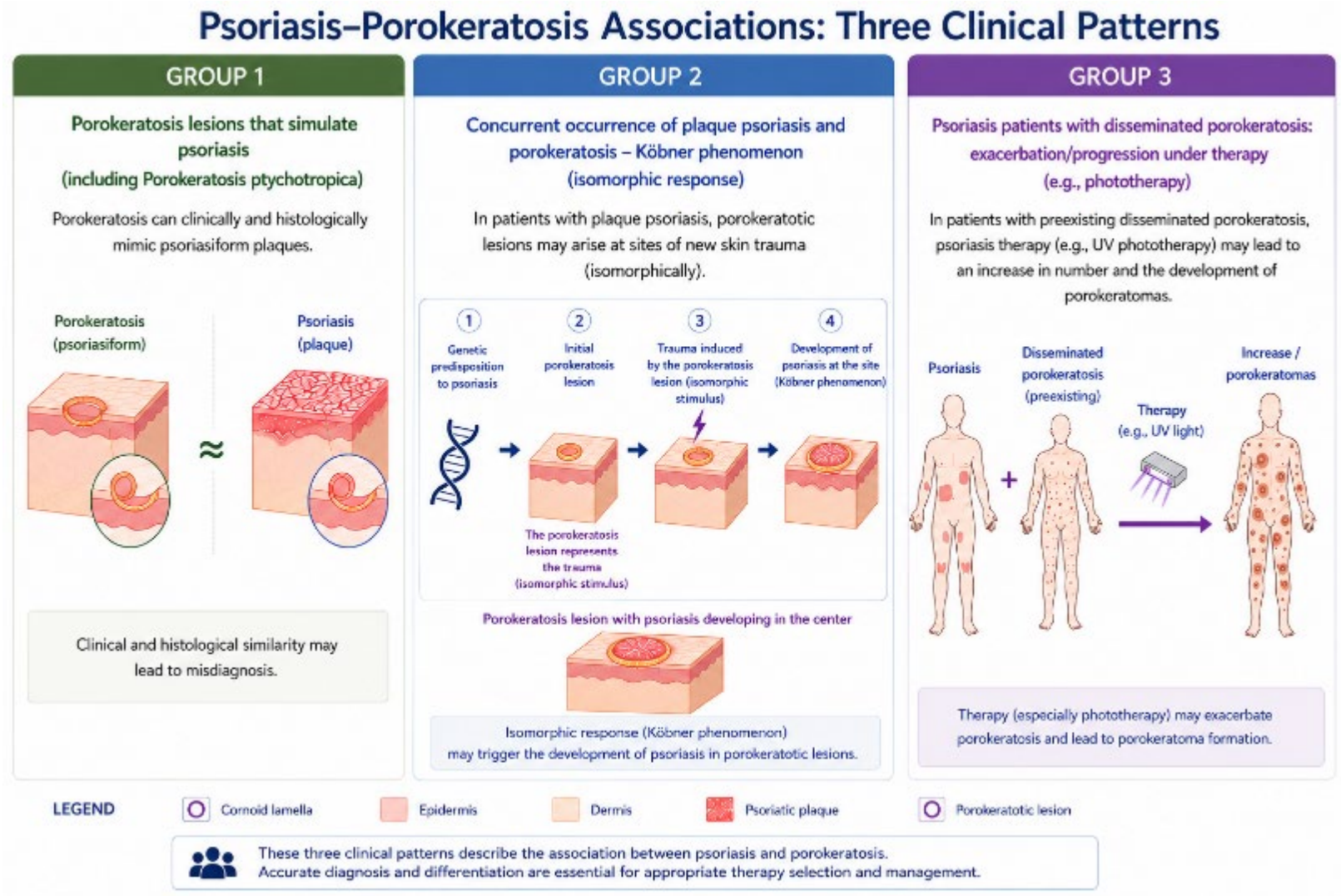
- Multiple porokeratomas with tumor-like lesions

TABLE 1 | Clinical data of all published cases of porokeratoma.

Case	Age	Sex	Duration	Location	Number of lesions	Concomitant diseases	Treatment	Regrowth (F/u, mo)
1 [2]	77	M	NG	Right intergluteal cleft	1	None	Complete excision in 10/11 cases. Cryotherapy in 1/11 cases	No (67)
2 [2]	77	F	6 months	Left posterior leg	1	None		No (66)
3 [2]	61	M	NG	Right lower lip	1	None		No (64)
4 [2]	55	M	NG	Right forearm	1	None		No (54)
5 [2]	37	M	2 months	Right hand	1	None		No (37)
6 [2]	31	M	5 years	Right thumb	1	None		No (37)
7 [2]	48	M	NG	Right posterior calf	1	None		No (30)
8 [2]	31	M	NG	Left chest	1	None		No (14)
9 [2]	74	F	NG	Left anterior thigh	1	None		No (8)
10 [2]	68	M	NG	Left buttock	1	None		No (3)
11 [2]	70	M	NG	Forehead	1	None		No (1)
12 [1]	61	M	1.5 years	Right shin	2	Ankylosing spondylarthritis	Excision	NG
13 [3]	78	M	4 years	Left buttock	6	Poliomyelitis, Ankylosing spondylarthritis	NG	NG
14 [4]	35	M	1 years	Both buttocks	1	None	Excision + acitretin 10 mg daily	No (15)
15 [5]	50	M	1 years	Inner aspect of the left knee	1	None	Total excision	No (6)
16 [6]	52	M	NG	Legs and pubis	1 (4 lesions, 3 were diagnosed as verruca vulgaris)	Chronic lymphoid leukemia, graft versus host disease	Excision	NG

Walsh SN et al. Porokeratoma. Am J Surg Pathol. 2007;31:1897-1901. **Zhang et al.** Multiple verrucous nodules and masses in an adolescent boy with porokeratoma: A case report and literature review. Pediatric Dermatology. 2025; 43:132-136.

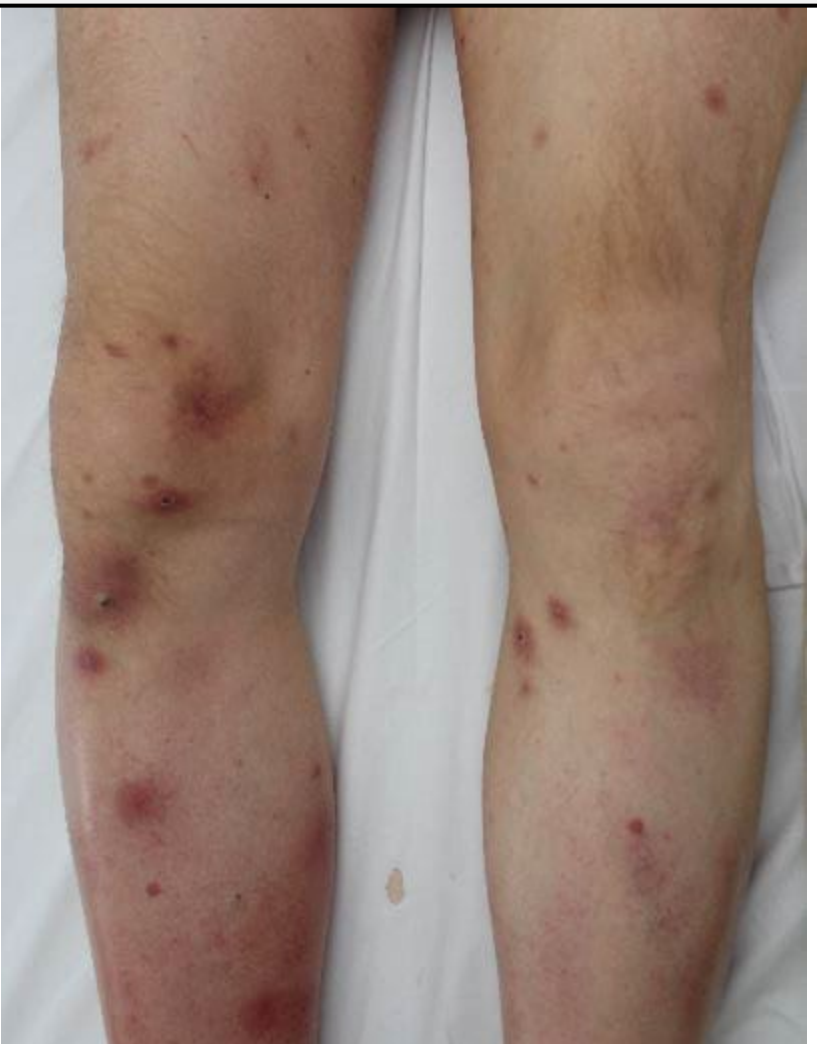
Psoriasis and Porokeratosis

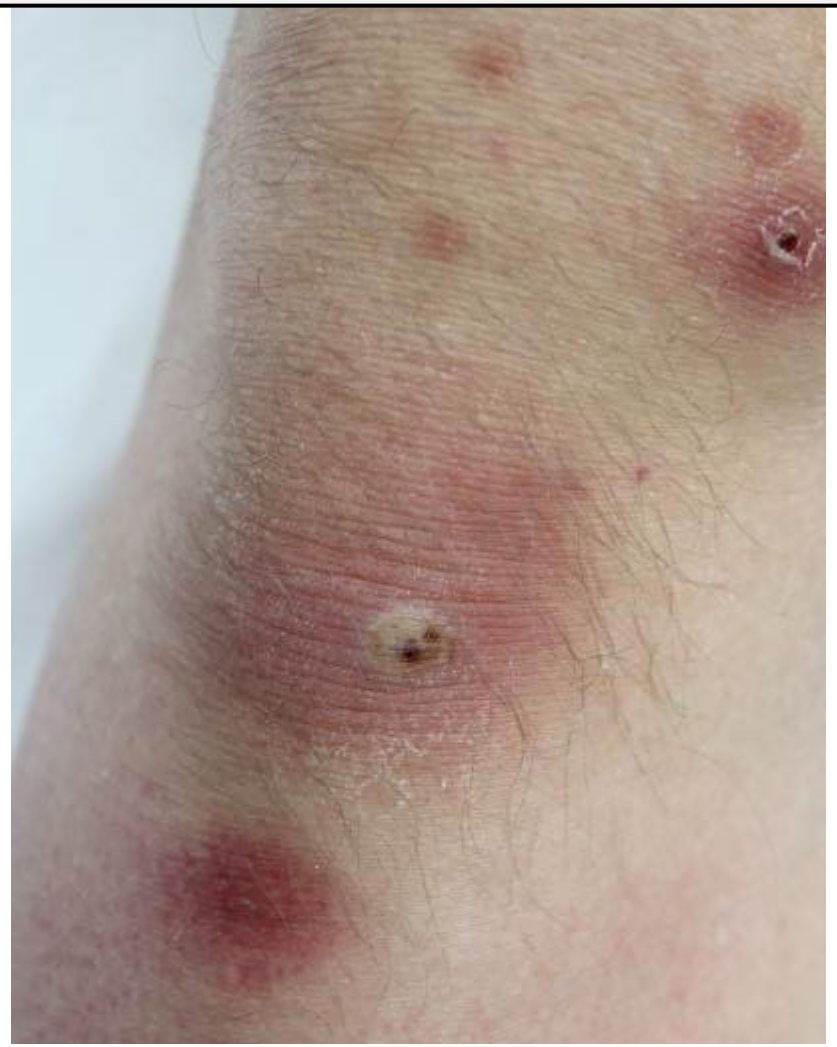


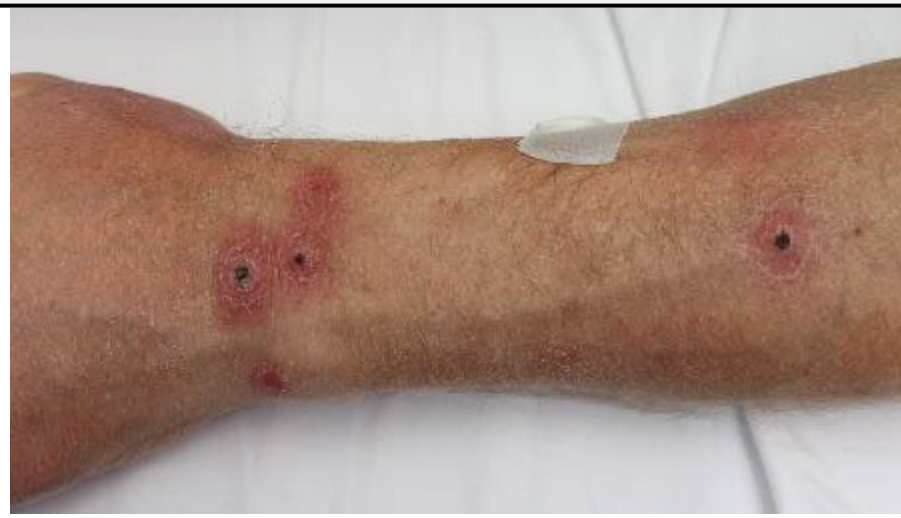
created with ChatGPT/OpenAI, 2026

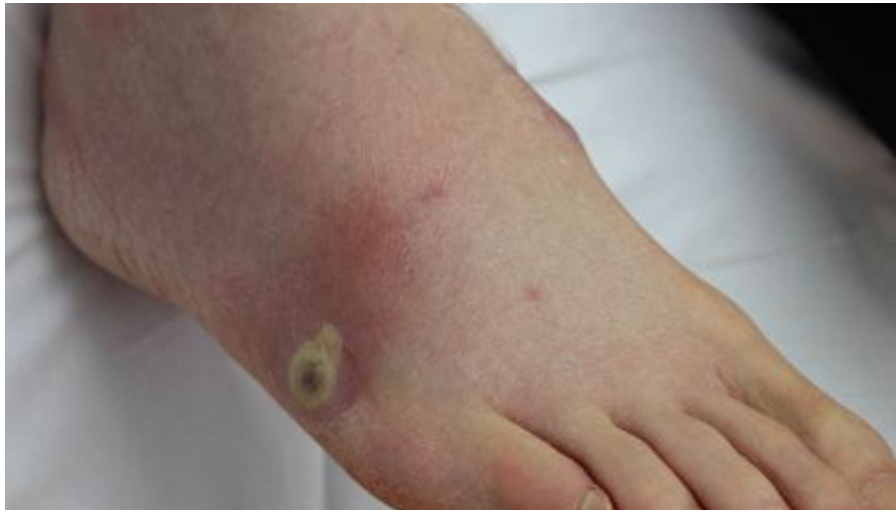
Case 7

Clinical presentation:



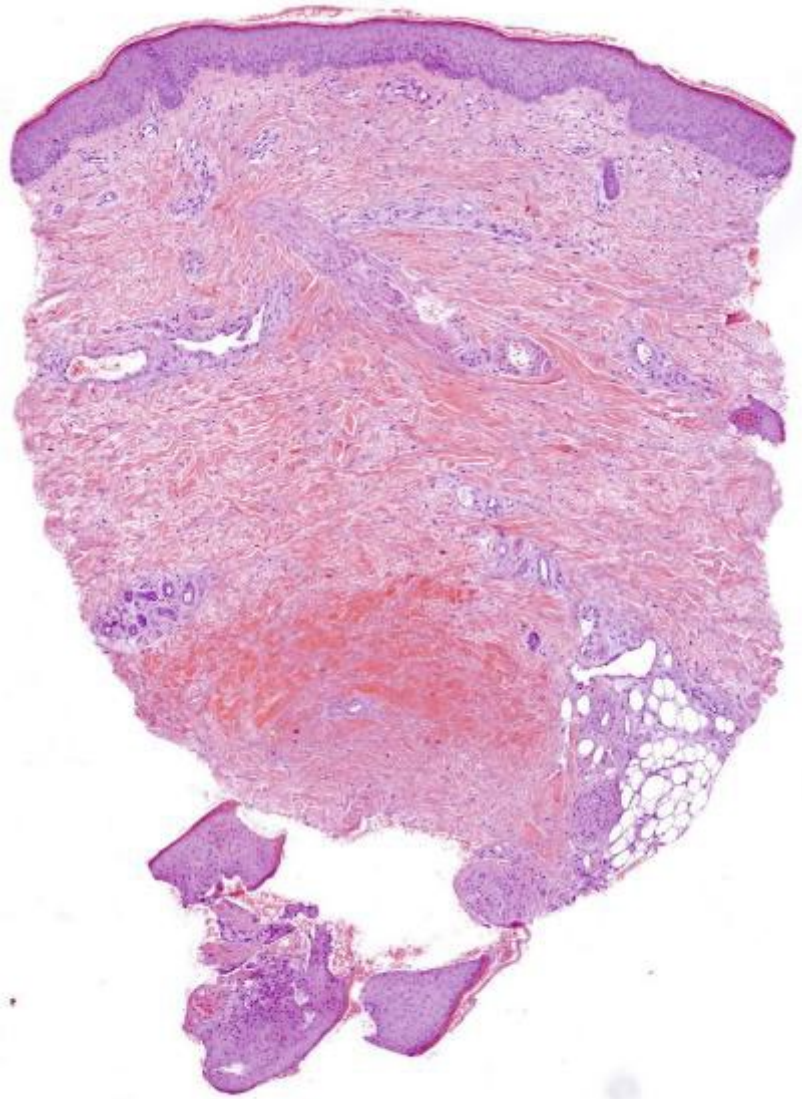


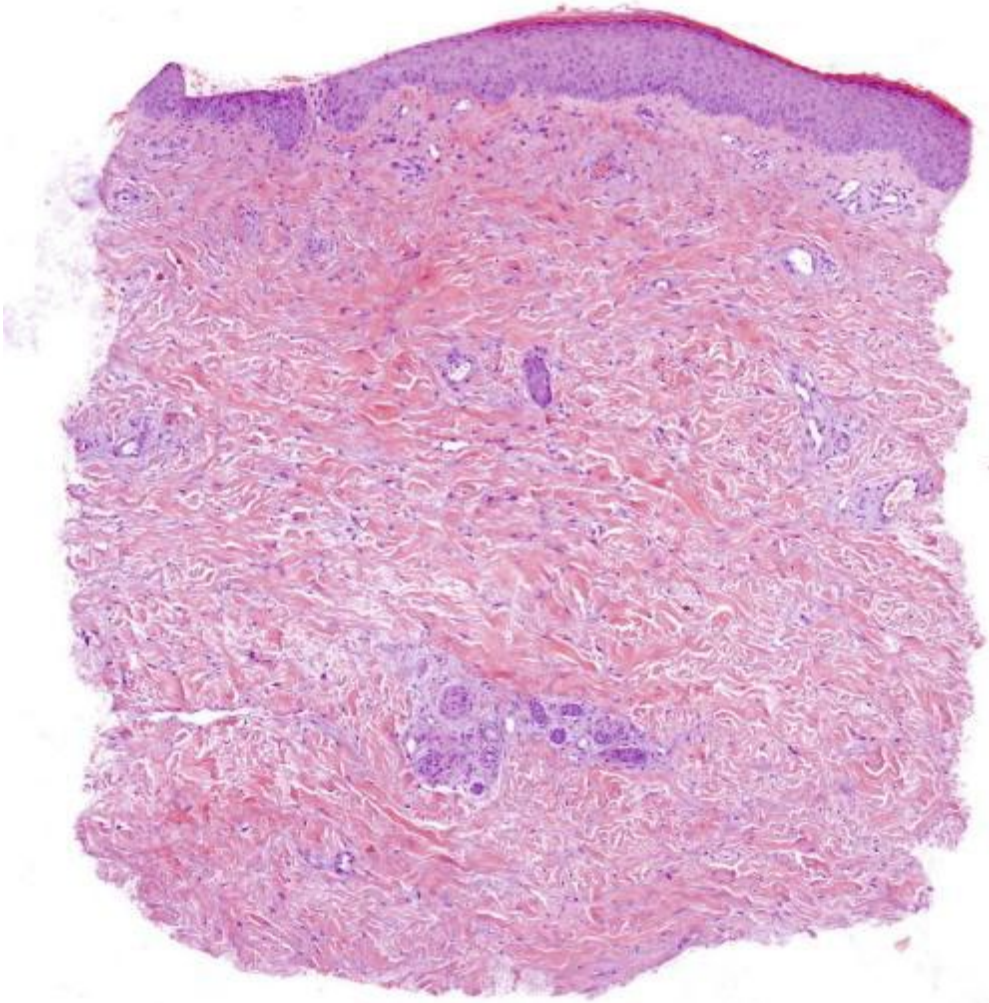


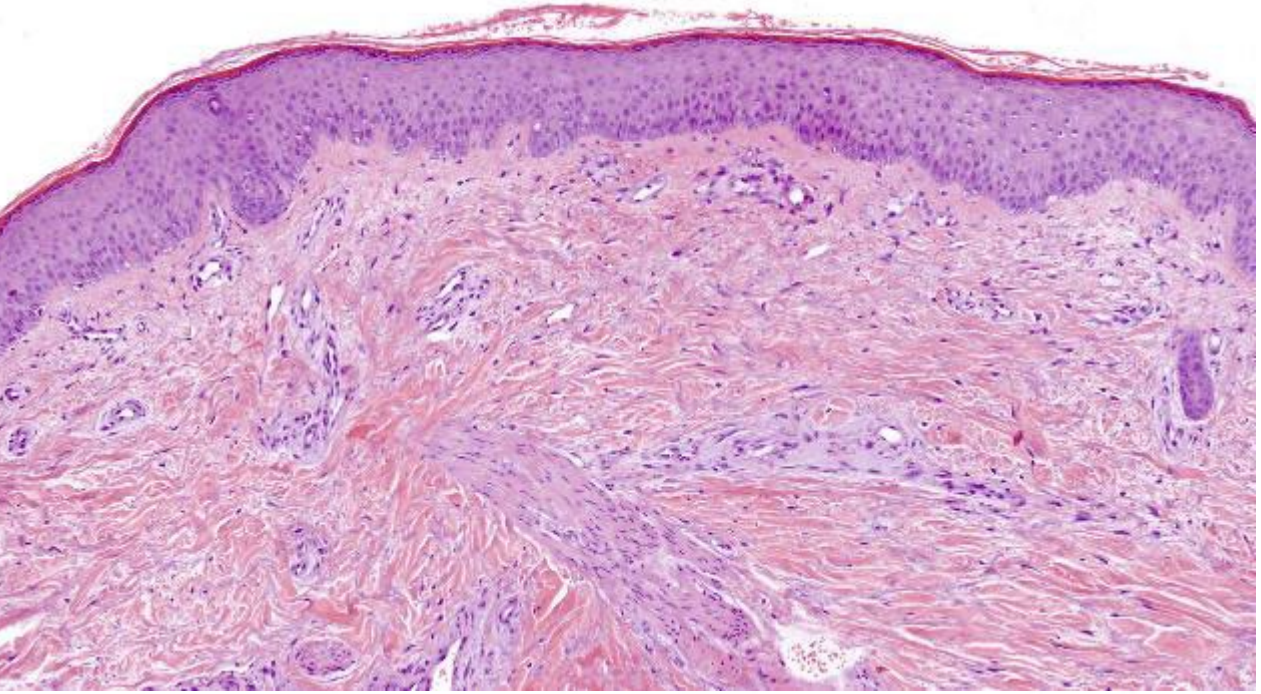


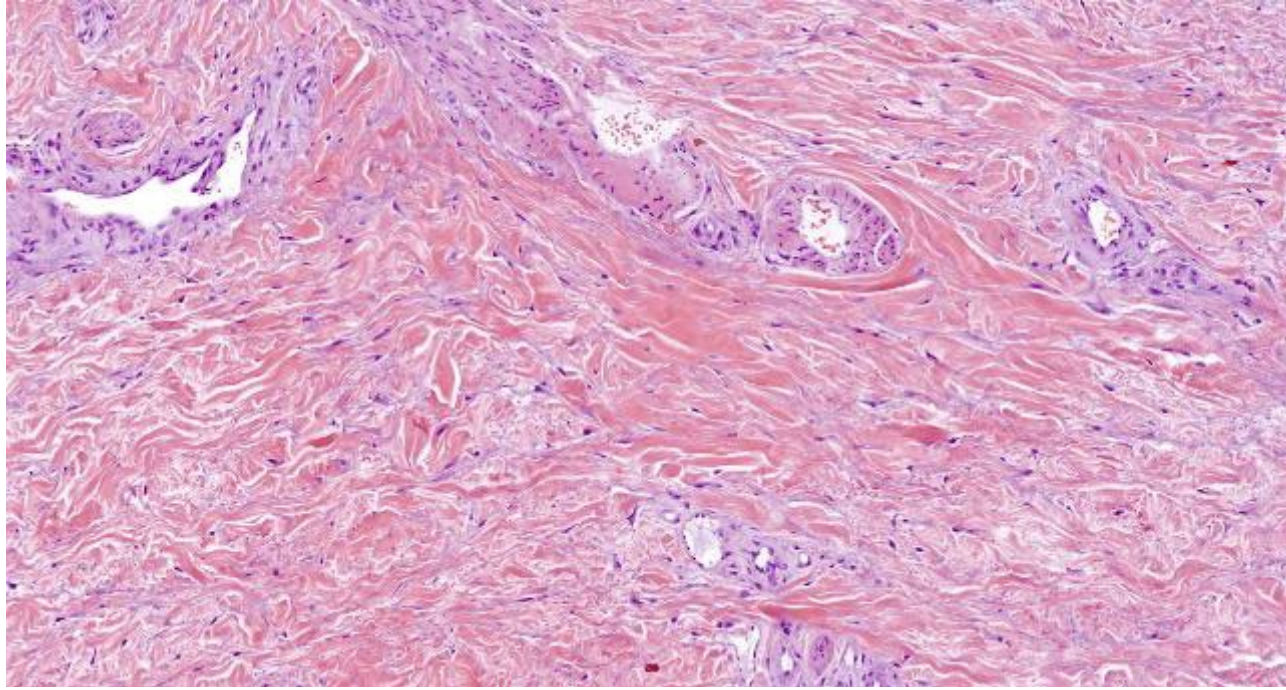
immunocompromised patient after allo-HSCT because of acute myelod leukemia with CBFM::MYH11 fusion

