Summer Academy 2026 Graz

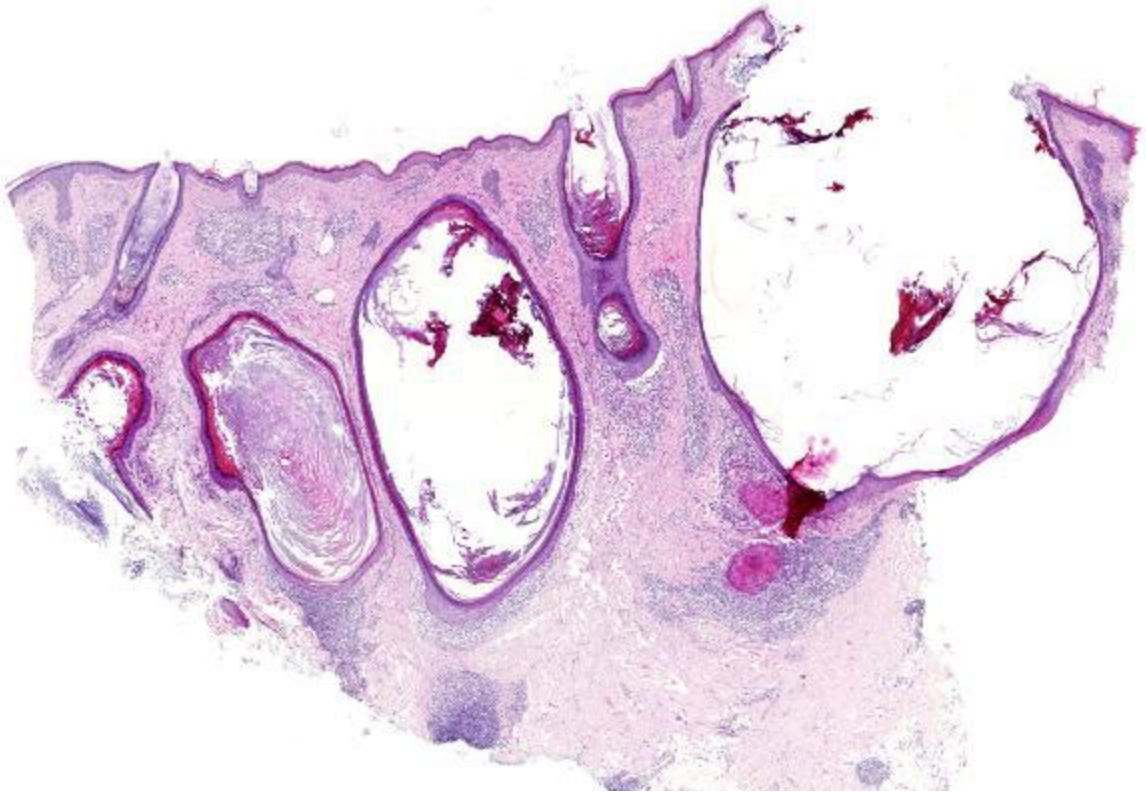
Cases from Zurich - a potpourri

Werner Kempf

Dept. of Dermatology
University Zürich

Kempf und Pfaltz
Histologische Diagnostik, Zürich
Switzerland

45-year-old woman: Grouped comedones on the right cheek since 6 months.



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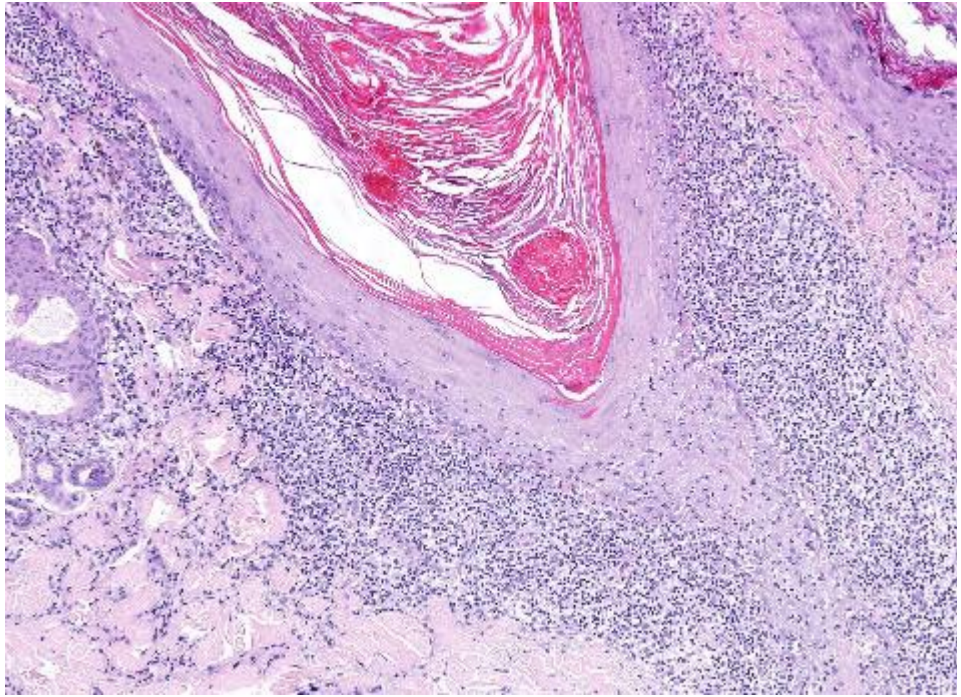
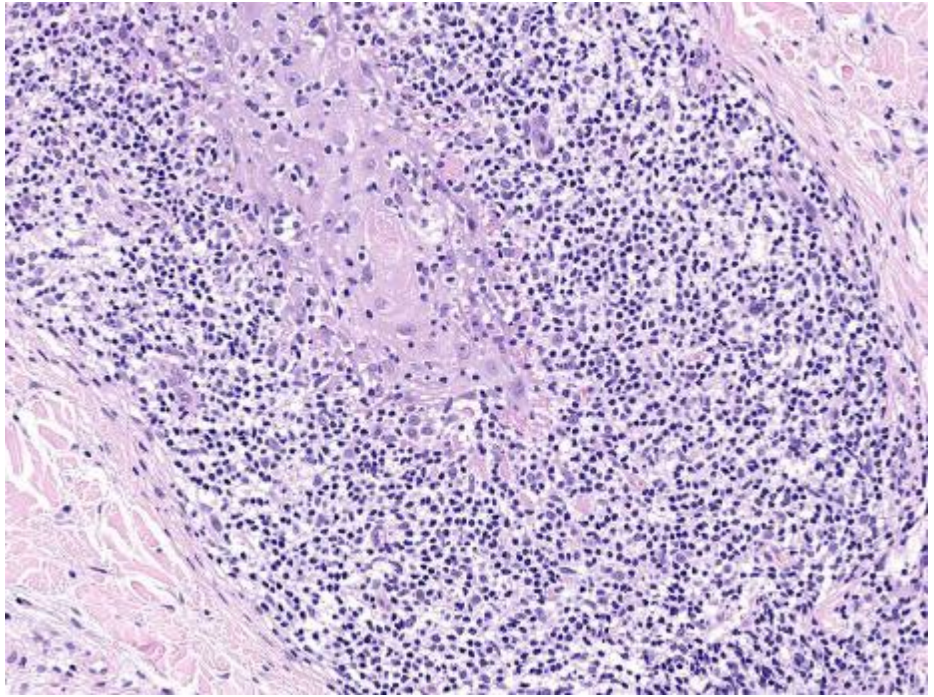
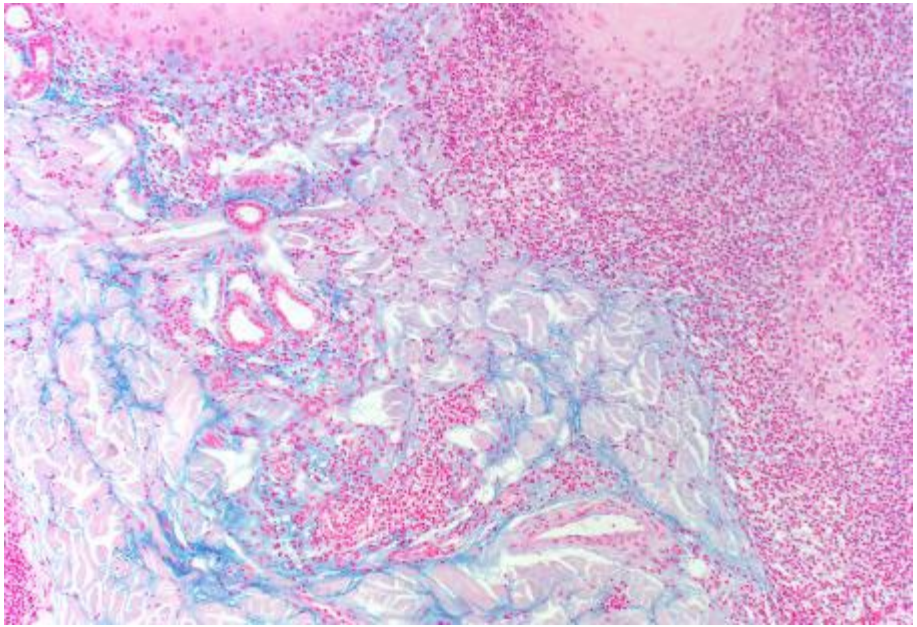
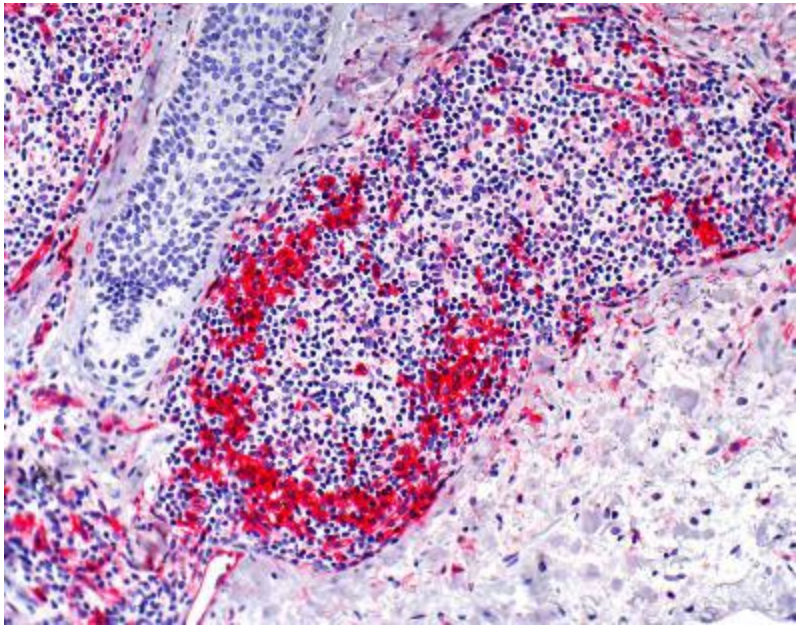






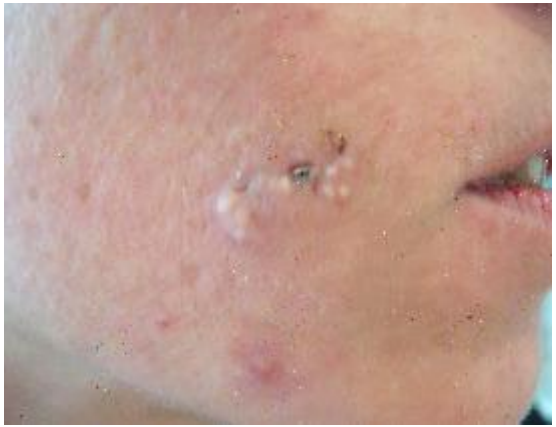
Alcian blue

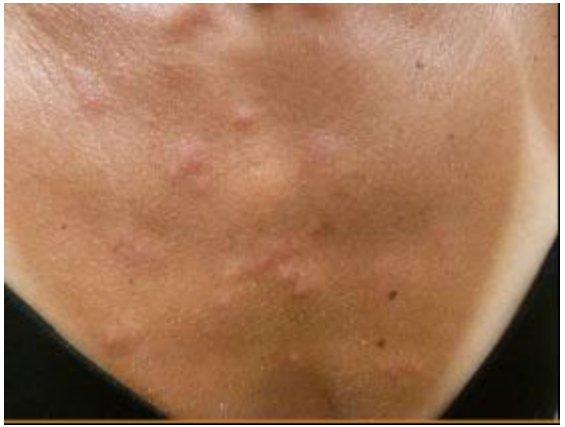


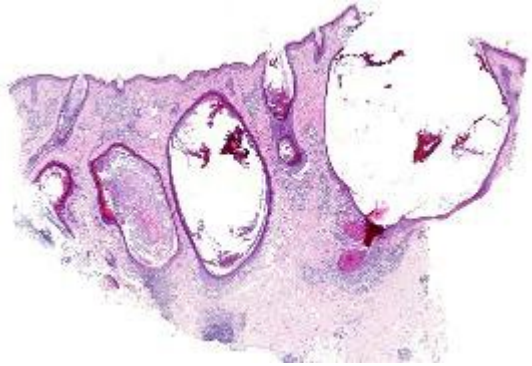
CD 123

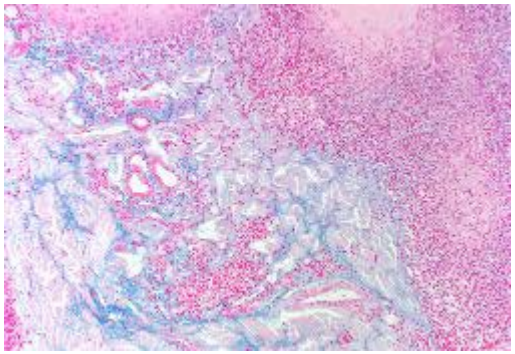
Direct immunofluorescence: Granular deposits of IgG, IgM and C3 along junctional zone

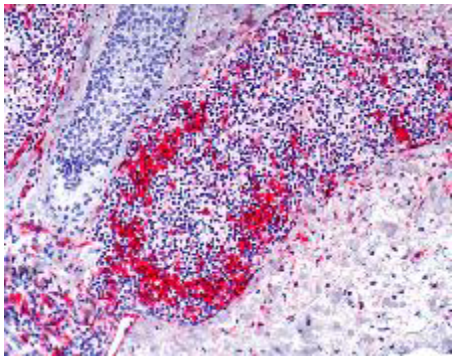
45-year-old woman: Grouped comedones on the right cheek since 6 months.







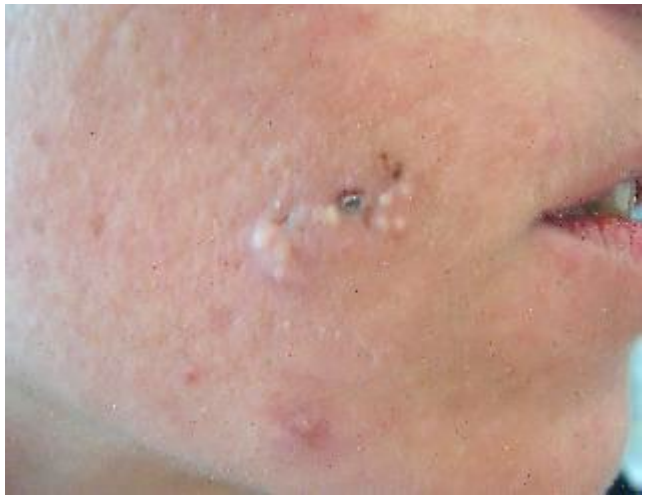


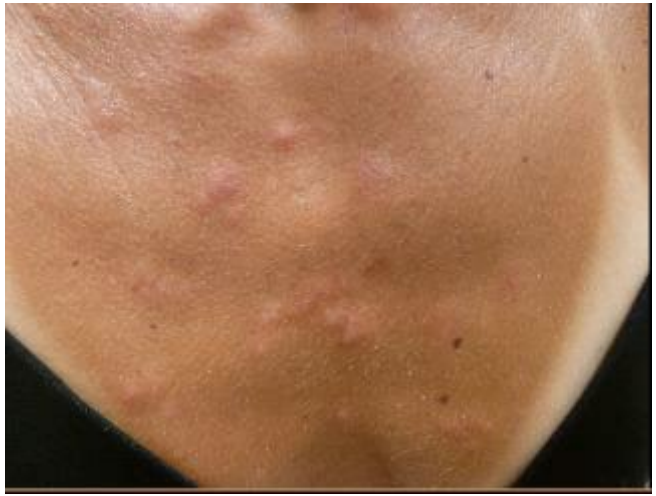


CD 123

Chronic cutaneous lupus erythematosus - comedonicus variant

Milia-studded plaque(s) or cystic lesions
Additional DLE lesions may be present syn- or metachronously





Tx: Excision, intralesional injection of steroids, hydroxychloroquine, retinoids

Surgical excision

Lupus comedonicus

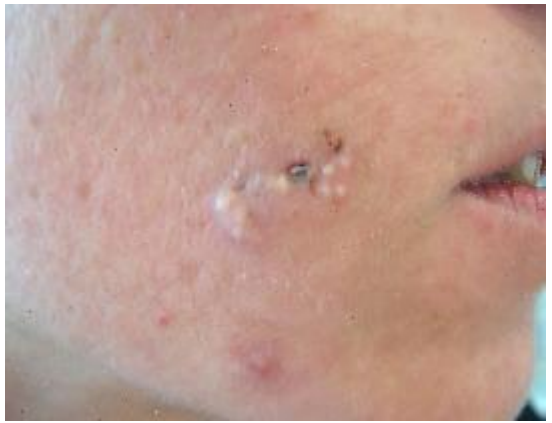
Table 1. Unusual subsets of cutaneous lupus erythematosus

Sontheimer R, et al. (1987)
- LE profundus
- LE hypertrophicus

Rowell N. (1992)
- LE tumidus
- LE telangiectaticus
- Annular atrophic plaques on the face
- LE gyratus repens
- LE bullosus
- Chilblain lupus
- Rowell's Syndrome
- LE profundus
- LE hypertrophicus et profundus

Braun-Falco O, et al. (1991)
- LE tumidus
- LE profundus
- Chilblain lupus
- LE hypertrophicus et profundus

Jablonska S, et al. (1993)
- LE edematosus
- LE tumidus
- LE hypertrophicus et verrucosus
- Chilblain lupus
- Annular atrophic plaques
- LE panniculitis



from: K.D. Pramatarov Clin Dermatol 2004

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Recurrent retroauricular cystic nodules: lichen planus follicularis tumidus

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[*]Authors contributed equally to this work

Affiliations: [1]School of Medicine, [2]Department of Pathology and Laboratory Medicine, [3]Department of Dermatology, University of California Davis, Sacramento, California, USA

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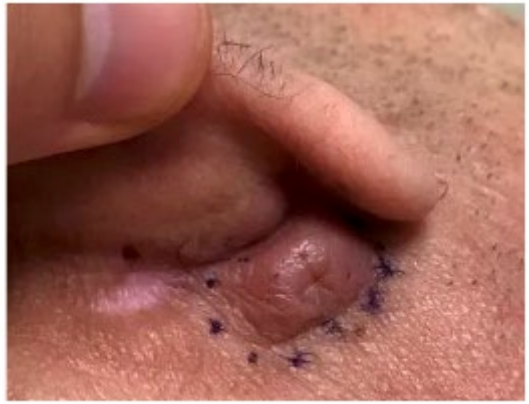
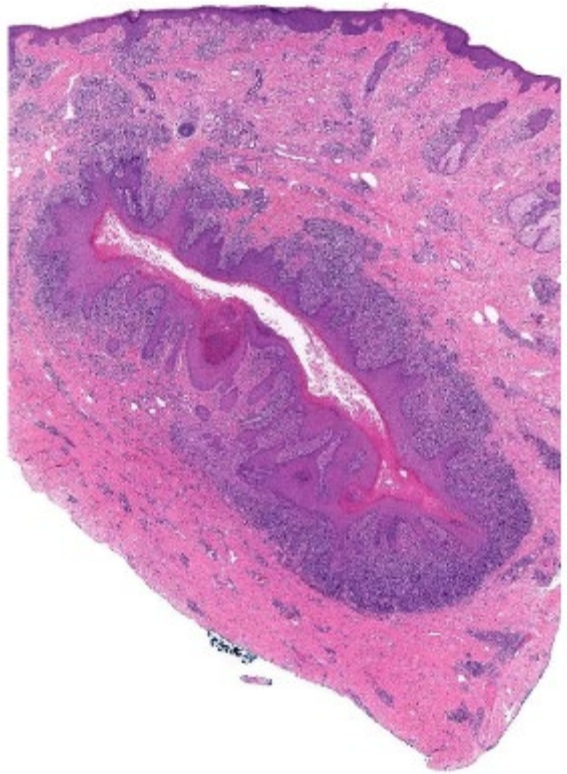
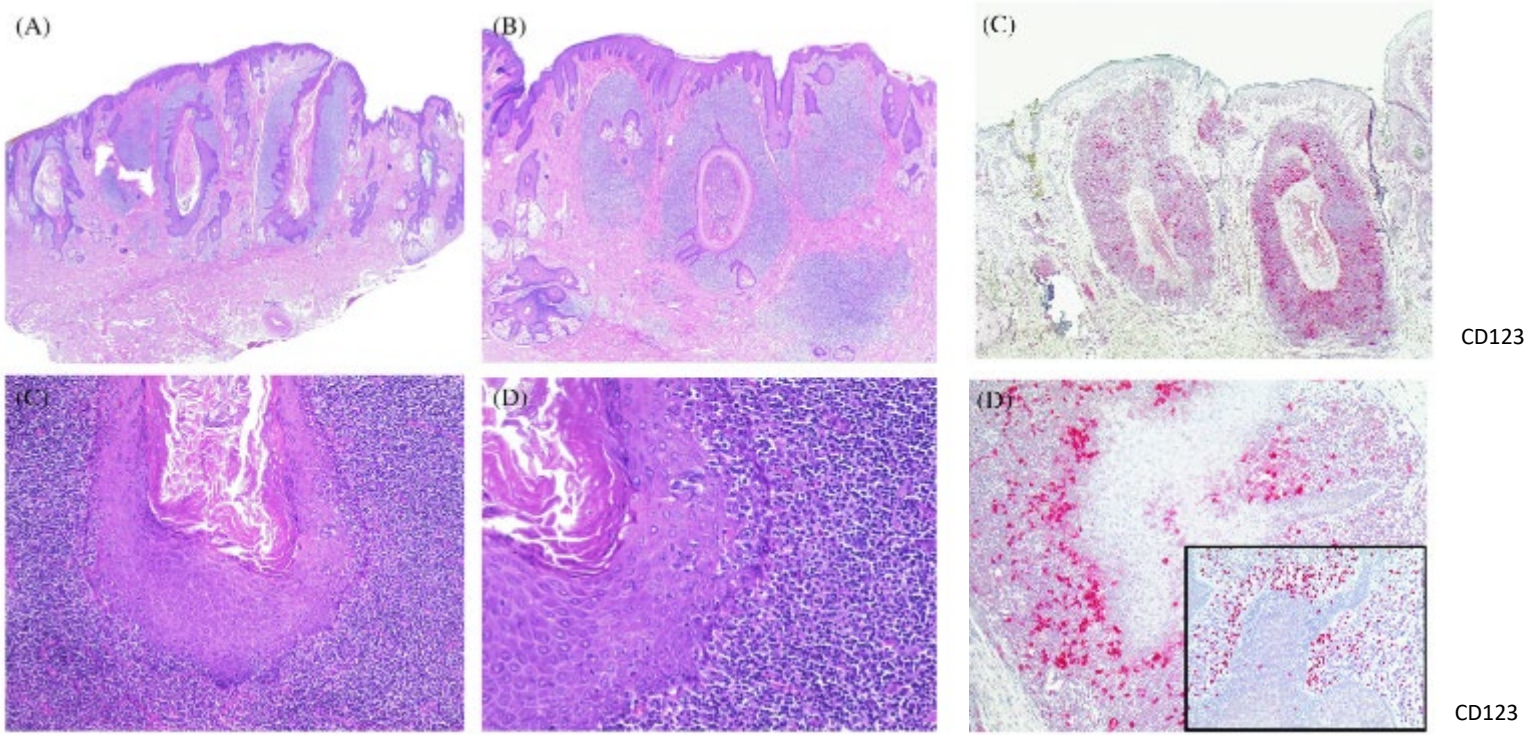




Figures from: Chau et al. Derm Online J 2018

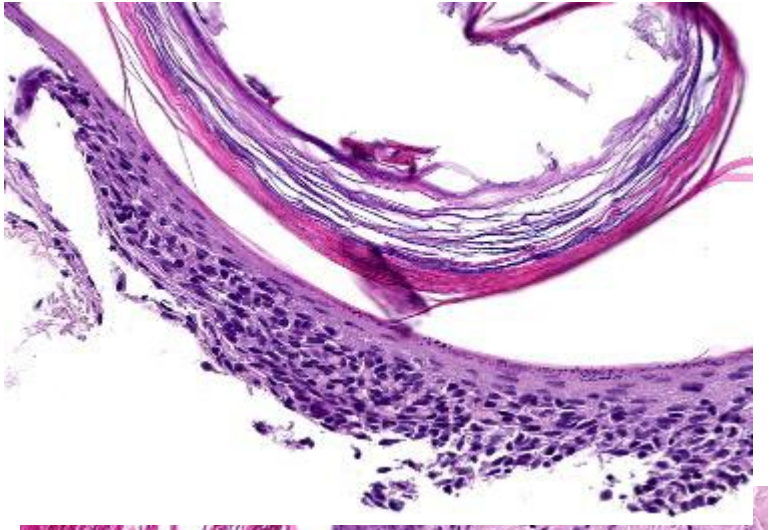
Lichen planus follicularis tumidus (LPFT) with cysts and comedones

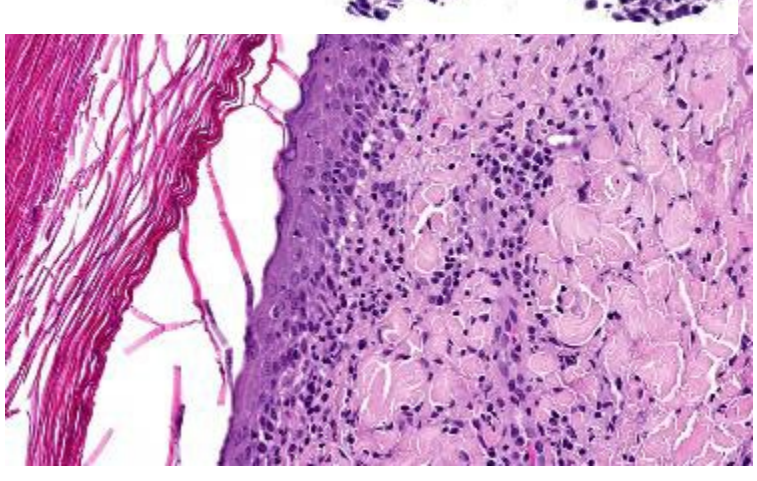
Very rare variant of LP most commonly involving retroauricular area, esp. in middle-aged females.



From: Saggini et al. J Cutan Pathol 2020

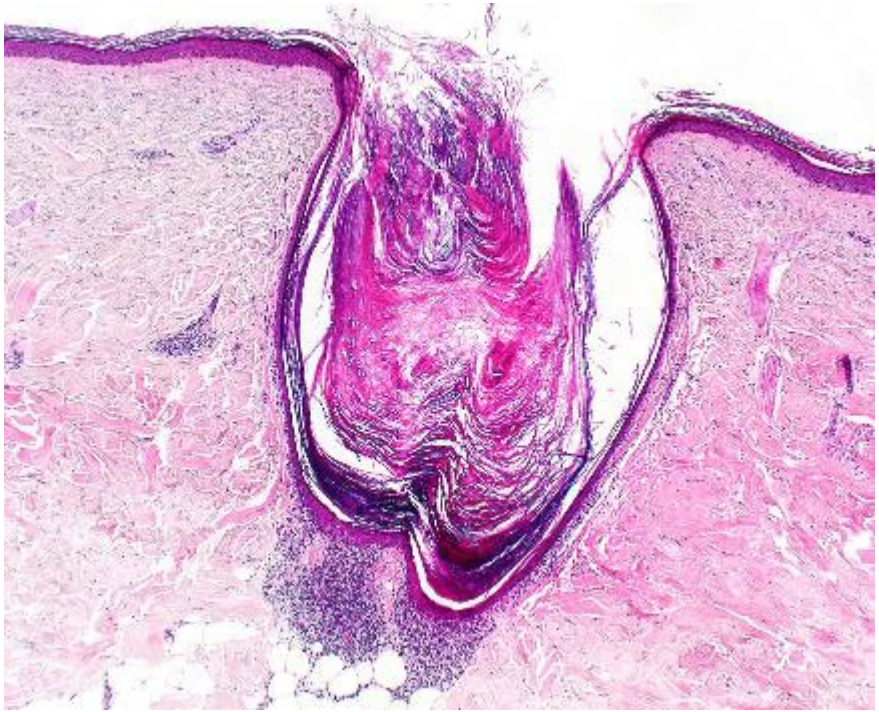
68-year-old woman with patchy alopecia around left ear and cystic lesions

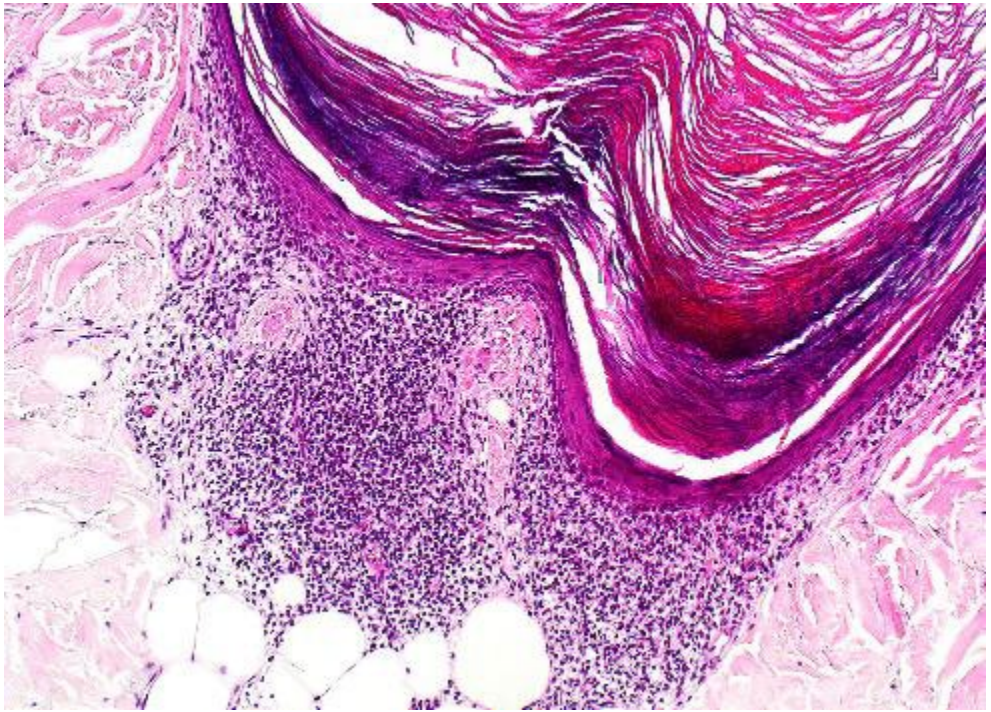




B17-113387

68-year-old woman: patchy alopecia around left ear and cystic lesions





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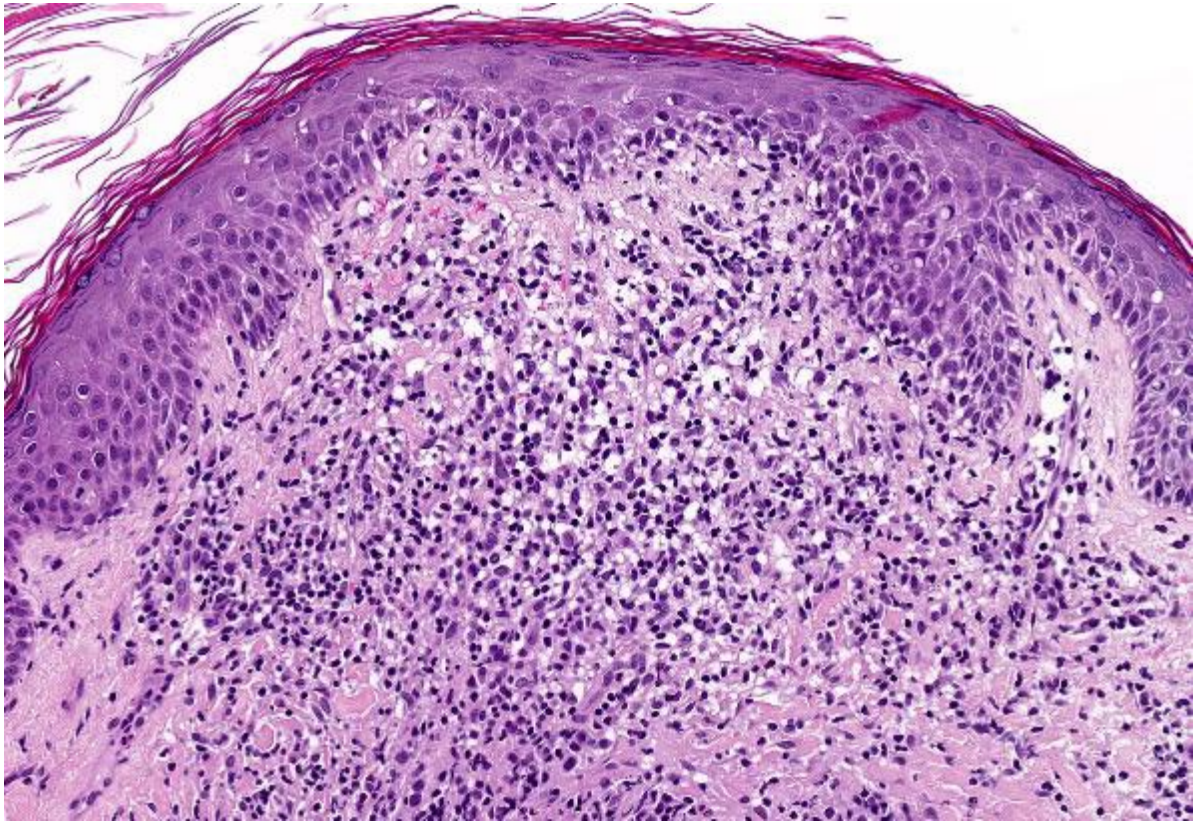

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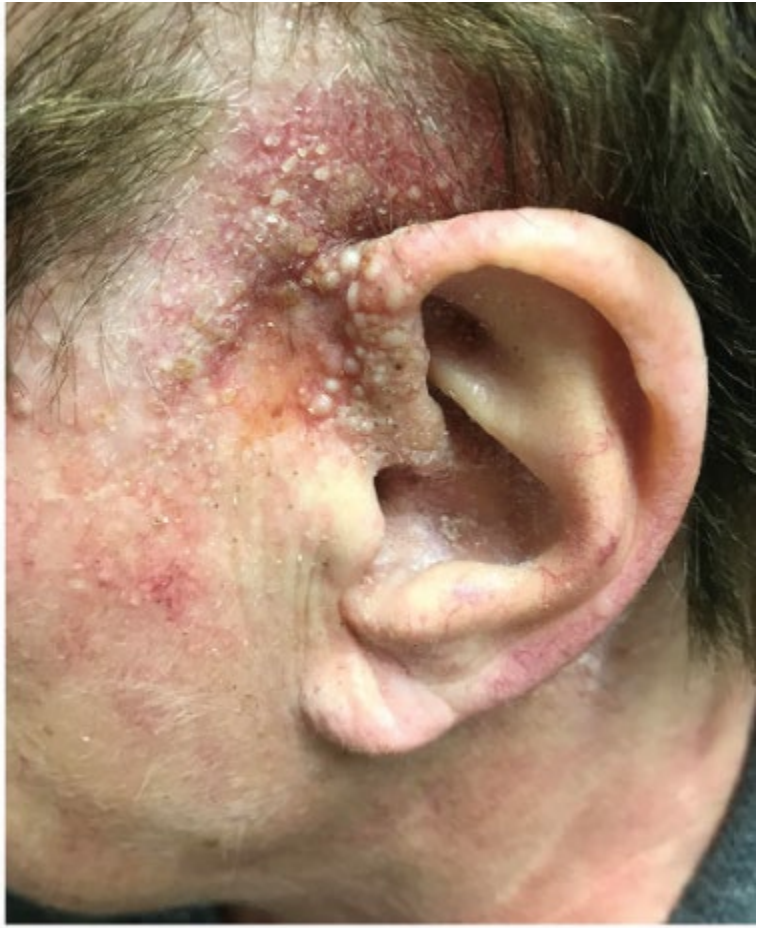

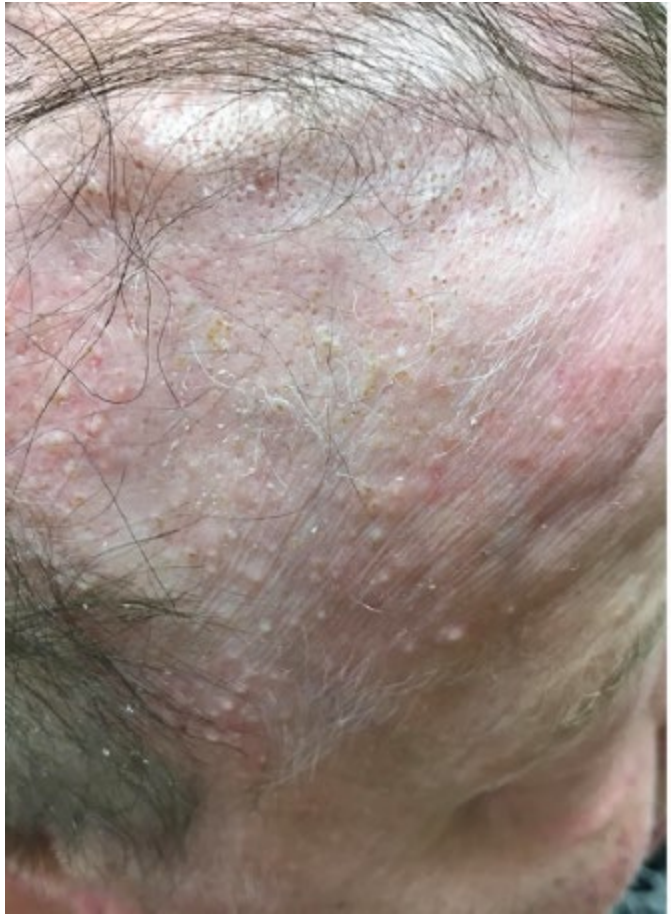

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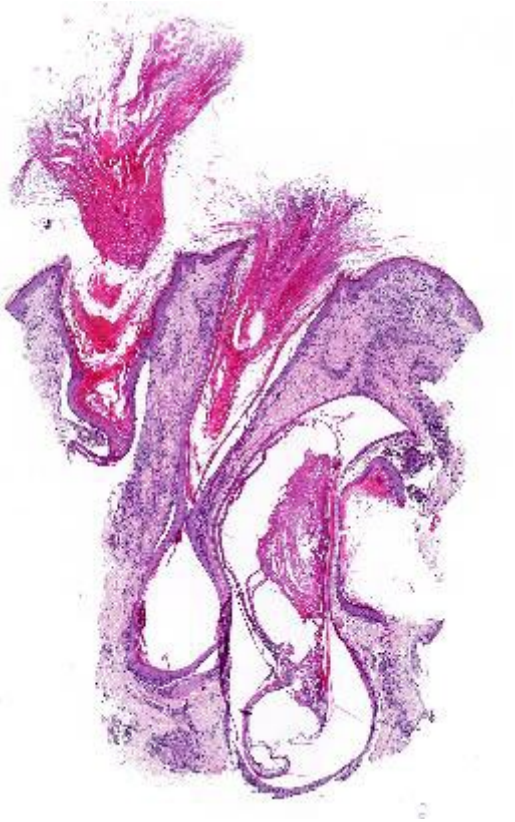
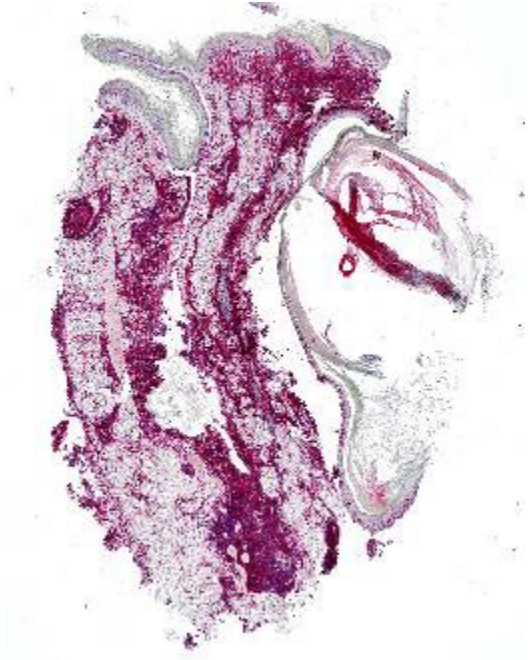
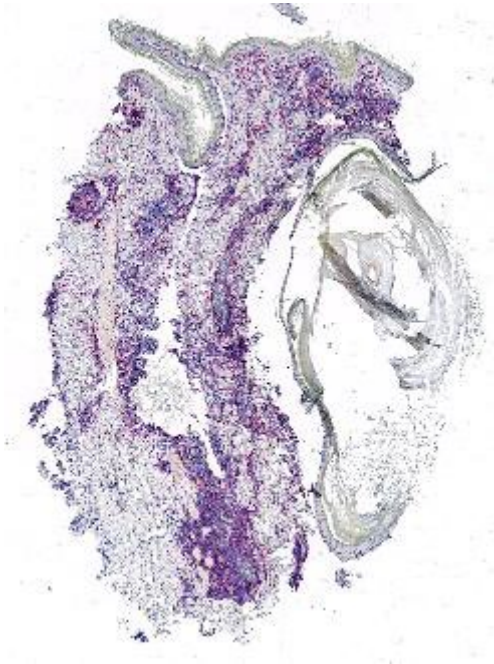


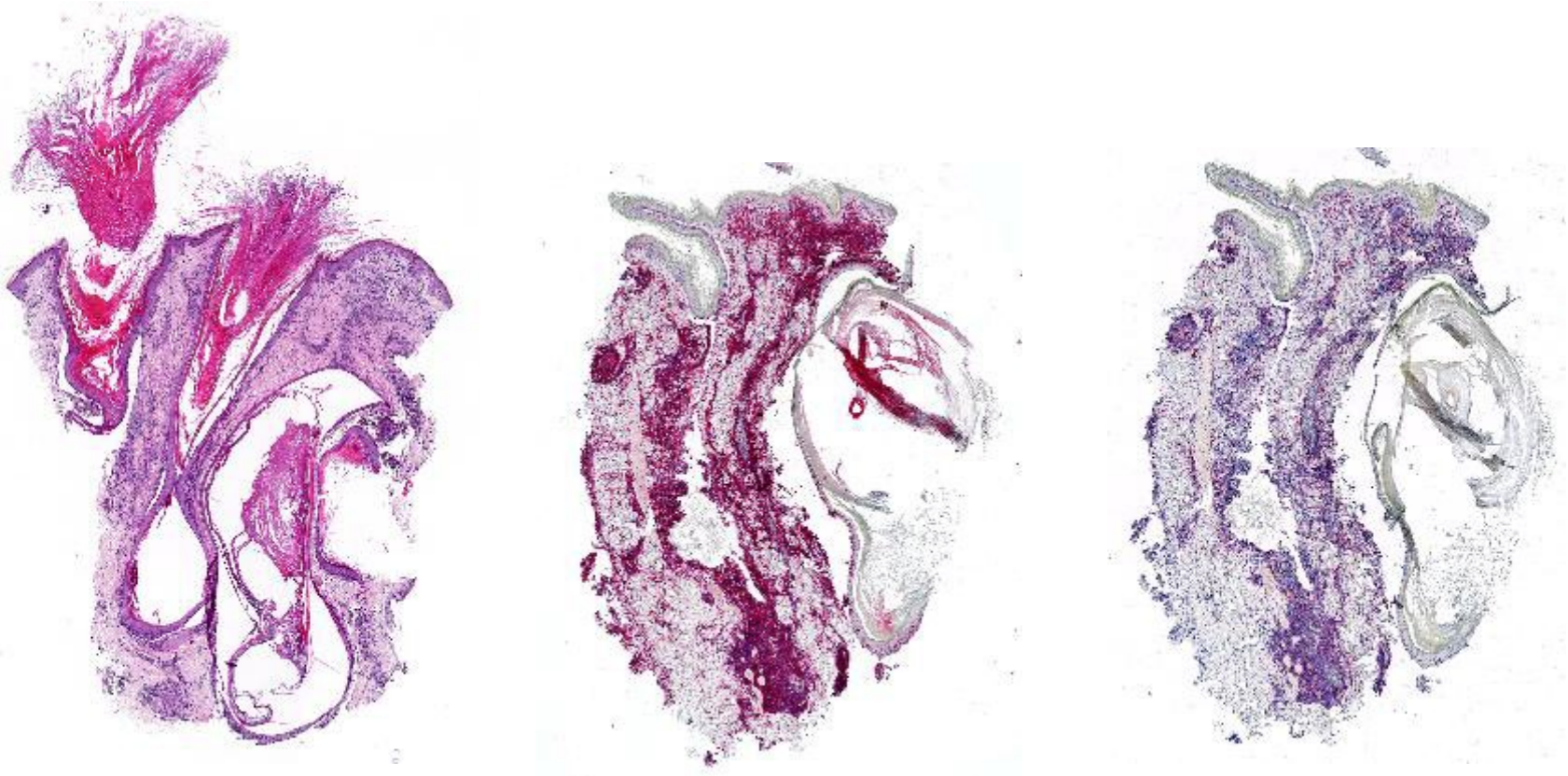


CD4



CD8

B17-113387



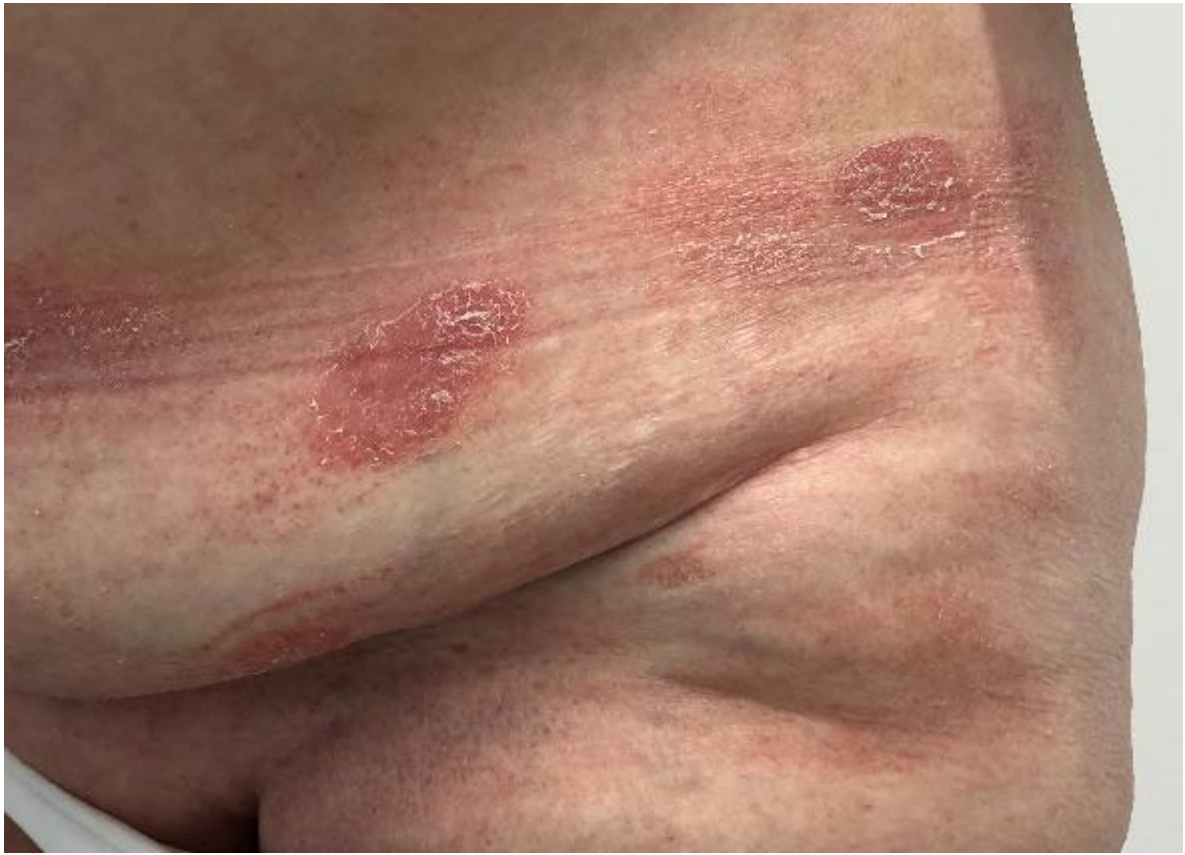
CD4

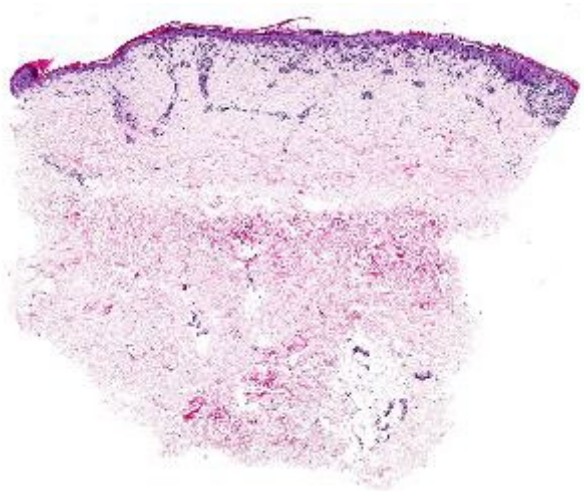
CD8

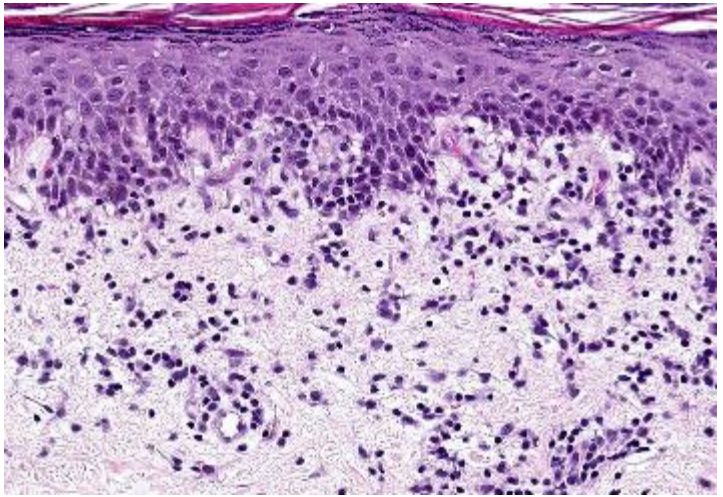
Monoclonal rearrangement of TCR genes with the same T-cell clone in two metachronous biopsies

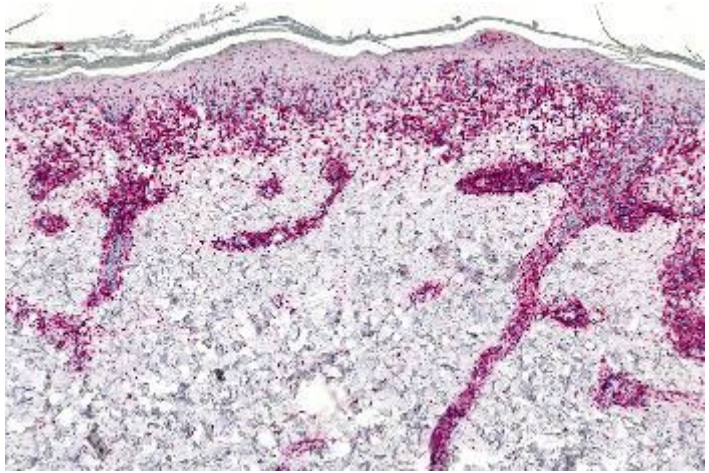
B17-113387

"Nummular atopic dermatitis" since several years









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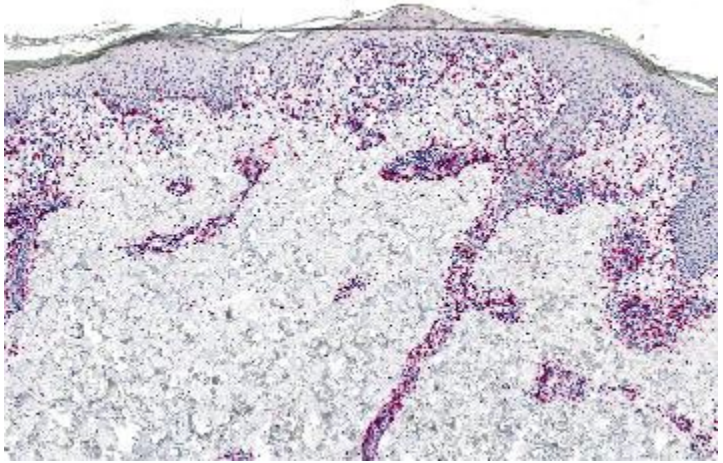
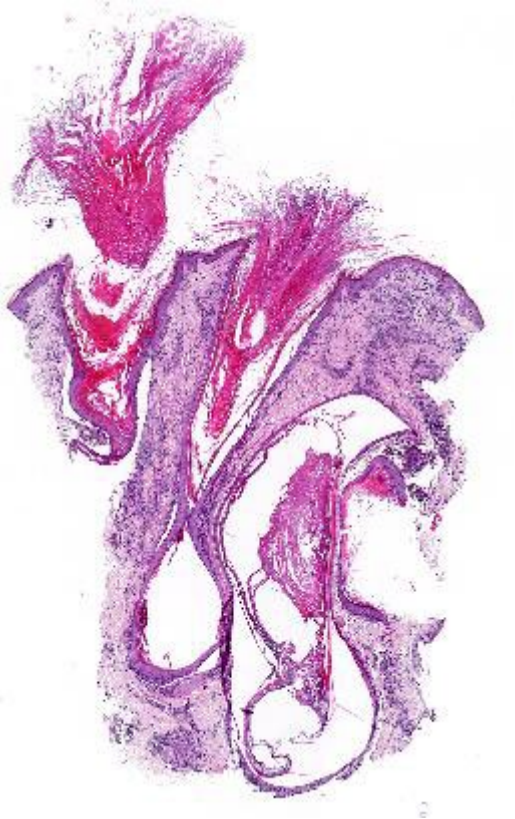
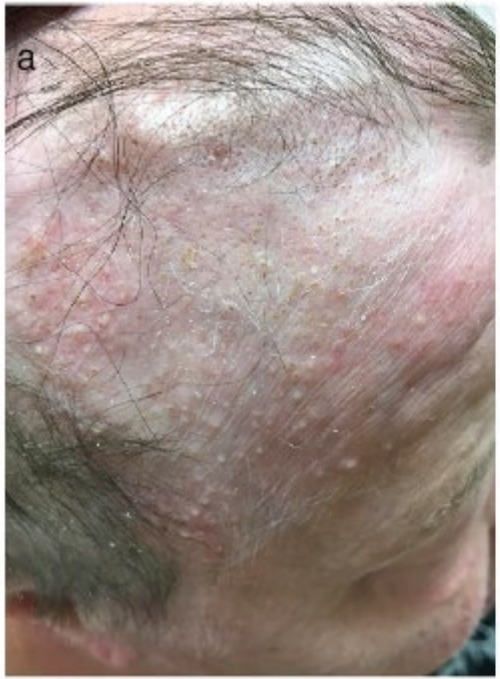