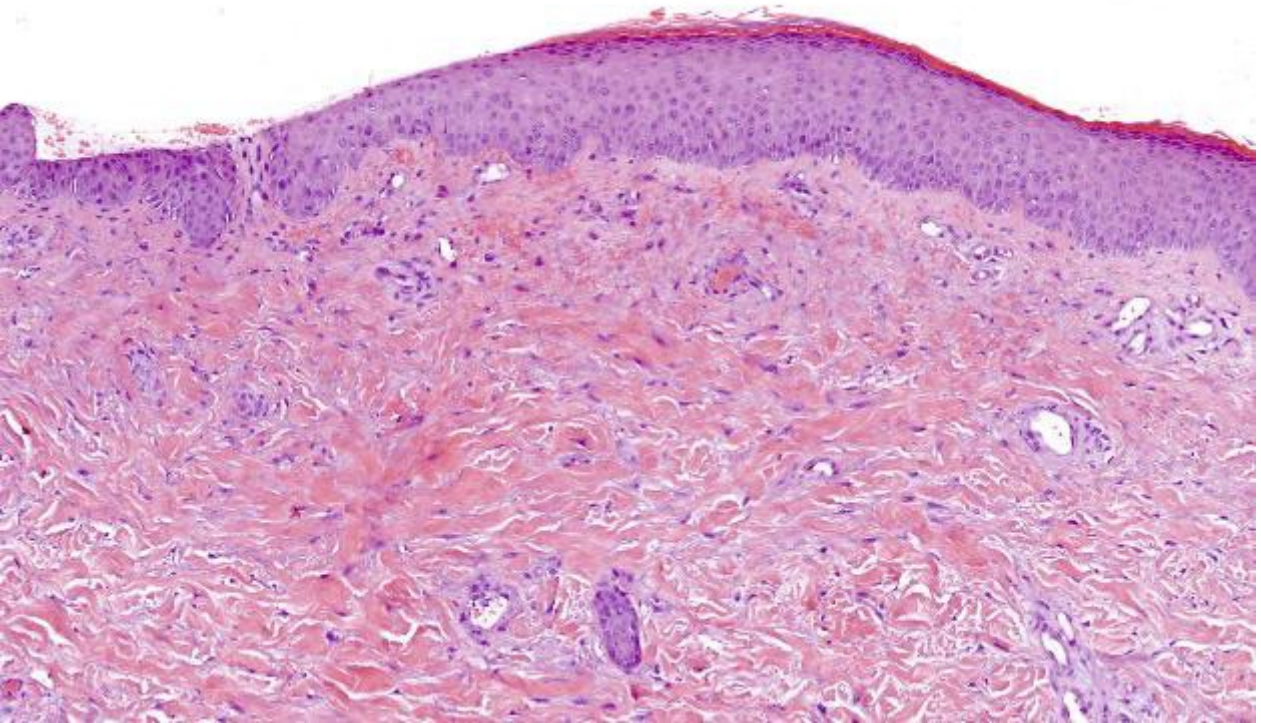
Histology:

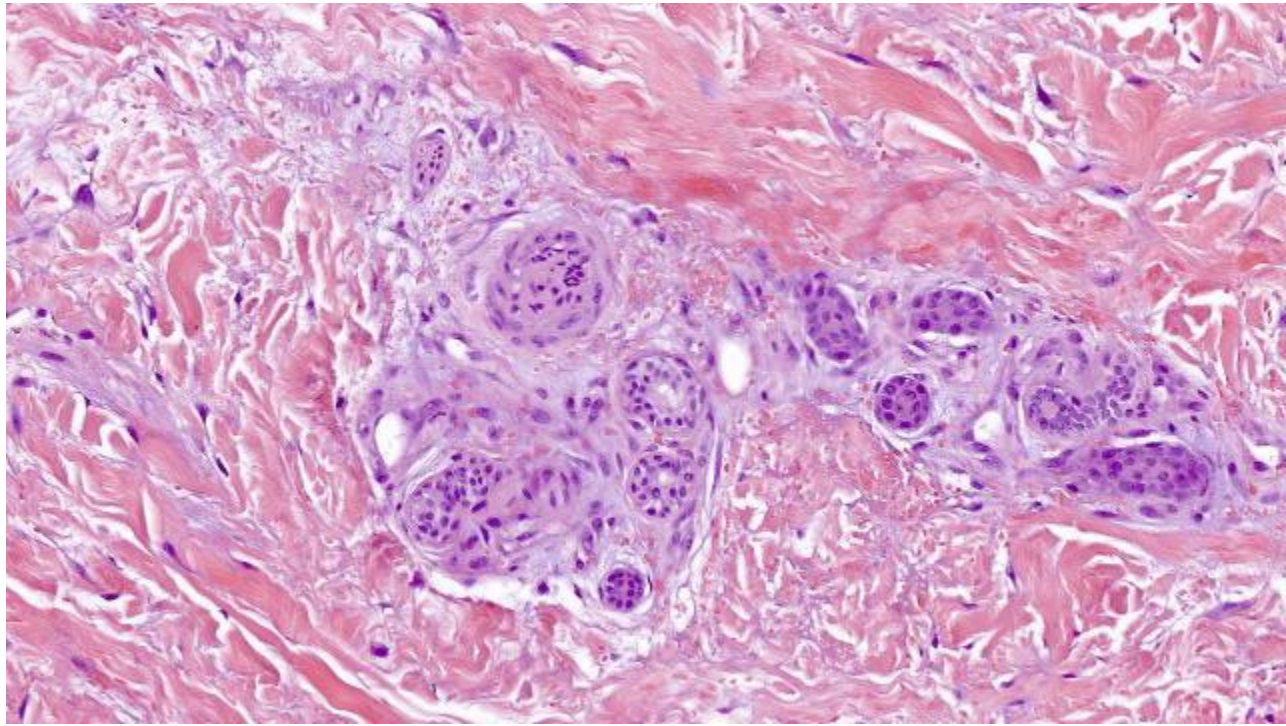




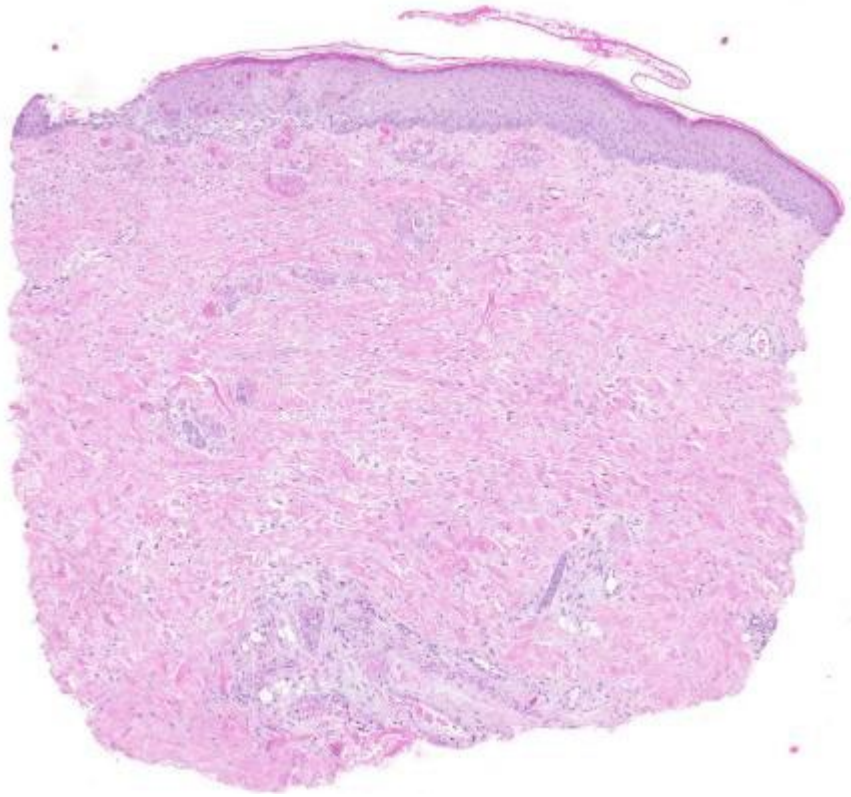


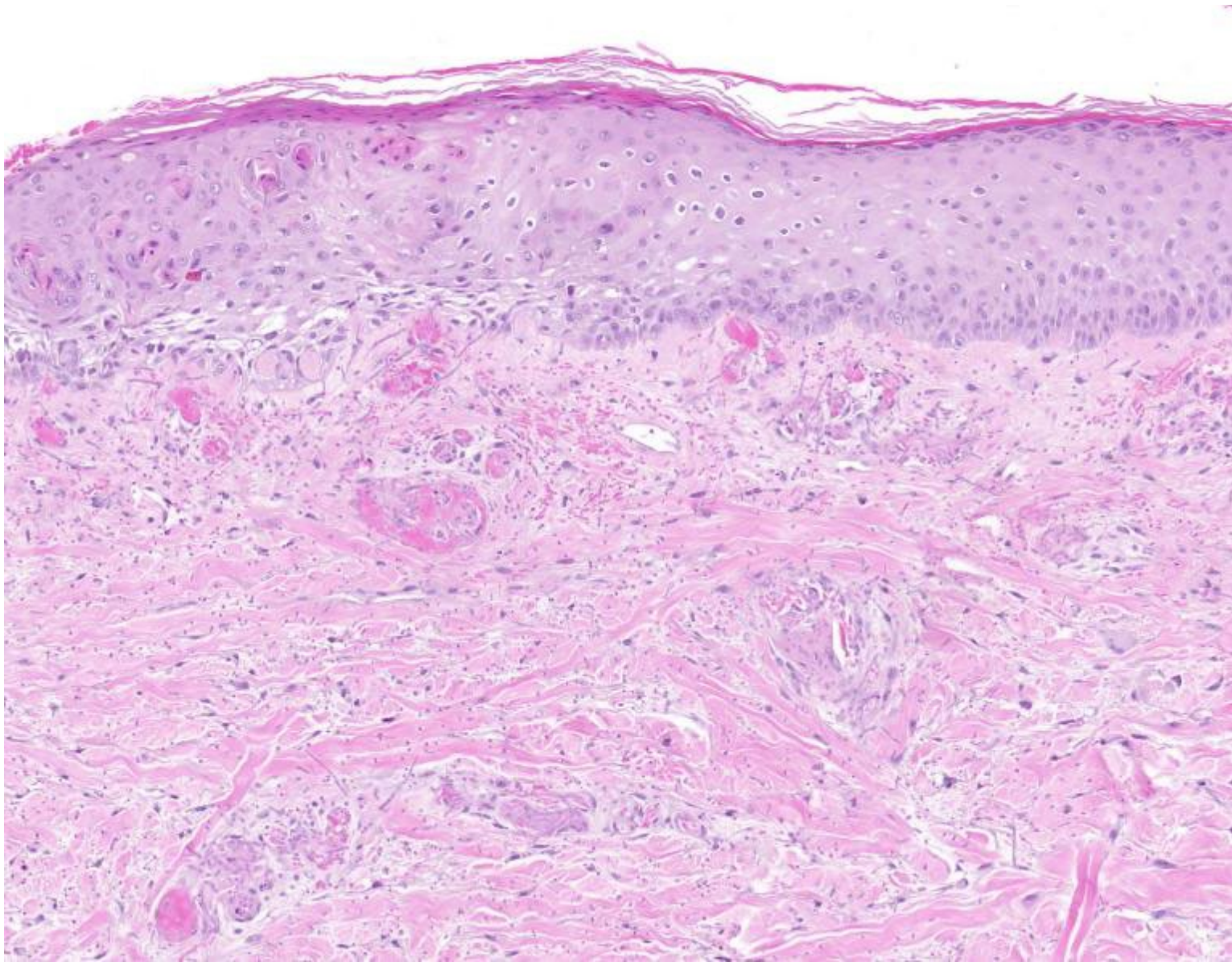


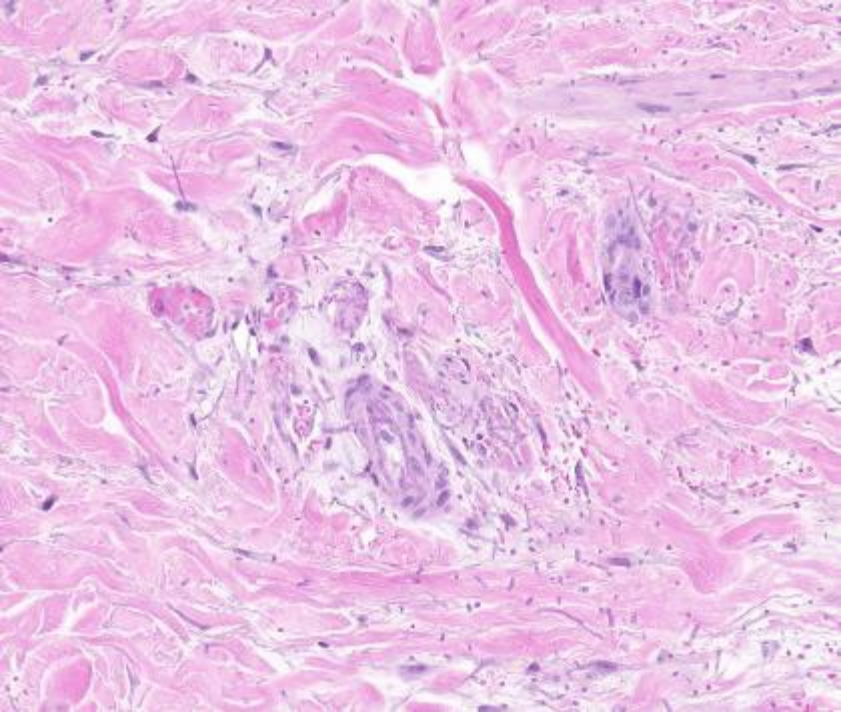
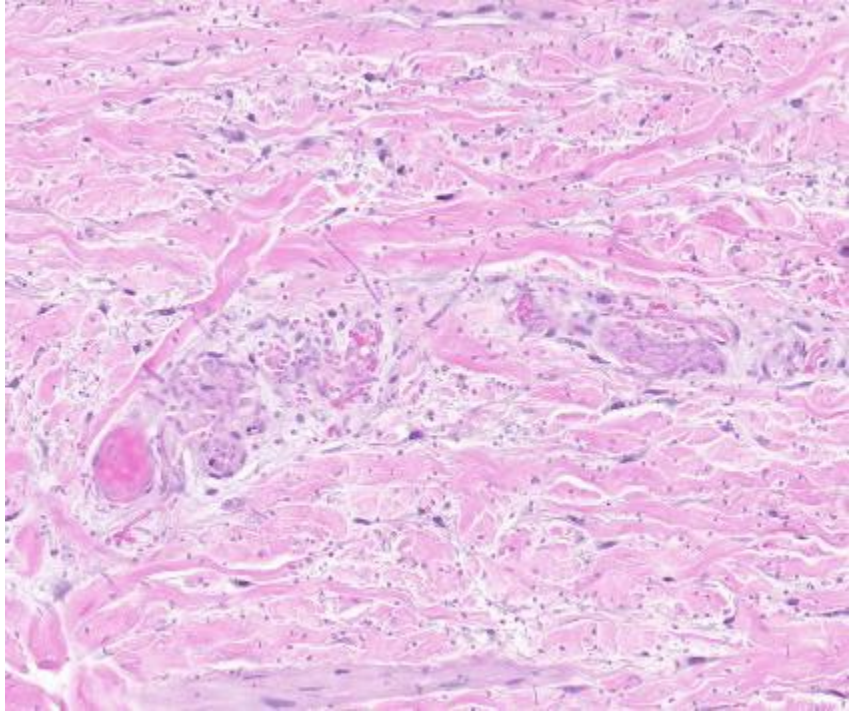
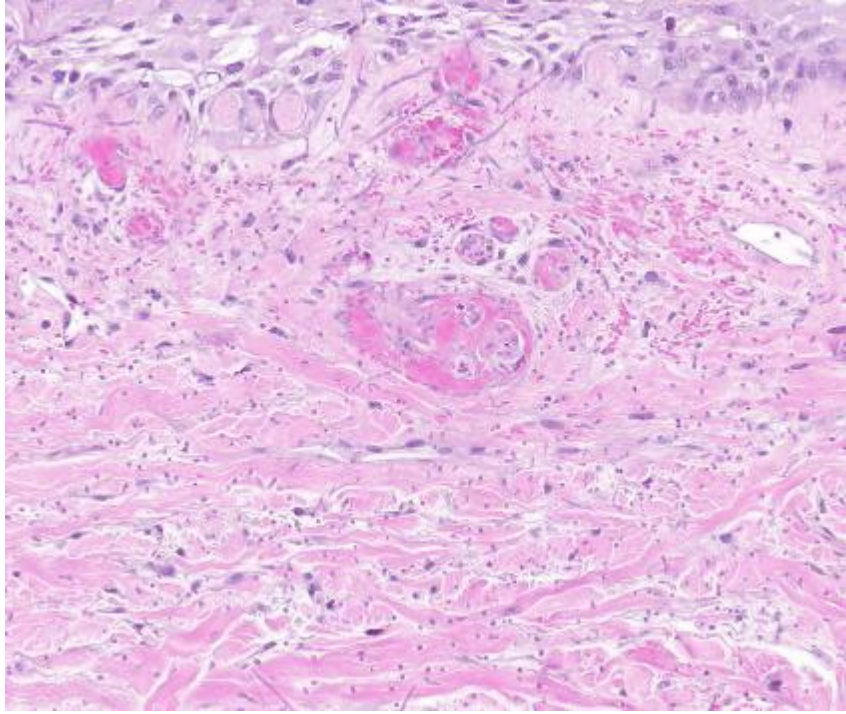
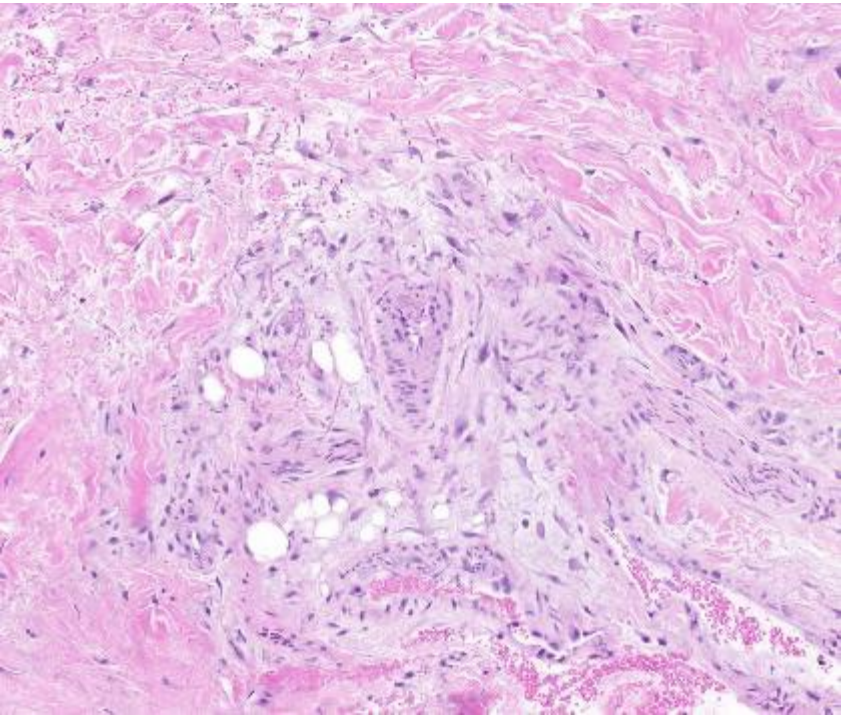
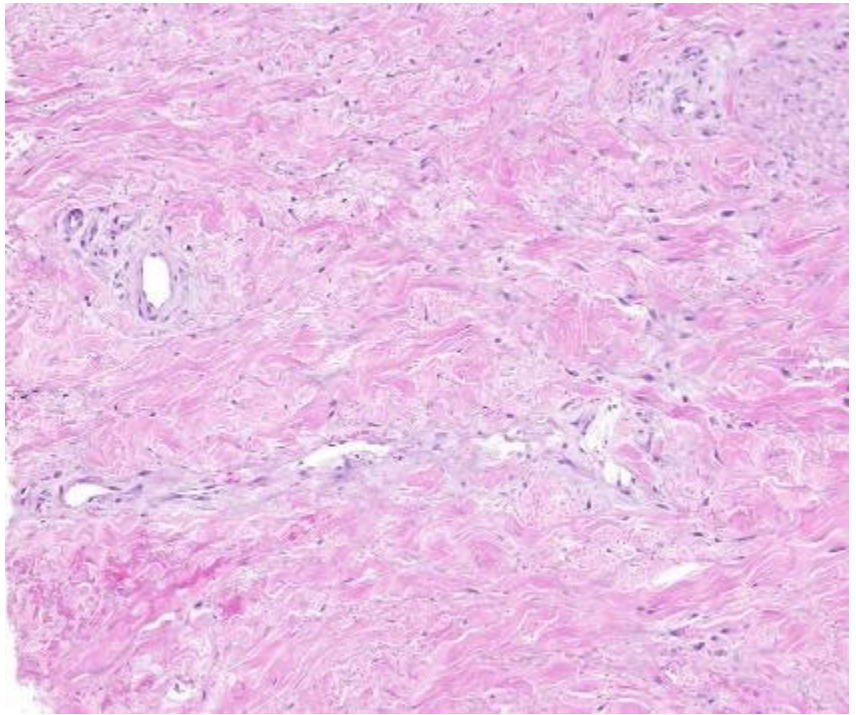
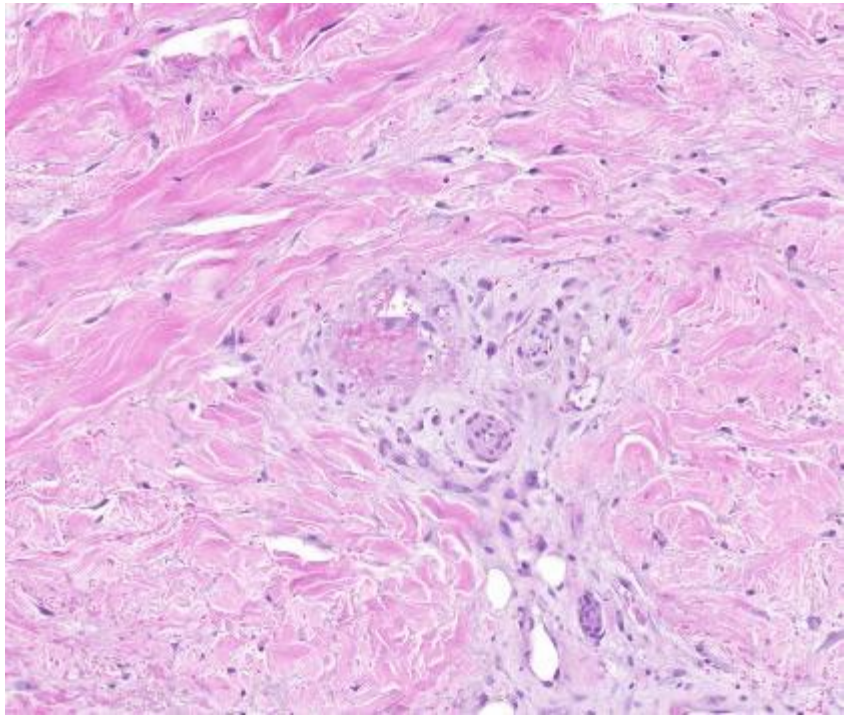


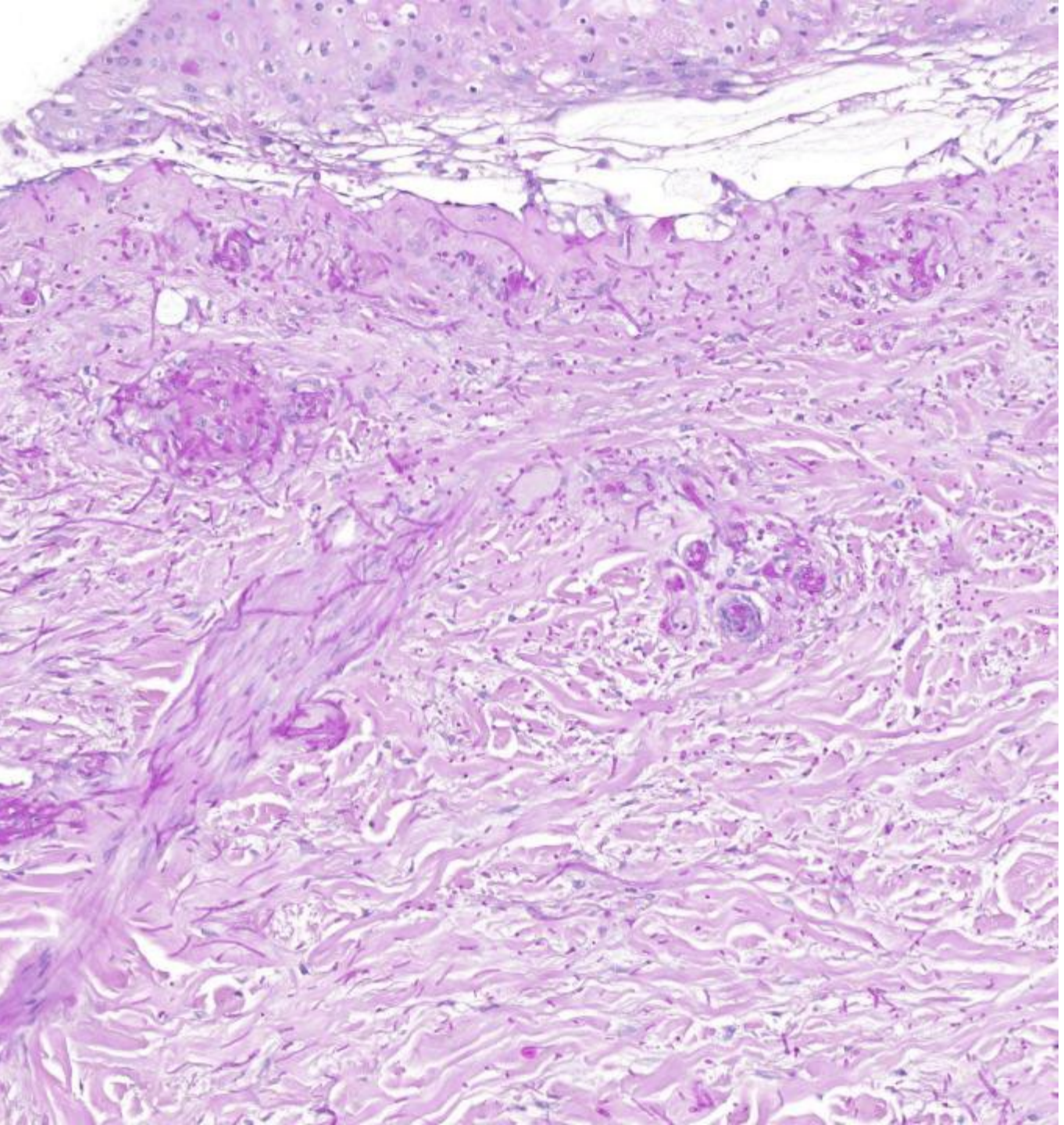


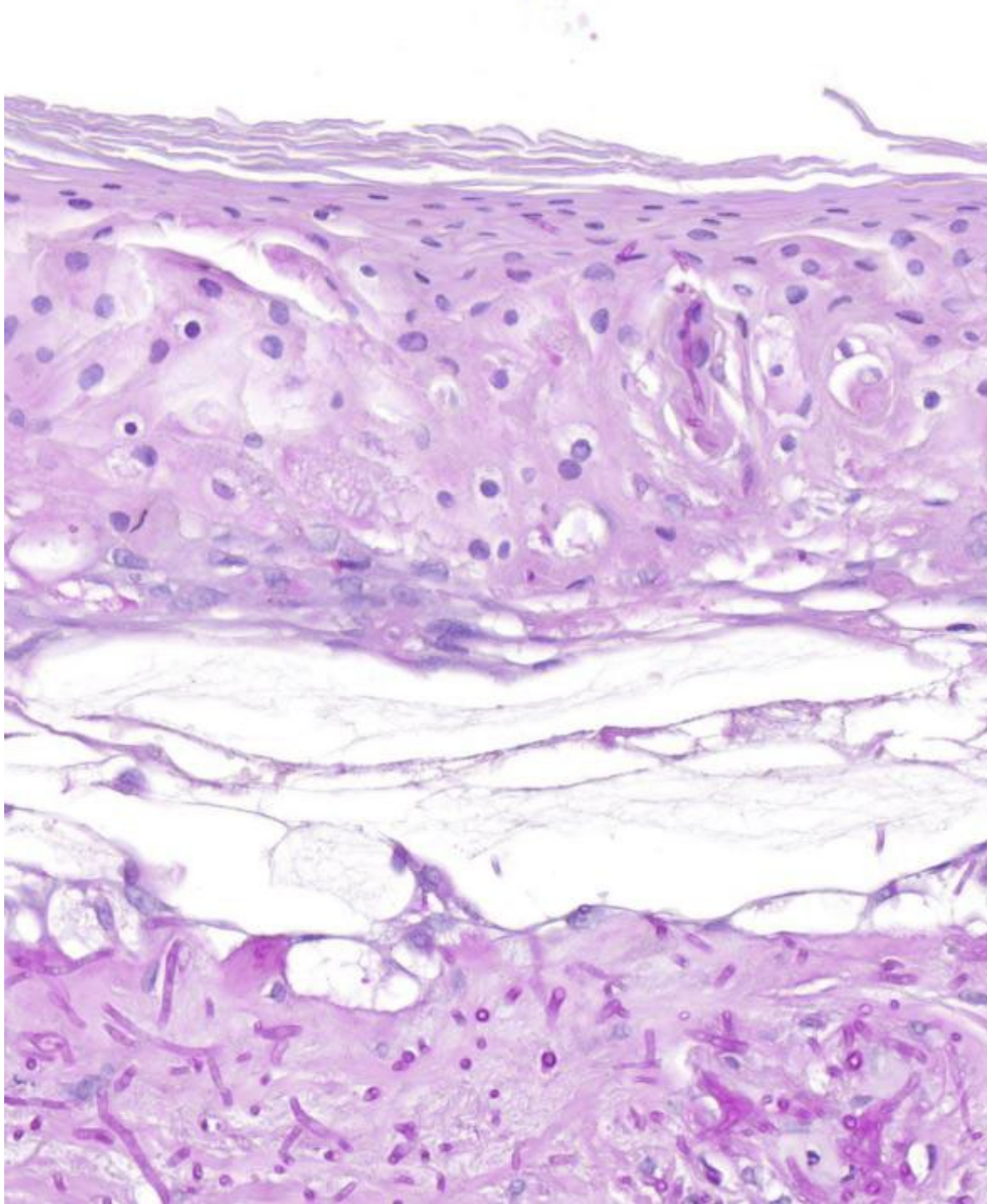
Histology – step sections:

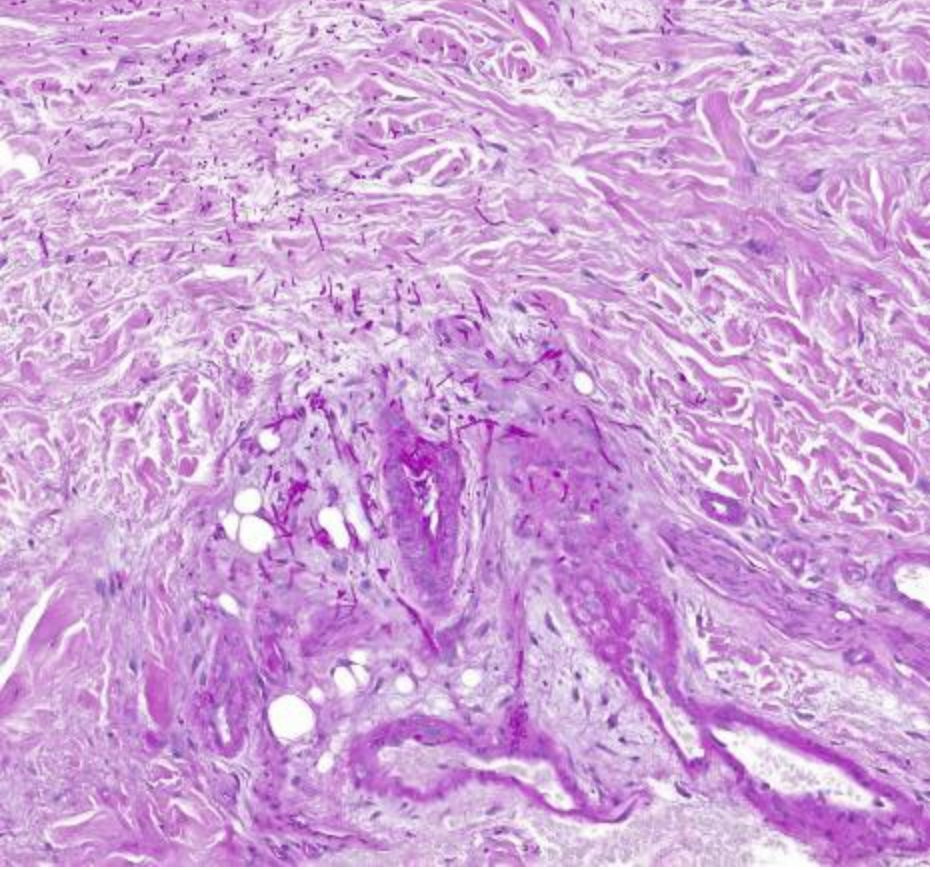
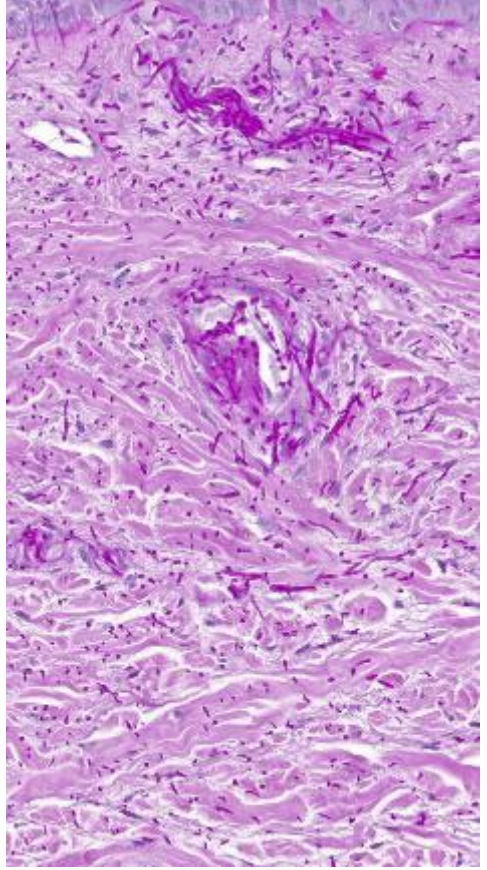
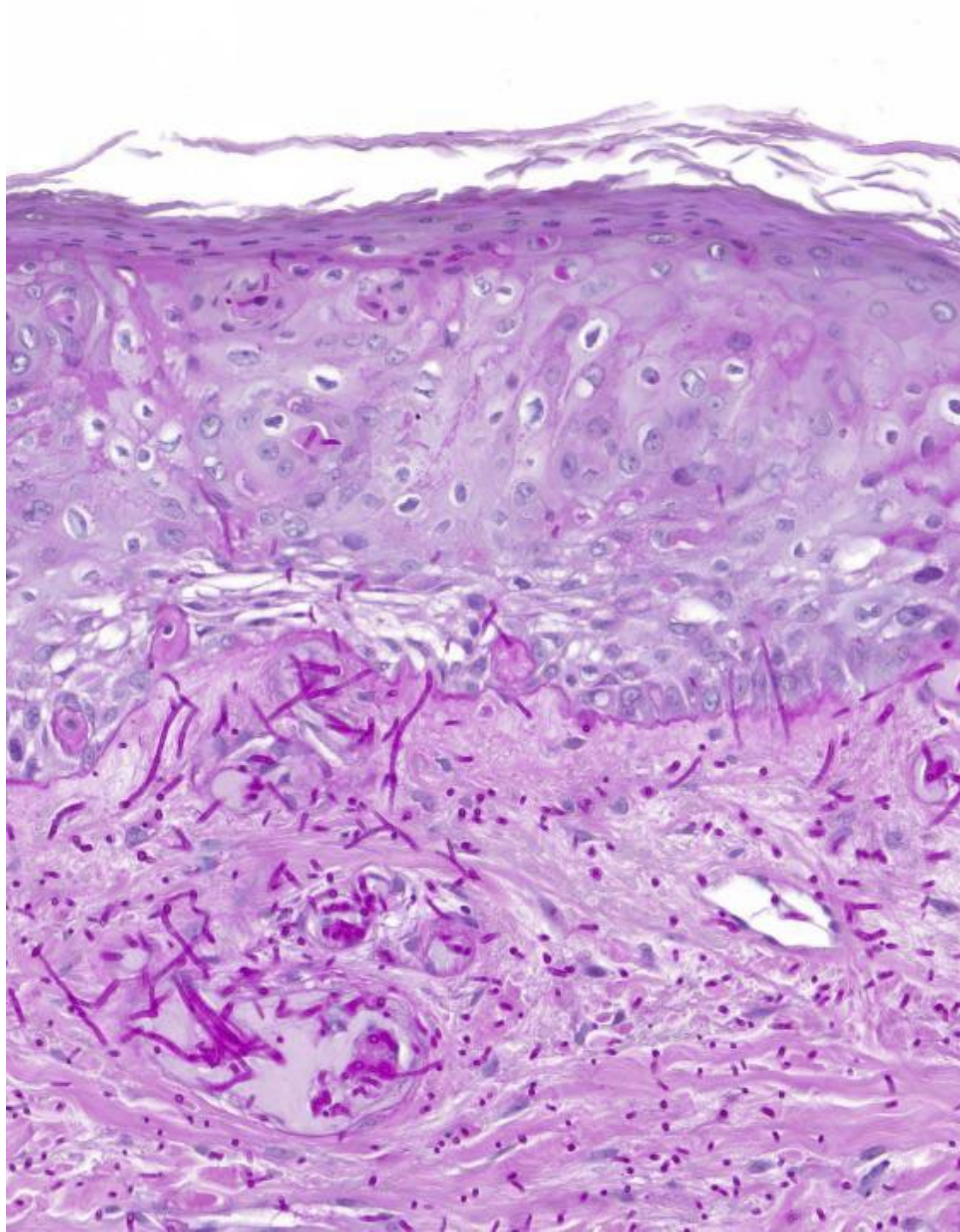
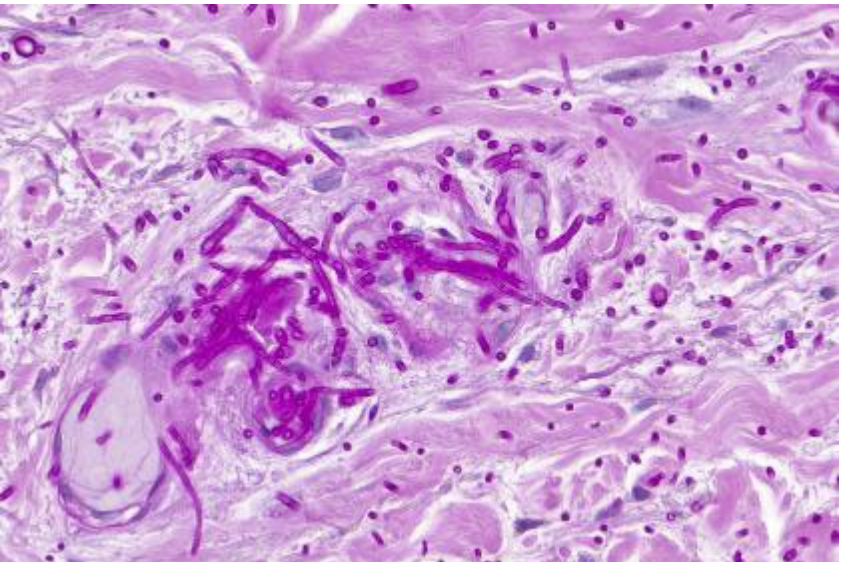
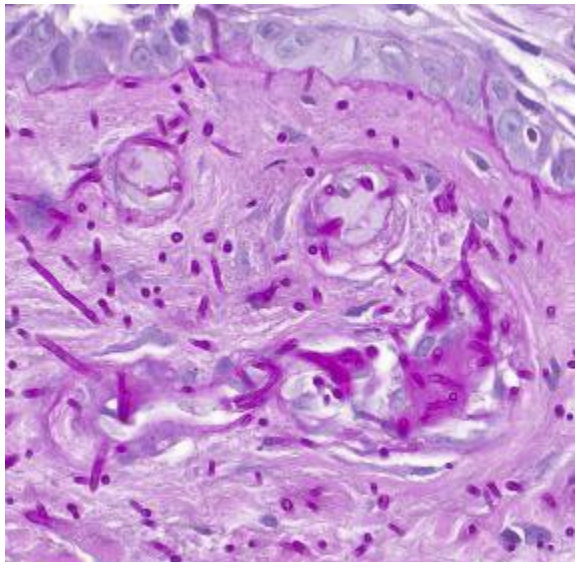


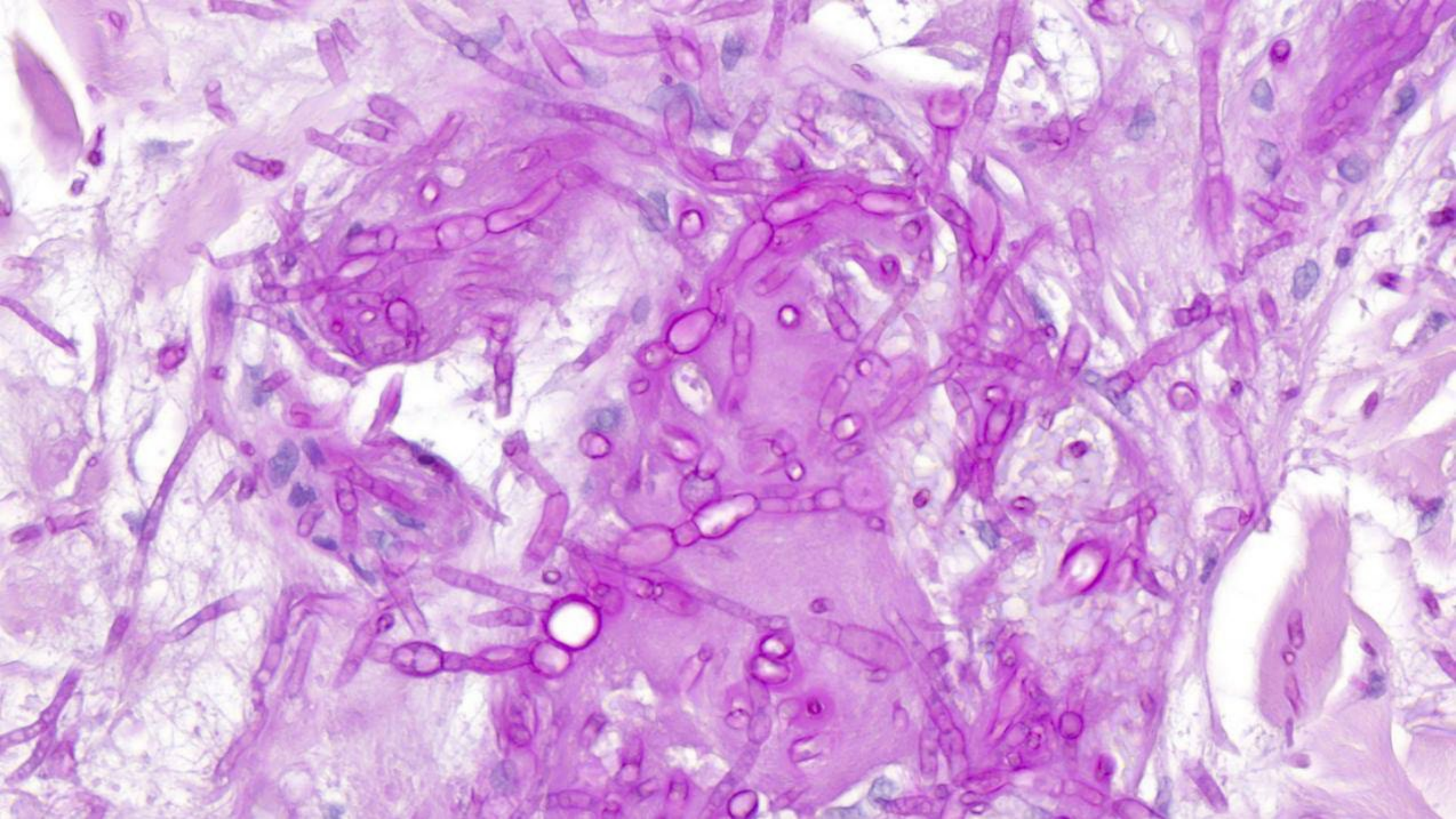








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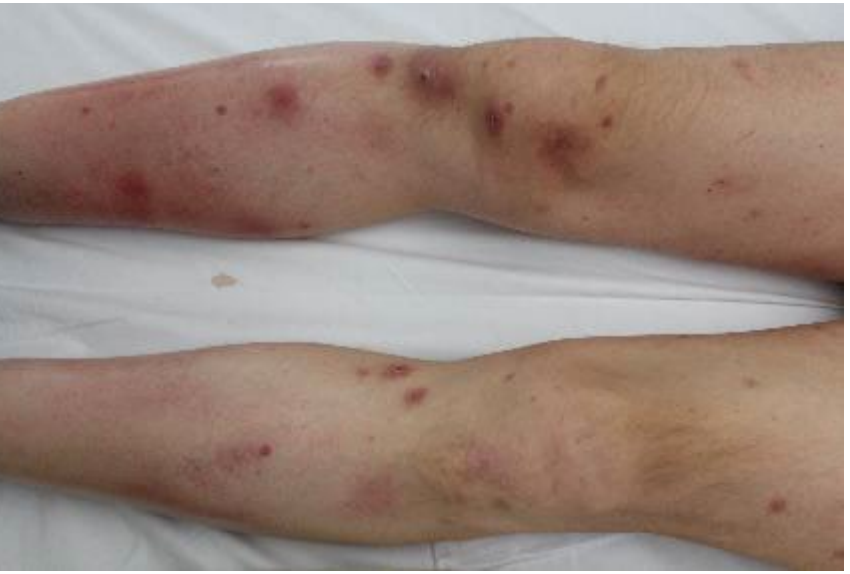
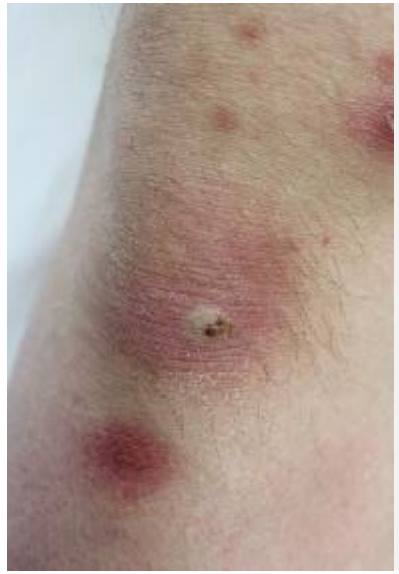
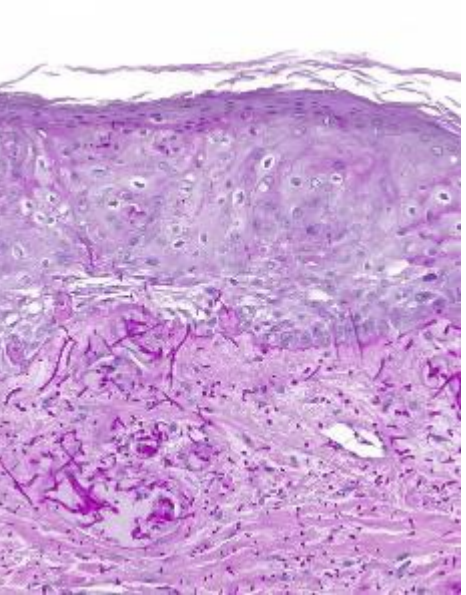
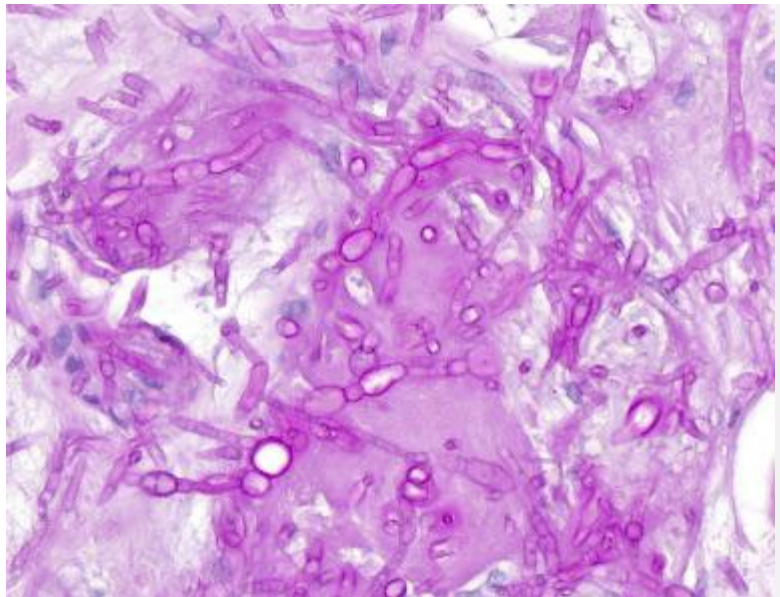




Clinical presentation:

- Necrotic papules
- Pustules
- Allogenic HSCT
- Neutropenia





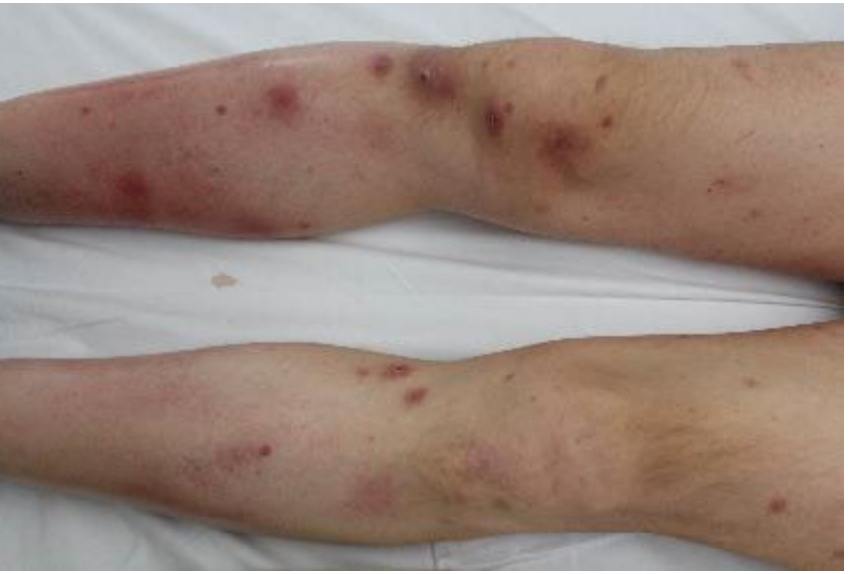
Histology:

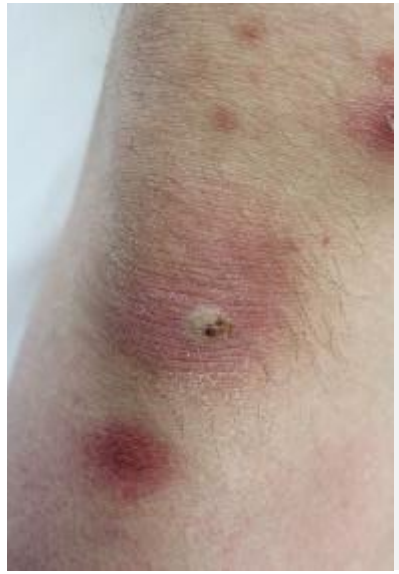
- Angioinvasive septated hyphae

diagnosis

?