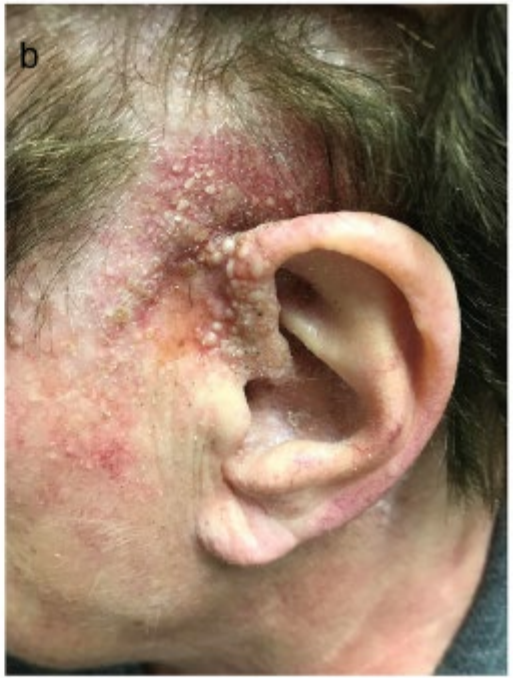
CD8

B17-52085

Folliculotropic mycosis fungoides with comedones and cysts



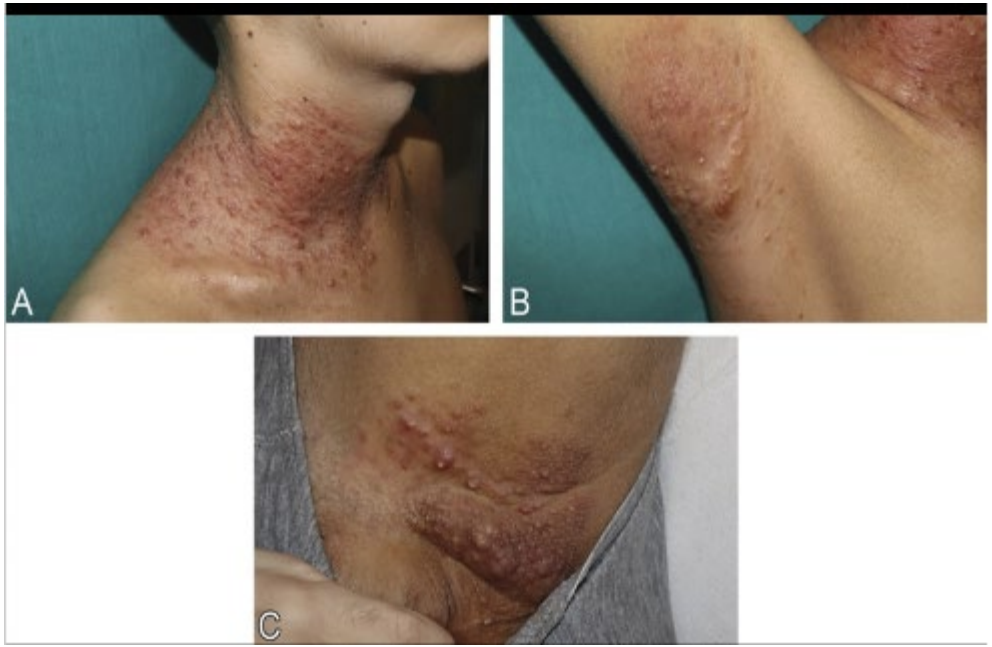


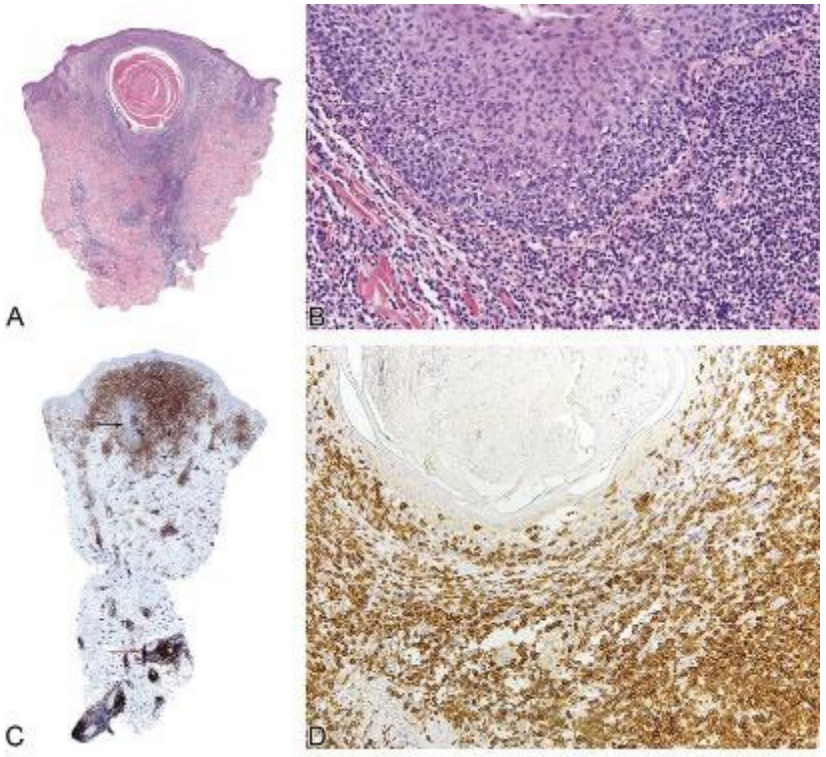


Iorizzo, Donghi, Kempf JDDG 2021

Folliculotropic Mycosis Fungoides With Flexural Eruptive Cysts and Comedones in a 14-Year-Old Boy

Juncal Roca-Ginés, MD, Clara Alfaro-Cervelló, MD, PhD,†‡ Javier Sánchez-Arraez, MD,* Ignacio Torres-Navarro, MD,* Miguel Á. Navarro-Mira, MD,* Carlos Monteagudo, MD, PhD,†‡ and Rafael Botella-Estrada, MD, PhD*§*





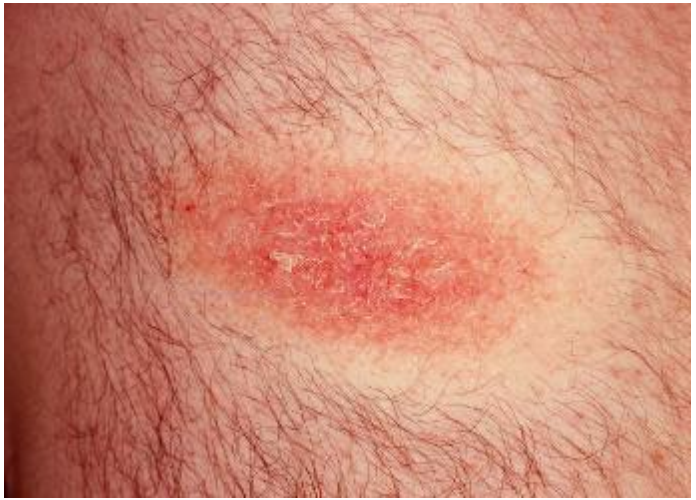
From: Roca-Ginés et al. Am J Dermatopathol 2021

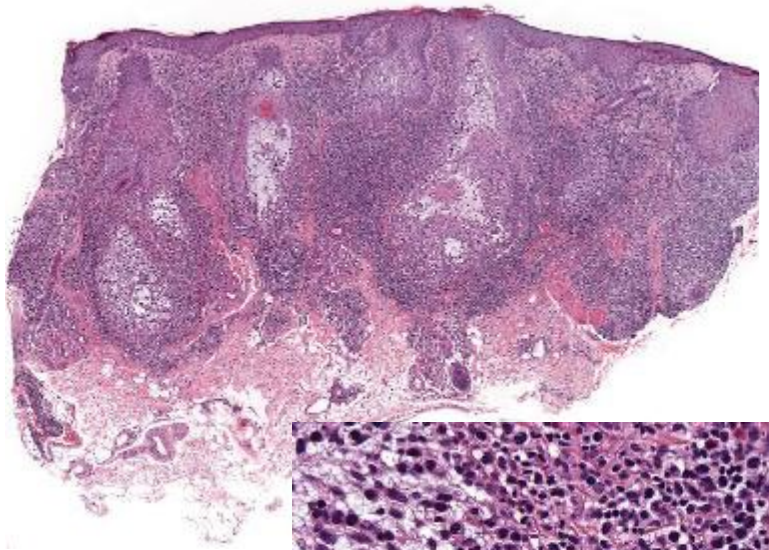
Folliculotropic mycosis fungoides

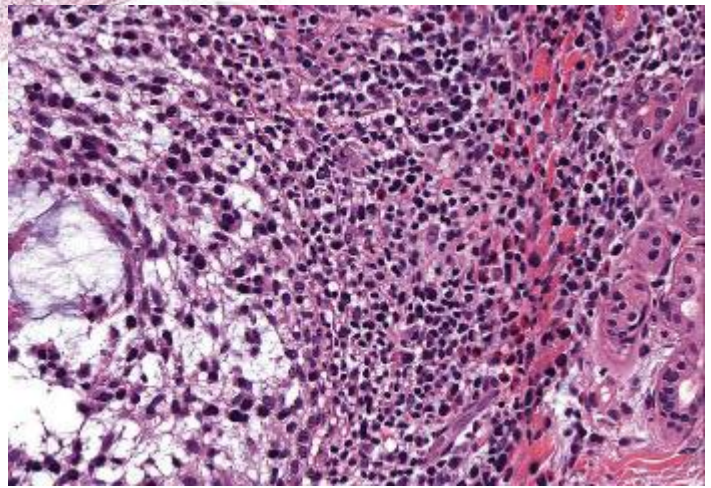
Patches and plaques with alopecia

Folliculotropic lymphocytic infiltrate

Mucinous degeneration of hair follicle epithelium: Follicular mucinosis (50%)

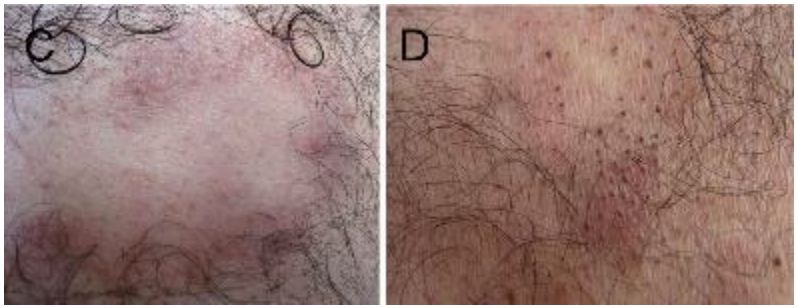


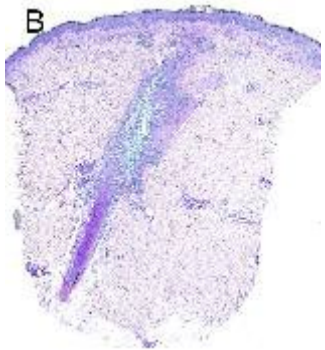




Folliculotropic MF – two prognostic forms

early / indolent

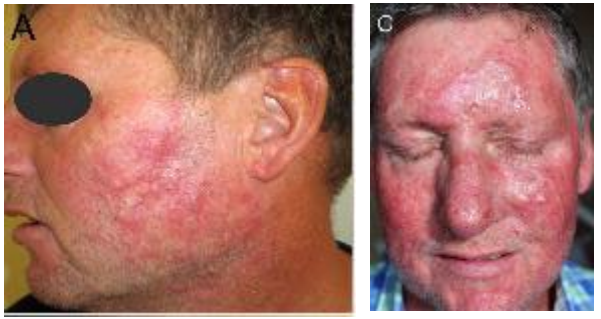


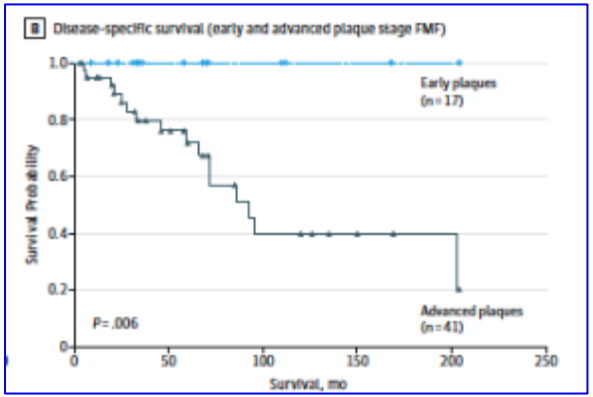


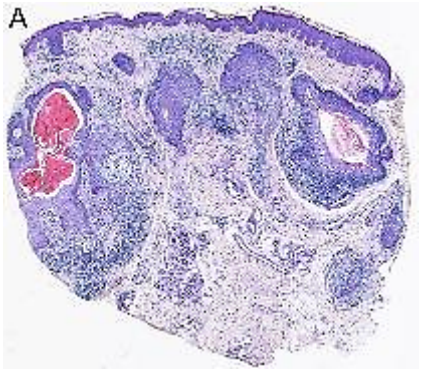
Patches / thin plaques, papules

10-y SR: 75%

advanced / aggressive







Thick plaques and tumors

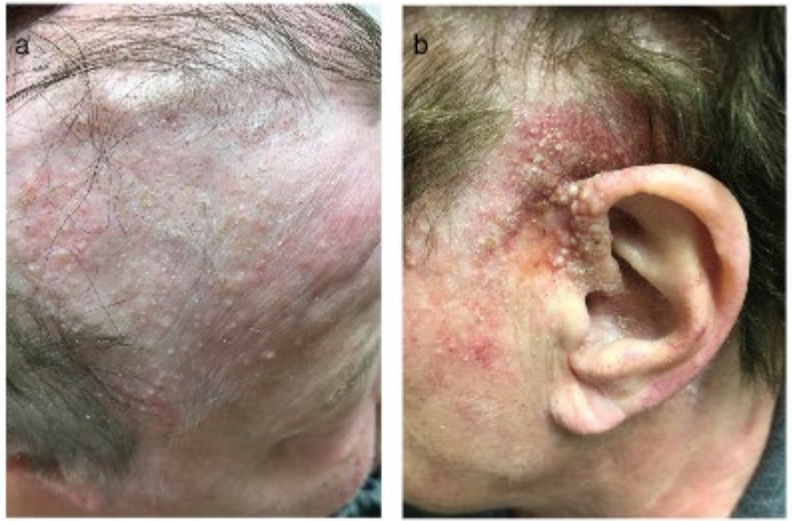
10-y-SR: 25%

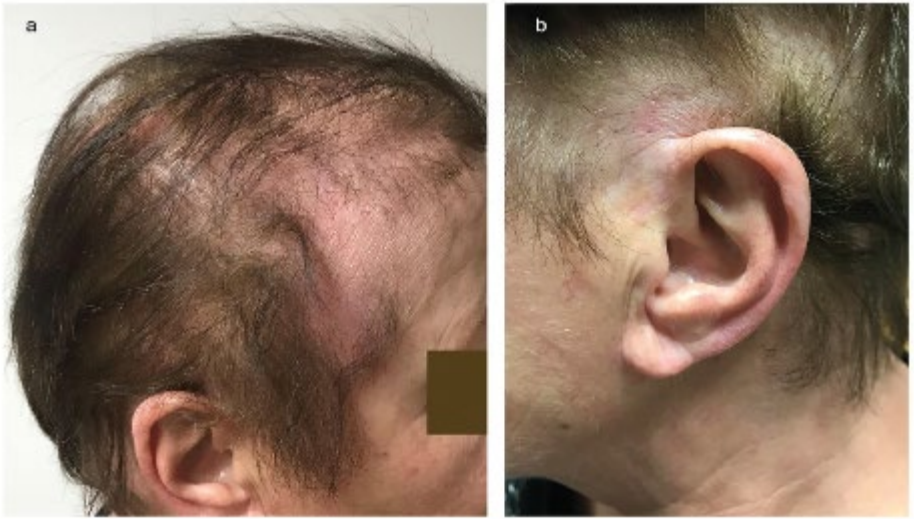
Hodak et al. J Am Acad Dermatol 2016

Van Santen et al. JAMA Dermatology 2016

Images: Mitteldorf et al. JDDG 2018

Folliculotropic MF with comedones and cysts

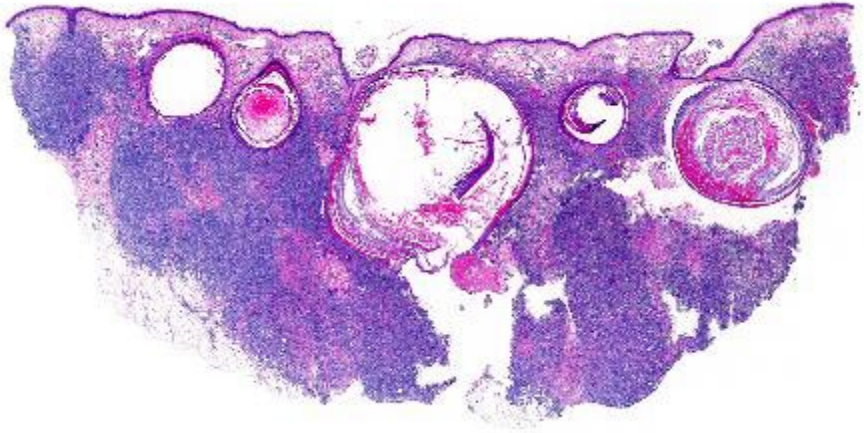


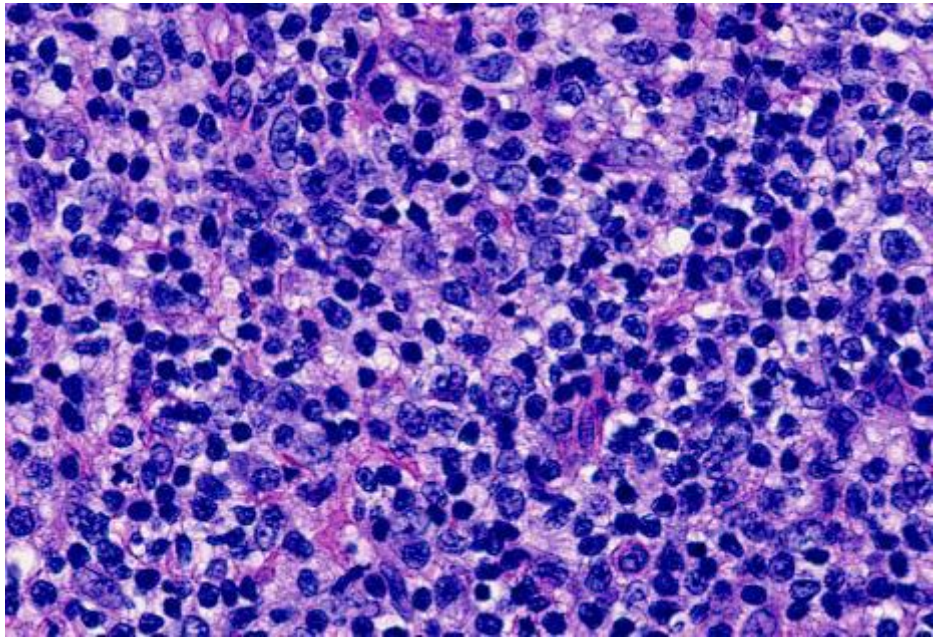


Response to bexarotene within 4 months.

Iorizzo, Donghi, Kempf JDDG 2021

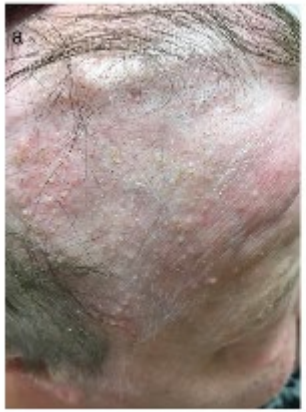
Progression to tumor stage (occiput) within 3 years

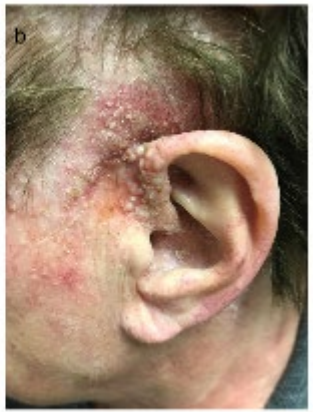


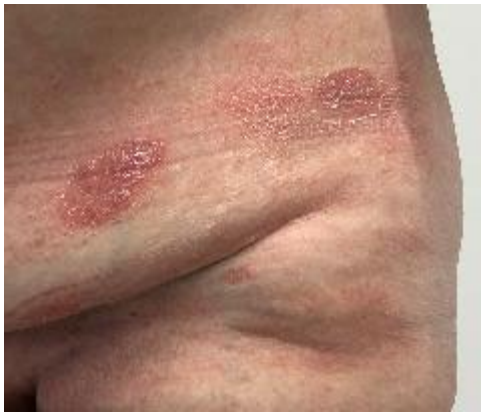


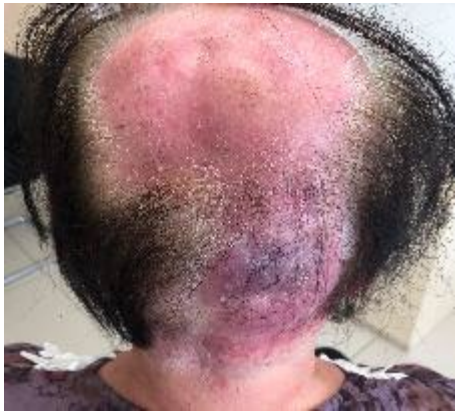
2020: Progression under treatment with bexarotene

2022: Death due to lymphoma progression



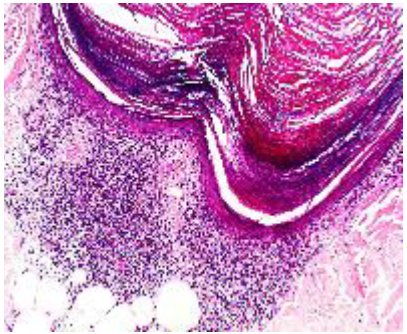


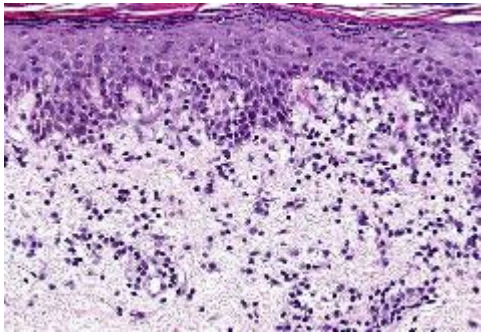


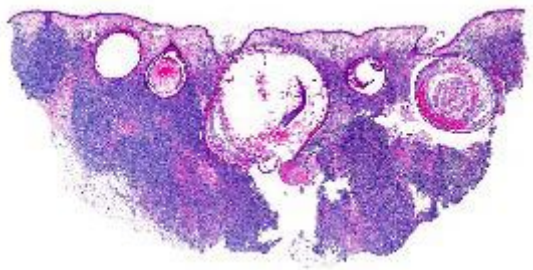


2017

2020

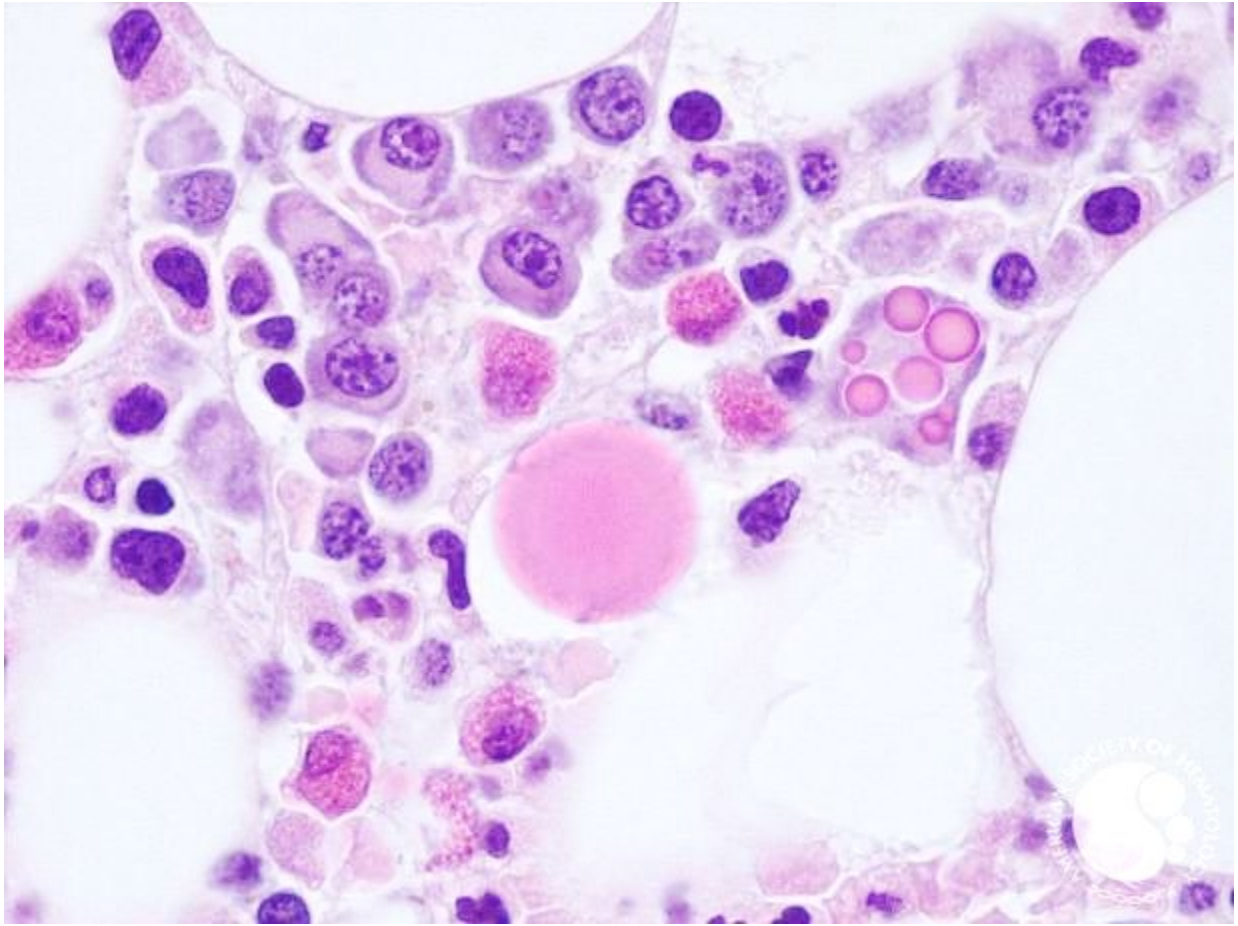






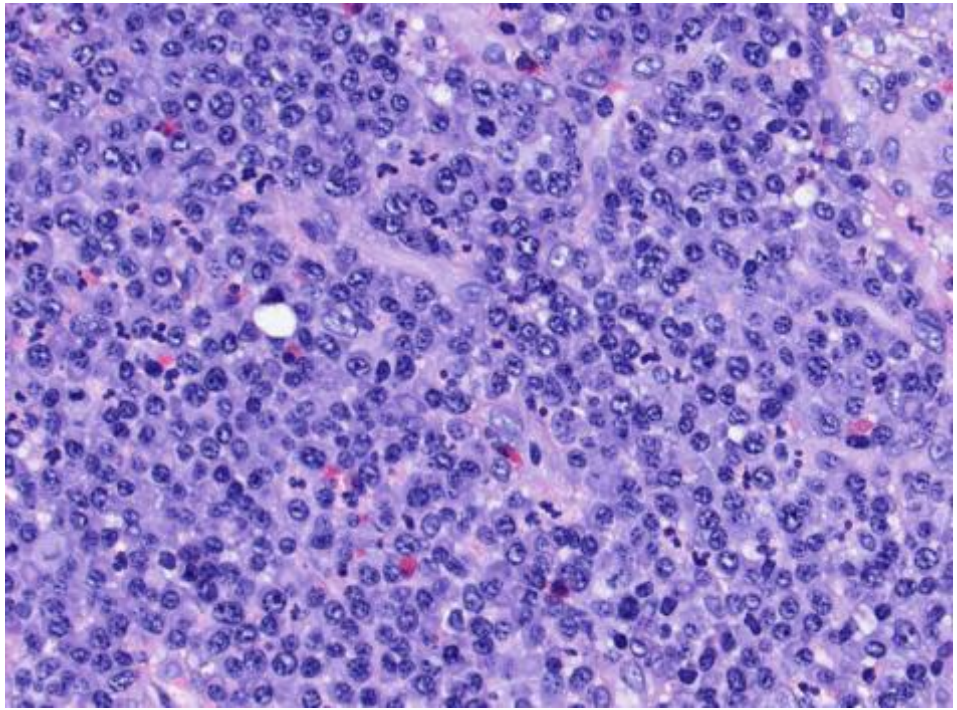
Differentialdagnosis

	LE comedonicus	LPP comedonicus	FMF with cysts
Cystic dilatation	++	++	++
Interface	++	++	(+)
Nuclear atypia	-	- / +	+
Mucin	++	-	-
PDC (CD123)	++ Cluster	+ (DEJ)	-
DIF	+ (IgM>IgG>IgA, C3 granular)	Cytoid bodies	-
Clonality (mulitple lesions)	- (?)	- (?)	+



From: Imagebank.hematology.org

Plasma cell rich infiltrates – DDx



The „6 L“ rule

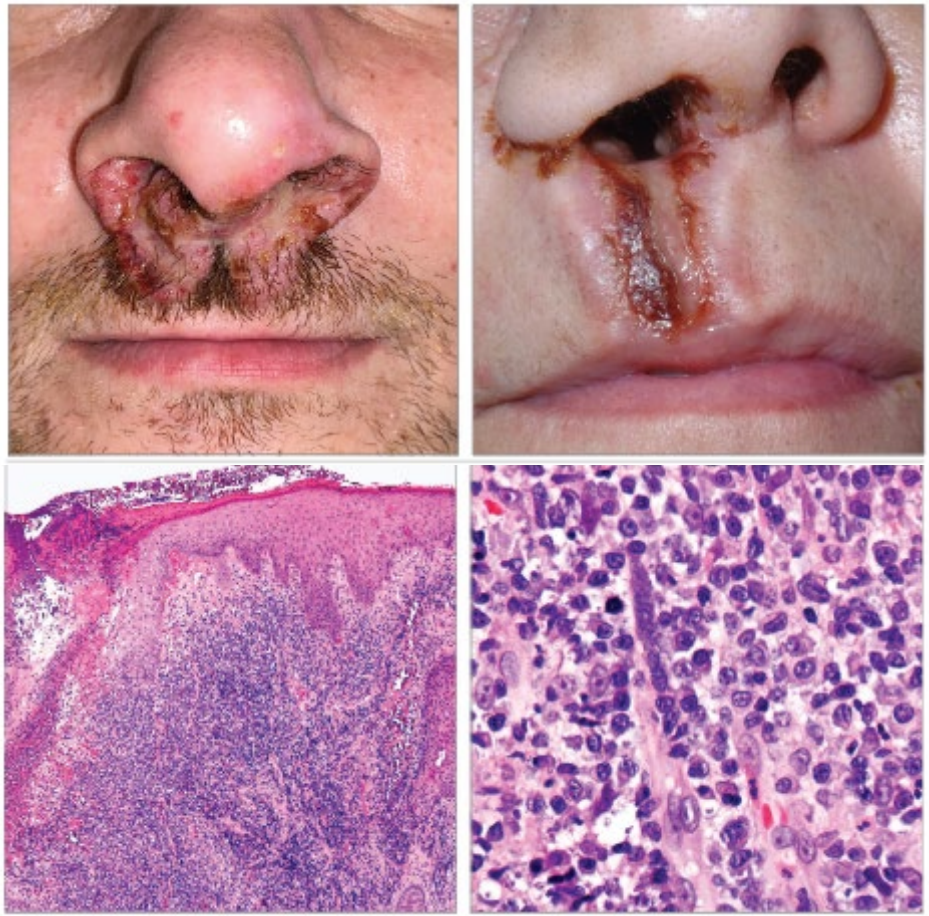
- Lues
- Lyme / borreliosis
- Leprosy
- Lupus vulgaris (Tbc)
- Leishmaniose
- Lymphomas

Cocaine-associated plasma cell orificial mucositis

Exsudative ulcerated plaques and septal or palate perforations (40%)

Dense inflammatory infiltrate of plasma cells without atypia and with eosinophils.

Viedma-Martinez et al. JAMA Dermatol 2024

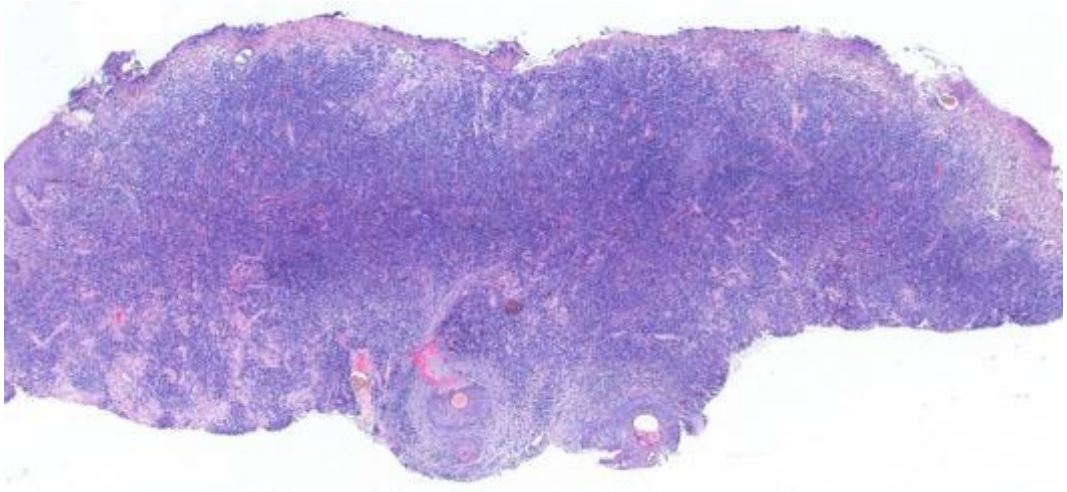


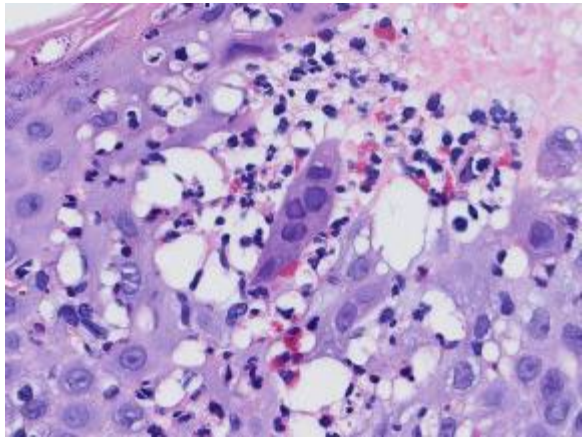
From: Viedma-Martinez et al. JAMA Dermatol 2024

Ulcerated tumor-like herpes infection

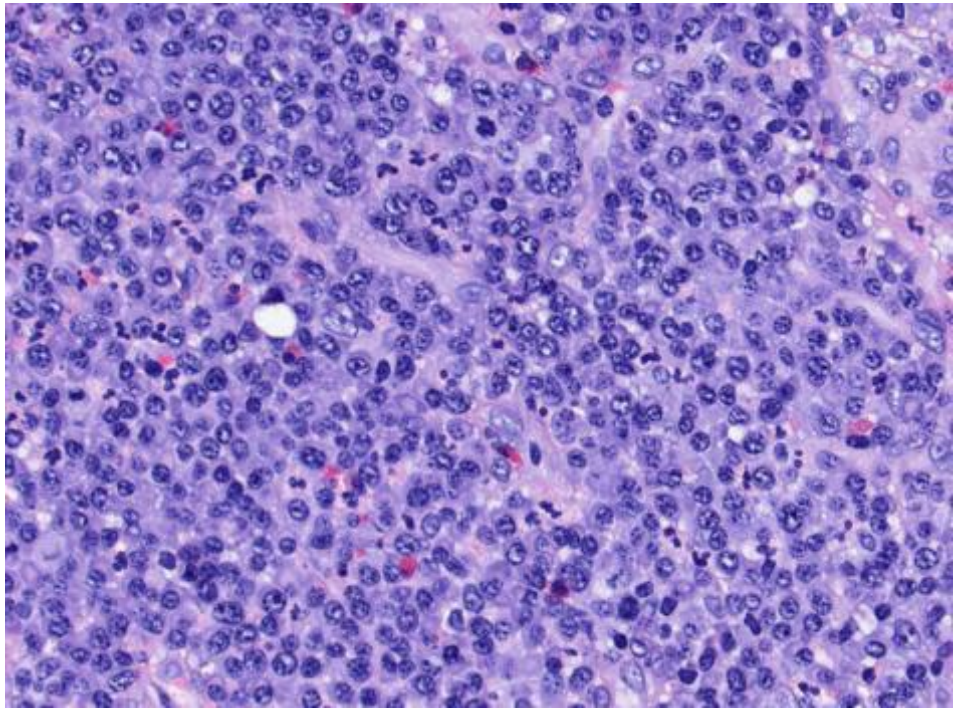
Immunosuppressed patients: HIV Infection, leukemia (B-CLL)

Roemer et al. JDDG 2008
Lautenschlager et al. Dermatology 2008





Plasma cell rich infiltrates – DDx



The „6 L“ rule

- Lues
- Lyme / borreliosis
- Leprosy
- Lupus vulgaris (Tbc)
- Leishmaniose
- **Lymphomas**

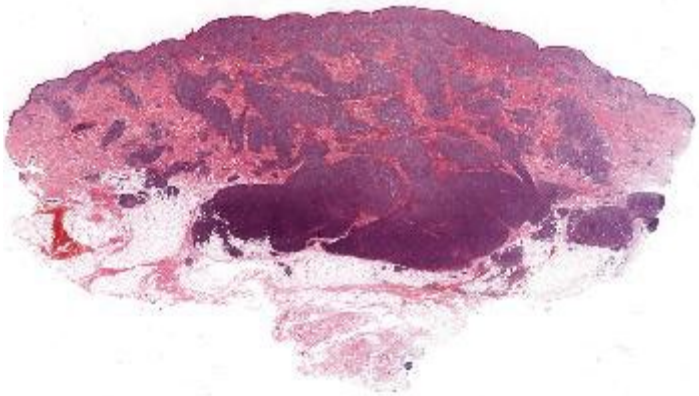
Primary cutaneous marginal zone lymphoma

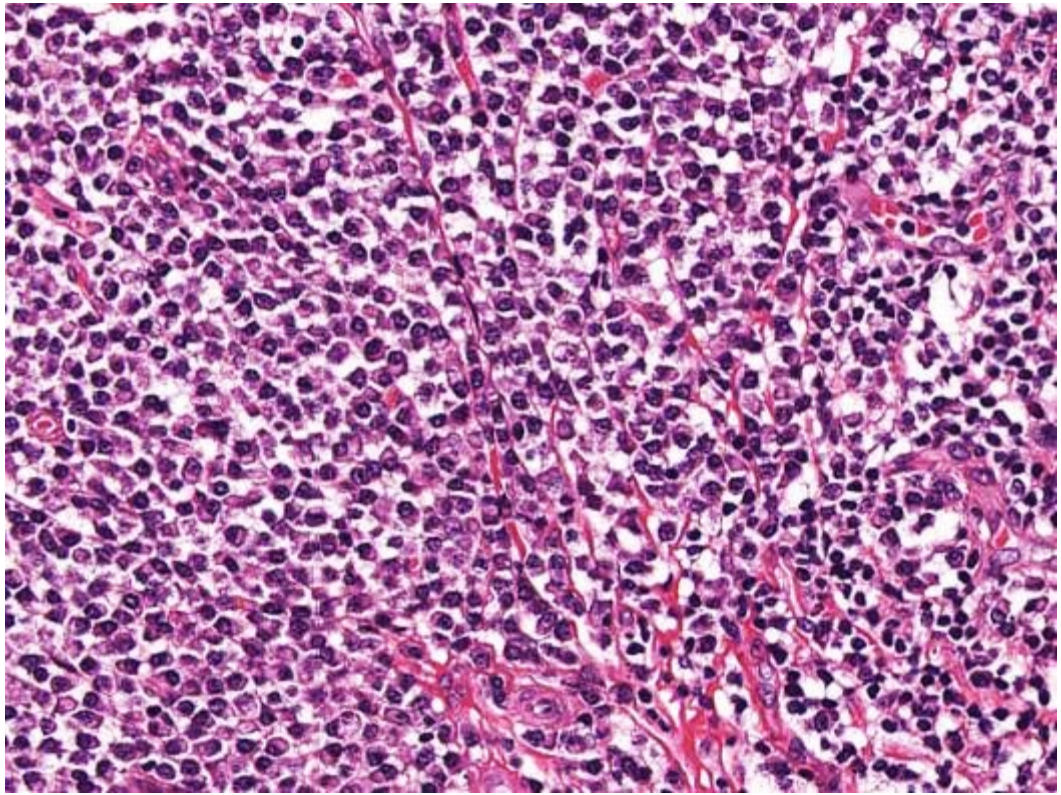
Plasma-cell rich variant ("Immunocytoma")

Intranuclear (Dutcher bodies)
and intracytoplasmic (Russell bodies)
inclusions

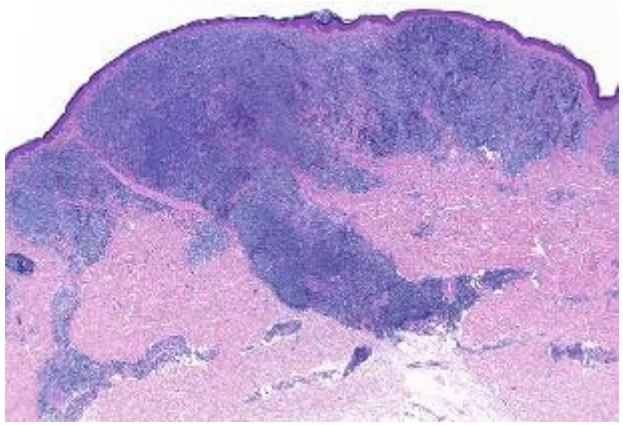
Association with Borrelia infection

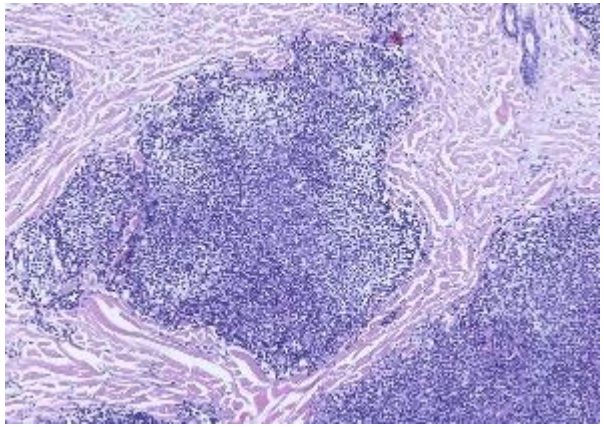
Cerroni et al. 1997
Goodlad et al. 2000

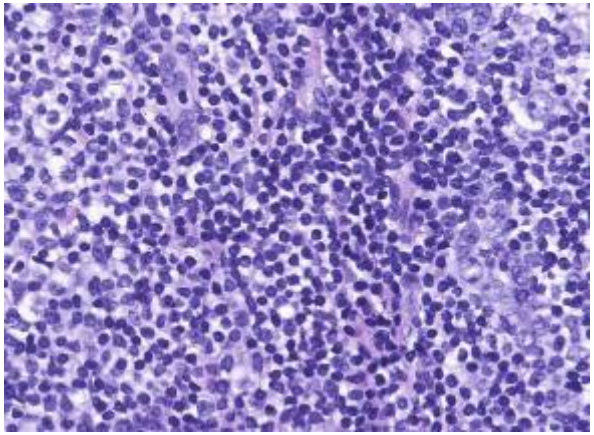


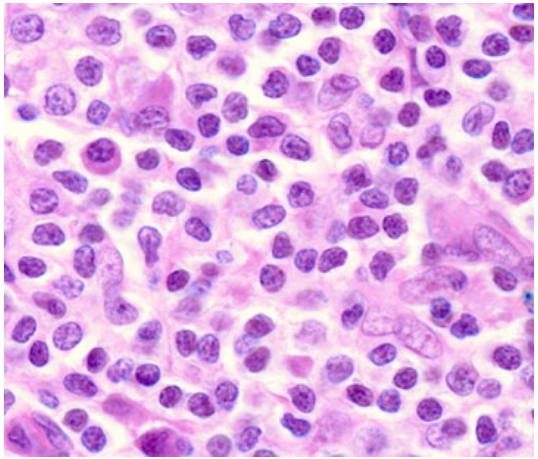


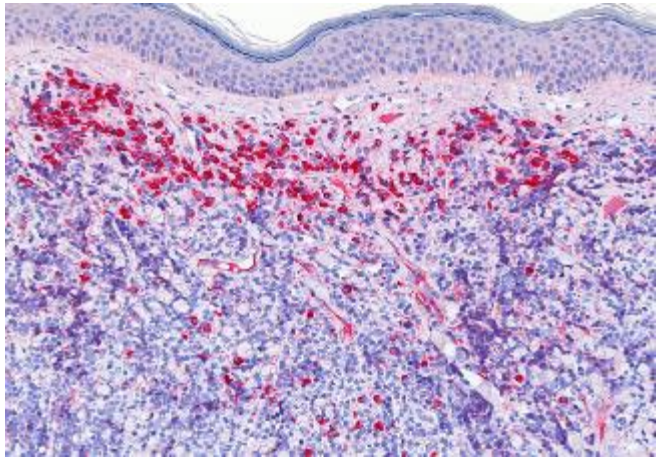
Primary cutaneous marginal zone lymphoma (WHO) / LPD (ICC)

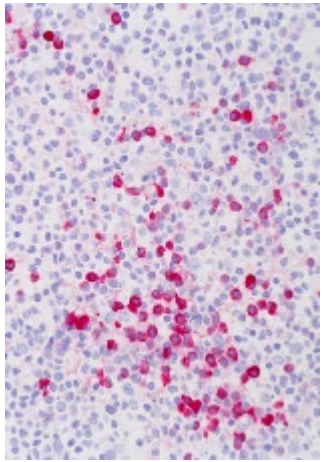


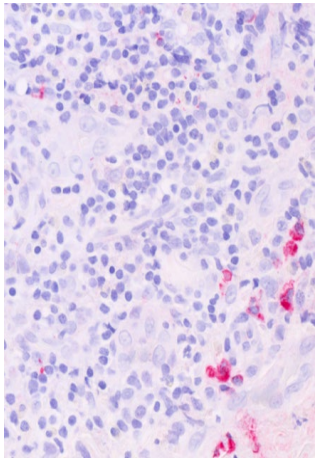








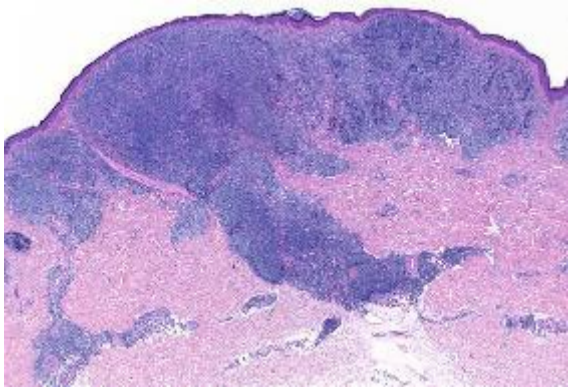




CD138 Ig kappa Ig lambda

Primary cutaneous marginal zone lymphoma - 2 subtypes

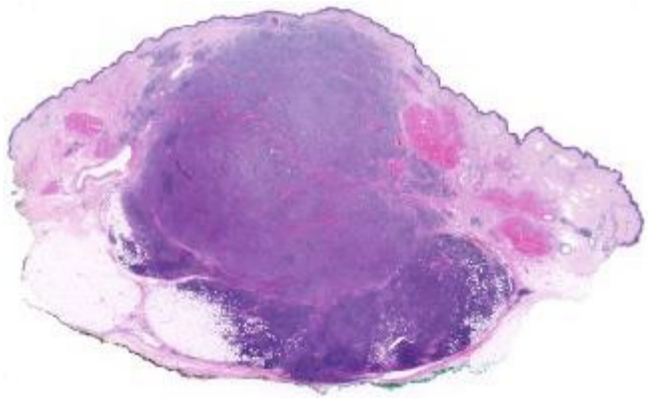
Heavy chain class-switched form (90%)



IgG+, IgA+, or IgE+

IgG4+ (20-40%)

Non class-switched form (10%)



IgM+

Recurrences (50%)

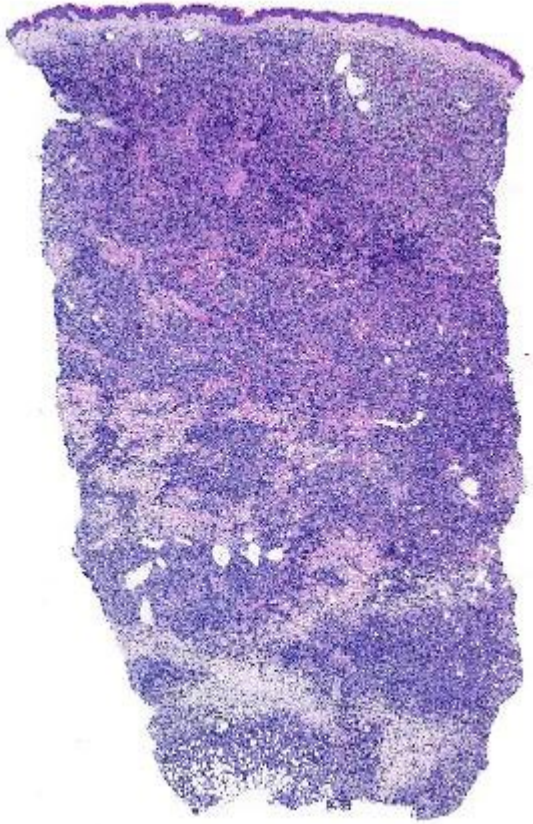
Extracutaneous involvement (< 4%)

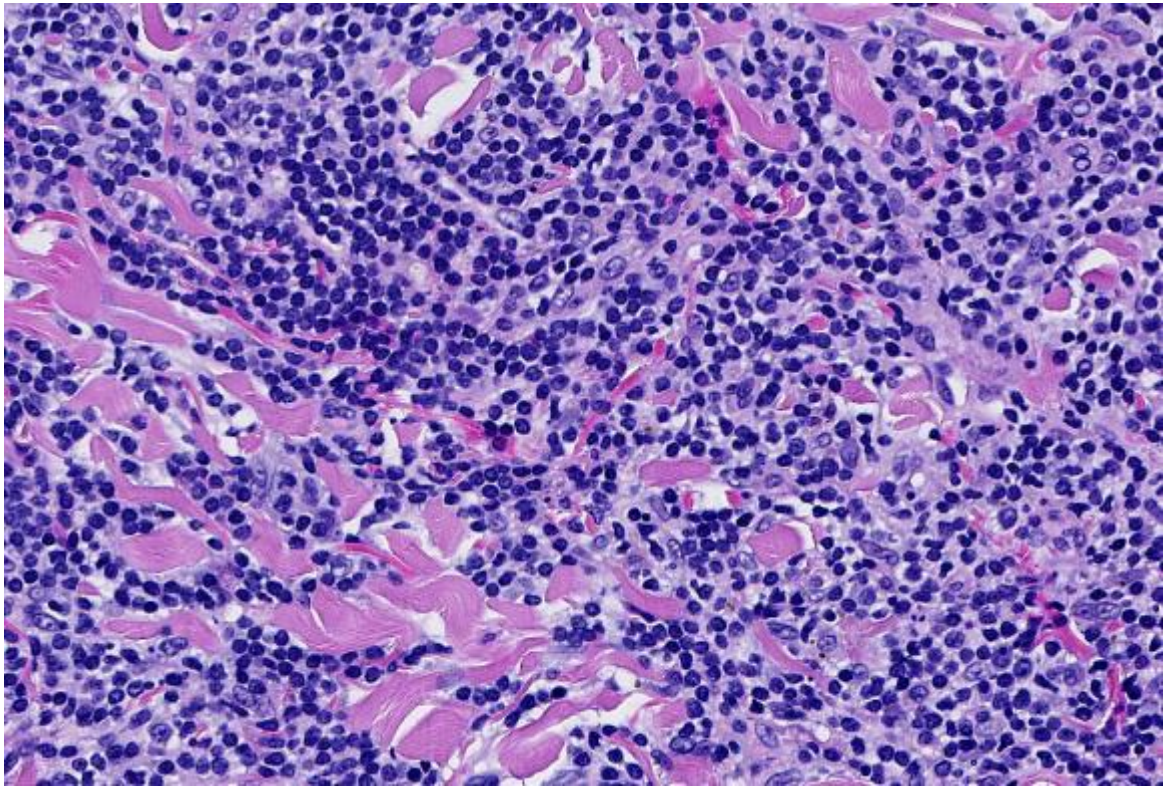
Excellent prognosis: 93-98% 5-year SR

Swerdlow Sem Diagn Pathol 2017

Carlsen et al J Cutan Pathol 2022

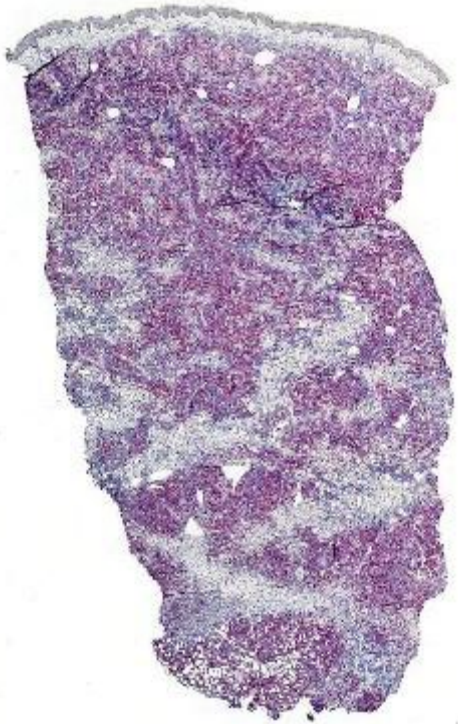
54-year-old man with infiltrated plaques on left thigh.



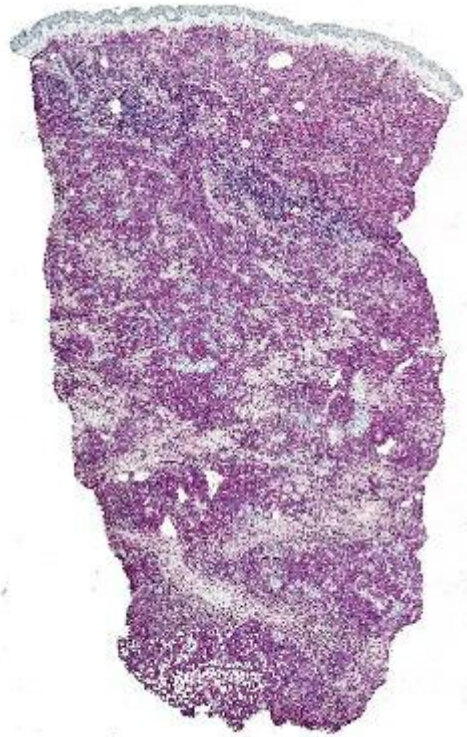


B23-110777

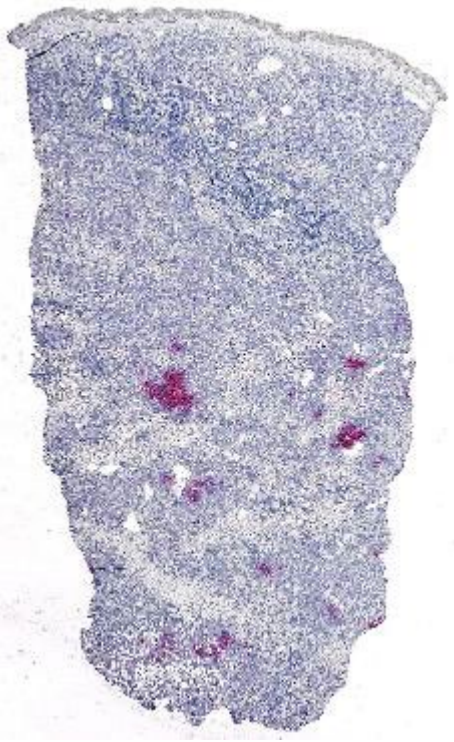
54-year-old man with infiltrated plaques on left thigh.