Summary of the findings:





Clinical presentation:

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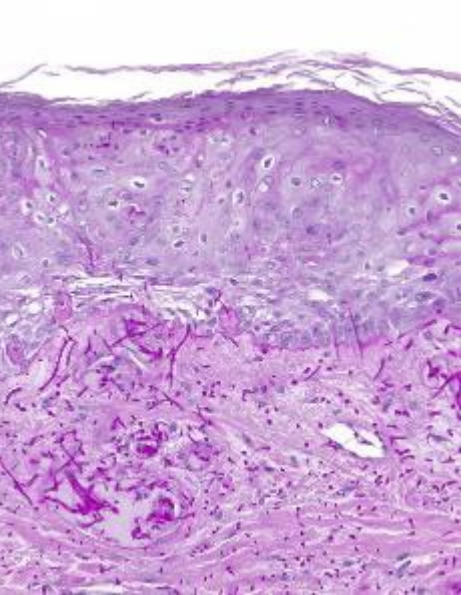
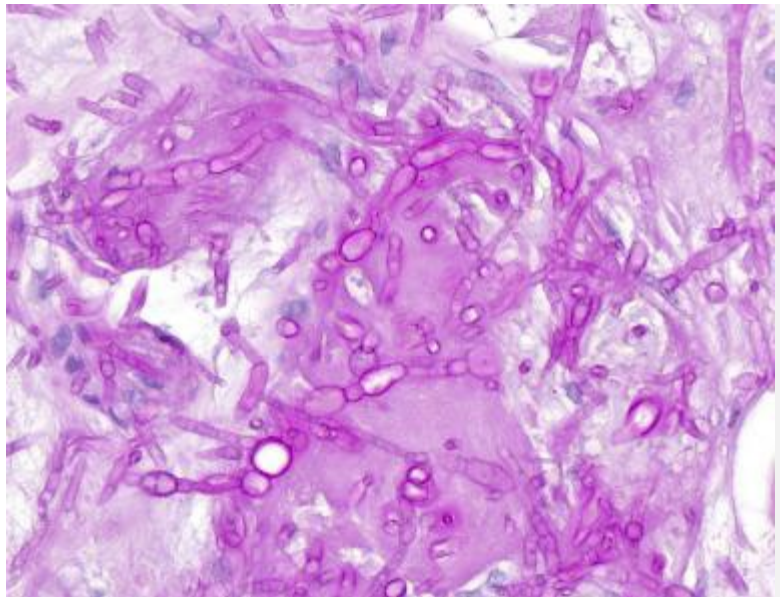




Histology:

- Angioinvasive septated hyphae

disseminated Fusarium infection
in an immuno-compromised patient

Fusarium infection in immunocompromised patients

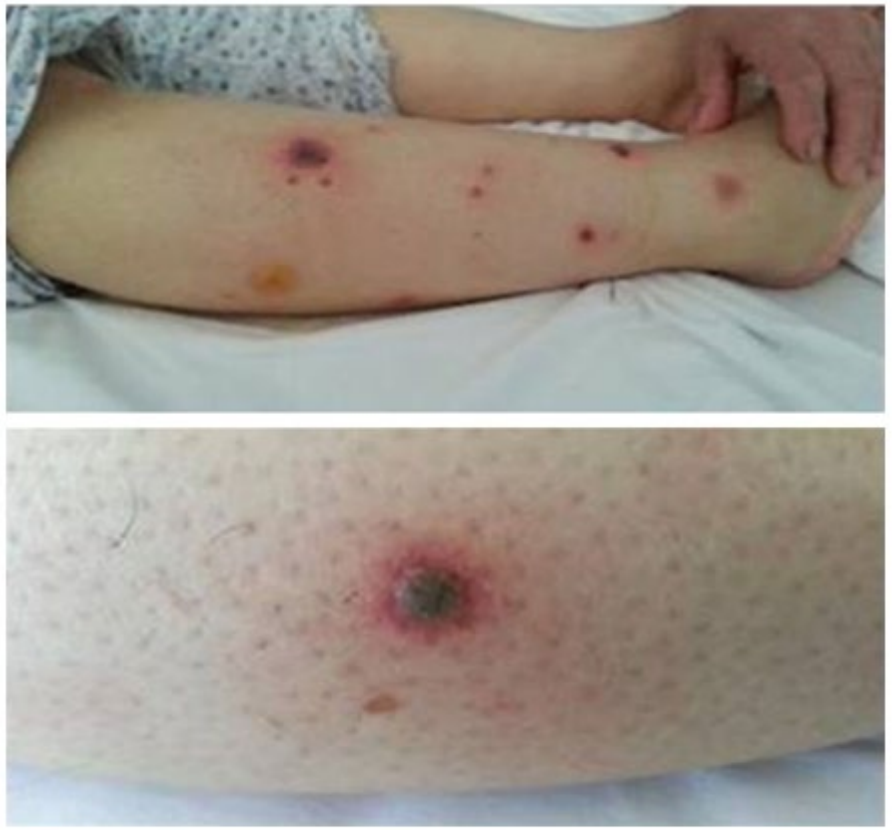
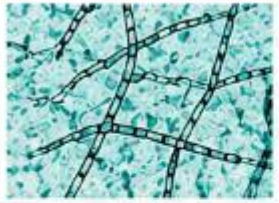
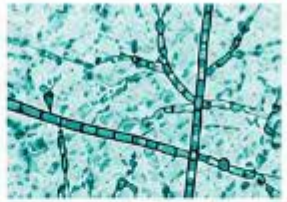
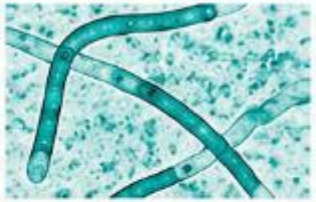
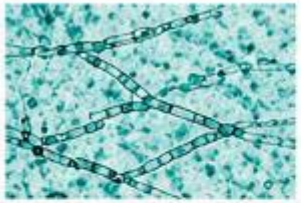
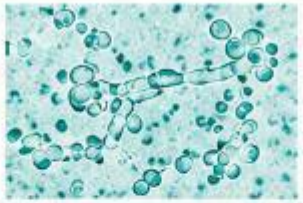
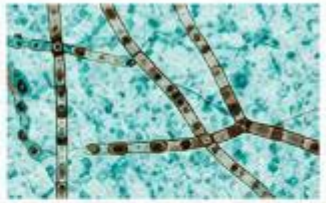
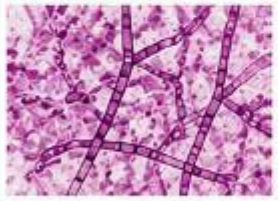
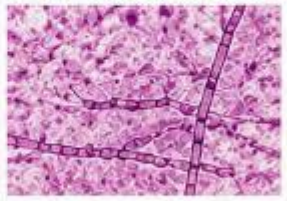
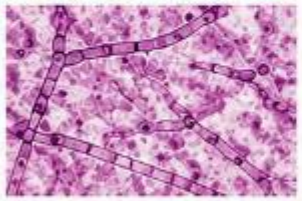
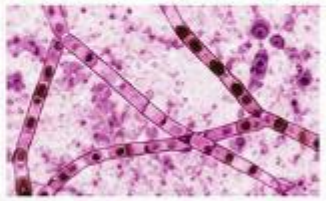
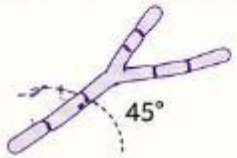
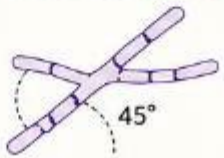
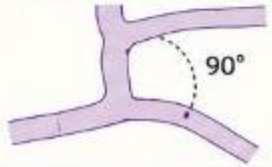
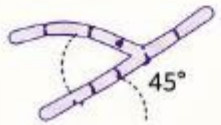
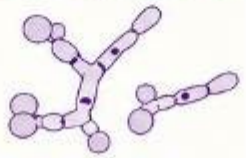


Fig. 1 Necrotic maculopapular skin lesions of the patient

- Fusarium spp. either penetrate the skin barrier or cause a pulmonary infection by airborne transmission
- Skin lesions are often the first sign of a disseminated infection and are commonly seen in the early stages of the disease.
- In 40 % blood cultures are positive, with skin lesions in 56 %
- Necrotic papular skin lesions are typical
- Especially found in patients with significant neutropenia.
- Fusarium species are refractory toward antifungals
  Treatment: Amphotericin B and voriconazole
- The prognosis of disseminated fusariosis is poor and is determined largely by the degree of immunosuppression

Ersal T et al. Fatal Disseminated Infection with Fusarium petroliphilum. Mycopathologia. 2014 Online

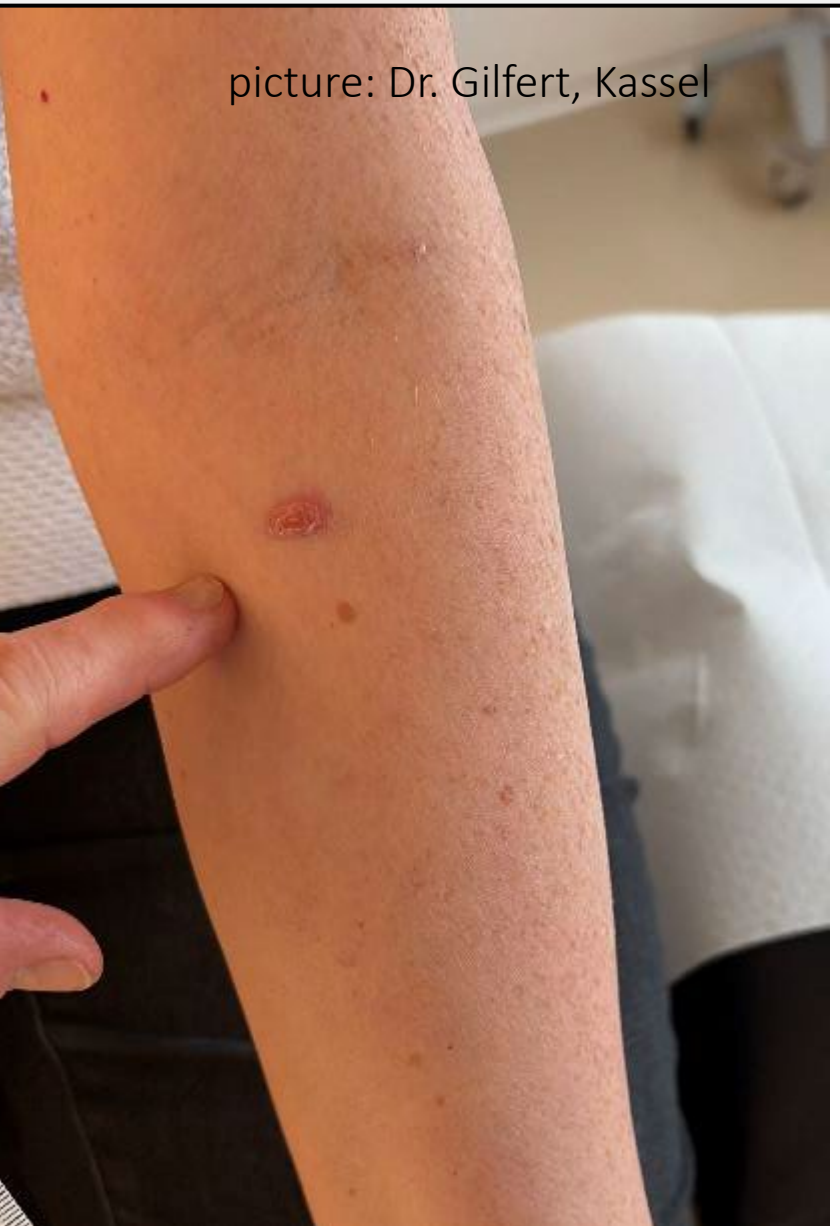
Pathogen	Aspergillus spp.	Fusarium spp.	Mucorales (e.g. Rhizopus, Mucor)	Scedosporium spp.	Candida spp.	Dematiaceous fungi (e.g. Alternaria)
Histology (GMS stain)						
Histology (PAS stain)						
Schematic representation						
Hyphal characteristics	Narrow (3–6 µm) septate regular dichotomous branching (~45°)	Narrow (3–8 µm) septate frequent dichotomous branching (~45°)	Broad (5–20 µm) rarely septate to aseptate irregular branching, often at right angles (~90°)	Narrow (2–6 µm) septate regular dichotomous branching (~45°)	Yeast cells (3–8 µm), budding, pseudohyphae	Narrow to moderately broad septate often pigmented (brown–black)
Typical risk patients	Neutropenia, AML, Hematopoietic stem cell transplantation, high-dose steroids	Severe neutropenia, AML, hematopoietic stem cell transplantation	Diabetes (especially ketoacidosis), neutropenia, iron overload, transplantation	Stem cell transplantation, neutropenia, chronic granulomatous disease	ICU, central venous catheters, parenteral nutrition, neutropenia	Transplantation, immunosuppression, traumatic inoculation
Typical clinical manifestations	Invasive pneumonia, sinusitis, CNS involvement, disseminated infection	Fungemia, disseminated infection, necrotic cutaneous lesions	Rhino-orbital-cerebral, pulmonary, cutaneous, disseminated	Pulmonary infections, cutaneous lesions, disseminated	Candidemia, disseminated candidiasis (liver, spleen, kidney, eye, endocardium)	Cutaneous lesions, disseminated infection, CNS involvement
Frequency	Very common	Moderately common	Common	Uncommon	Very common	Uncommon
Key points	Most common angioinvasive mold; halo sign in the lung common	Often positive blood cultures; characteristic necrotic skin lesions	Marked angioinvasion and necrosis; broad, pauciseptate hyphae with right-angle branching	Histology similar to Aspergillus; often resistant to many antifungals	Yeasts with budding and pseudohyphae; angioinvasion rare, more microabscesses	Pigmented (brown–black) hyphae; phaeohyphomycosis possible

GMS = Gomori Methenamine Silver; PAS = Periodic Acid–Schiff; AML = Acute Myeloid Leukemia; HSCT = Hematopoietic Stem Cell Transplantation; ICU = Intensive Care Unit; CNS = Central Nervous System

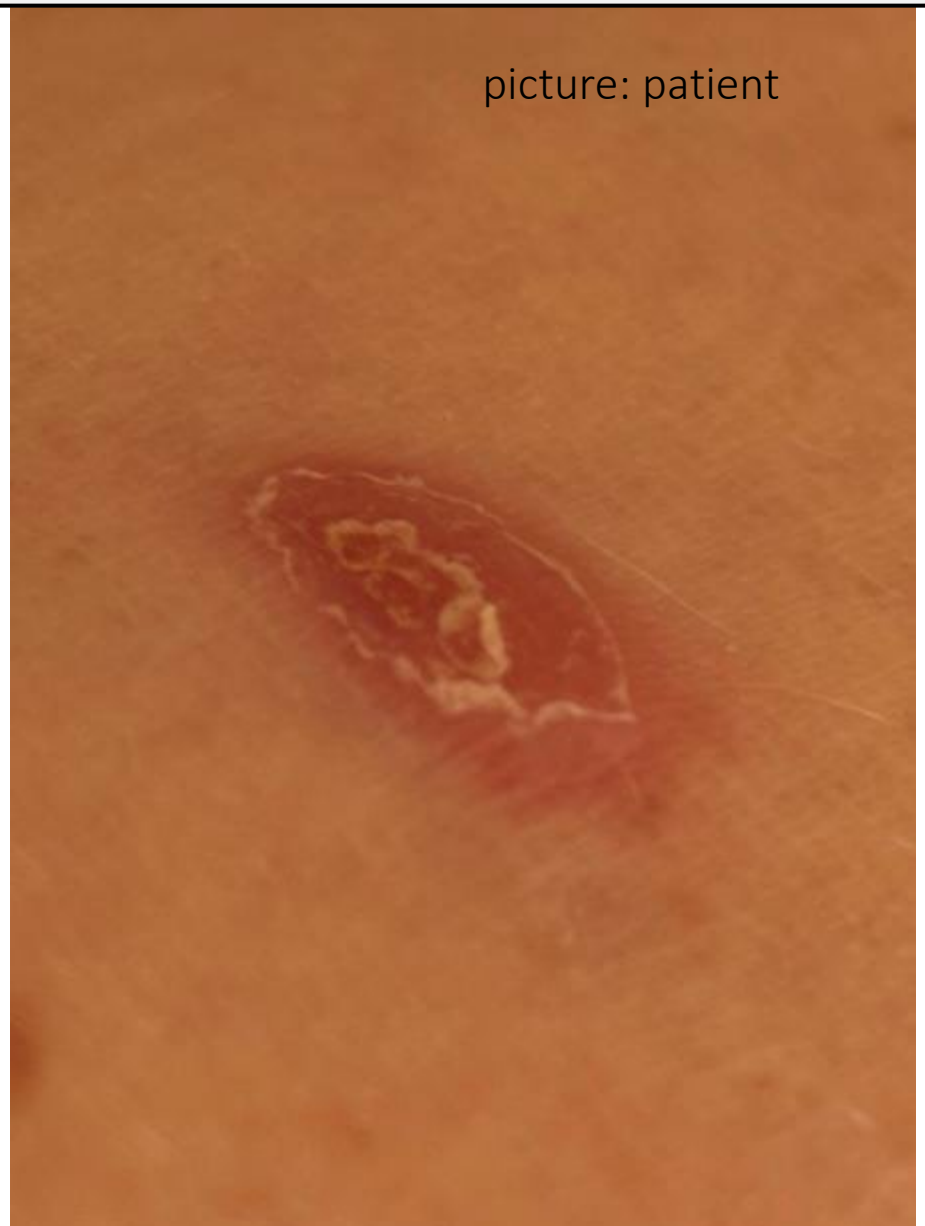
created with ChatGPT/OpenAI, 2026

Case 8

Clinical presentation:



picture: Dr. Gilfert, Kassel



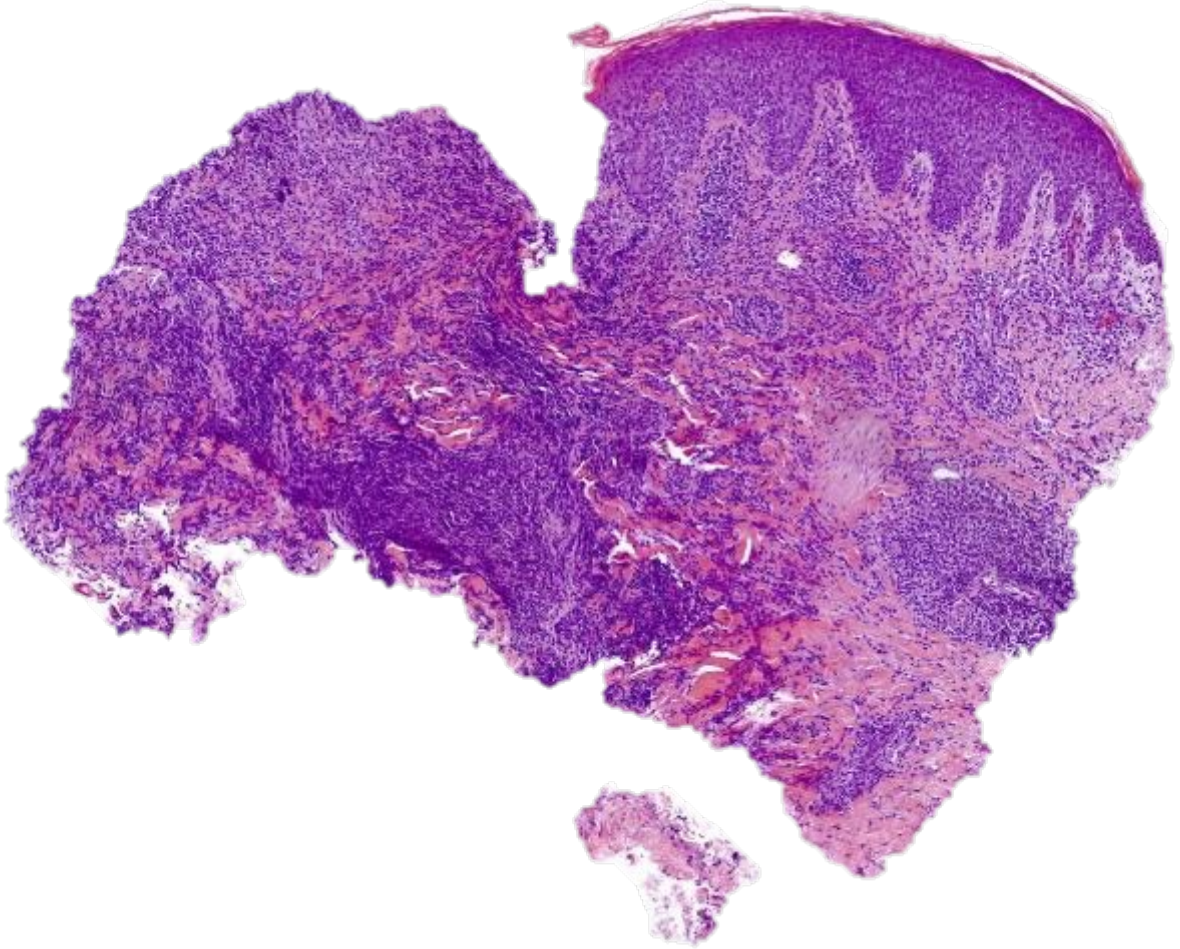
picture: patient

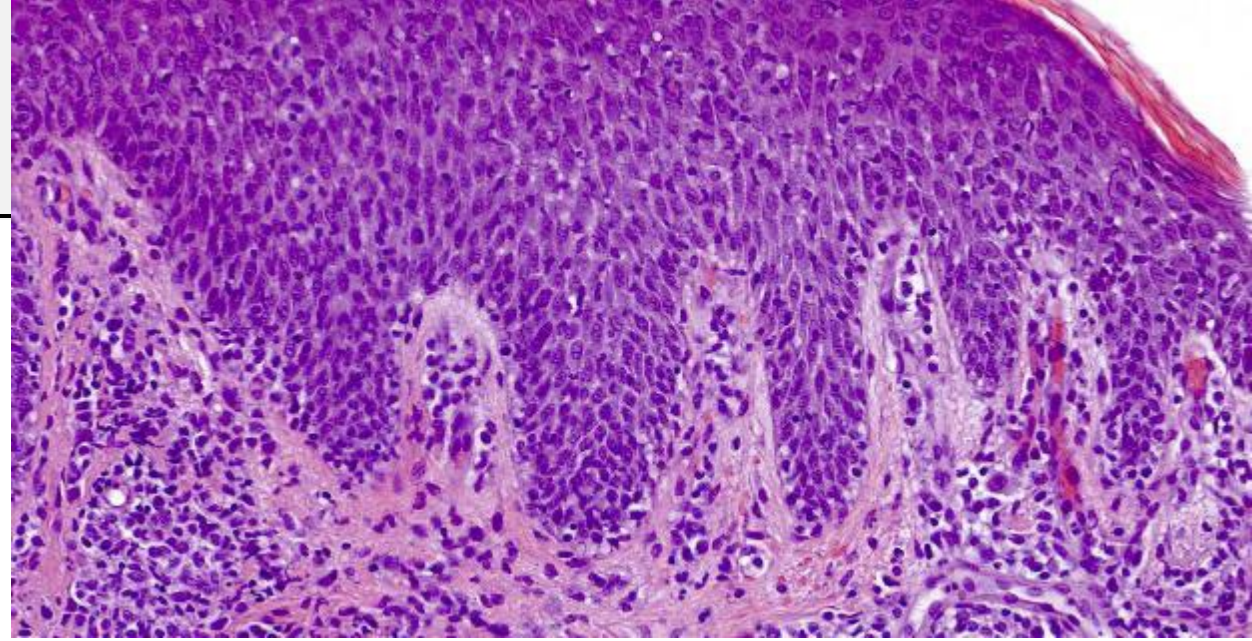
- 49-year-old female patient
- Night sweats and hot flashes
- No fever
- No weight loss
- Otherwise healthy

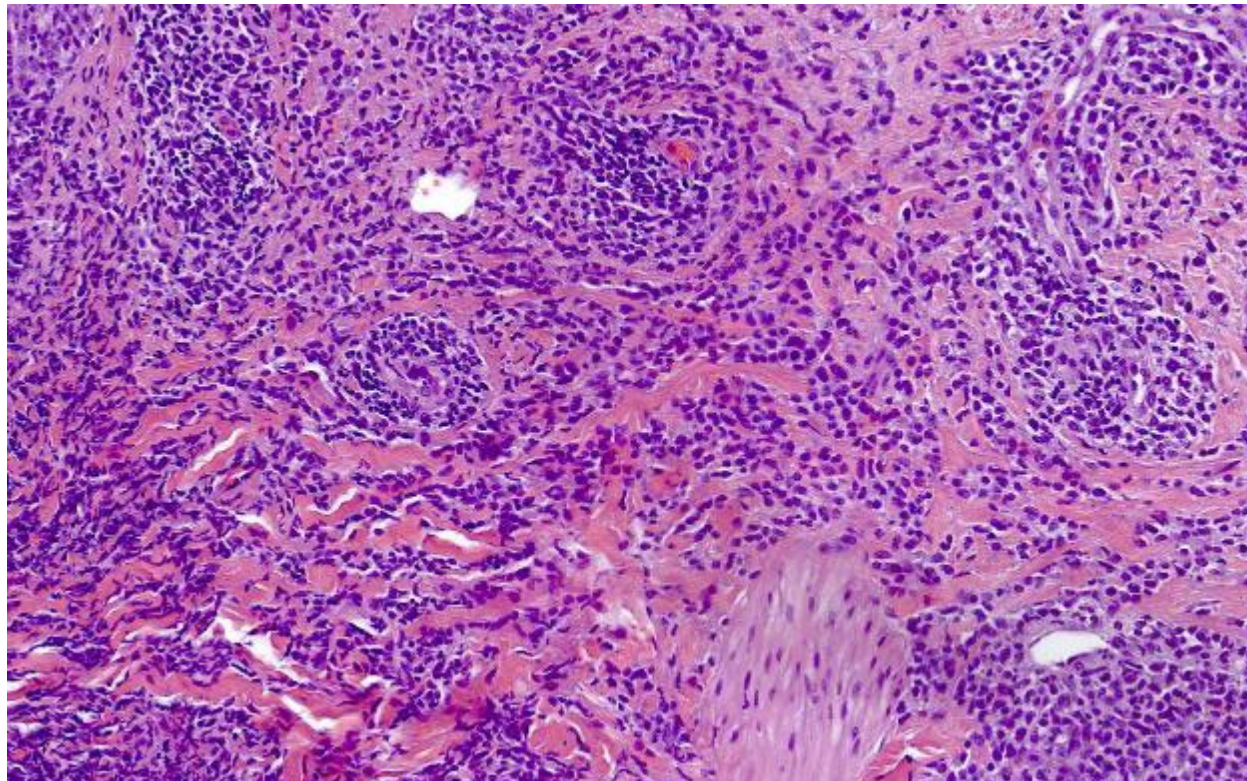
- Skin eruption occurred four weeks ago
- She suspected first an insect-bite
- Only partial remission but no complete regression

- Consultation of a dermatologist: suspected diagnosis: Insect-bite?

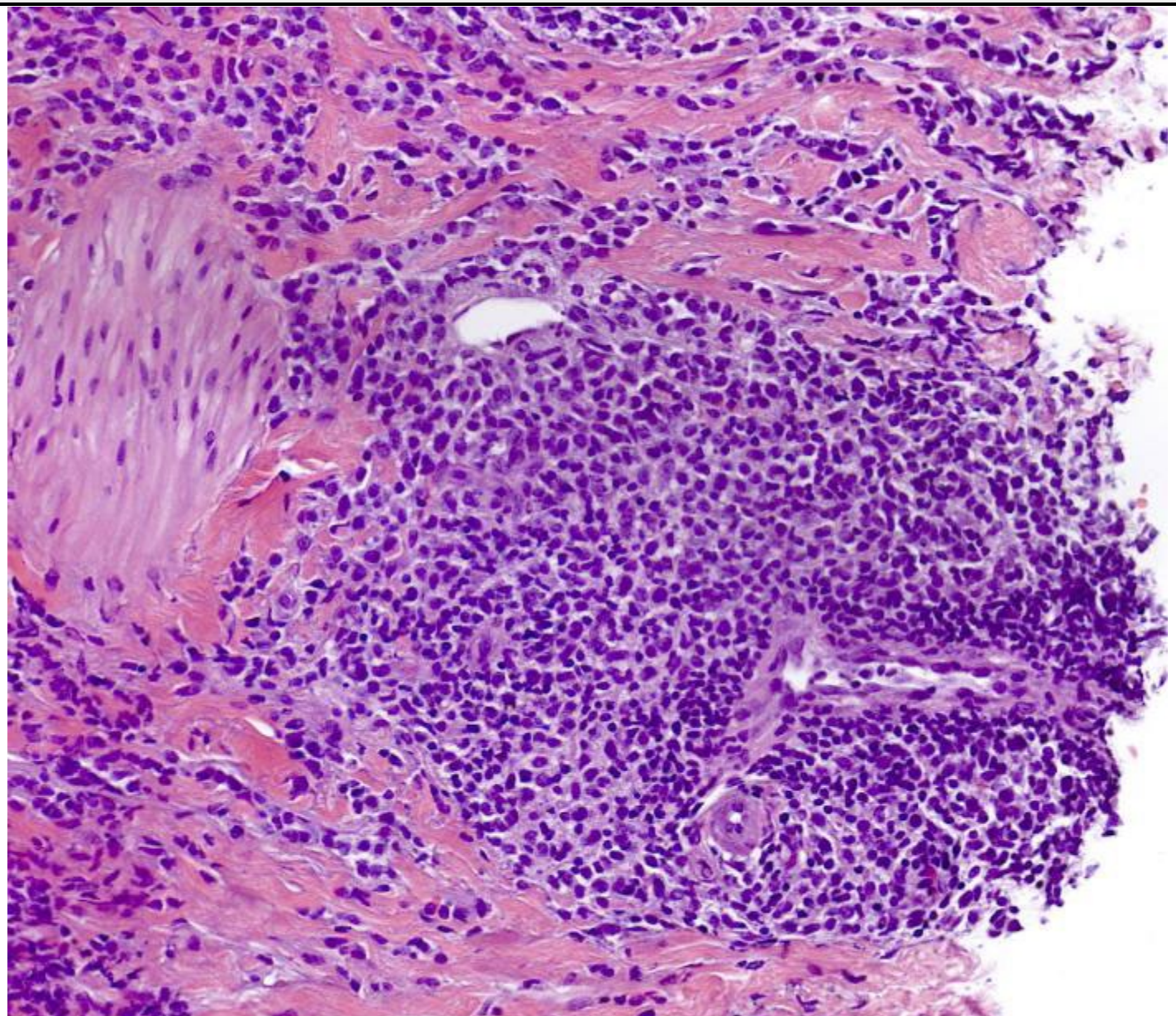
Histology:

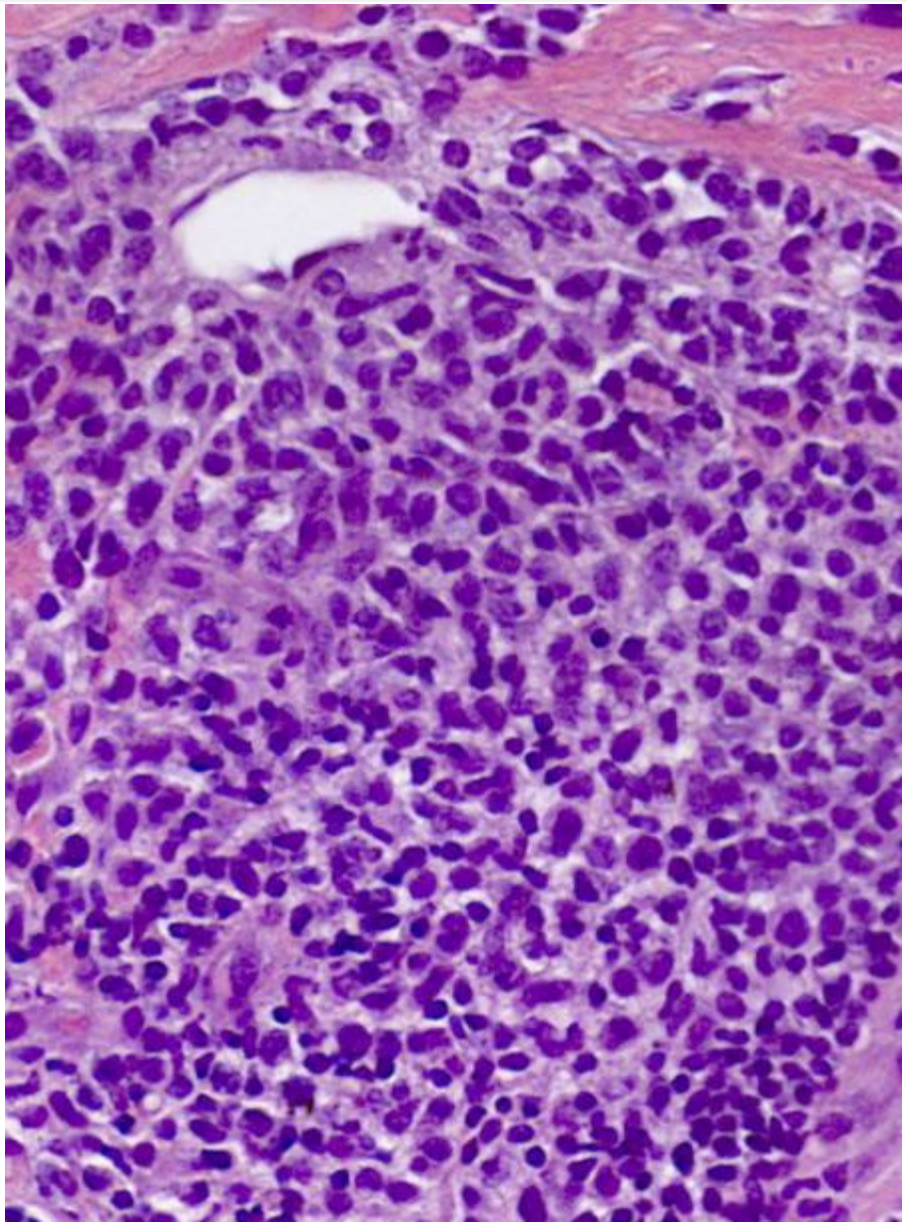


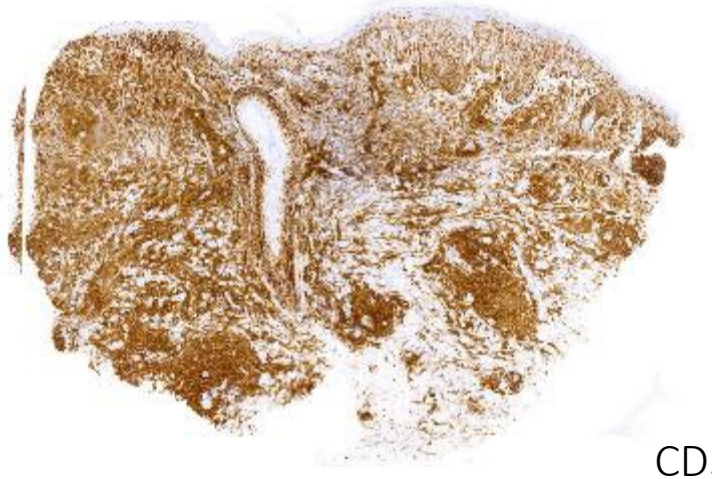




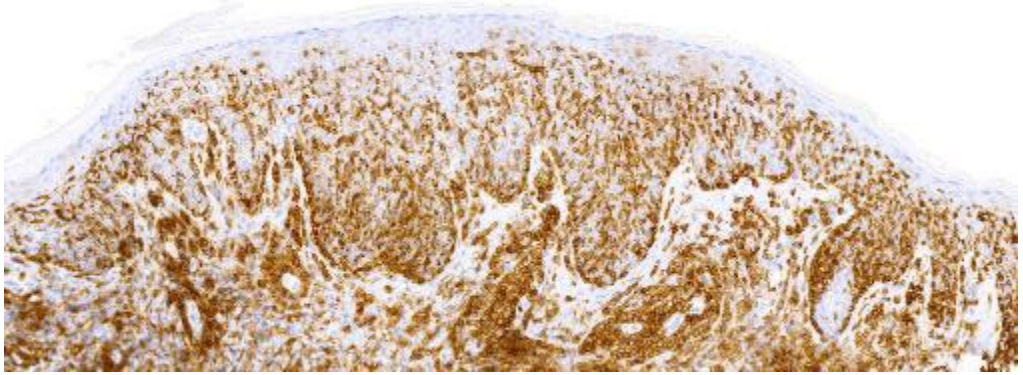
Histology:

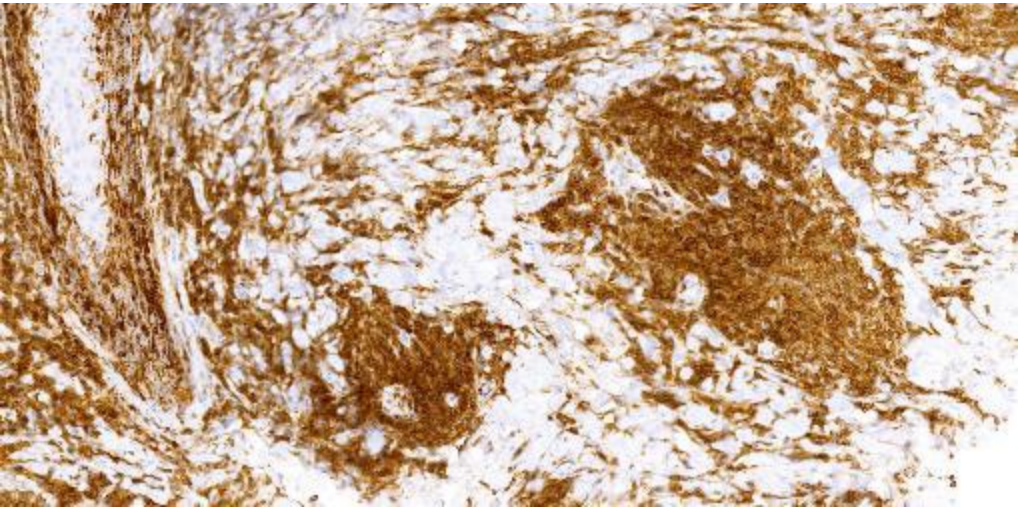


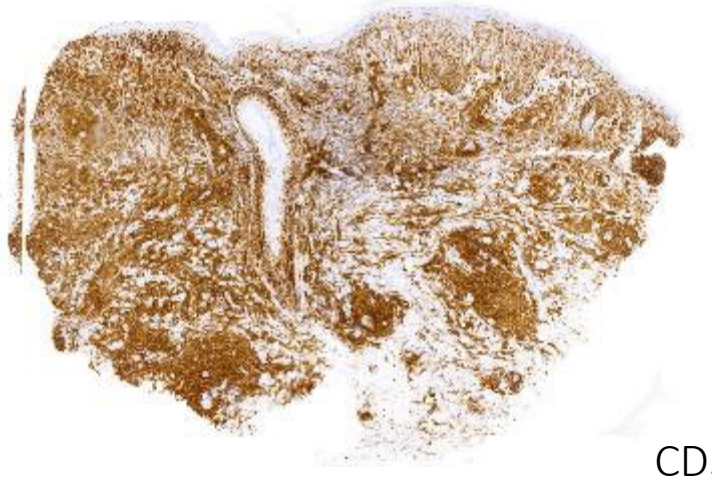




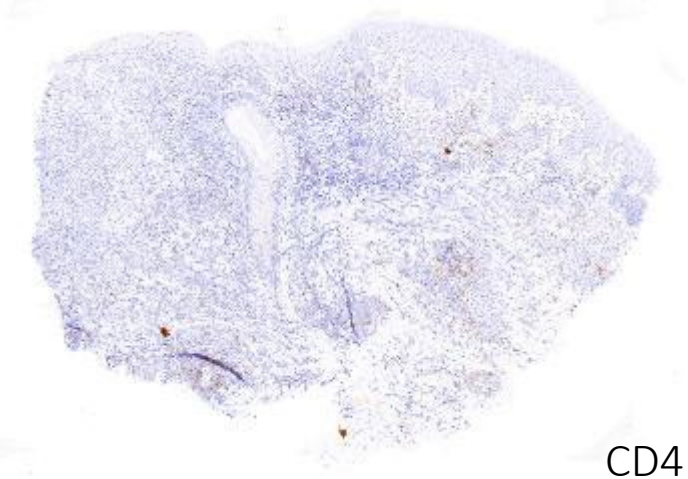
CD3



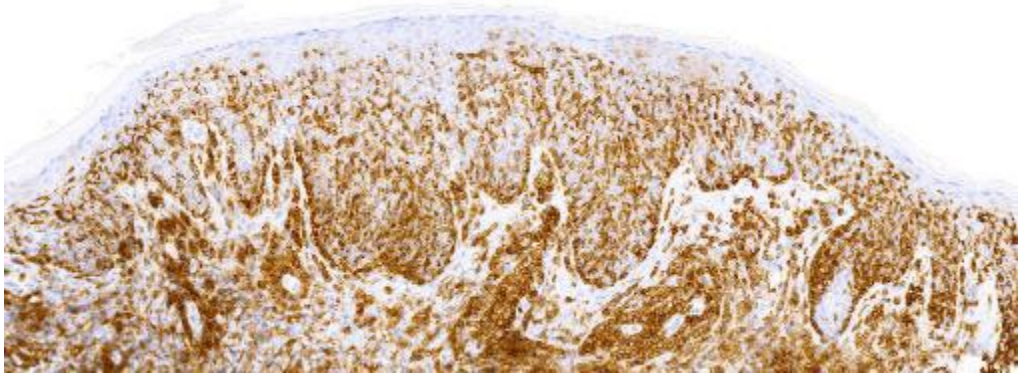


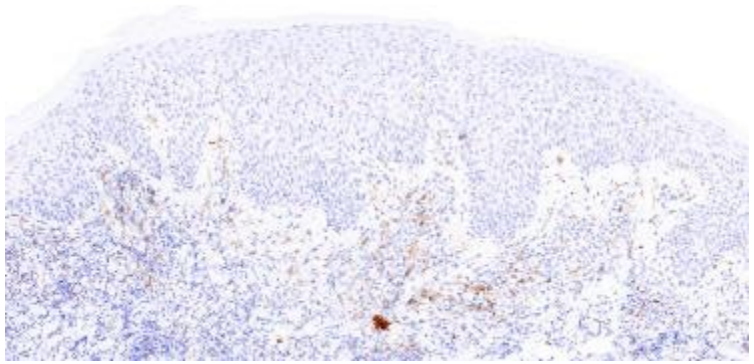


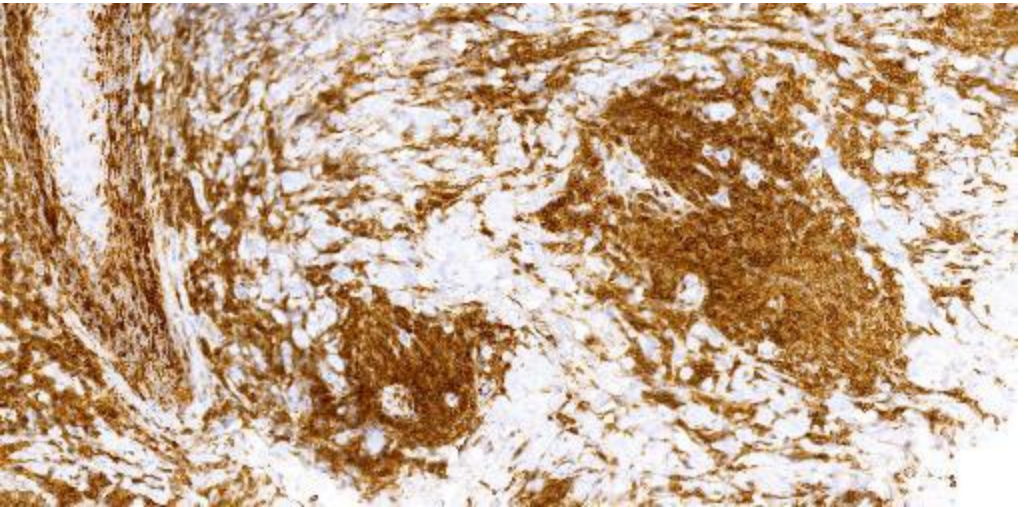
CD3

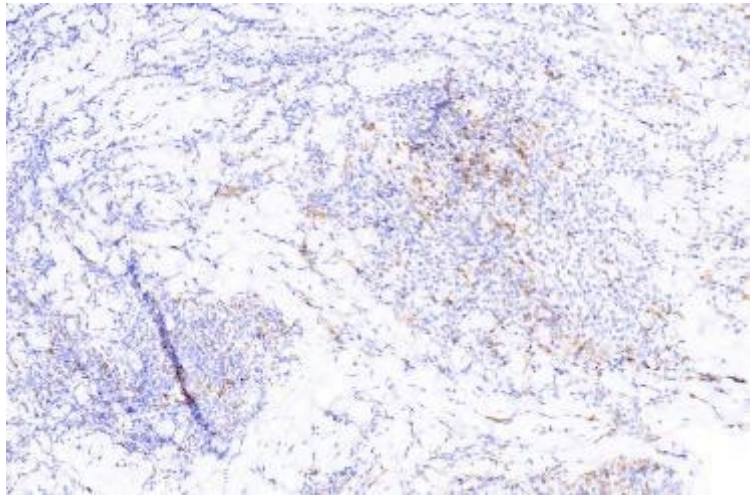


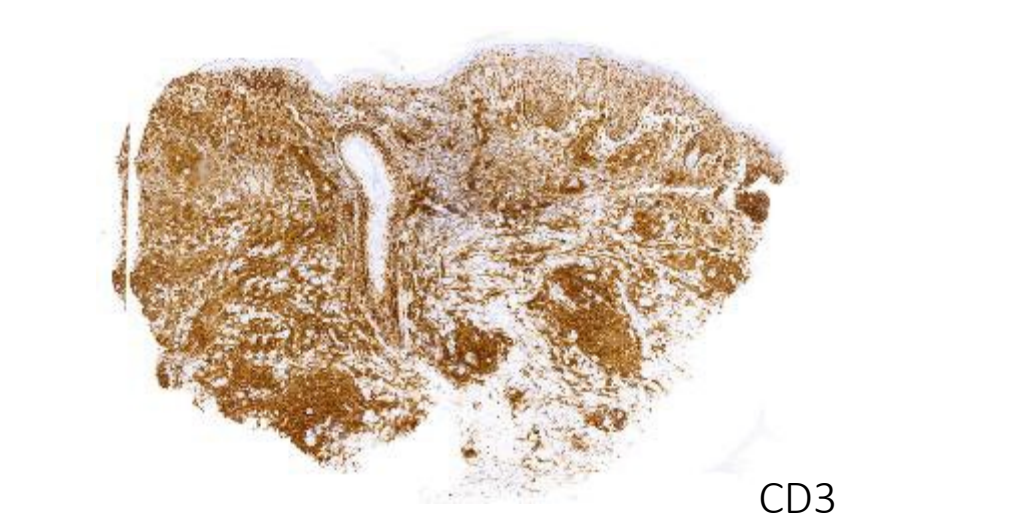
CD4











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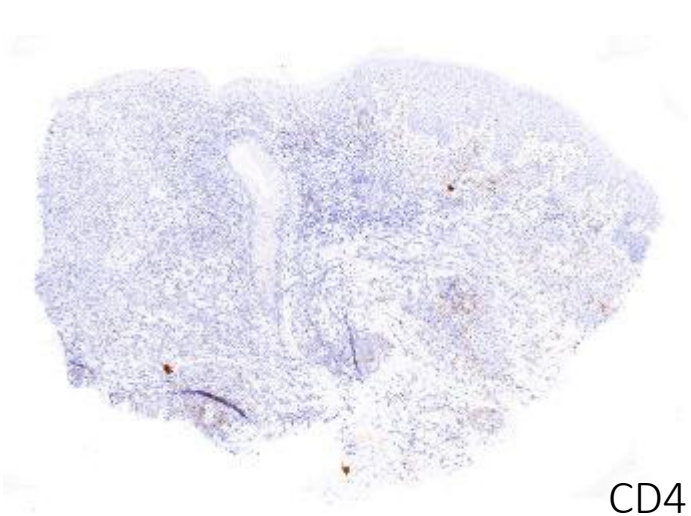
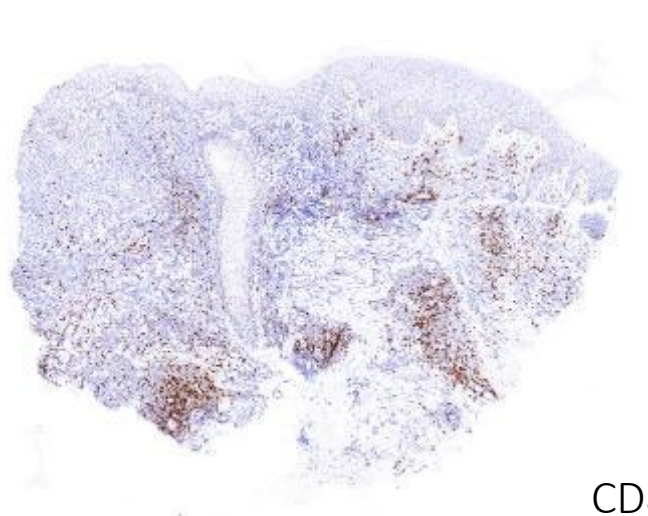


CD4



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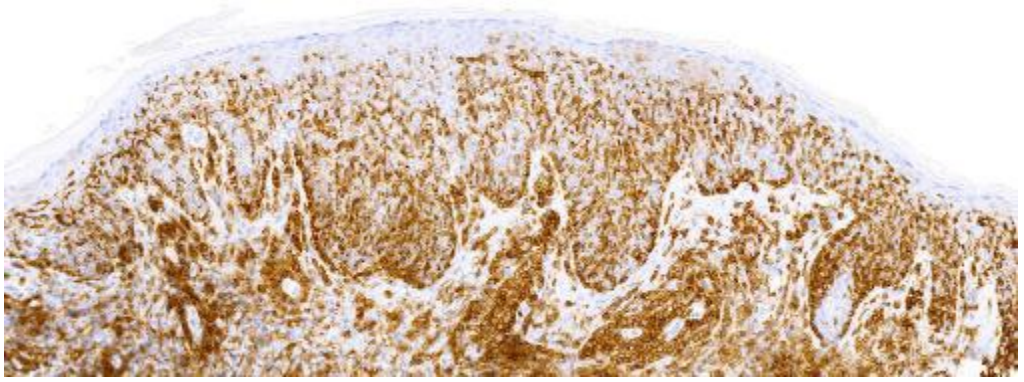