CD3

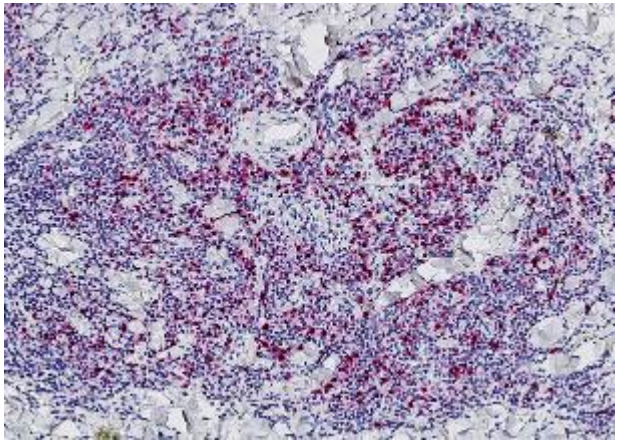


CD20

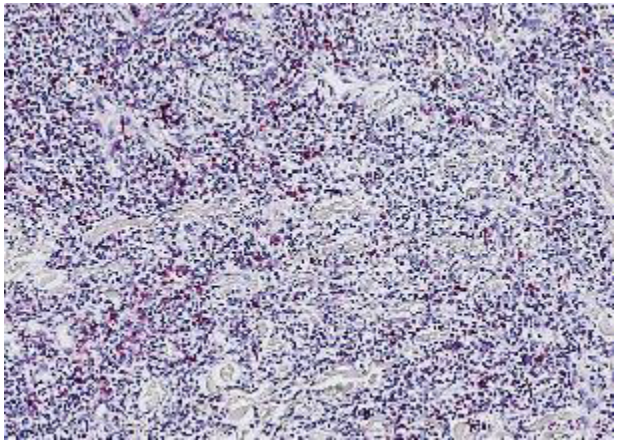


CD21

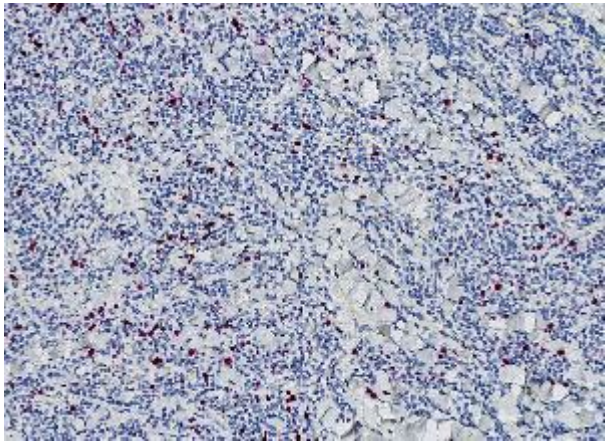
B23-110777



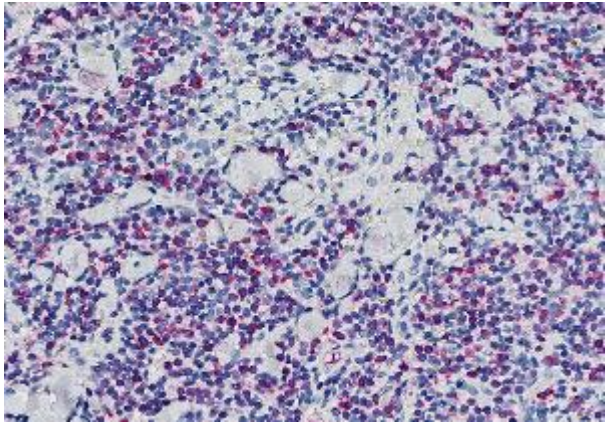
MUM-1



Ig kappa (monotypic)



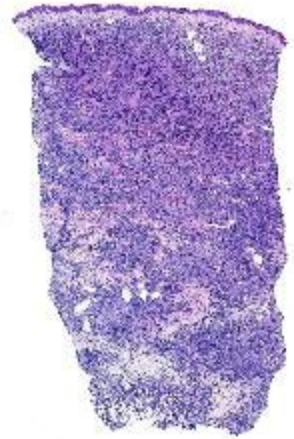
Ki67

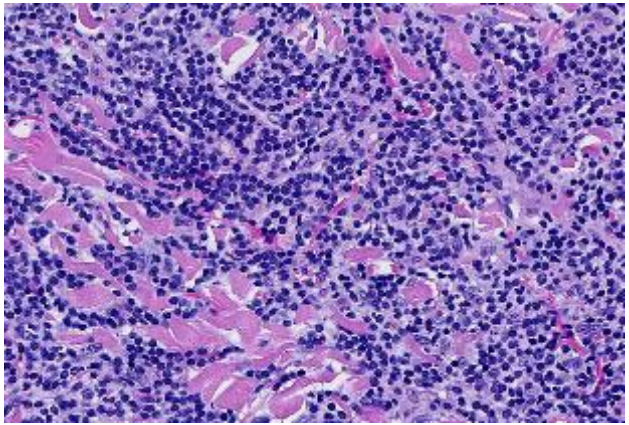


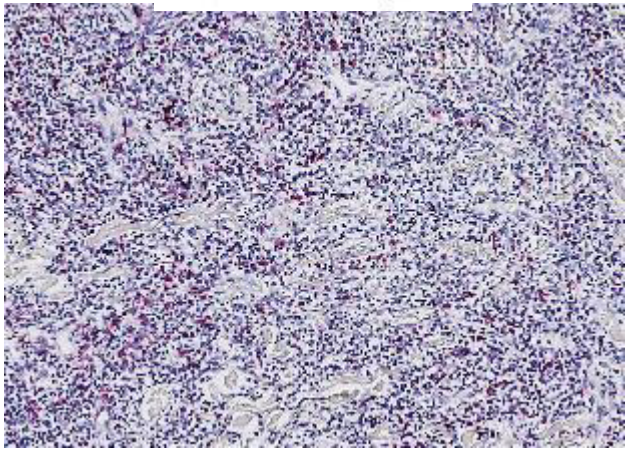
IgM

B23-110777

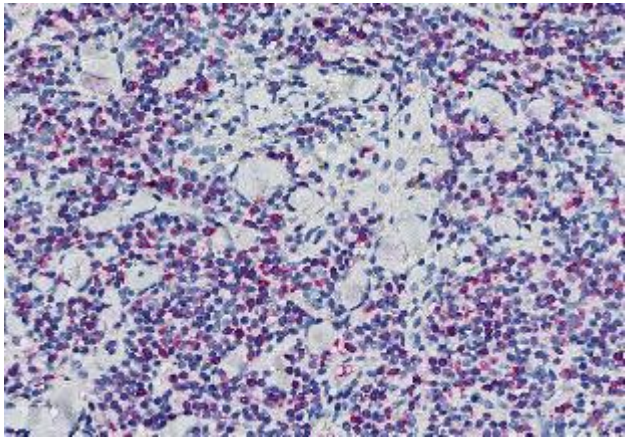
Non class-switched form of cutaneous marginal zone lymphoma?







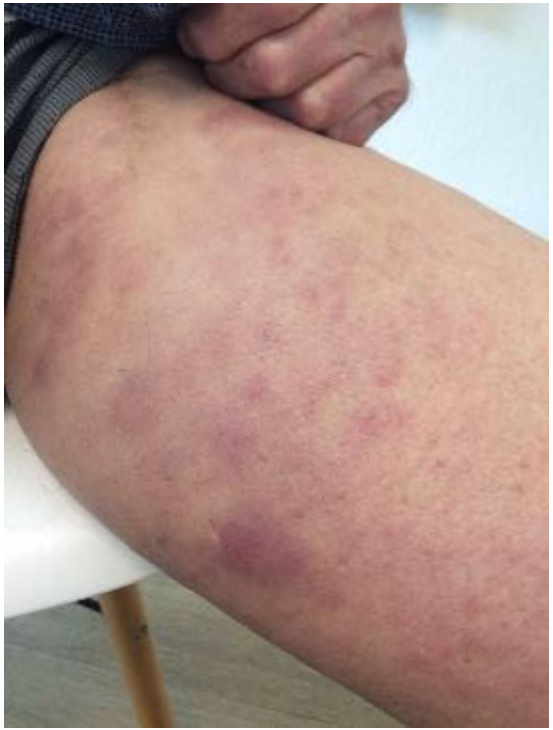
Ig kappa

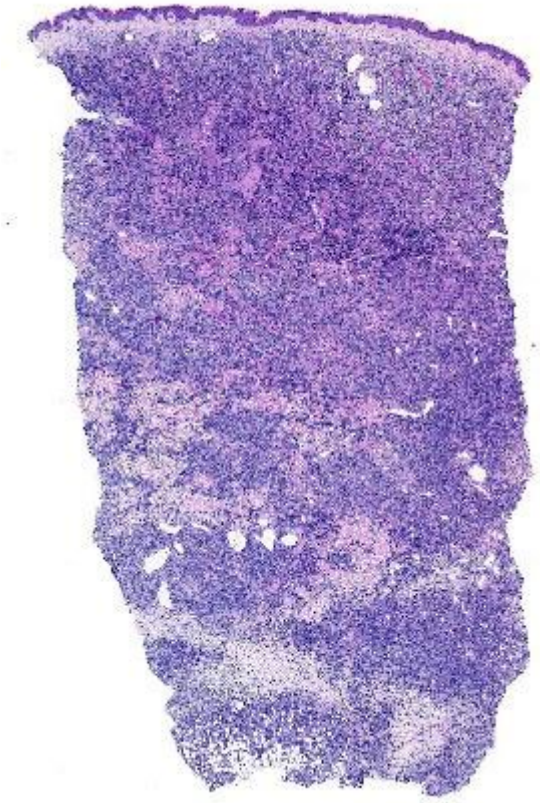


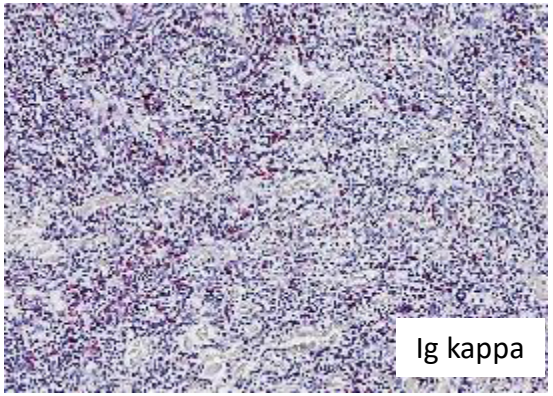
IgM

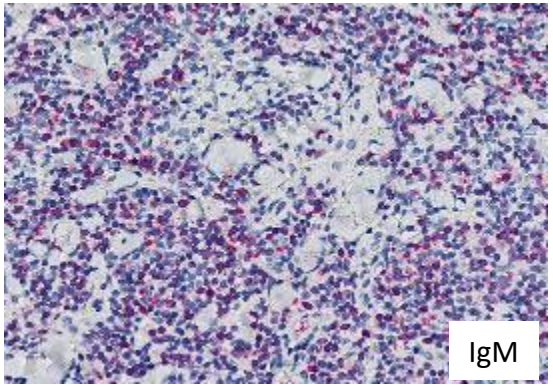
B23-110777

54-year-old man with infiltrated plaques on left thigh.



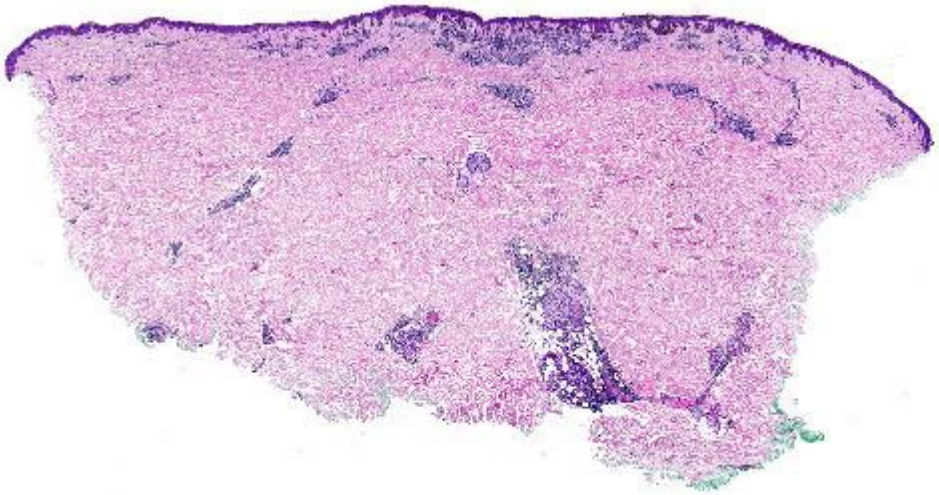


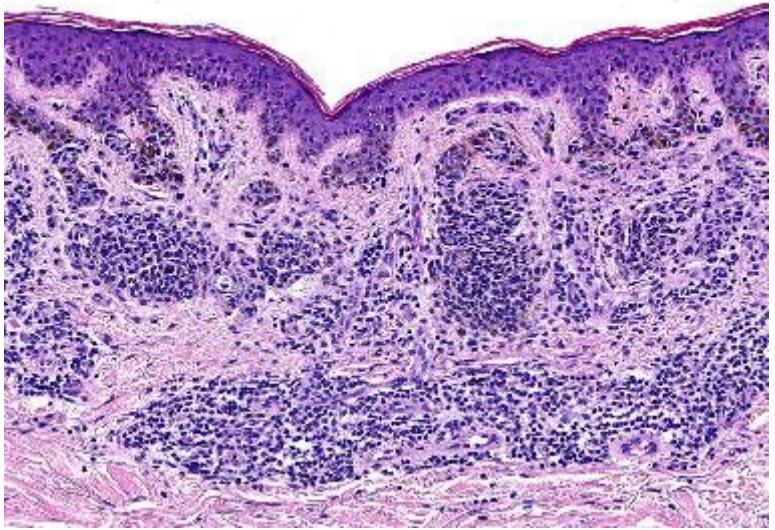


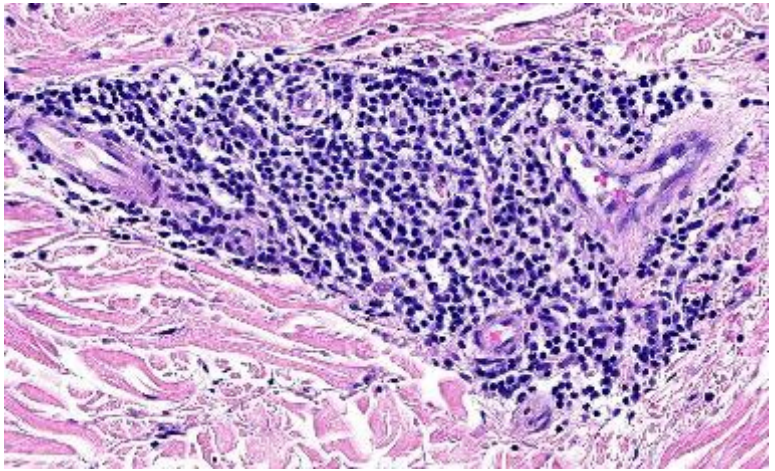


B23-110777

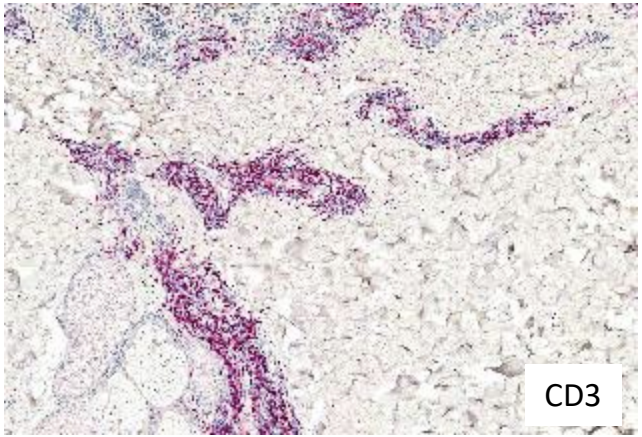
81-year-old man with atypical nevus (face)

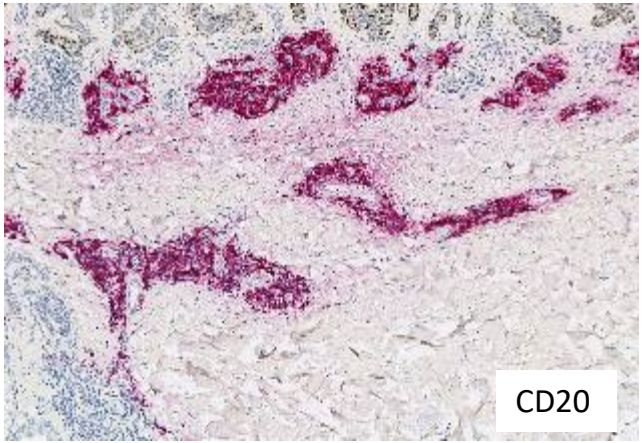


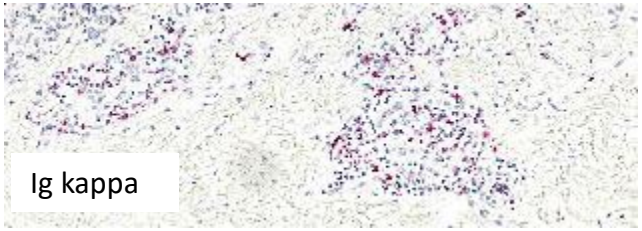


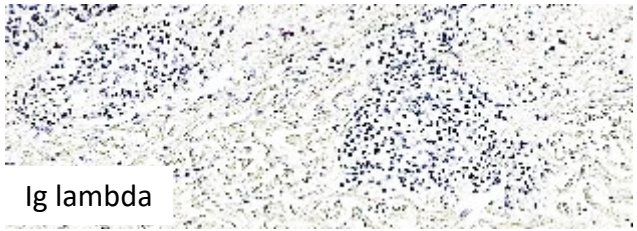


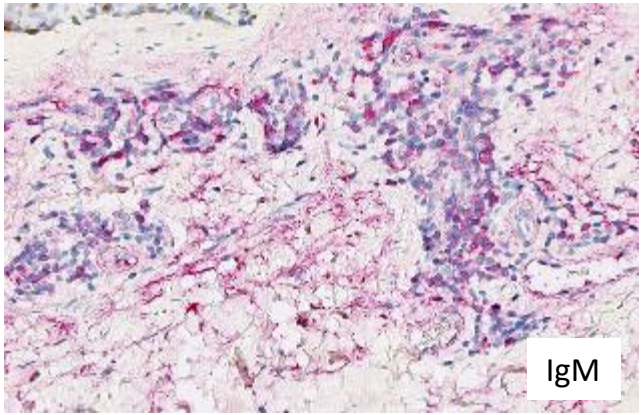
B22-47425











B22-47425

Waldenström macroglobulinemia (WM)

From: Kyle and Rajkumar Blood 2008

Mature B-cell neoplasm (WHO HAEM 5th) - Lymphoplasmacytic lymphoma (LPL)
Subtypes: IgM-type LPL; Waldenström macroglobulinaemia (IgM-type LPL/WM)
"WM is LPL, but not every LPL is WM"

> WM = LPL bone marrow involvement - Blood: IgM component

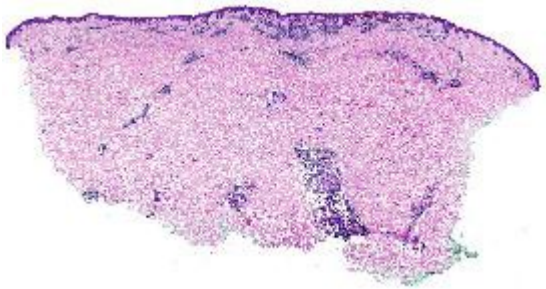
Men, white, ≥ 65 years of age

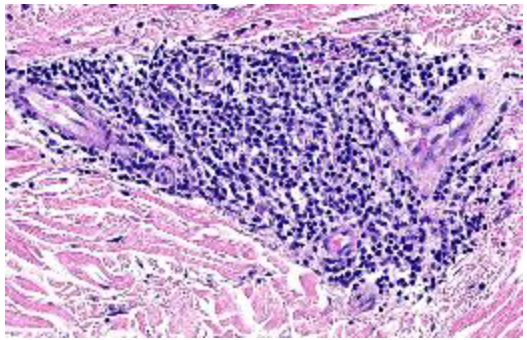
In WM constitutional symptoms (fatigue), lymphadenopathy, splenomegaly, and IgM paraprotein-related symptoms due to hyperviscosity (blurred vision, epistaxis, headache, shortness of breath), neuropathy.

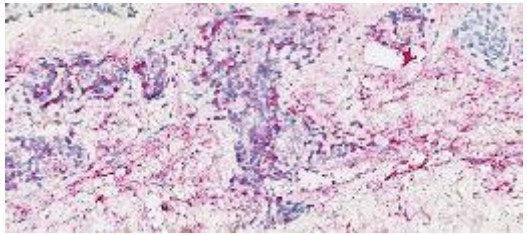
Driver mutation is MYD88 p.L265P in 93–97% of LPL/WM cases Ngo VN et al. Nature 2011

Lymphoma infiltrates in the skin and other mucosal sites can be difficult to distinguish from MZL and may require mutation testing for MYD88p.L265P (WHO HAEM 5)

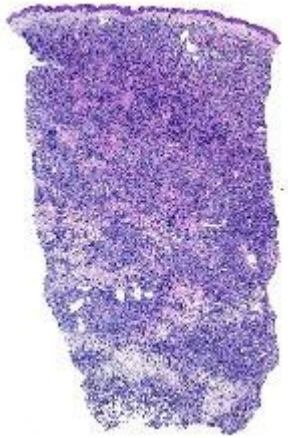
Waldenström macroglobulinemia

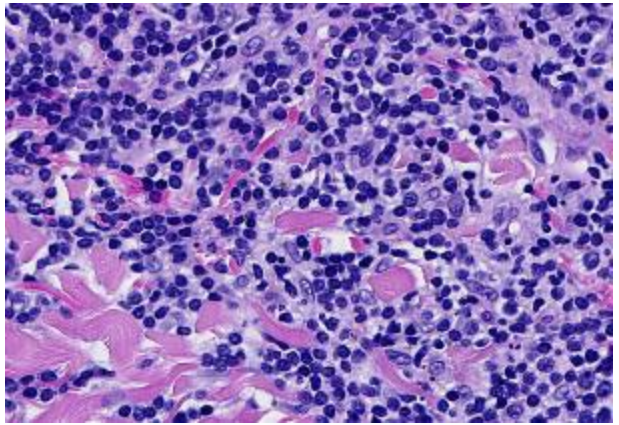


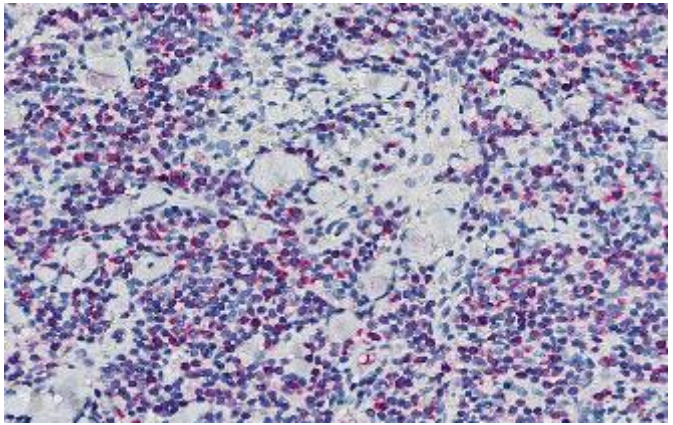




IgM

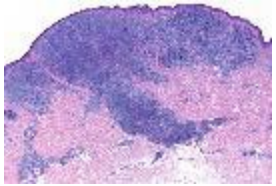
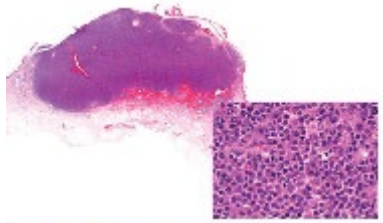






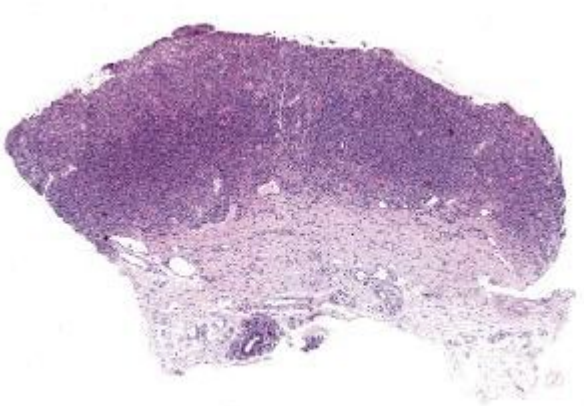
IgM

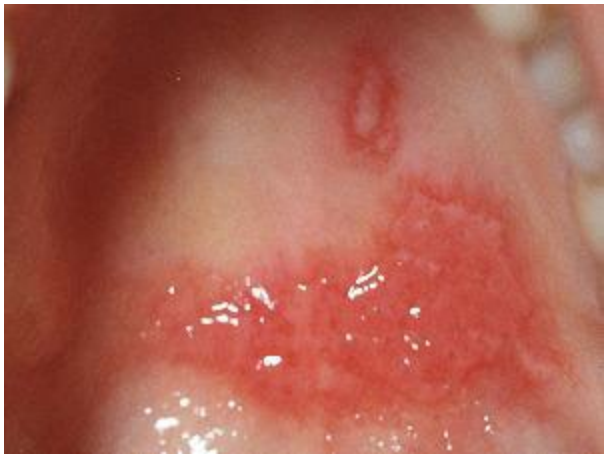
Monotypic plasma cell infiltrates

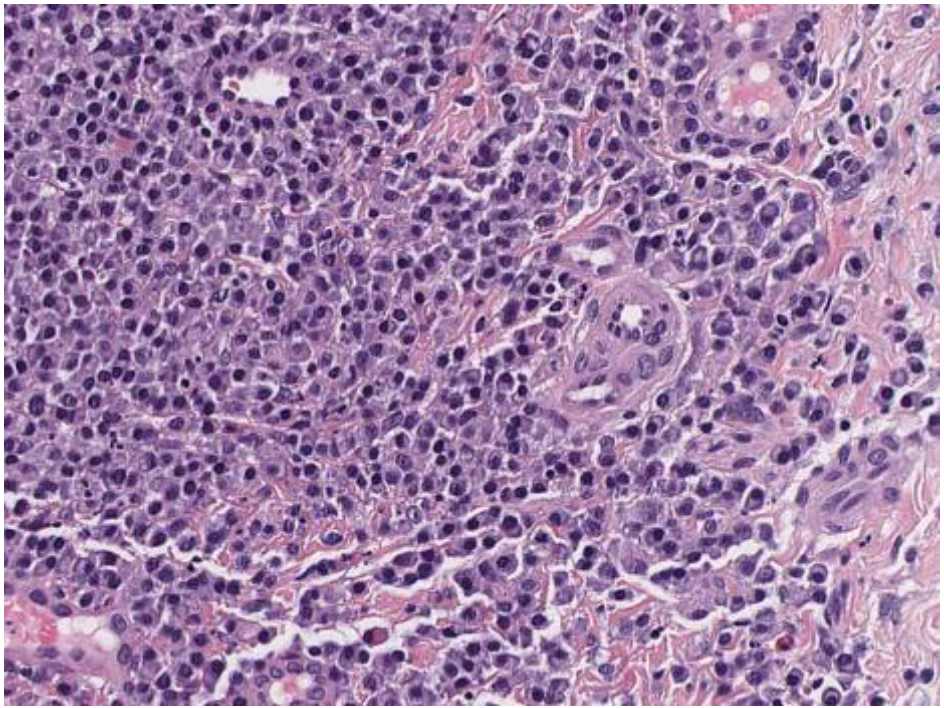
Cutaneous marginal zone lymphoma/LPD	Waldenström macroglobulinemia	Lymphoplasmacytic lymphoma (LPL)	Plasma-cell myeloma (Syn: multiple myeloma)	Plasmacytoma
				
IgG - IgG4 (20-40%) IgM (subset)	  IgM Spike	IgM 	 IgG / IgM	

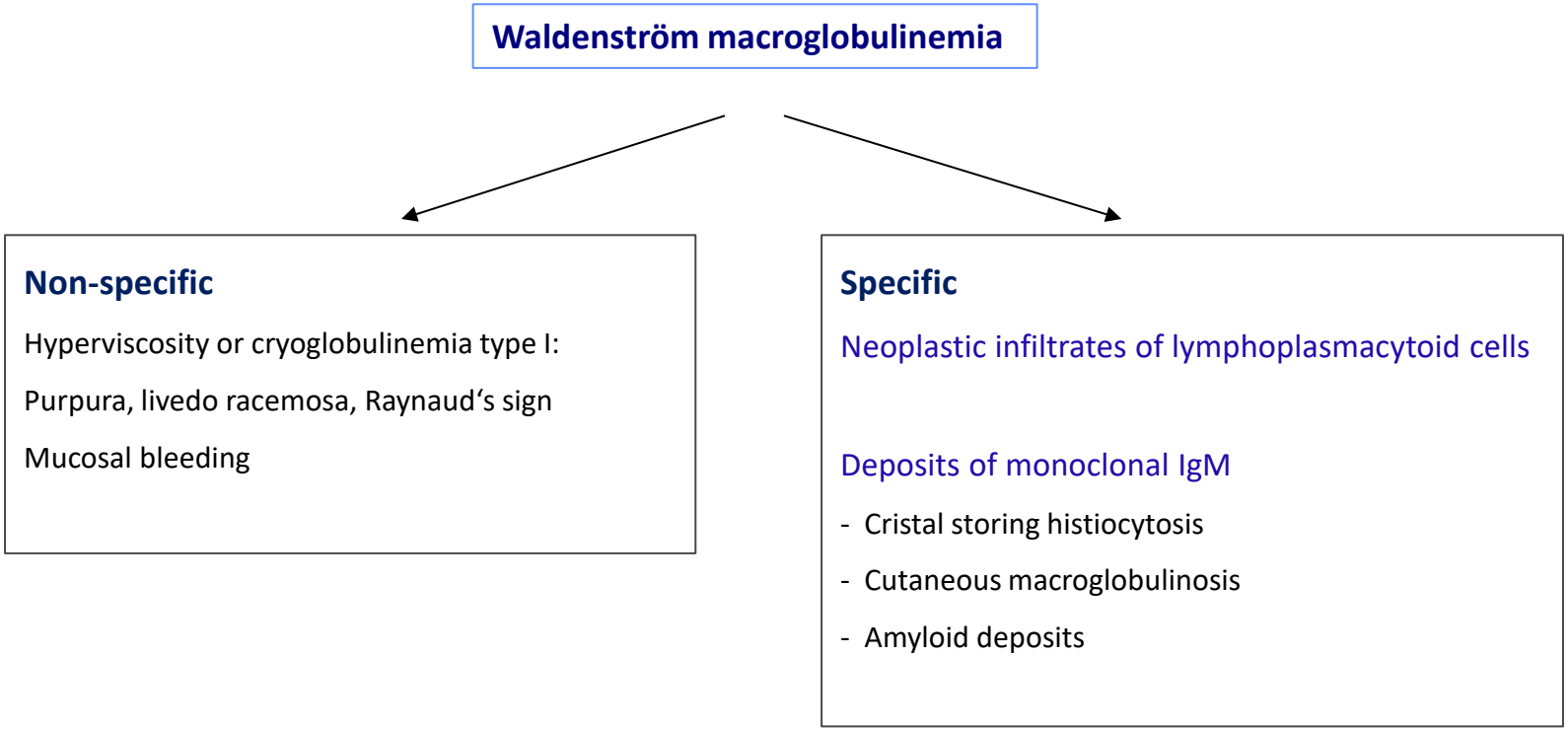
MYD8 8p. L295P mutation

64-year-old lady with multiple myeloma and oral ulcerations









Camp BJ & Magro C. J Cutan Pathol 2012

Cutaneous macroglobulinosis

Isolated deposits of monoclonal IgM in the skin

Tichenor RE et al. Arch Dermatol 1978

Skin-colored papules, rarely nodules

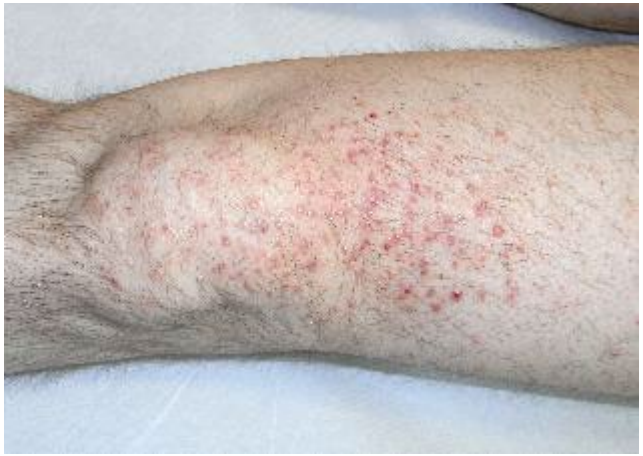
Knees, buttocks, extensor surfaces of arms and legs

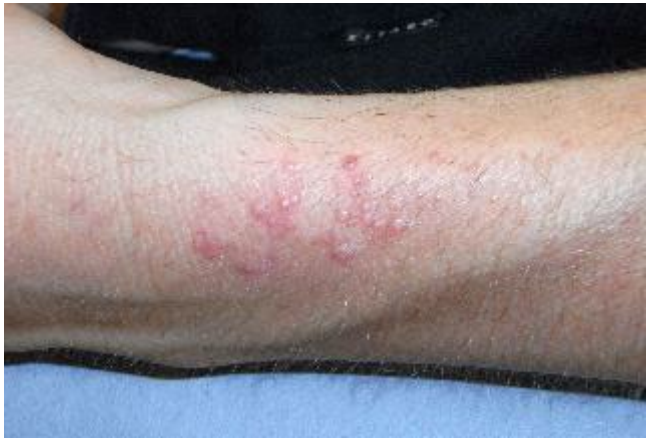
Prior, concurrently of after diagnosis of Waldenström.