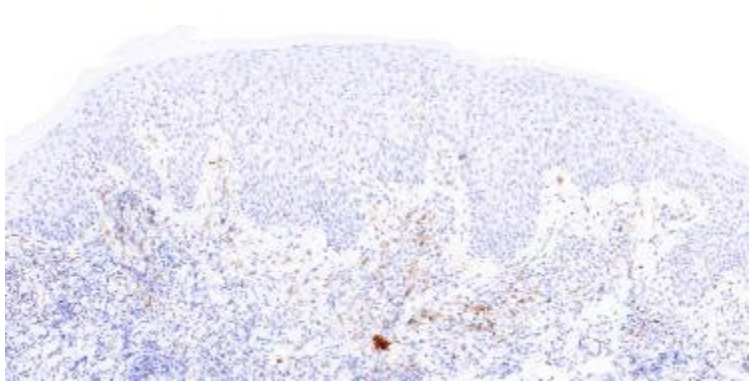

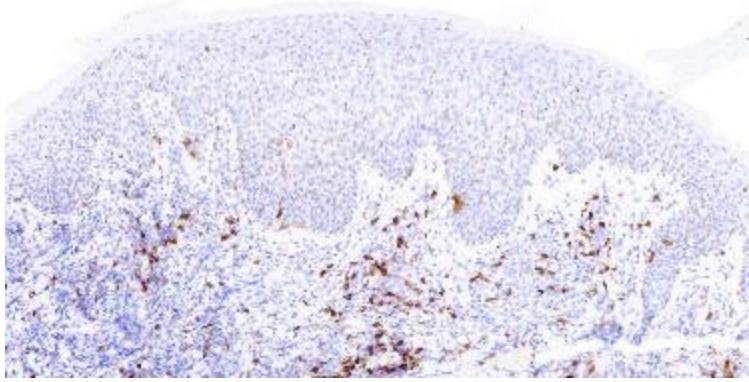

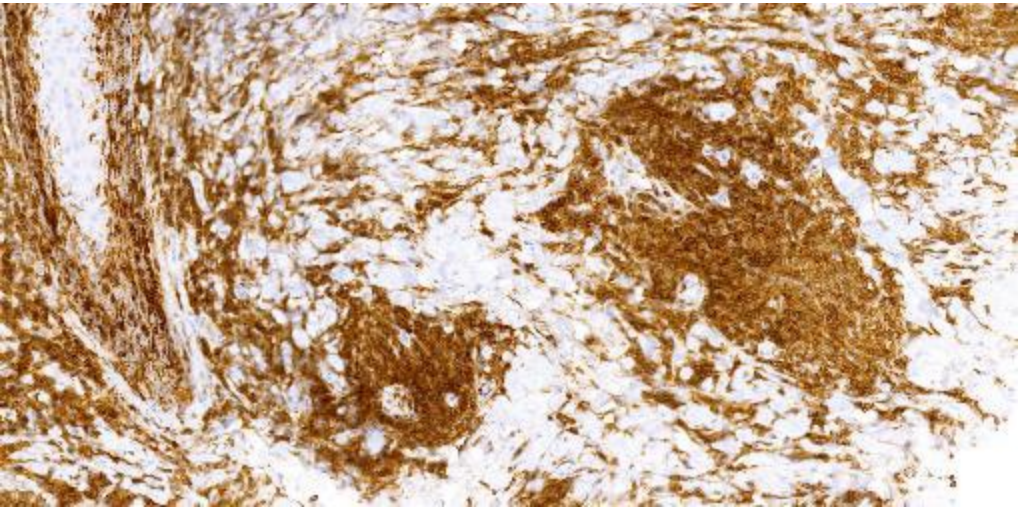

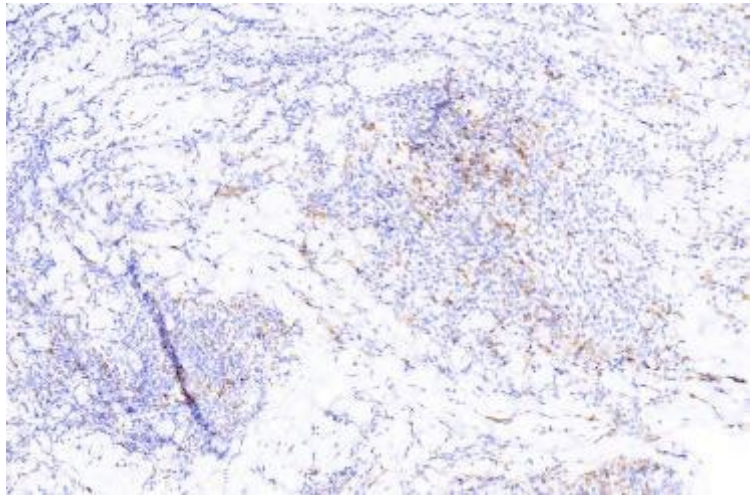

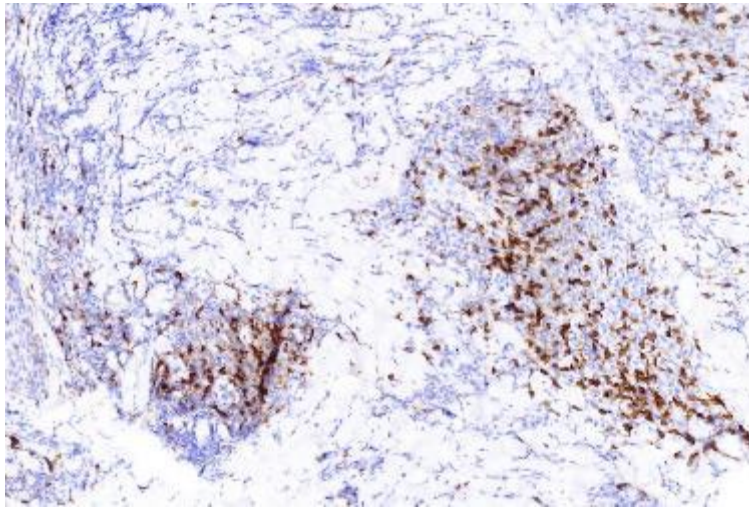

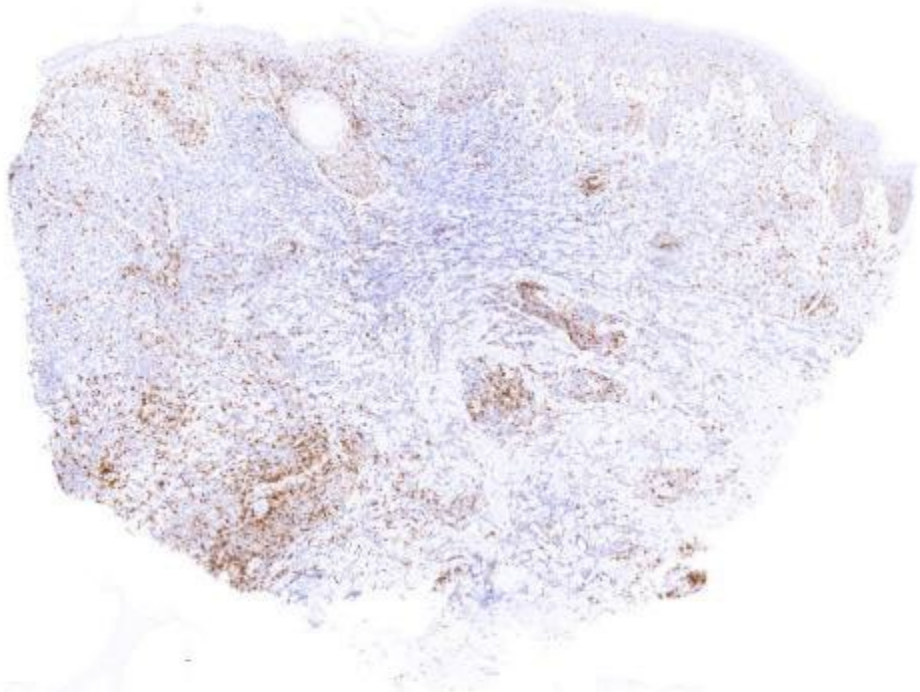

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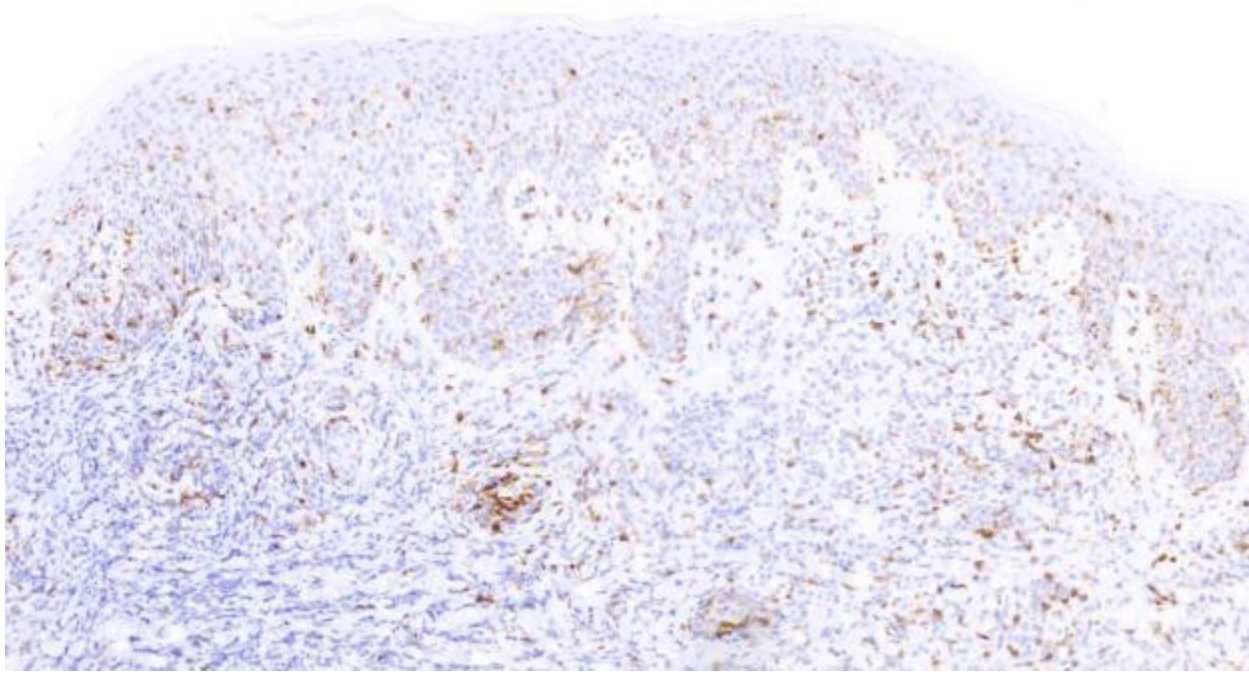
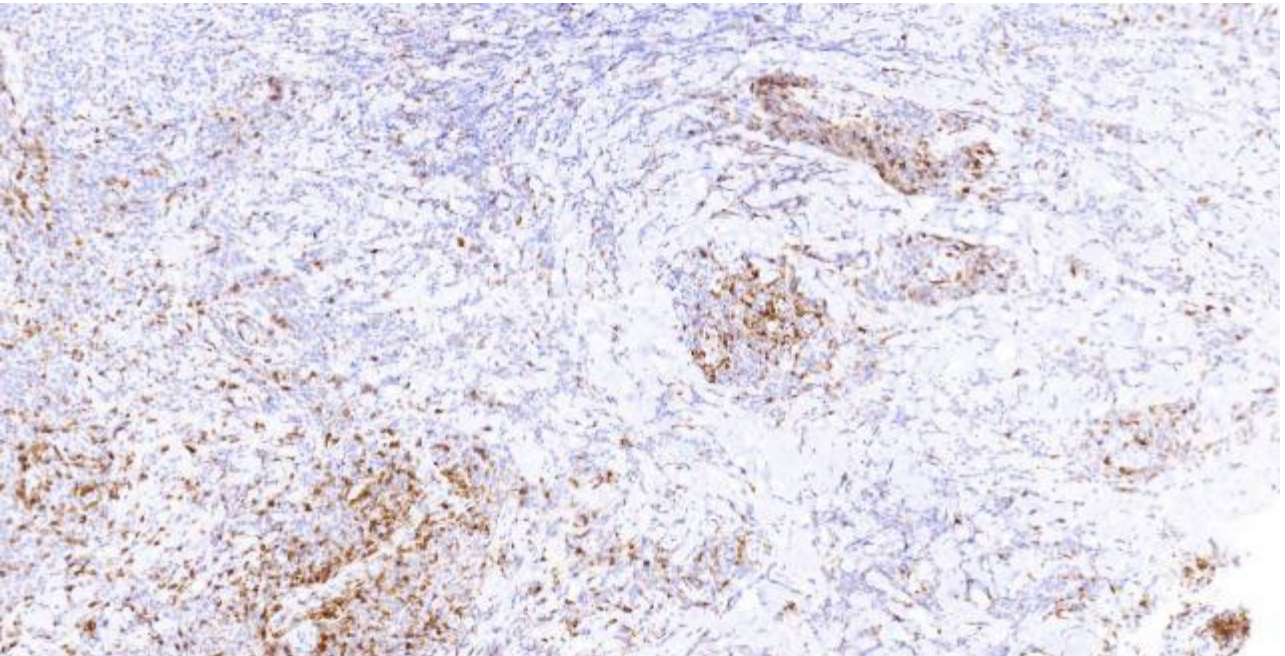
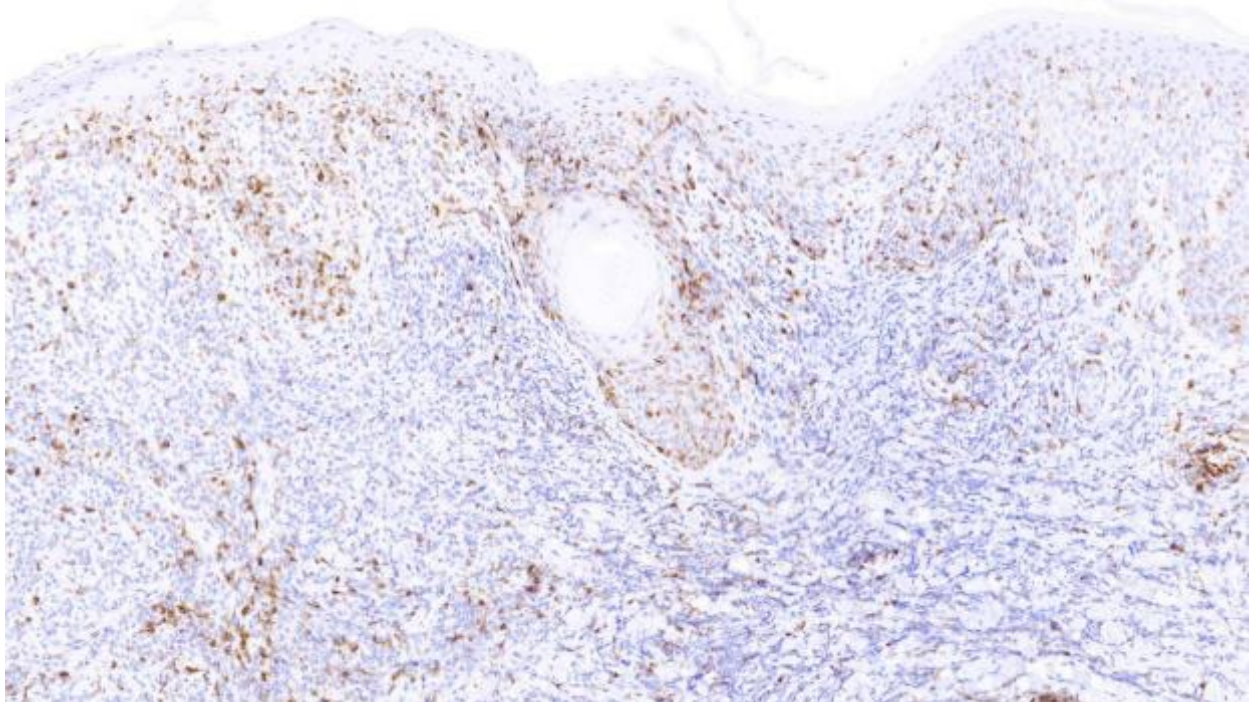
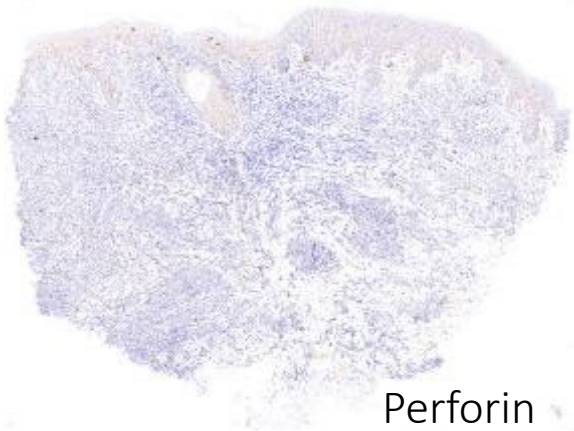


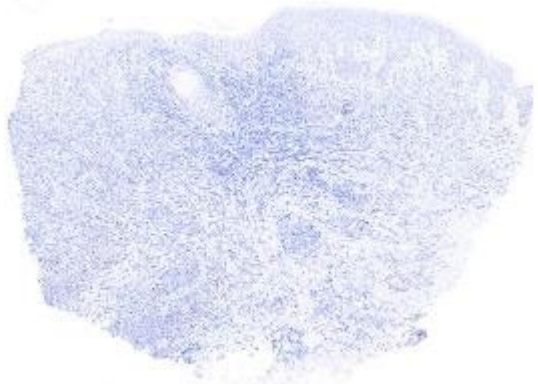




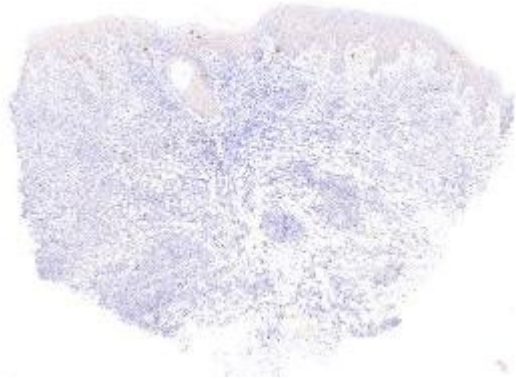
Immunohistochemistry:



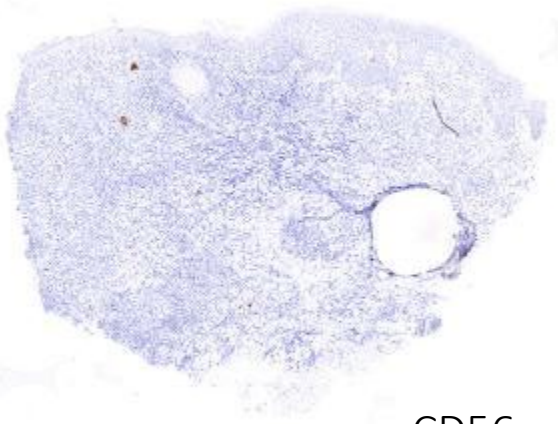
Perforin



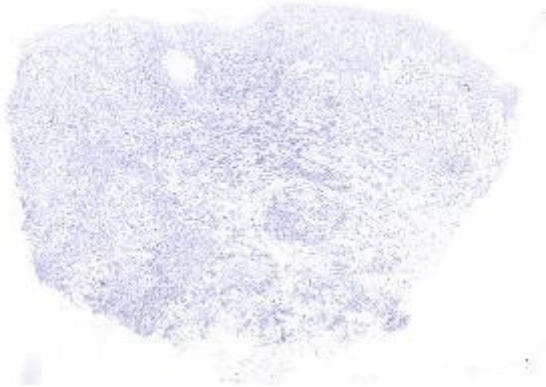
Granzyme B



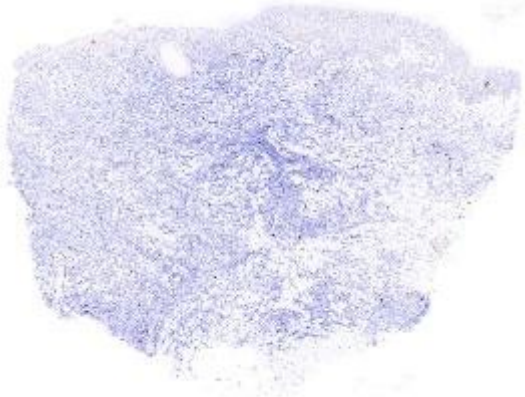
TIA1



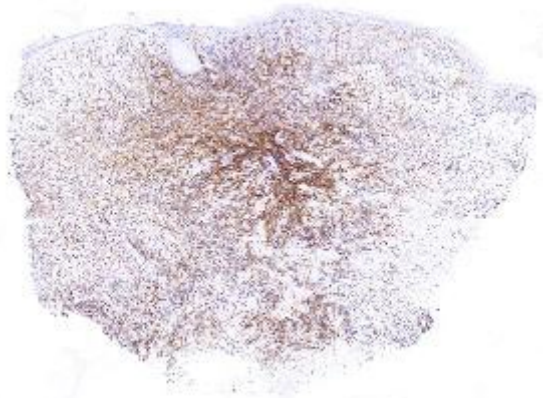
CD56



PD1

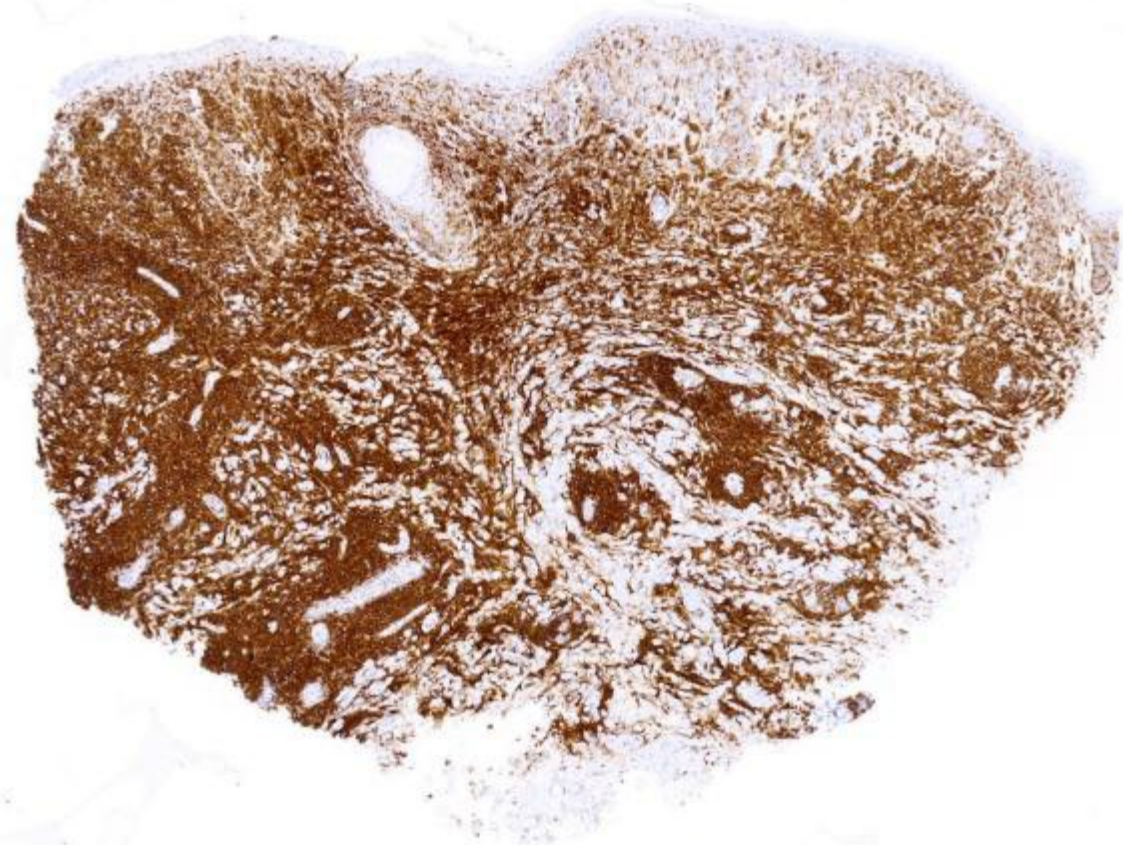


TCRgamma

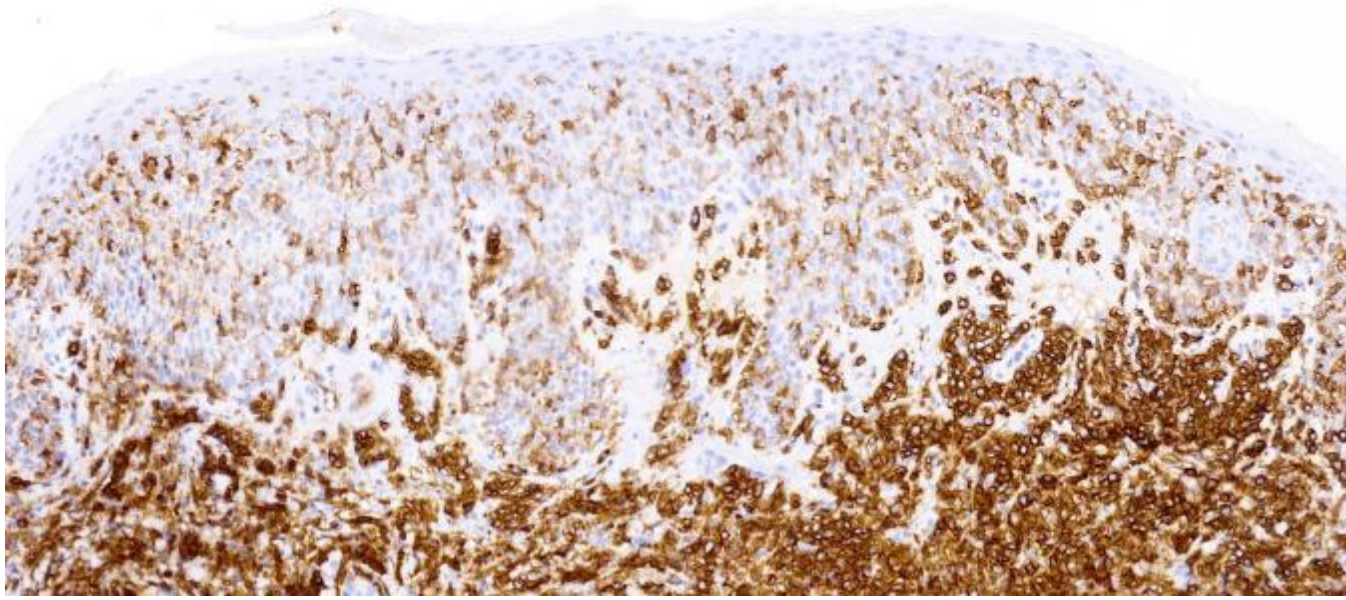


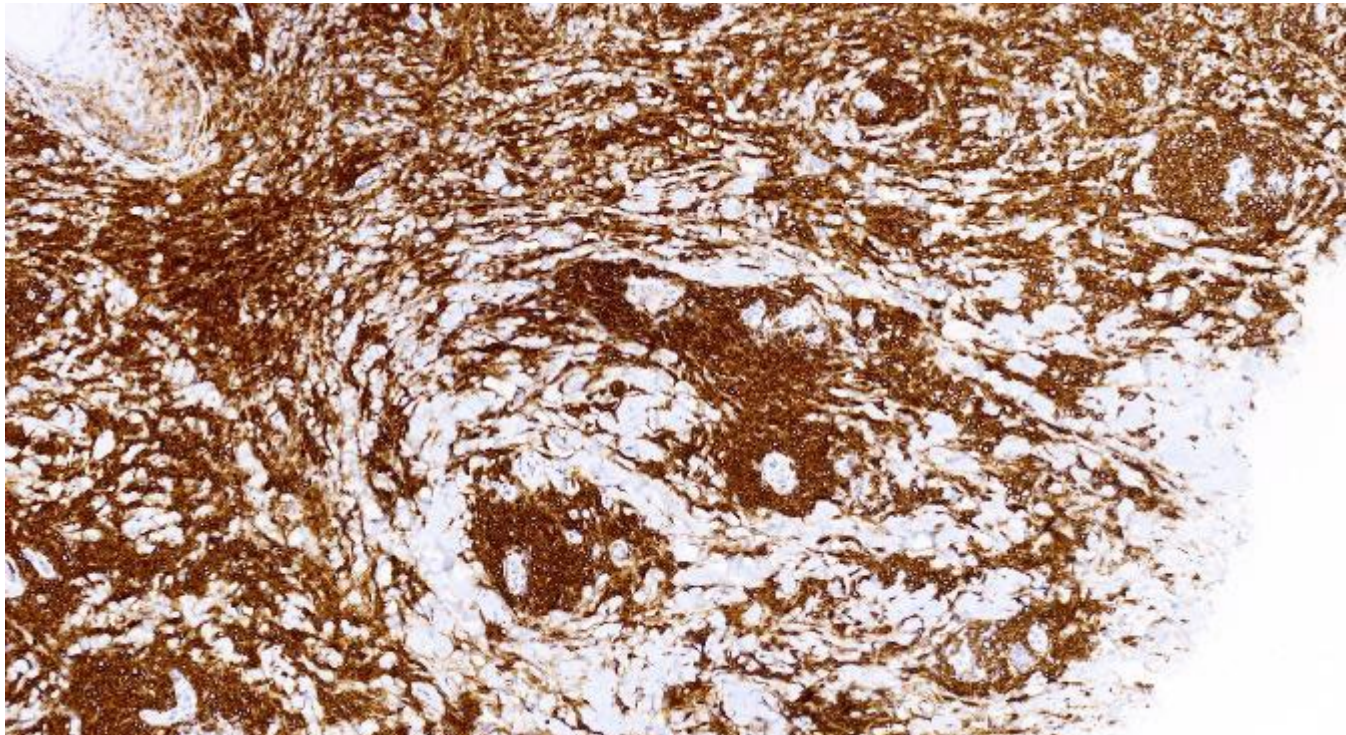
TCRbeta

Ki67

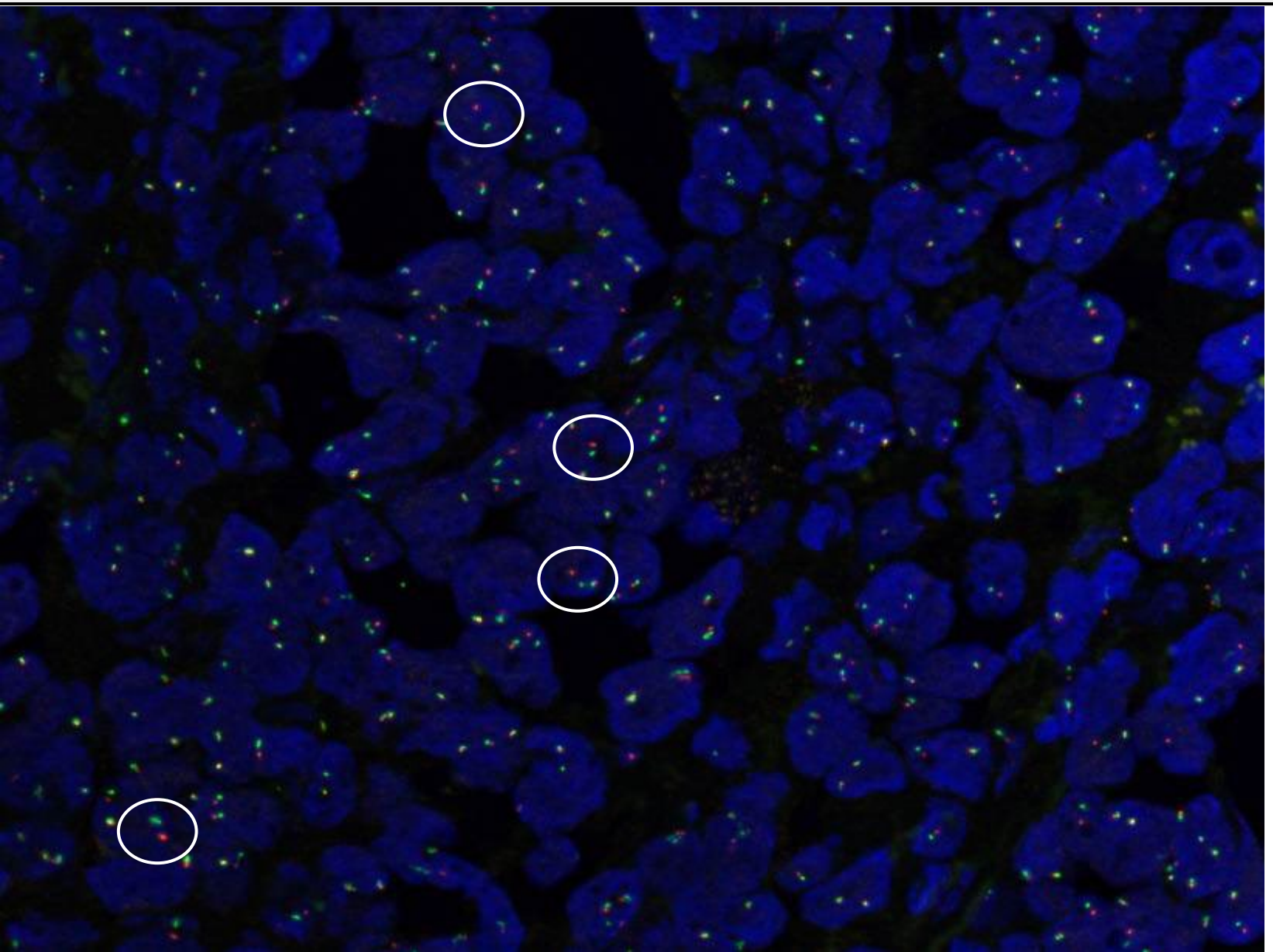


CD30





FISH: DUSP22-IRF4 Rearrangement



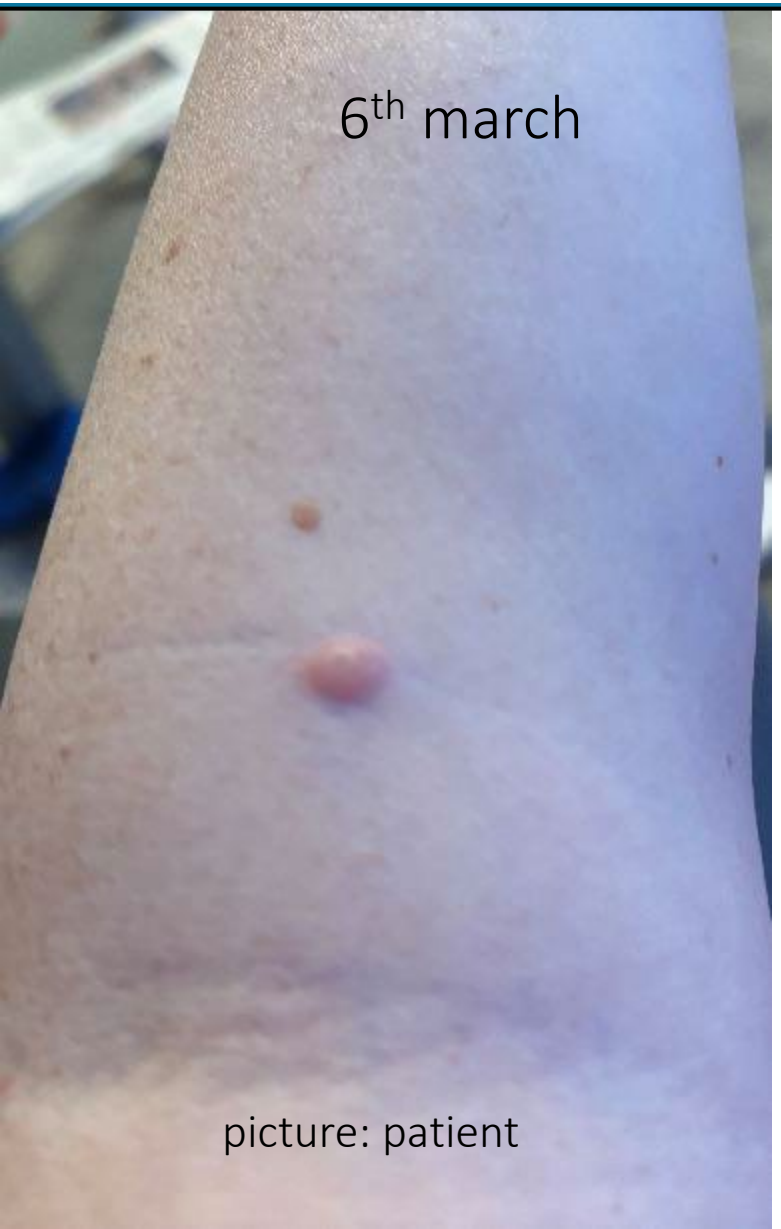
FISH break apart probe of IRF4

intact fusion signal = yellow

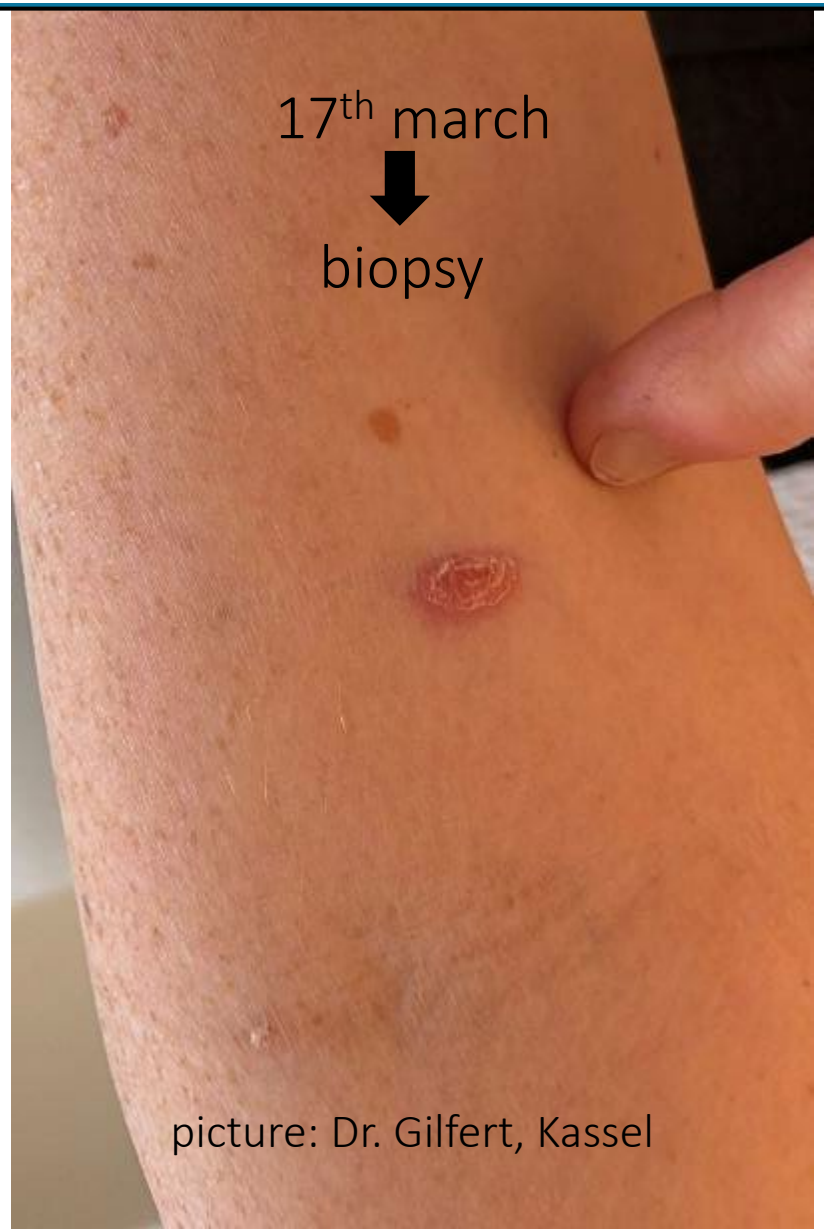
abnormal split signal
red = telomeric
green = centomeric

→ green and red signals indicate separation of IRF4

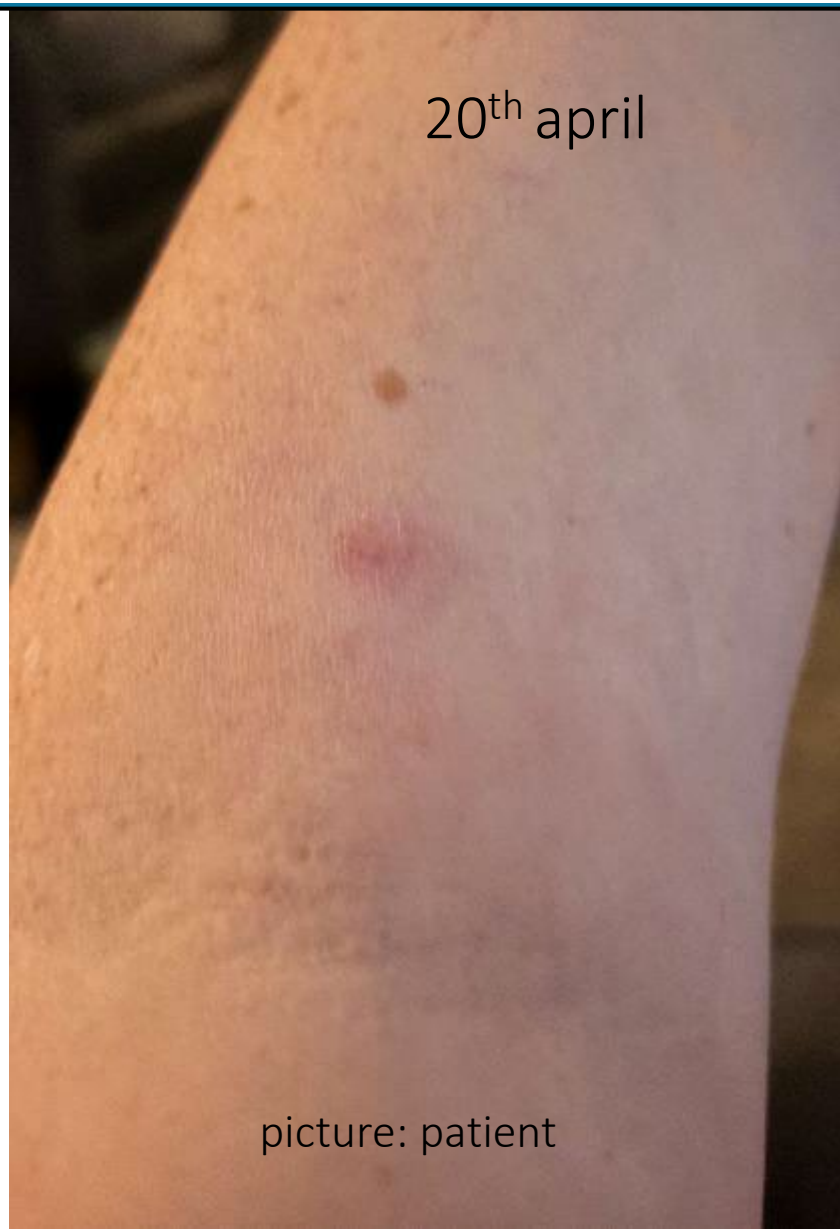
Clinical presentation:



picture: patient

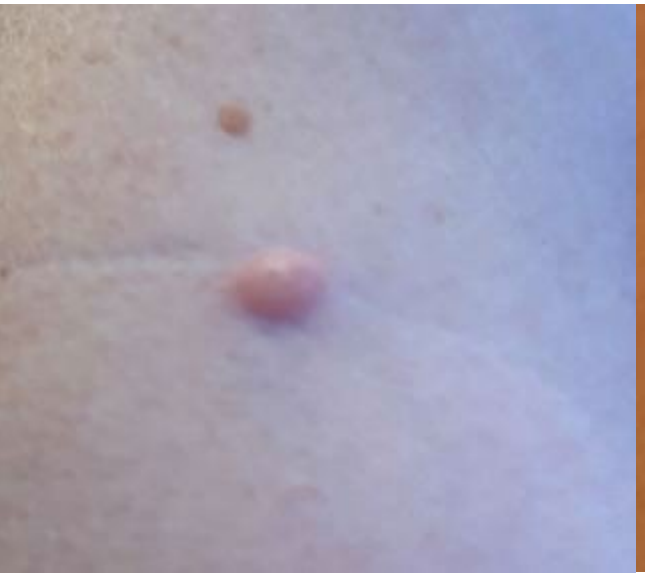


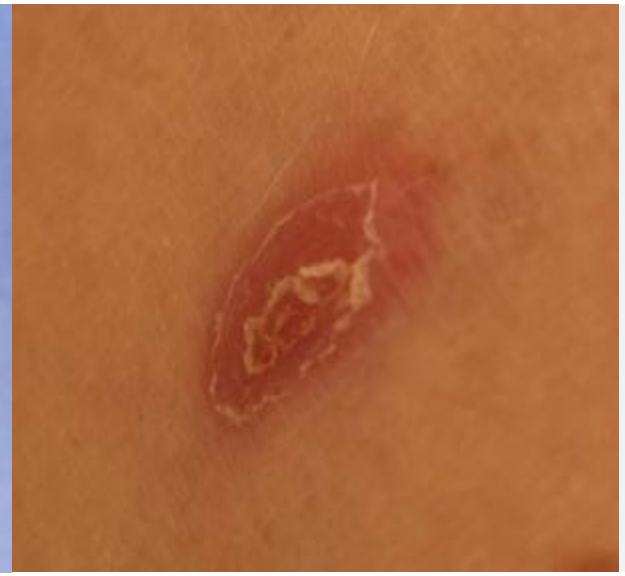
picture: Dr. Gilfert, Kassel



picture: patient

Summary of the findings:



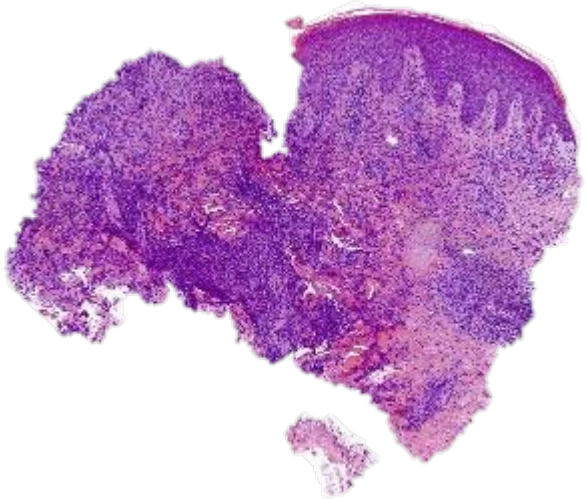


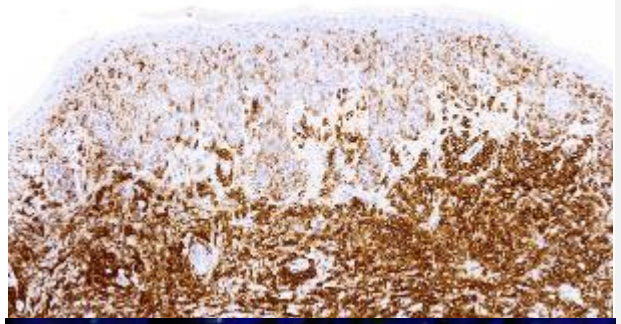
Clinical presentation:

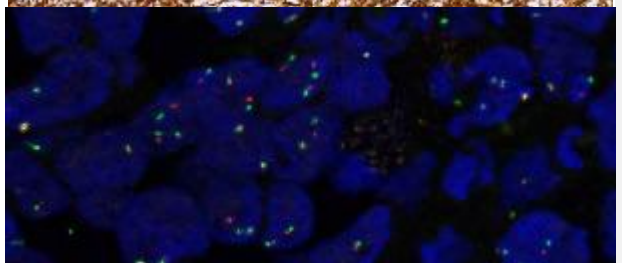
- Solitary plaque / nodule
- No ulceration
- Spontaneous regression
- Restricted to the skin
- Staging negative

diagnosis

?



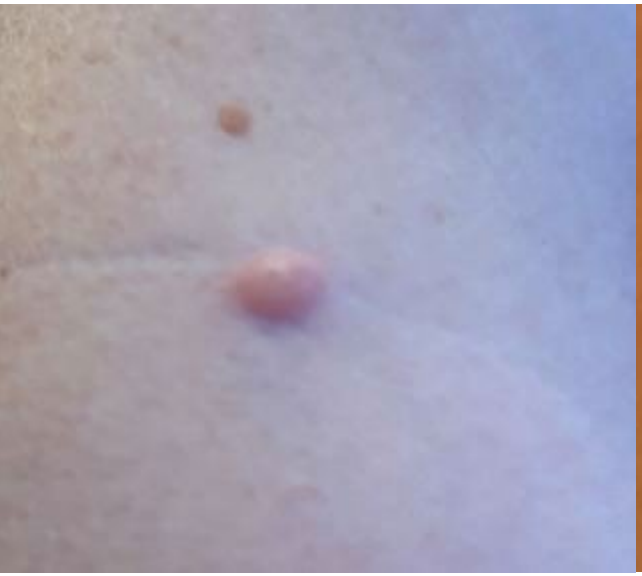


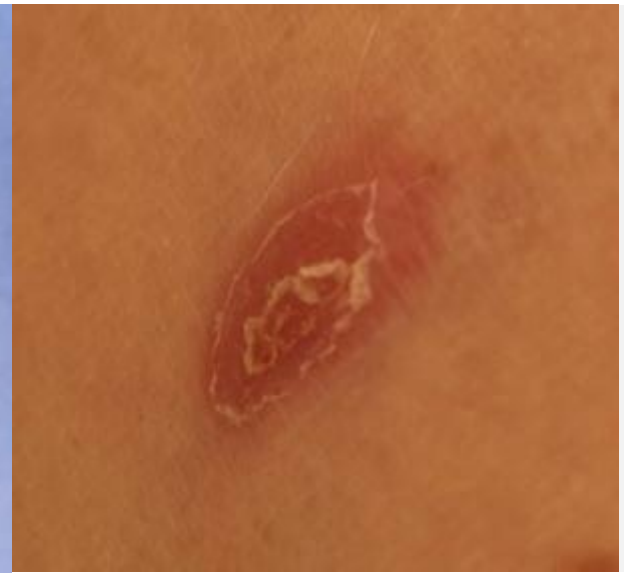


Histology:

- Biphasic pattern: small intraepidermal (pagetoid) and larger dermal lymphocytes
- CD3+, CD4-, CD8-, CD7-
- CD30+ biphasic
- TRCa/b +, TCR g/d-, cytotox.+/-
- DUSP22-IRF4 rearrangement

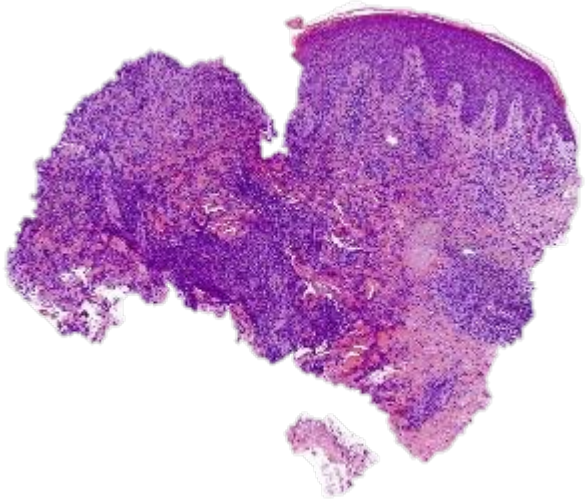
Summary of the findings:

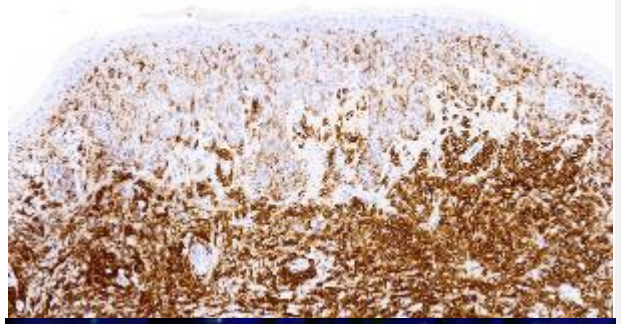


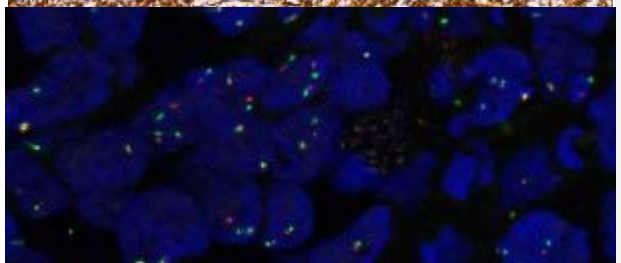


Clinical presentation:

- Solitary plaque / nodule
- No ulceration
- Spontaneous regression
- Restricted to the skin
- Staging negative







Histology:

- Biphasic pattern: small intraepidermal (pagetoid) and larger dermal lymphocytes
- CD3+, CD4-, CD8-, CD7-
- CD30+ biphasic
- TRCa/b +, TCR g/d-, cytotox.+/-
- DUSP22-IRF4 rearrangement

DUSP22-IRF4 rearranged CD30+ primary cutaneous lymphopro-liferative disorder

Literature: DUSP22 in LyP

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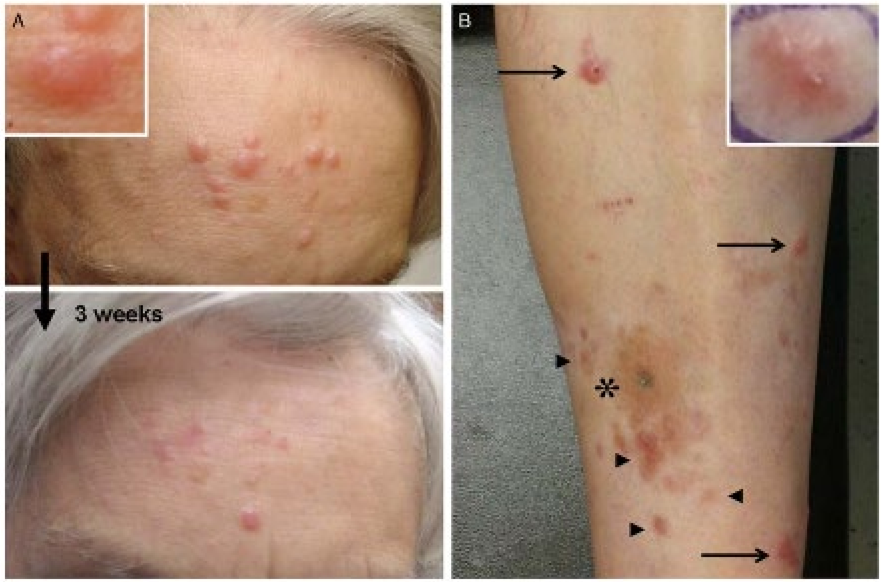
(*Am J Surg Pathol* 2013;37:1173–1181)



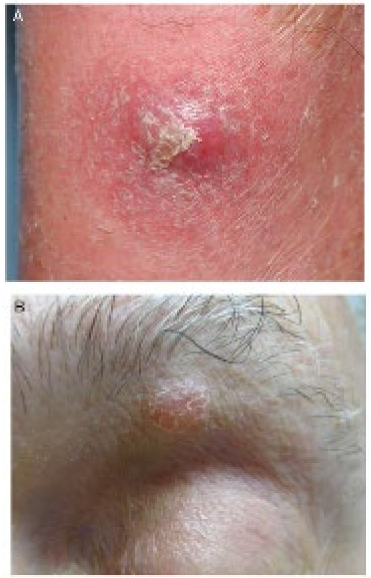


TABLE 2. Clinical, Histologic, Immunophenotypic, and Genetic Features of 11 Patients With LyP

	Case 1	2	3	4	5	6	7	8	9	10	11
Clinical presentation											
Age at onset (y)	84	71	71	67	78	82	74	75	68	88	67
Sex	F	M	M	M	M	M	M	M	M	M	F
Site	Forehead	Chest	Shin	Forehead	Scapula	Chest	Back	Axilla	Arm	Arm	Back
Histologic features											
Periadnexal spread	Present	Present	Absent	Present	Present	Present	Present	Present	Absent	Present	Absent
Eosinophils	Absent	Absent	Absent	Absent	Present	Focal	Absent	Absent	Absent	Absent	Absent
Pathologic diagnosis	CD30+ TLPD	LyP	CD30+ TLPD	CD30+ TLPD	CD30+ TLPD	CD30+ TLPD	CD30+ TLPD	CD30+ TLPD	LyP	CD30+ TLPD	LyP
Immunophenotype											
ALK	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Negative
CD3	Positive	Weak	Negative	Positive	Positive	Positive	Positive	Positive	Positive	Positive	Positive
CD4	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Positive	Negative	Negative	Partial
CD8	Negative	Negative	Negative	Weak	Positive	Positive	Negative	Positive	Negative	Partial	Negative
CD15	Focal	Positive	Negative	Positive	Positive	Focal	Negative	Focal	NA	Negative	NA
CD30	D > E	D > E	Positive	Positive	Positive	D > E	D > E	Positive	Positive	D > E	Positive
Ki-67	> 80%	> 80%	> 80%	NA	NA	> 80%	> 80%	> 80%	NA	60%-80%	NA
TCR-β	Positive	Weak	Positive	Positive	NA	Positive	Positive	Positive	Negative	Positive	NA
TIA-1	Negative	Positive	Positive	Negative	NA	Negative	Negative	Negative	Negative	Negative	Negative
Genetics											
6p25.3 rearrangement	Present	Present	Present	Present	Present	Present	Present	Present	Present	Present	Present
Follow-up											
Treatment	None	Excision	Radiation	Radiation	Excision, cryo	Radiation	Radiation	None	None	Excision	None
Spontaneous resolution	Yes	—	—	—	—	—	—	Yes	Yes	—	Yes
Skin recurrence	No	No	Yes	No	Yes	No	No	No	Yes	Yes	Yes
Disseminated skin disease	No	No	No	No	No	No	No	No	No	No	No
Extracutaneous disease	No	No	No	No	No	No	No	No	No	No	No
Status last follow-up	NED	NED	LyP, same site	NED	NED	NED	NED	NED	NED	NED	NED
Months since onset	21	150	12	7	21	30	36	10	104	22	20
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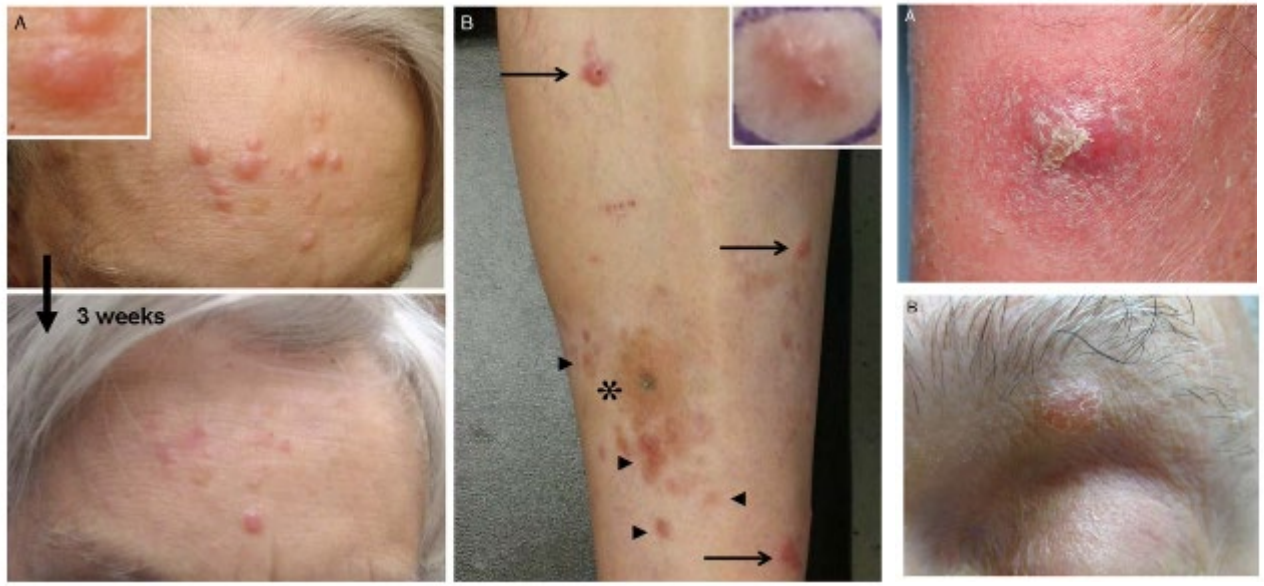

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Periadnexal spread	Present	Present	Absent	Present	Present	Present	Present	Present	Absent	Present	Absent
Eosinophils	Absent	Absent	Absent	Absent	Present	Focal	Absent	Absent	Absent	Absent	Absent
Pathologic diagnosis	CD30⁻ TLPD	LyP	CD30⁺ TLPD	CD30⁻ TLPD	CD30⁺ TLPD	CD30⁺ TLPD	CD30⁺ TLPD	CD30⁻ TLPD	LyP	CD30⁺ TLPD	LyP
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CD8	Negative	Negative	Negative	Weak	Positive	Positive	Negative	Positive	Negative	Partial	Negative
CD15	Focal	Positive	Negative	Positive	Positive	Focal	Negative	Focal	NA	Negative	NA
CD30	D > E	D > E	Positive	Positive	Positive	D > E	D > E	Positive	Positive	D > E	Positive
Ki-67	> 80%	> 80%	> 80%	NA	NA	> 80%	> 80%	> 80%	NA	60%-80%	NA
TCR-β	Positive	Weak	Positive	Positive	NA	Positive	Positive	Positive	Negative	Positive	NA
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6p25.3 rearrangement	Present	Present	Present	Present	Present	Present	Present	Present	Present	Present	Present
Follow-up											
Treatment	None	Excision	Radiation	Radiation	Excision, cryo	Radiation	Radiation	None	None	Excision	None
Spontaneous resolution	Yes	–	–	–	–	–	–	Yes	Yes	–	Yes
Skin recurrence	No	No	Yes	No	Yes	No	No	No	Yes	Yes	Yes
Disseminated skin disease	No	No	No	No	No	No	No	No	No	No	No
Extracutaneous disease	No	No	No	No	No	No	No	No	No	No	No
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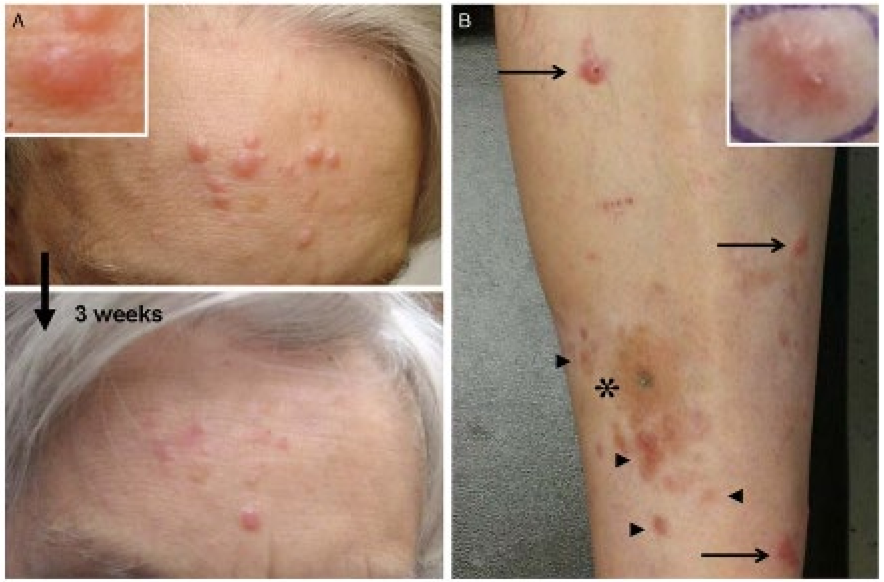

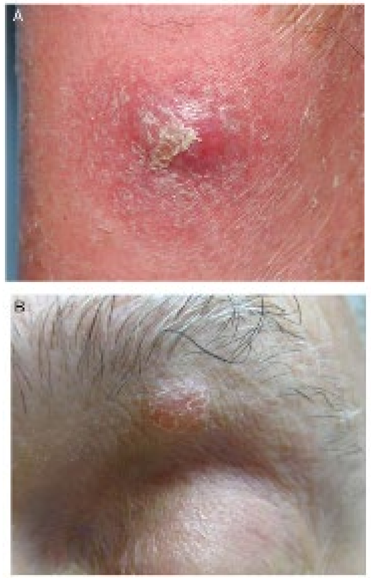

Am J Surg Pathol • Volume 37, Number 8, August 2013

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Periadnexal spread	Present	Present	Absent	Present	Present	Present	Present	Present	Absent	Present	Absent
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Pathologic diagnosis	CD30+ TLPD	LyP	CD30+ TLPD	CD30+ TLPD	CD30+ TLPD	CD30+ TLPD	CD30+ TLPD	CD30+ TLPD	LyP	CD30+ TLPD	LyP
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CD30	D > E	D > E	Positive	Positive	Positive	D > E	D > E	Positive	Positive	D > E	Positive
Ki-67	> 80%	> 80%	> 80%	NA	NA	> 80%	> 80%	> 80%	NA	60%-80%	NA
TCR-β	Positive	Weak	Positive	Positive	NA	Positive	Positive	Positive	Negative	Positive	NA
TIA-1	Negative	Positive	Positive	Negative	NA	Negative	Negative	Negative	Negative	Negative	Negative
Genetics											
6p25.3 rearrangement	Present	Present	Present	Present	Present	Present	Present	Present	Present	Present	Present
Follow-up											
Treatment	None	Excision	Radiation	Radiation	Excision, cryo	Radiation	Radiation	None	None	Excision	None
Spontaneous resolution	Yes	—	—	—	—	—	—	Yes	Yes	—	Yes
Skin recurrence	No	No	Yes	No	Yes	No	No	No	Yes	Yes	Yes
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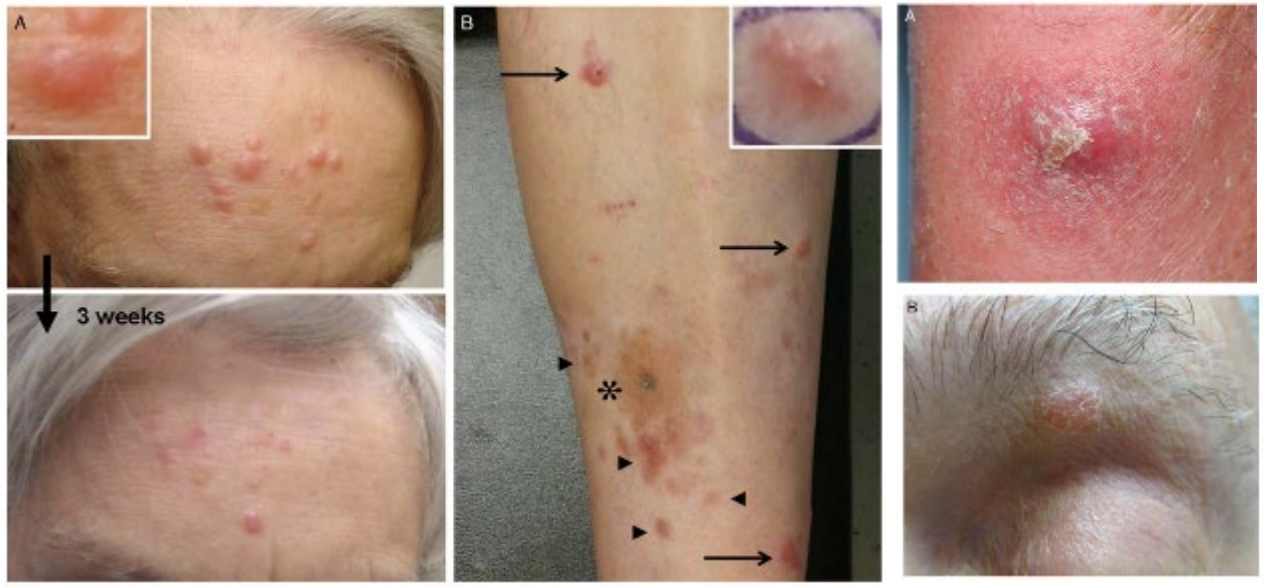

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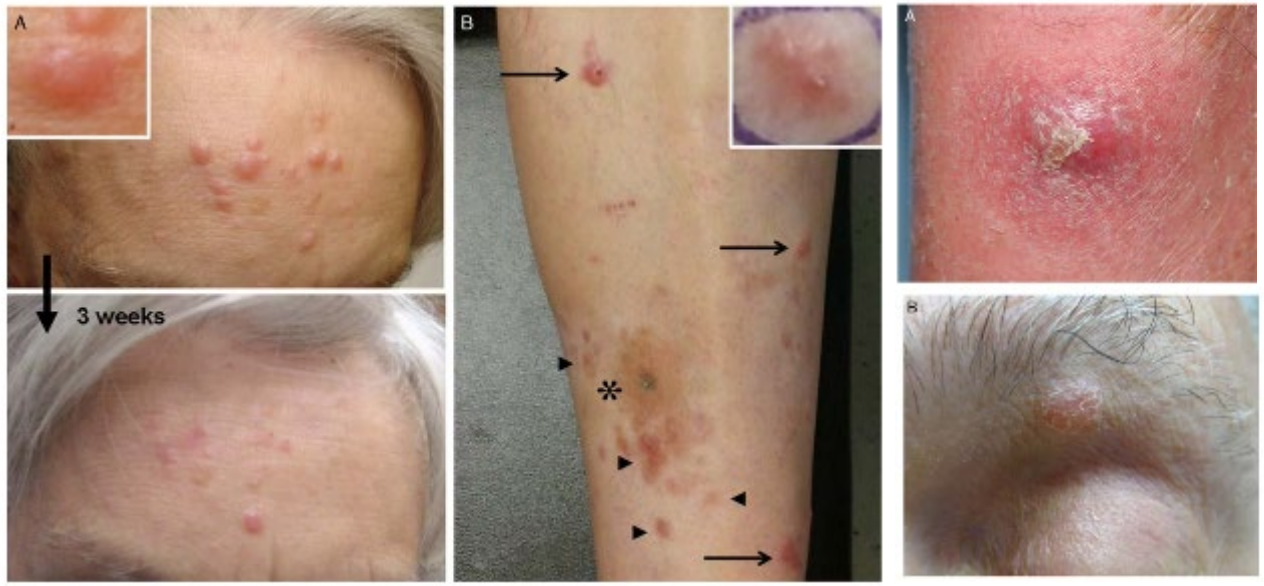

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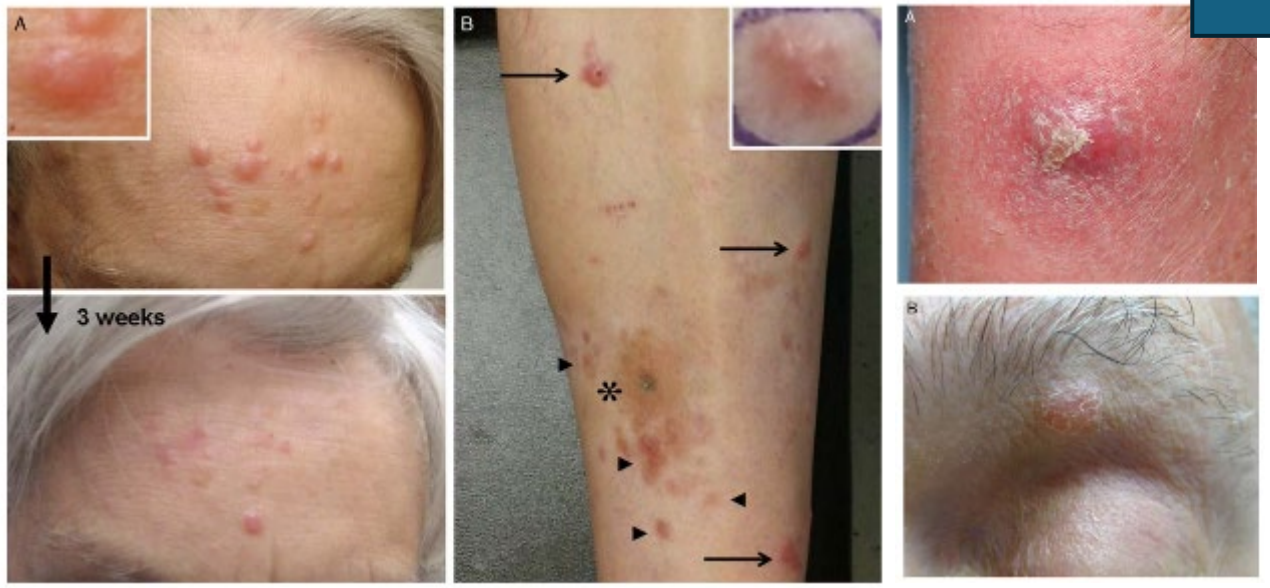

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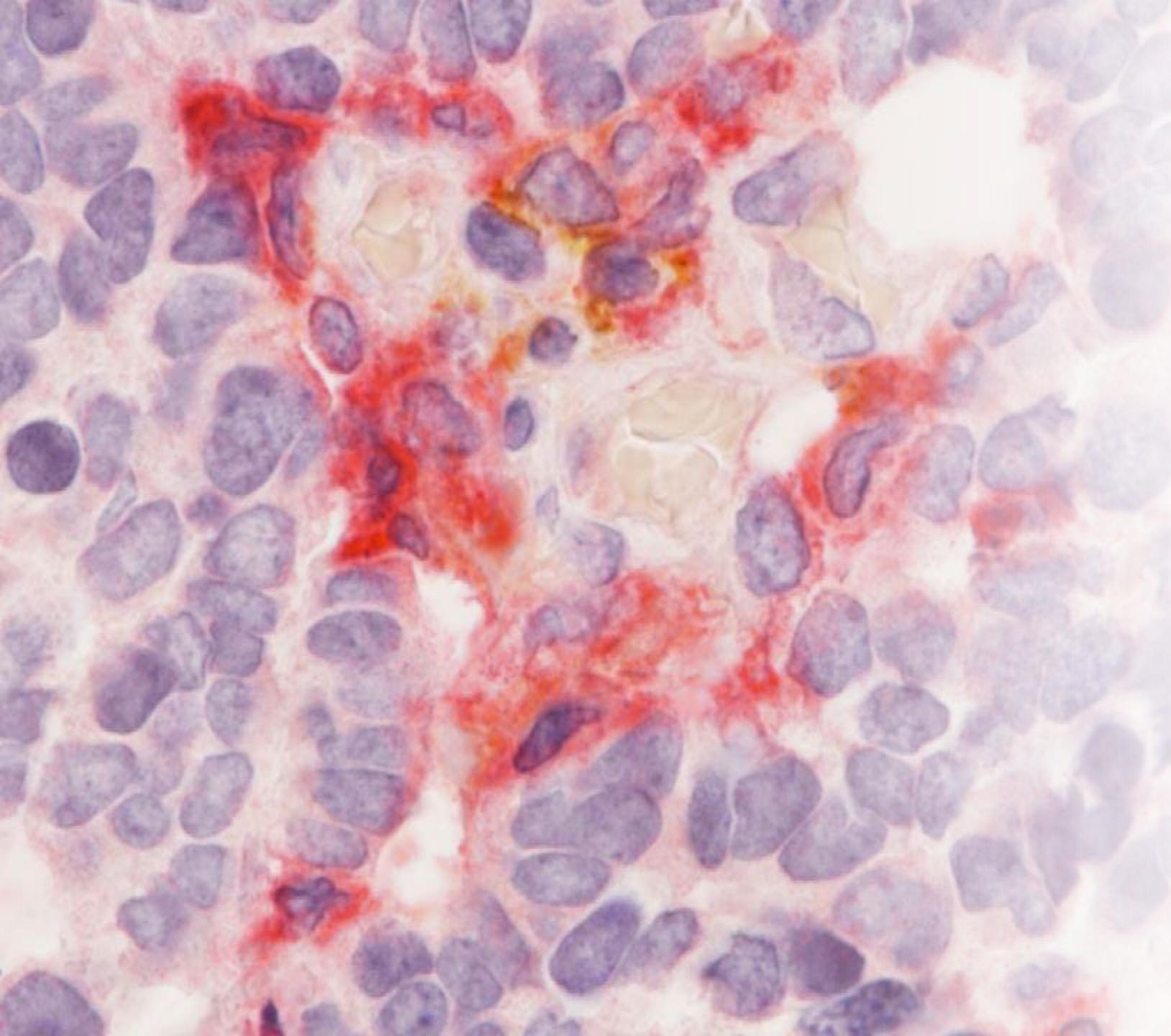
Eosinophils	Absent	Absent	Absent	Absent	Present	Focal	Absent	Absent	Absent	Absent	Absent
Pathologic diagnosis	CD30 LyP	LyP	CD30 LyP/ TLPD	CD30 LyP/ TLPD	CD30 LyP/ TLPD	CD30 LyP/ TLPD	CD30 LyP/ TLPD	CD30 LyP/ TLPD	LyP	CD30 LyP/ TLPD	LyP
Immunophenotype											
ALK	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Negative
CD3	Positive	Weak	Negative	Positive	Positive	Positive	Positive	Positive	Positive	Positive	Positive
CD4	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Positive	Negative	Negative	Partial
CD8	Negative	Negative	Negative	Weak	Positive	Positive	Negative	Positive	Negative	Partial	Negative
CD15	Focal	Positive	Negative	Positive	Positive	Focal	Negative	Focal	NA	Negative	NA
CD30	D > E	D > E	Positive	Positive	Positive	D > E	D > E	Positive	Positive	D > E	Positive
Ki-67	> 80%	> 80%	> 80%	NA	NA	> 80%	> 80%	> 80%	NA	60%-80%	NA
TCR-β	Positive	Weak	Positive	Positive	NA	Positive	Positive	Positive	Negative	Positive	NA
TIA-1	Negative	Positive	Positive	Negative	NA	Negative	Negative	Negative	Negative	Negative	Negative
Genetics											
6p25.3 rearrangement	Present	Present	Present	Present	Present	Present	Present	Present	Present	Present	Present
Follow-up											
Treatment	None	Excision	Radiation	Radiation	Excision, cryo	Radiation	Radiation	None	None	Excision	None
Spontaneous resolution	Yes	–	–	–	–	–	–	Yes	Yes	–	Yes
Skin recurrence	No	No	Yes	No	Yes	No	No	No	Yes	Yes	Yes
Disseminated skin disease	No	No	No	No	No	No	No	No	No	No	No
Extracutaneous disease	No	No	No	No	No	No	No	No	No	No	No
Status last follow-up	NED	NED	LyP, same site	NED	NED	NED	NED	NED	NED	NED	NED
Months since onset	21	150	12	7	21	30	36	10	104	22	20
Months disease free	20	26	–	5	3	22	32	9	2	22	8

ALK indicates anaplastic lymphoma kinase; cryo, cryotherapy; D > E, positive staining stronger in dermal component than in epidermal component; F, female; M, male; NA, not available; NED, no evidence of disease.

- 16 cases reported, LyP subtype (WHO 5th ed.)
- Older patients, male predominance
- Solitary or localized, not generalized
- No ulceration
- Spontaneous regression common
- CD4-/CD8-, CD4-/CD8+, CD30+, TCRb > TCRg
- Indolent course



Thank you!

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