Not a prognostic sign

Camp BJ & Magro C. J Cutan Pathol 2012

Spicknall KE et al. J Cutan Pathol 2013

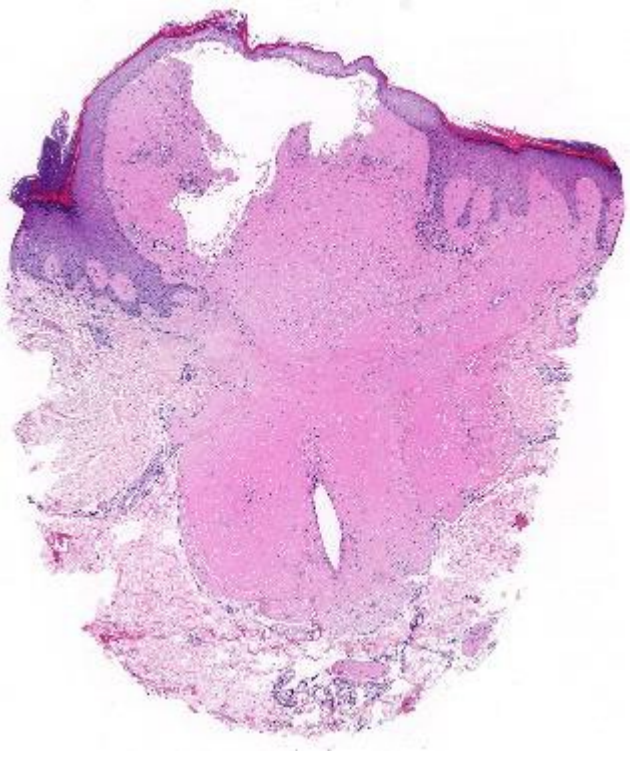


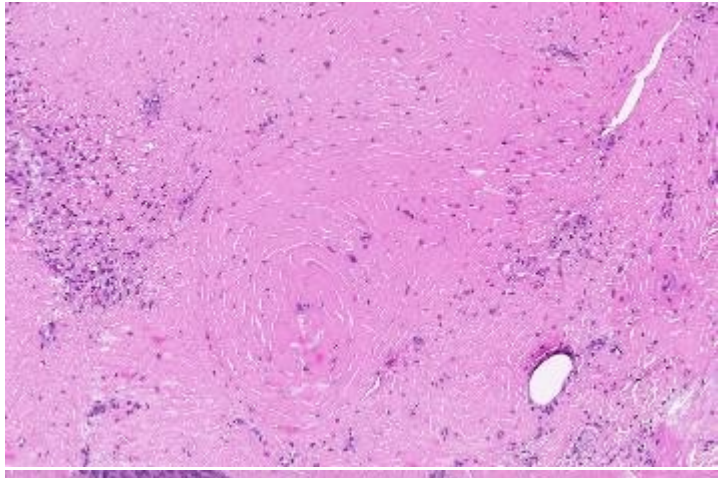


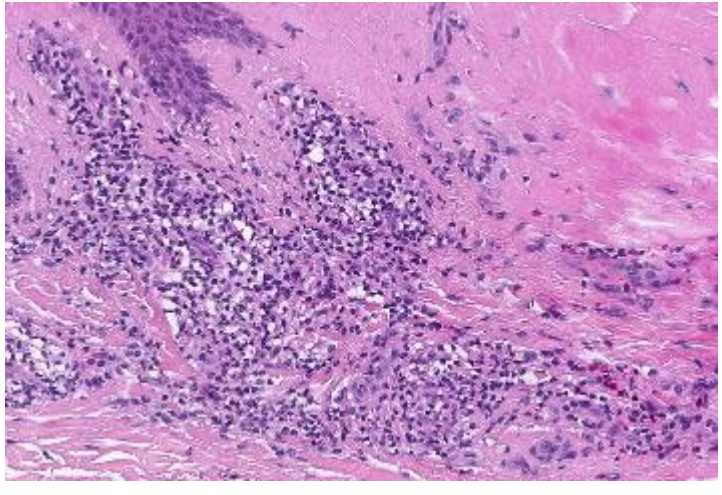
By courtesy Dr. F. Kind, Schaan

Cutaneous macroglobulinosis

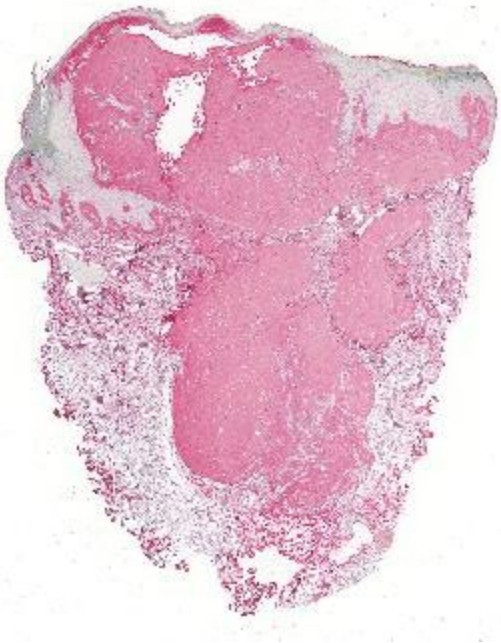
Dense dermal deposits of an clefted eosinophilic material
esp. around hair follicles and eccrine glands.
PAS and IgM positive



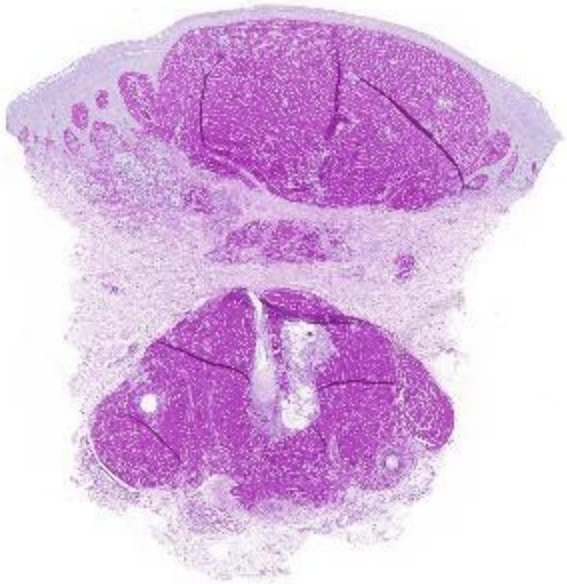




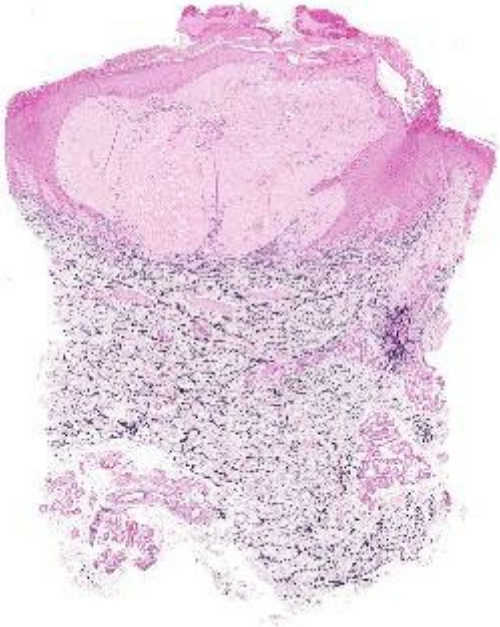
Cutaneous macroglobulinosis



IgM



PAS



Elastica

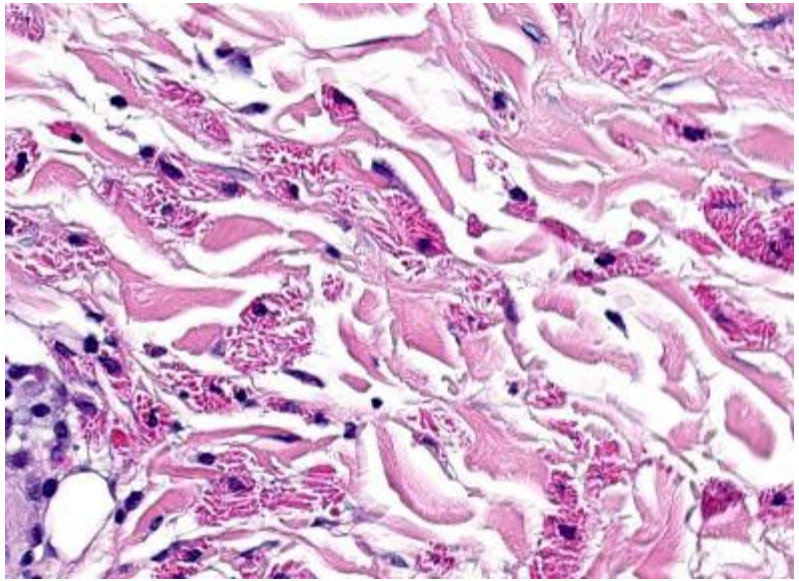
Laboratoty tests: Waldenstrom macroglobulinemia (IgM gammopathy, kappa)

Crystal storing histiocytosis

Intracytoplasmatic non-birefringent eosinophilic crystals in histiocytes

Localized (various organs) or generalized (bone marrow)

Skin: First manifestation or preceding underlying neoplasm

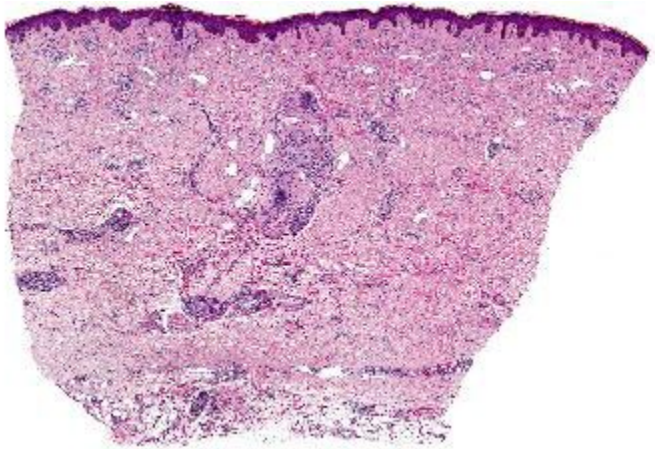


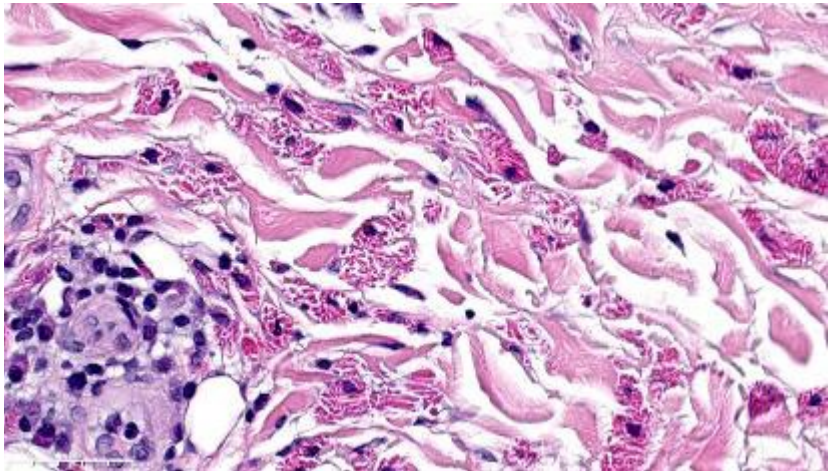
Lymphoproliferative disorders

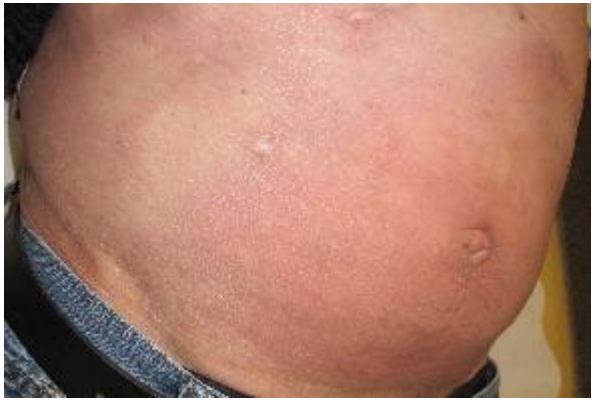
- Waldenström macroglobulinemia (WM)
- Lymphoplasmacytic lymphoma (LPL)
- Multiple myeloma
- MGUS
- Other B-cell neoplasms

Jenkins RE et al. Arch Dermatol. 1994
Harada M et al. Hum Pathol. 1996
Pock L et al. Int J Dermatol 2006
Dogan S et al. Head Neck Pathol 2012
Kanagal-Shamanna R et al. Histopathology 2015

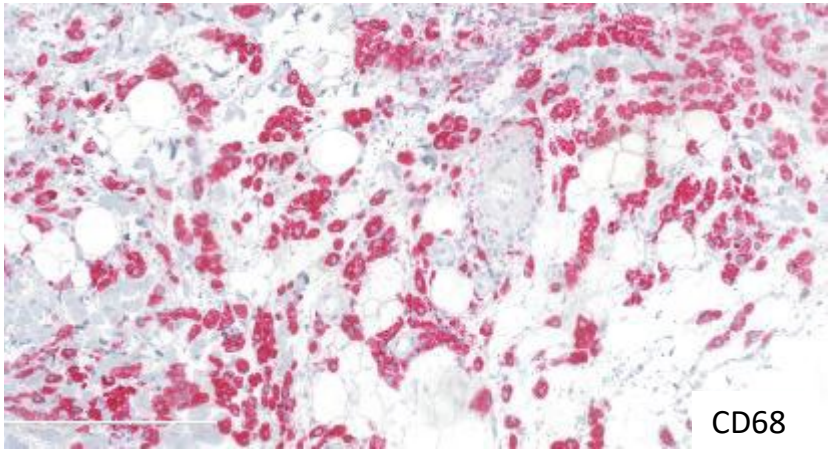
Crystal storing histiocytosis





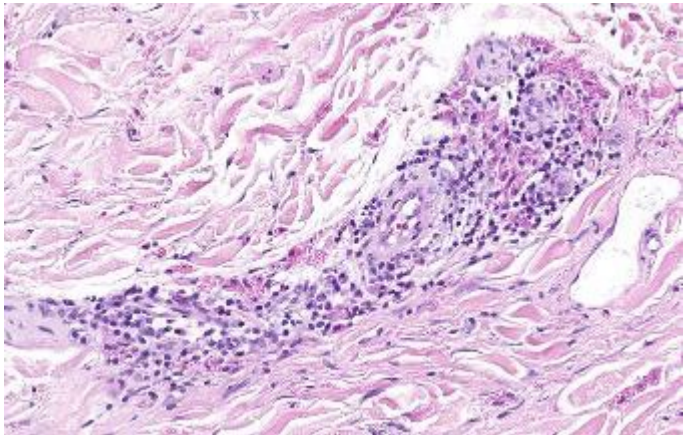


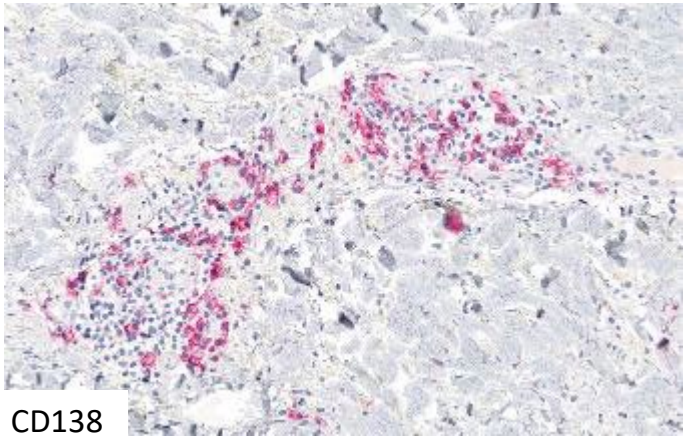
Fried I, Buslau M, Kempf W JAMA Dermatol 2016



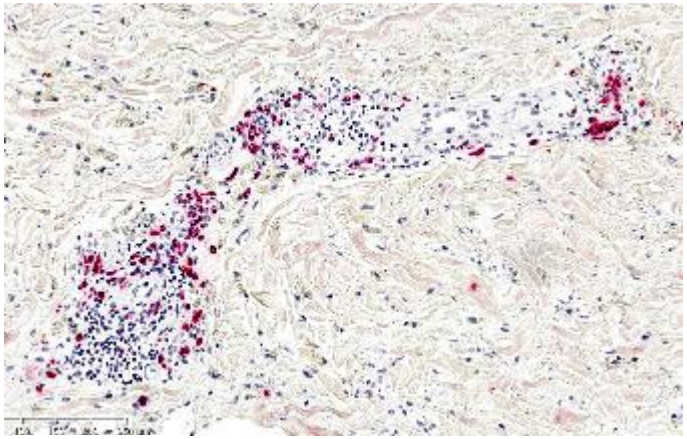
CD68

Crystal storing histiocytosis

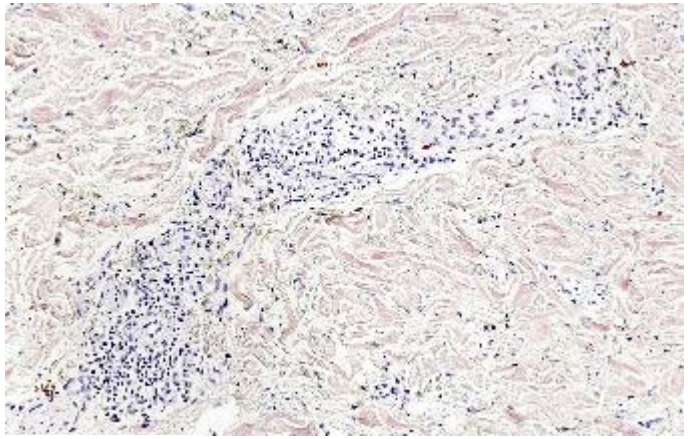




CD138

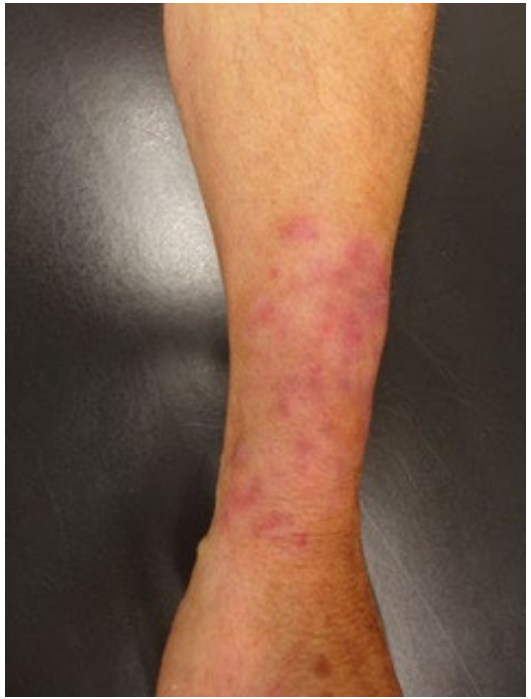


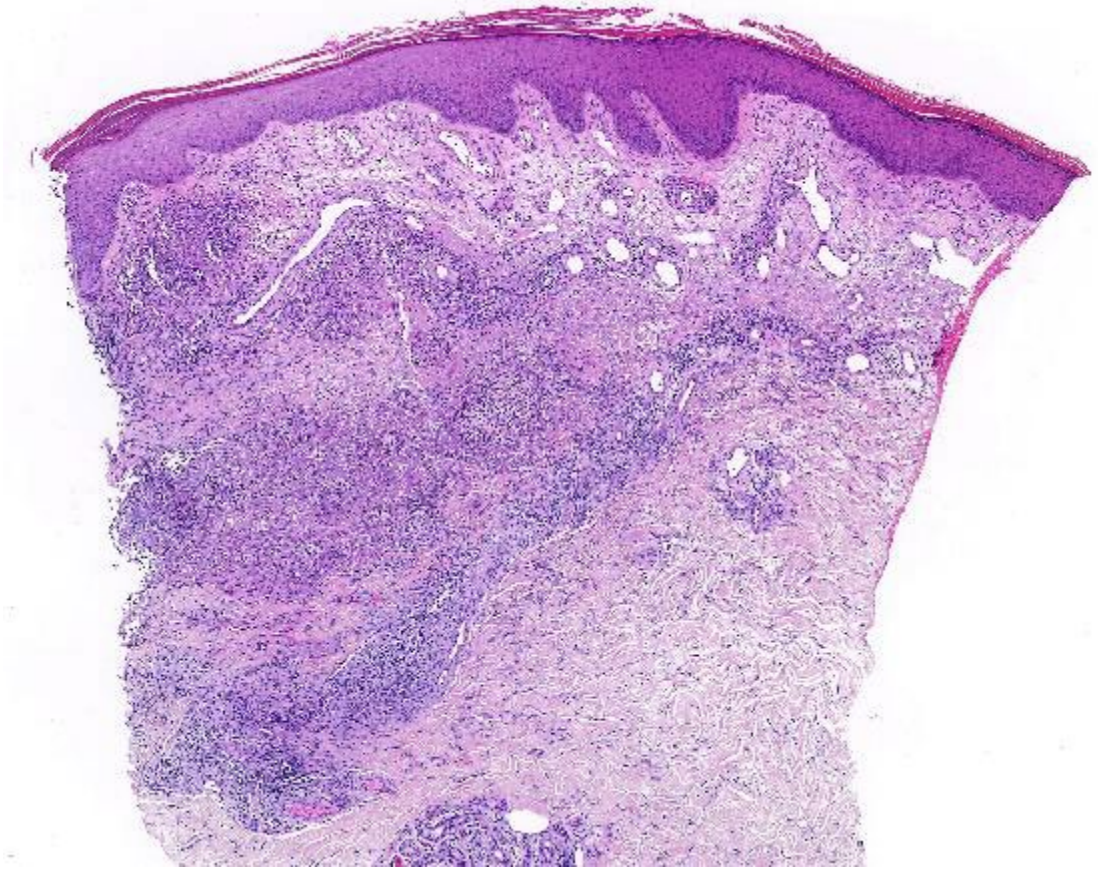
Ig kappa

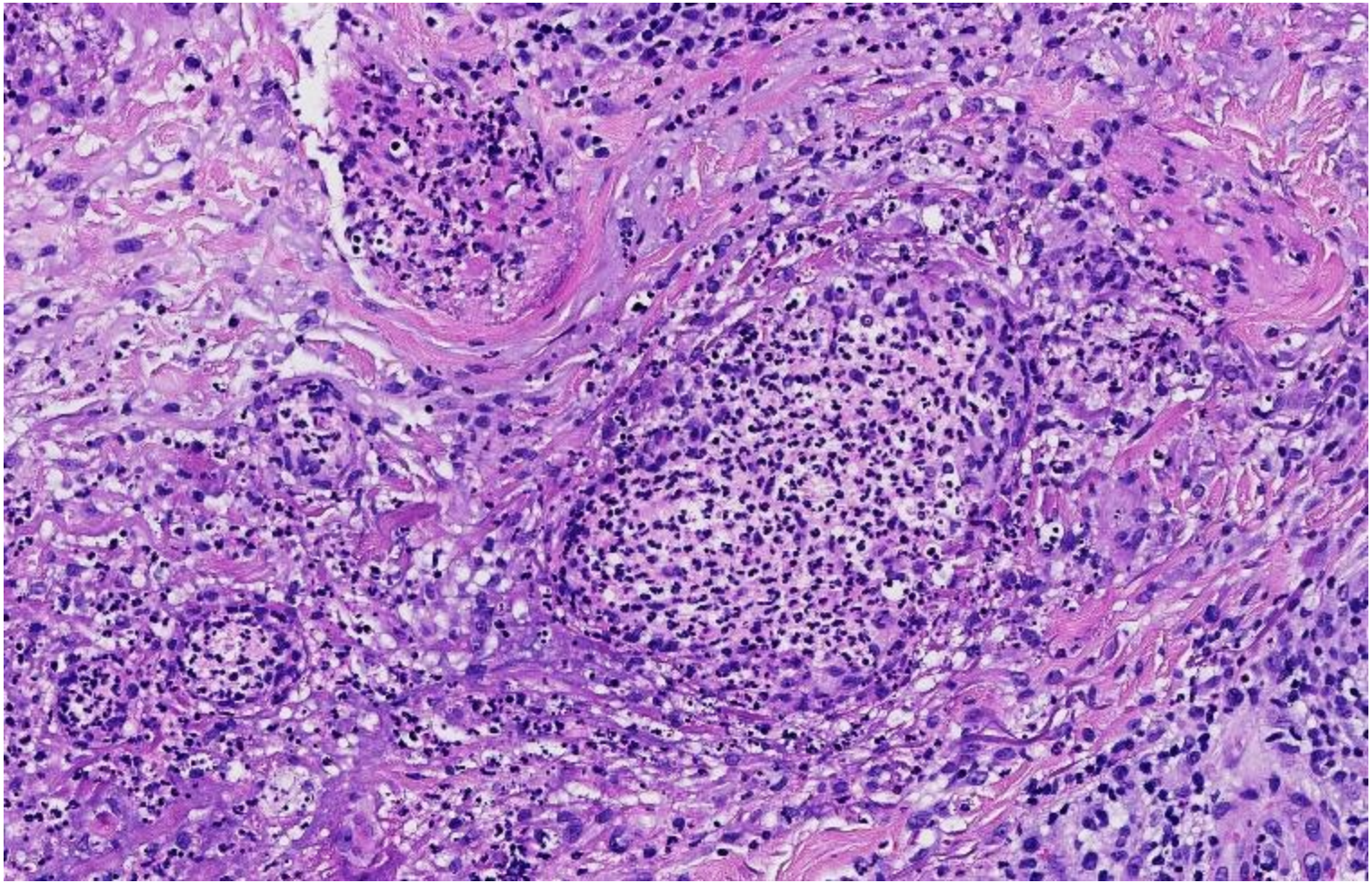


Ig lambda

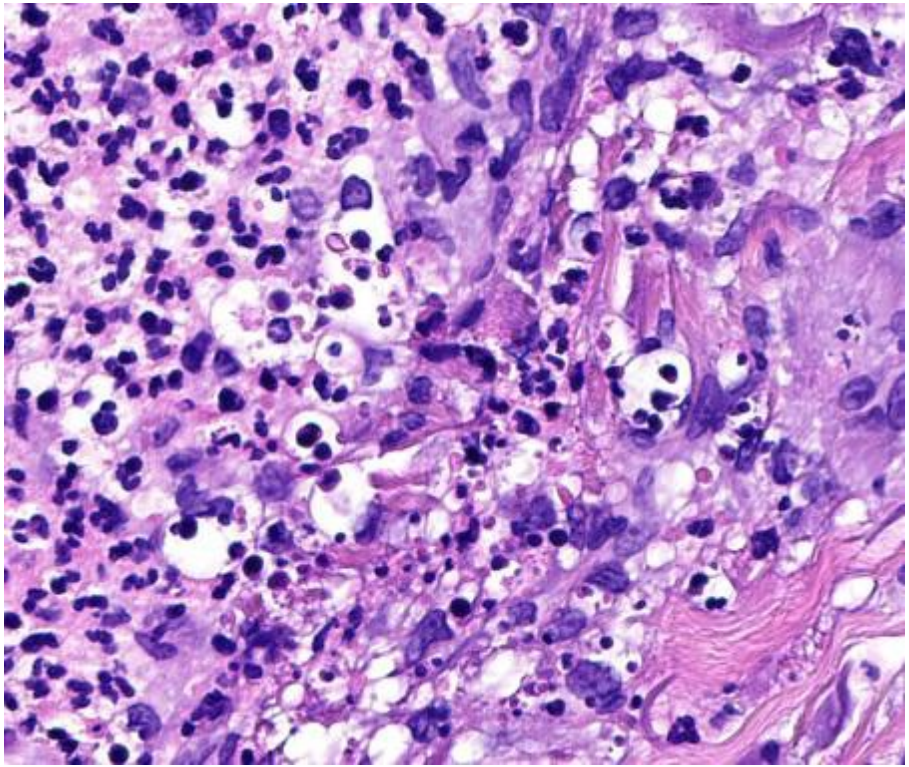
67-year-old man with papular lesions on left forearm after vacation on Crete. Mycobacterial or fungal infection?

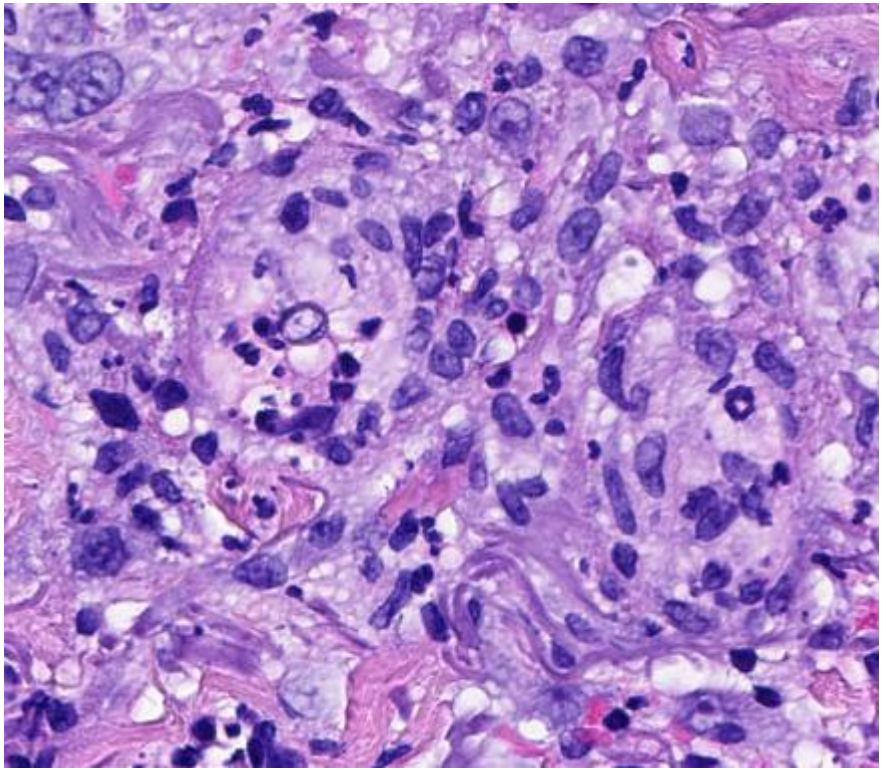




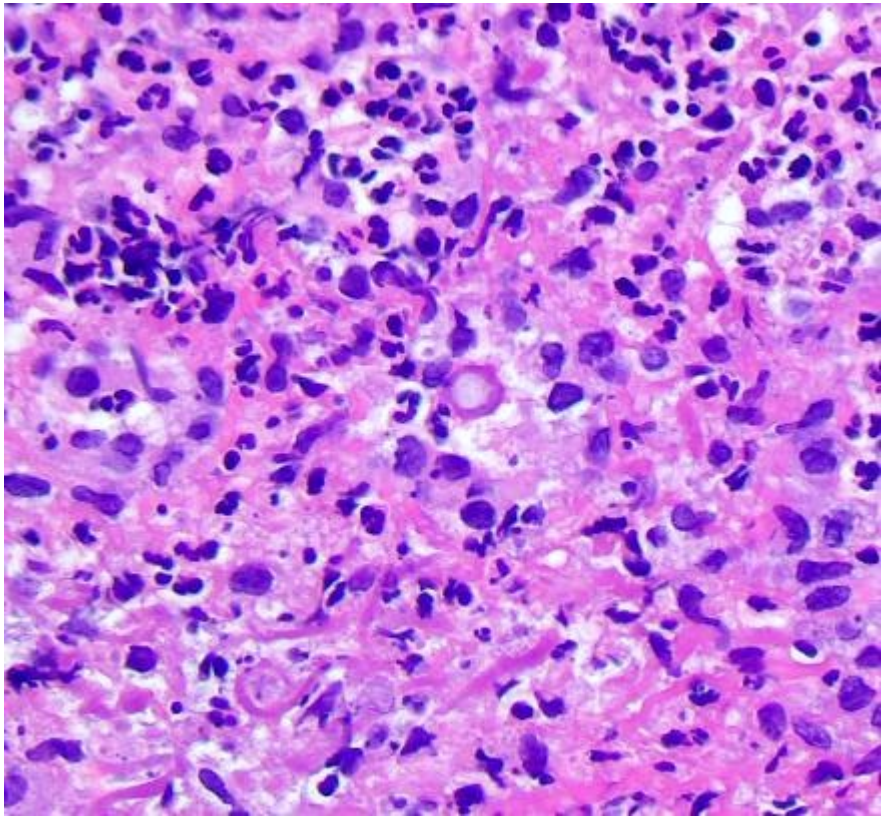


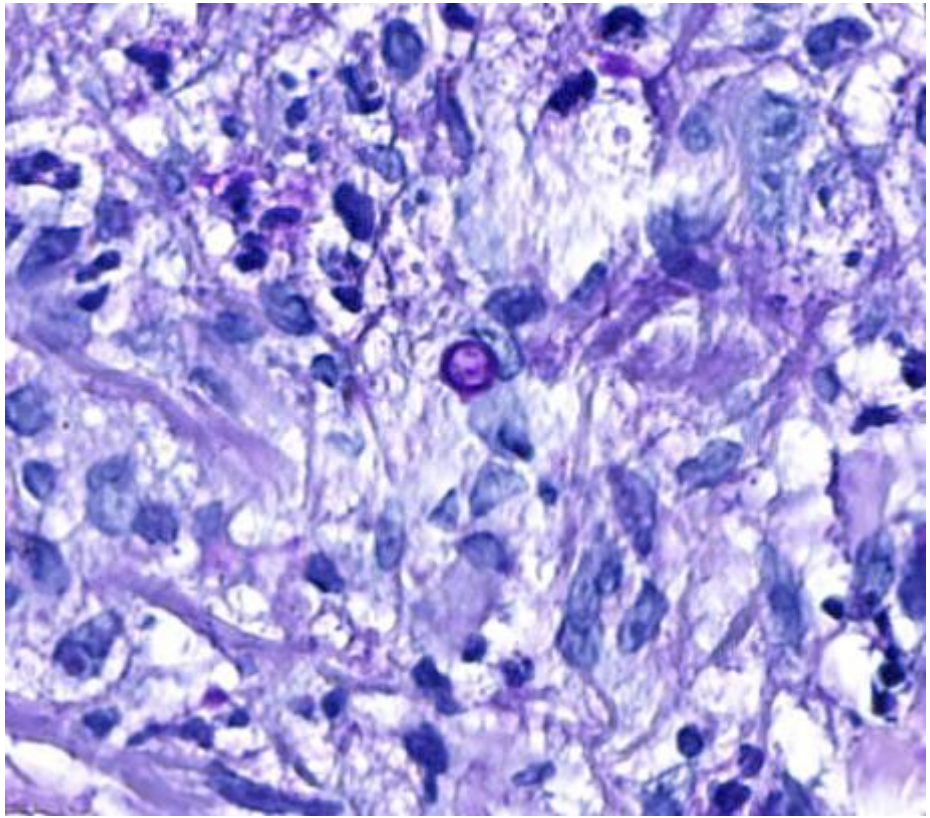
B 25-122189





B 25-122189

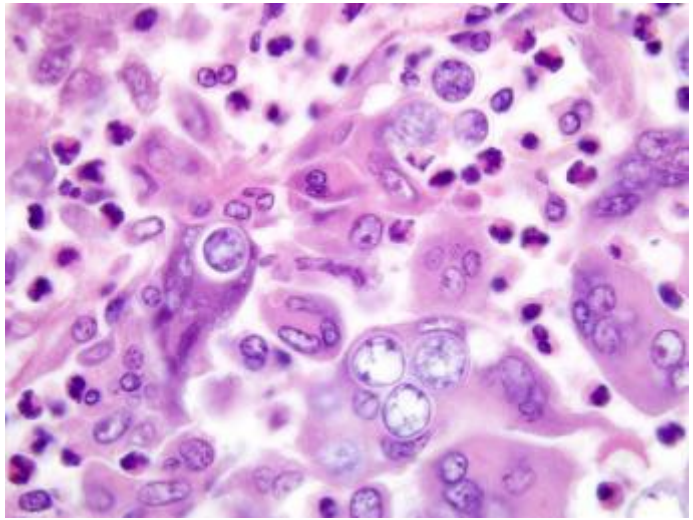




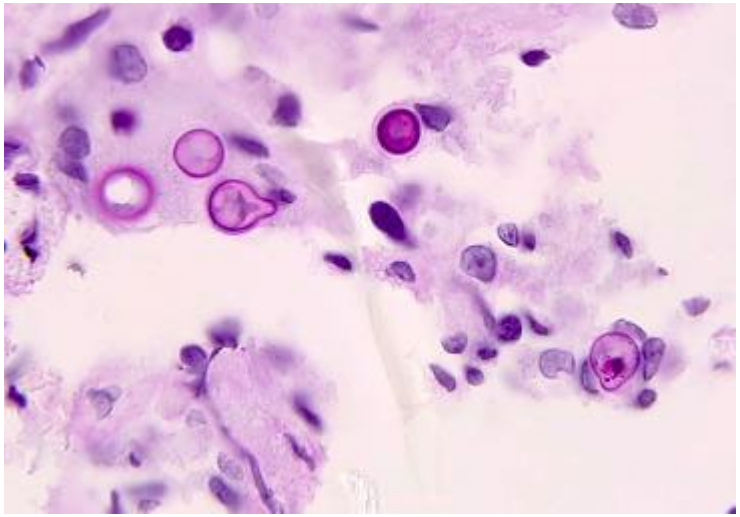
PAS

B 25-122189

Blastomyces dermatitidis



From: Pathology Outlines



From: The Joint Pathology Center, WSC Conf 2025-26

North American blastomycosis:

Dimorphic fungus, systemic disease caused by Blastomyces dermatitidis, mainly affects dogs and humans


CAUTION
WET FLOOR
CAUTION
SLIPPERY WHEN WET

Trichophyton rubrum mimicking blastomyces dermatitidis

Broad spectrum PCR and sequencing:
Trichophyton rubrum

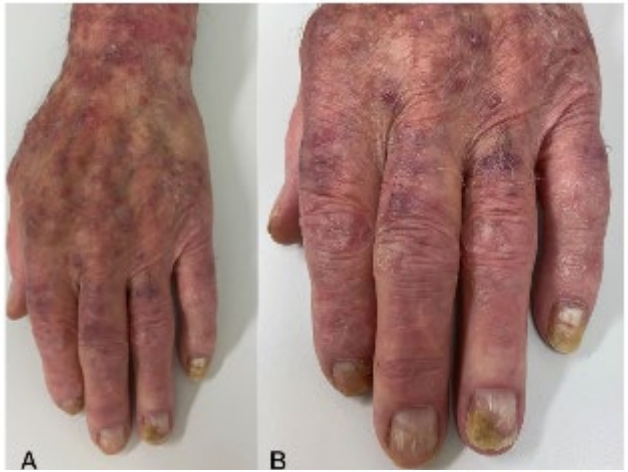
Complete remission after 3 weeks
of topical ketoconazol cream.

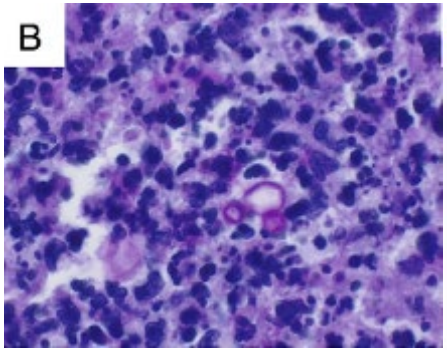
Trichophyton rubrum:
Invasive dermal infection, esp. in
Immunocompromised patients,
suppurative features

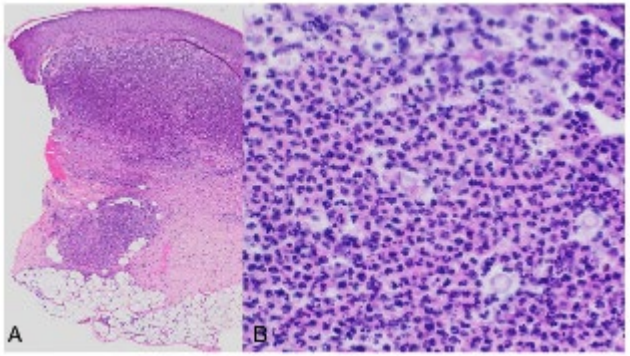
Tr. rubrum may resemble blastomycses
Dermatophyte dimorphism

Indicator of underlying immunodeficency
(primary or secondary)

Squeo et al. J Am Aacd Dermatol 1998
Lillis et al. J Cutan Pathol 2010
Nazarian et al. J Cutan Pathol 2019
Forster & Mühleisen J Cutan Pathol 2024

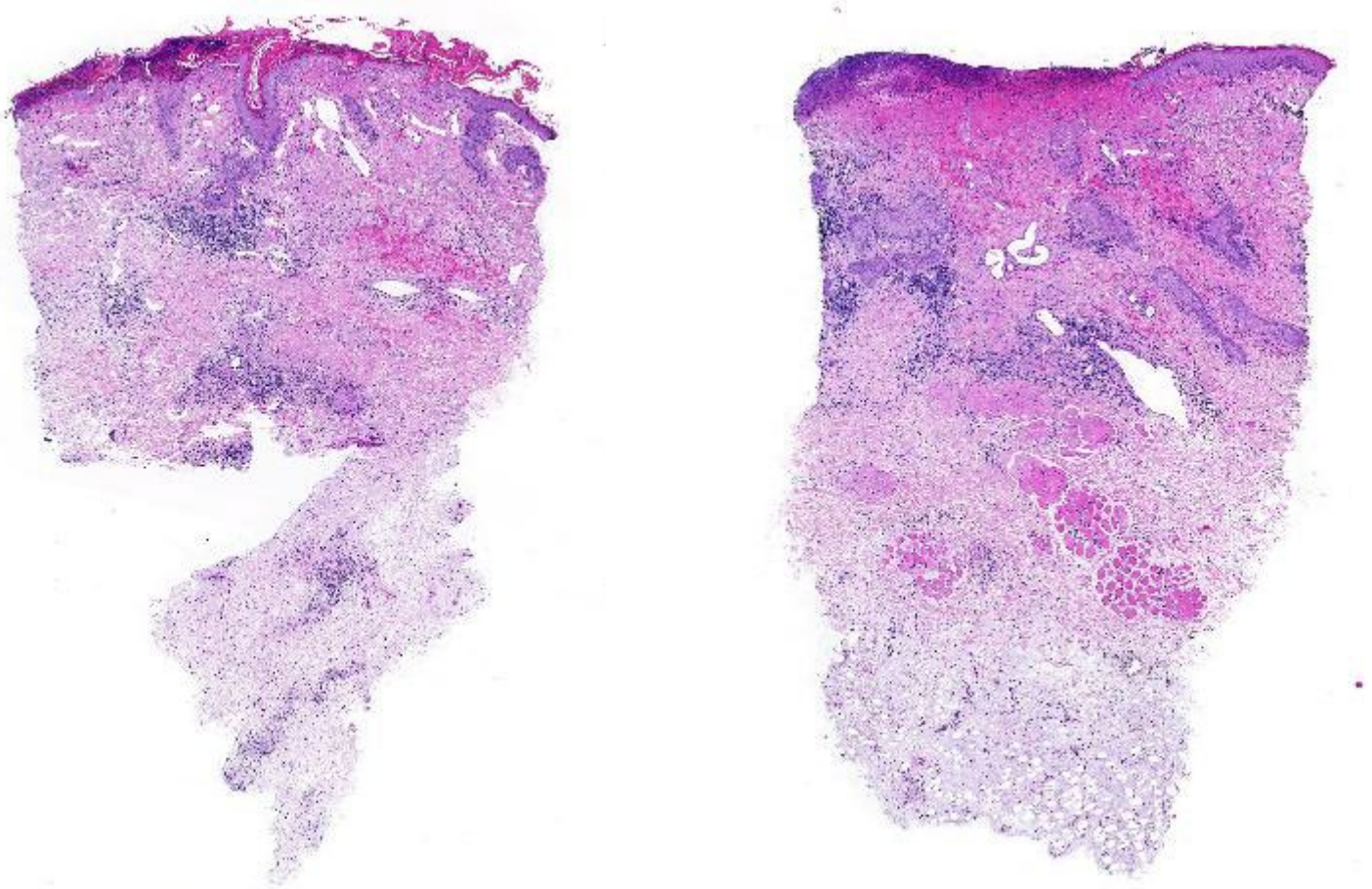


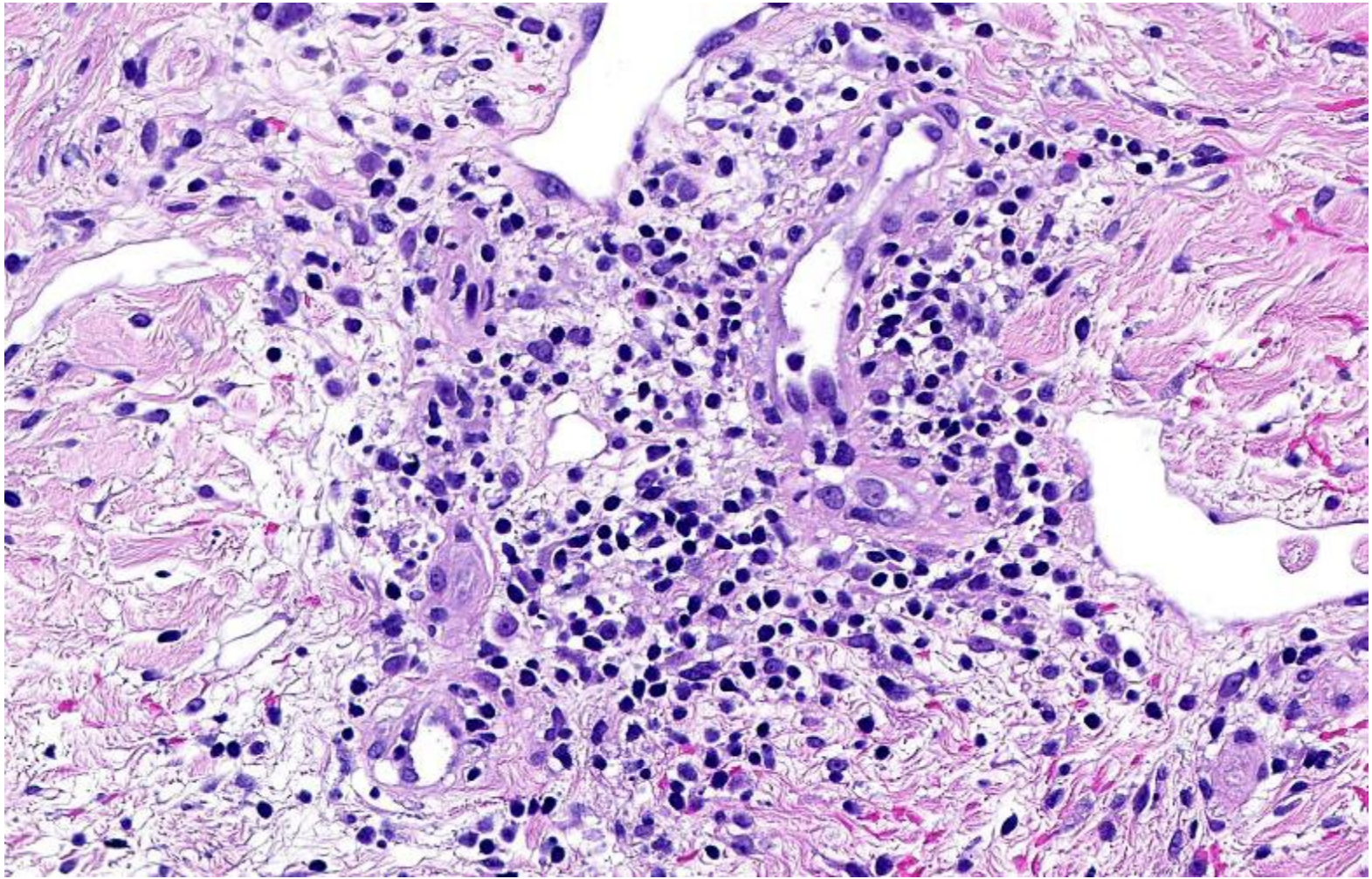


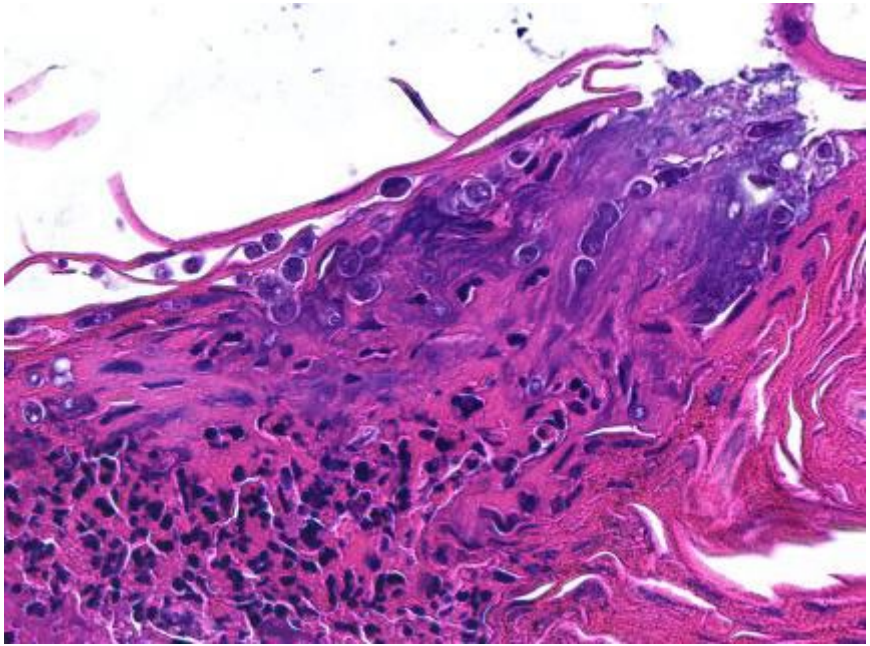


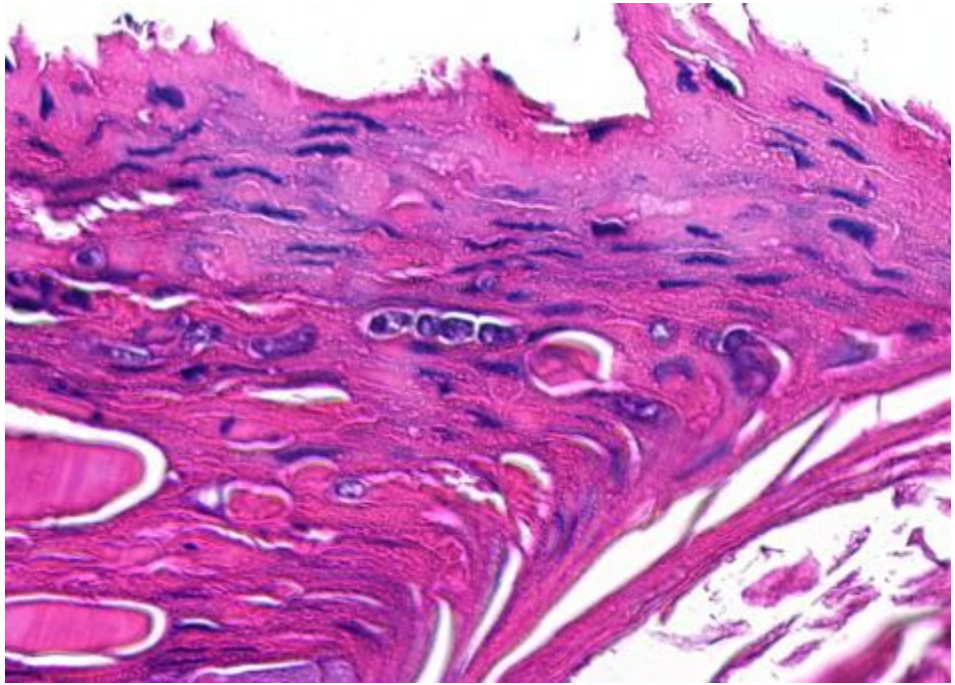
From: Figures 1 and 2
Forster AM & Mühleisen B J Cutan Pathol 2024

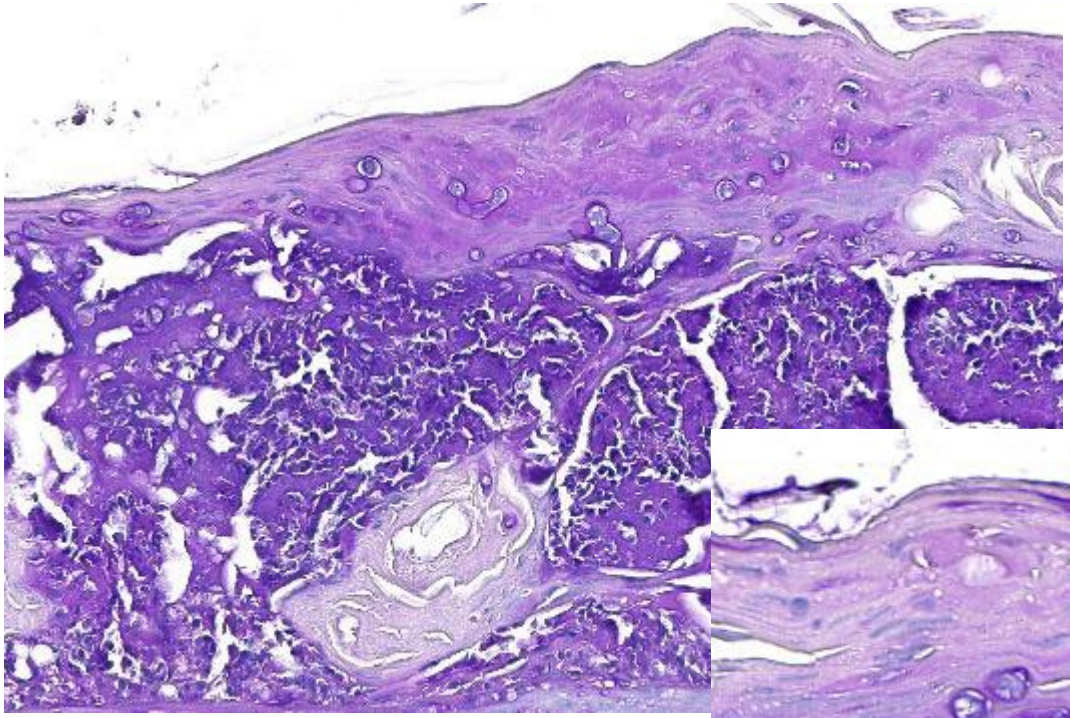
70-year-old man with hyperkeratotic lesions on the nose DDx: Lupus erythematosus, Rosacea

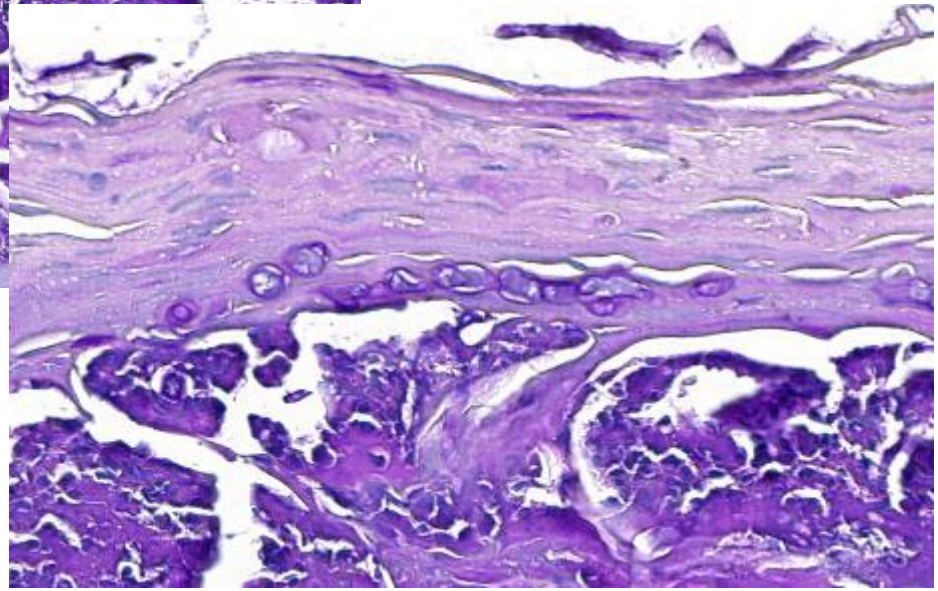




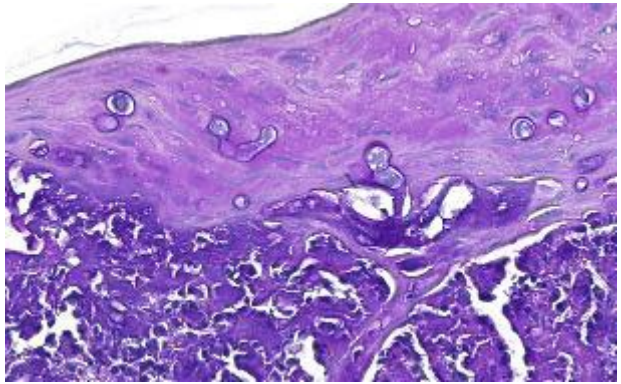


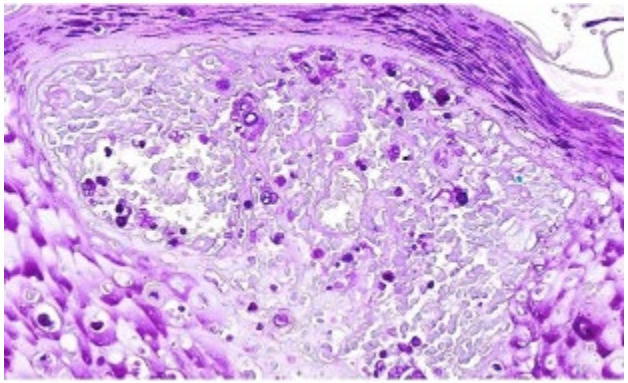






Fungal infection vs. Algae (Prototheca sp.)





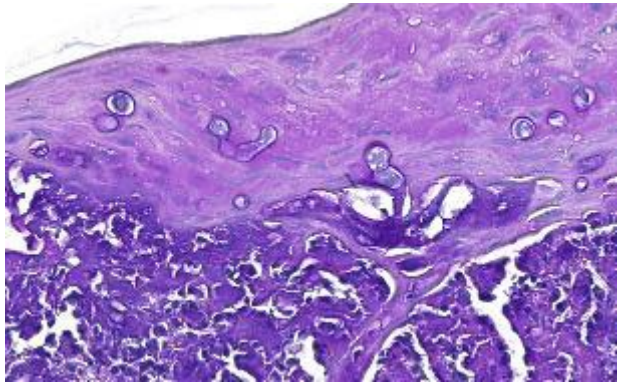
From: Li et al. Braz J Infect Dis 2025

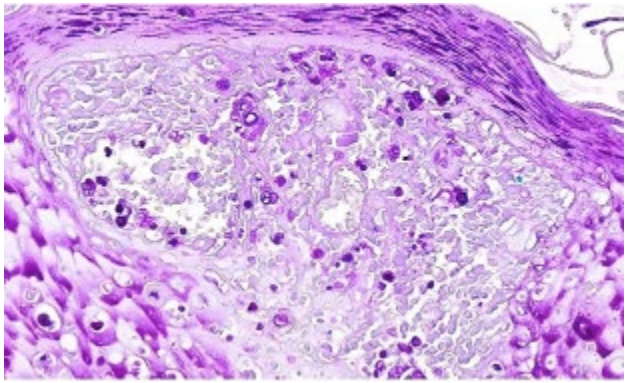
Fungal culture: no growth

Fungal PCR: negative

?

Fungal infection vs. Algae (Prototheca sp.)



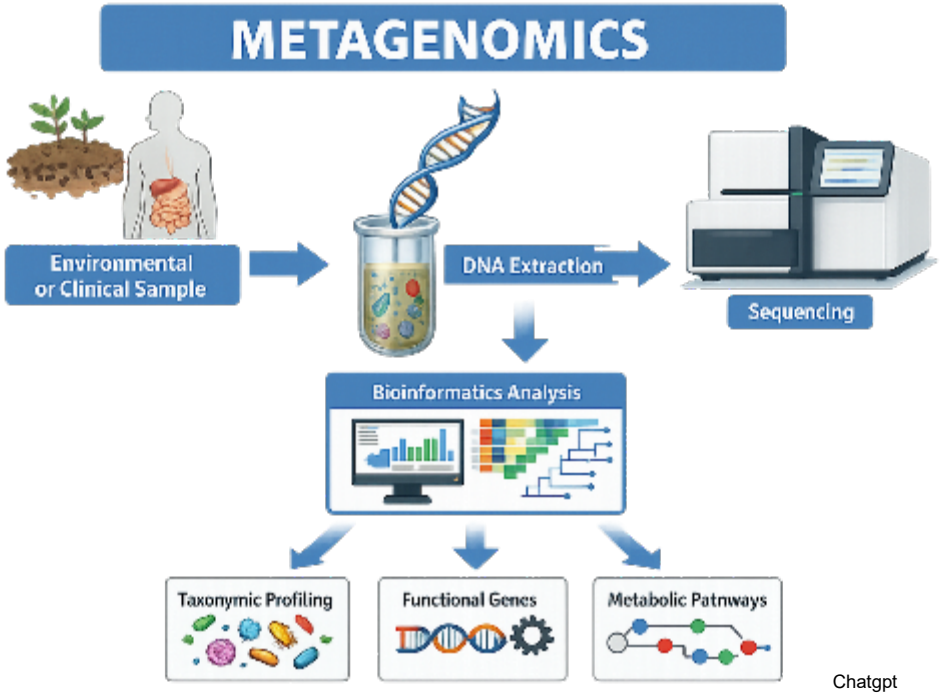


From: Li et al. Braz J Infect Dis 2025

Fungal culture: no growth

Fungal PCR: negative

→ Metagenomics



Chatgpt

Metagenomics:

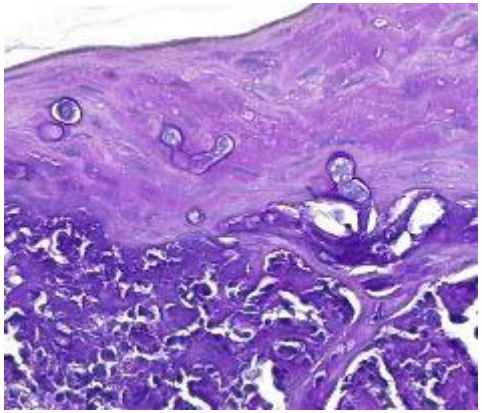
Sequencing-based analysis of all microbial DNA in a sample

Substraction of human DNA
➔ microbial DNA

➔ Sequence analysis
Identification of microbes by sequence similarity

Metagenomics

Universitätsspital Basel
Pathologie



Whole Genome Sequencing und taxonomisches Profiling der non-human Reads mittels CLC Genomics Workbench und CLC Microbial Module.

Ergebnis der Untersuchung auf Pathogene:
DNA-Viren: Keine pathogenen DNA-Viren detektiert.
Bakterien: Keine pathogenen Bakterien detektiert.
Fungi: Alternaria spp.
Protozoa: Kein Pathogen detektiert.
Taxonomie:
Ascomycota/Dothideomycetes/Pleosporales/Pleosporaceae/Alternaria/Alternaria/sp./MG1

Prof. Dr. K. Mertz, Dept. Pathology, Univ. Basel

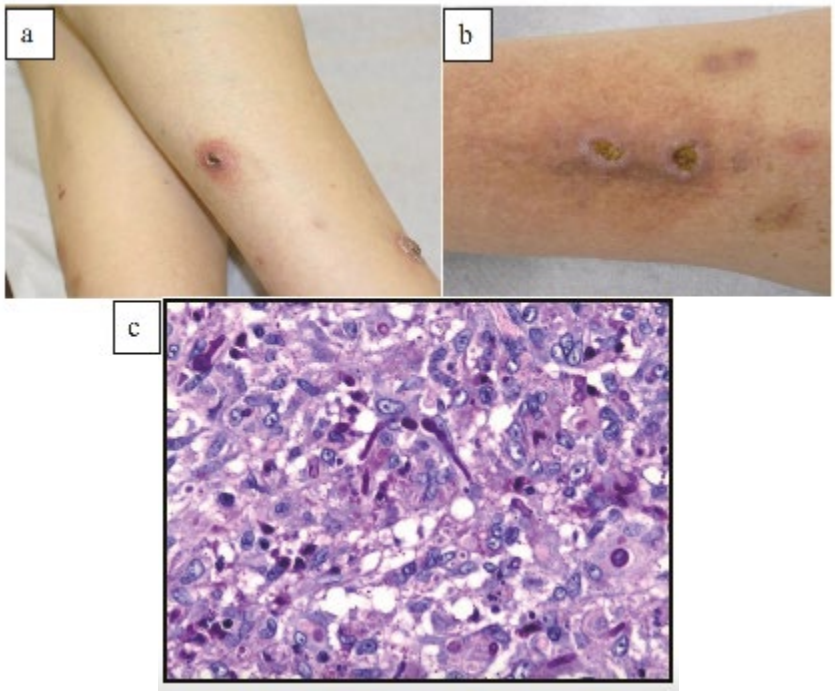
Cutaneous Alternaria infection

Broad clinical spectrum:
Papules, nodules, ulcers
erythematosus patches
verruciform lesions
vegetating tumors

Therapy:
Surgical excision and itraconazole
(from 100 to 600 mg daily - 2.8 m

Relapses possible even after
prolonged treatment

-> long-term clinical follow-up

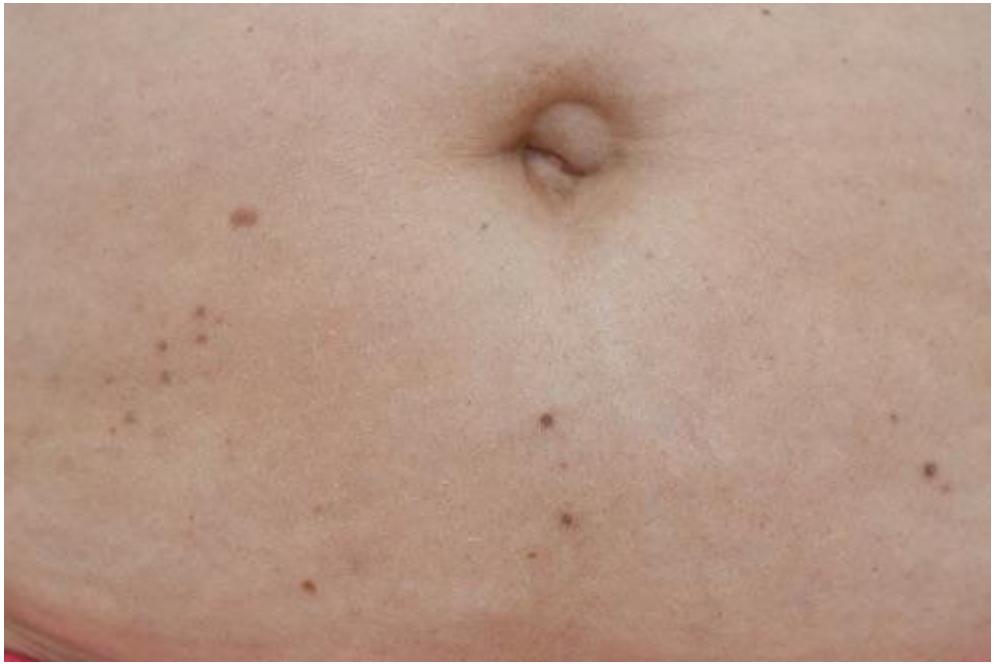


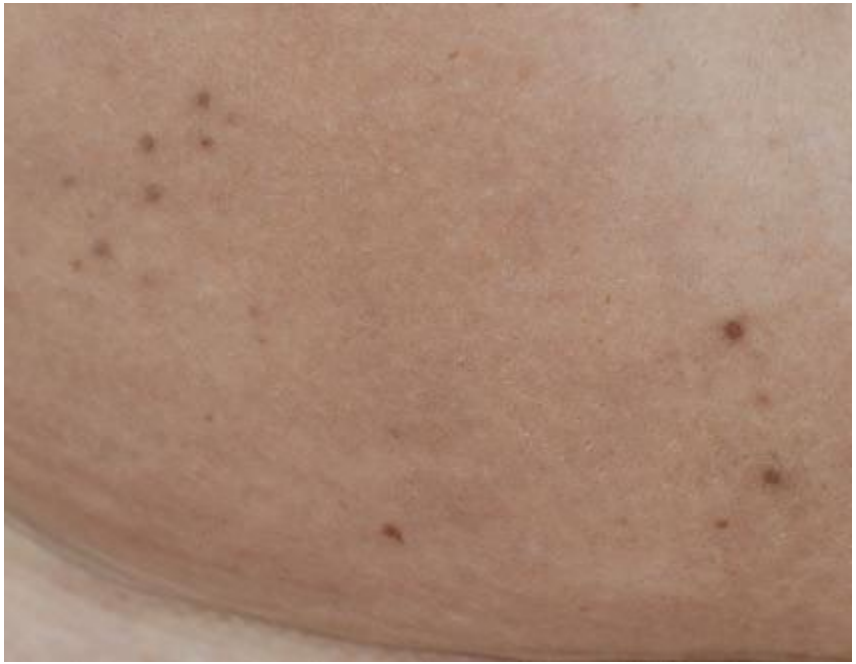
Gilaberte M et al. J Am Acad Dermatol. 2005
Ara M et al. Acta Derm. Venereol 2006
Lopes L et al. Healthcare 2013

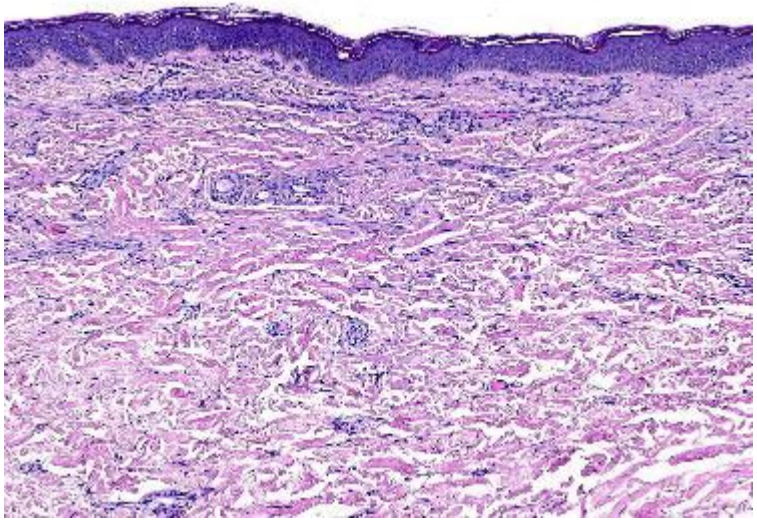
Fig. 2 from Lopes L et al. Healthcare 2013

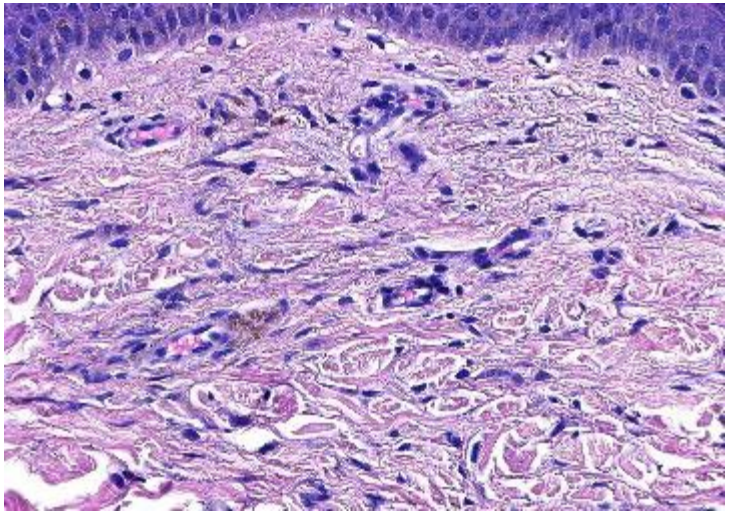
Liu Bolin

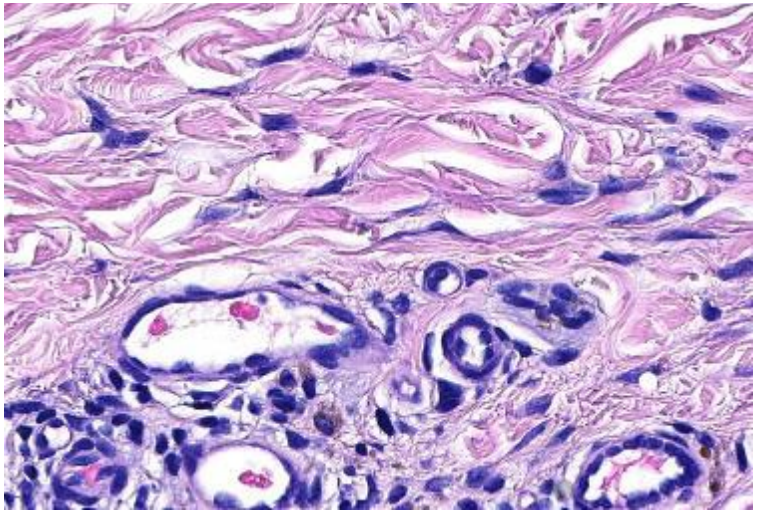
53-year-old woman with sudden onset of brownish papules on the abdomen

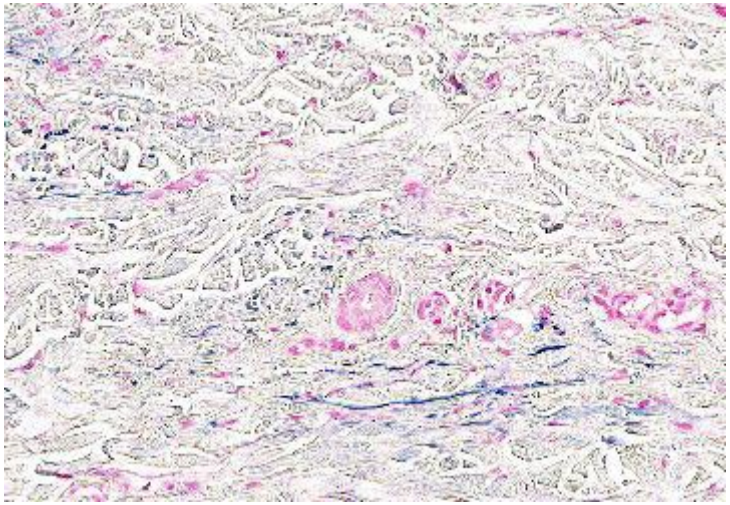








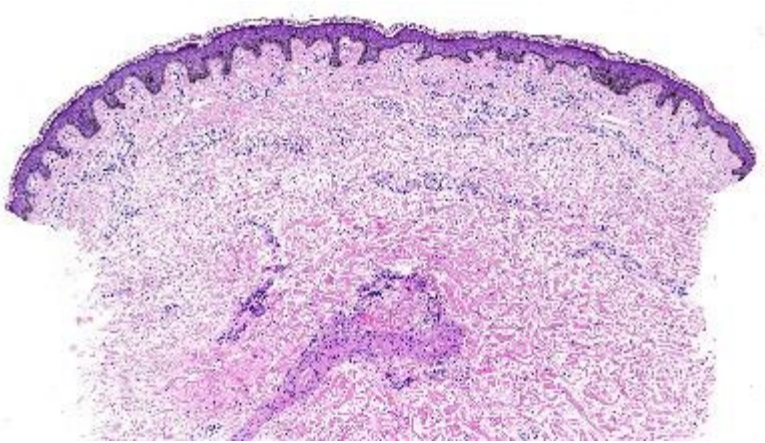


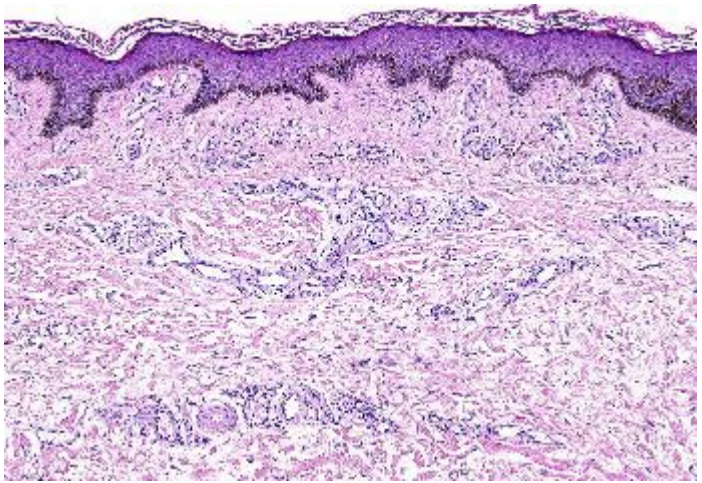


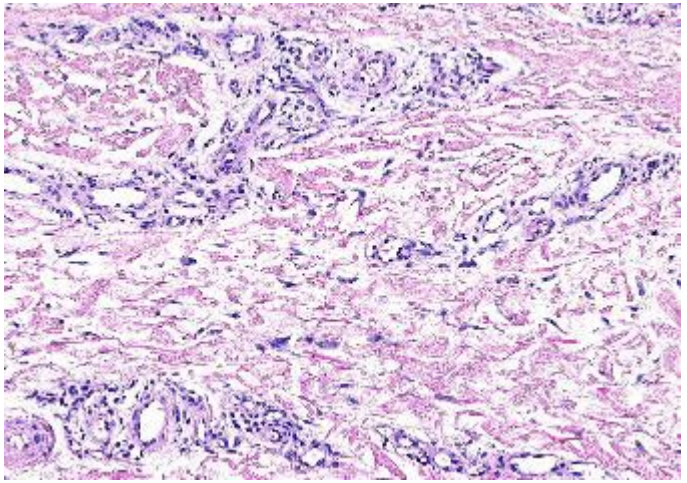
Iron

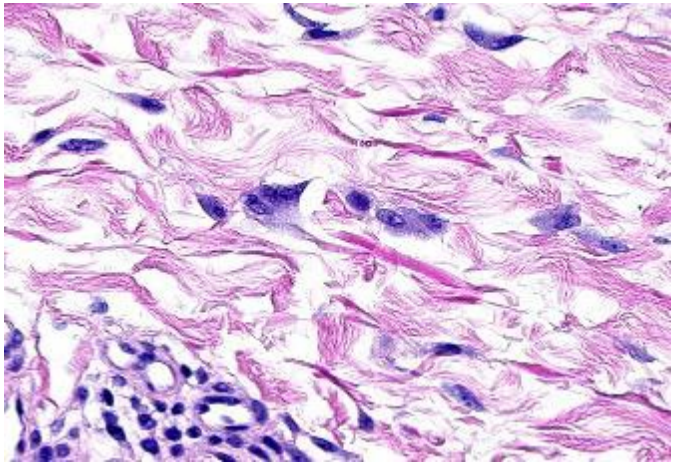
B23-68072

69-year-old woman with grouped papules on the left thigh



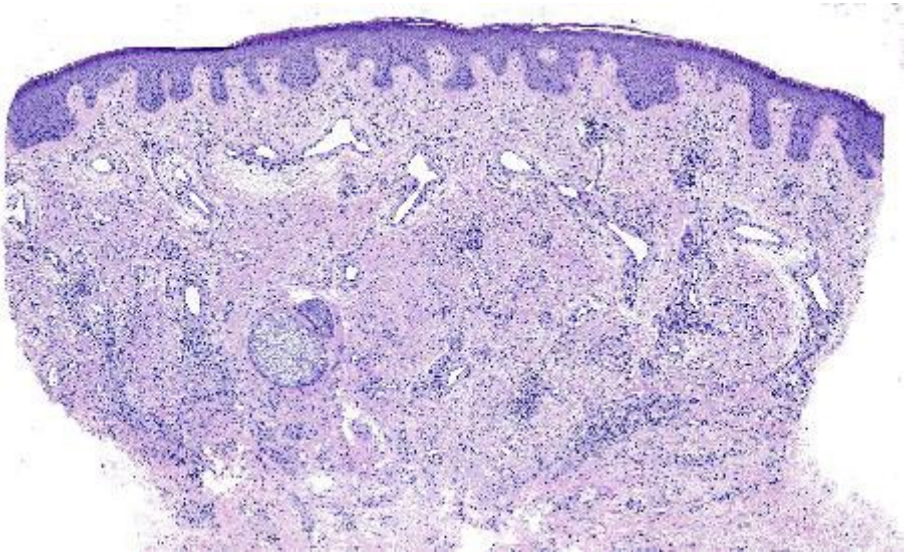


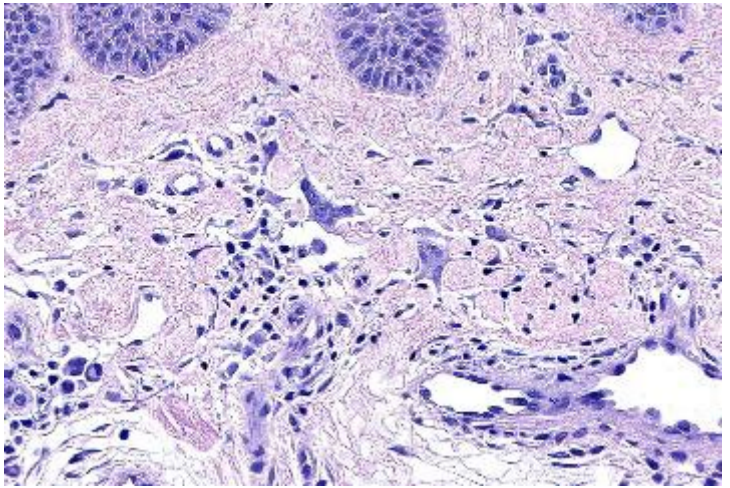


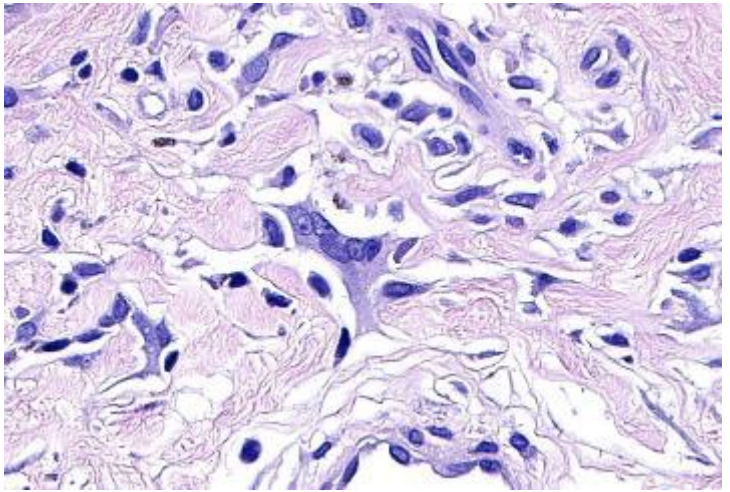


B26-49960

62-year-old man with papules on the back

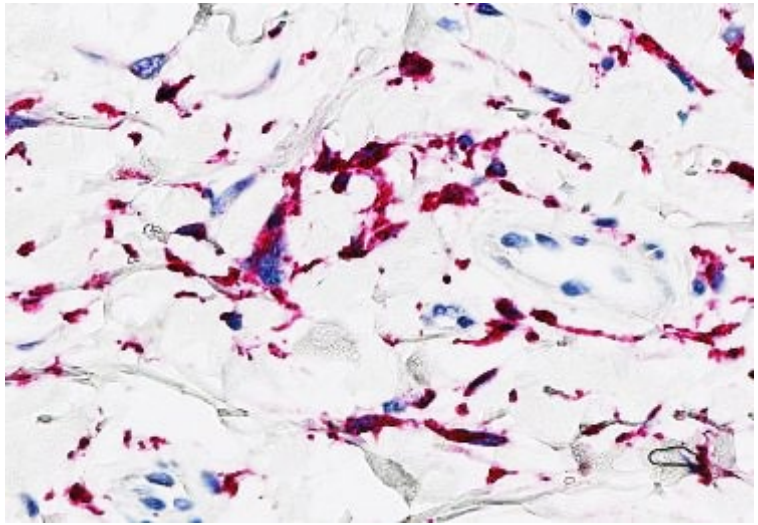


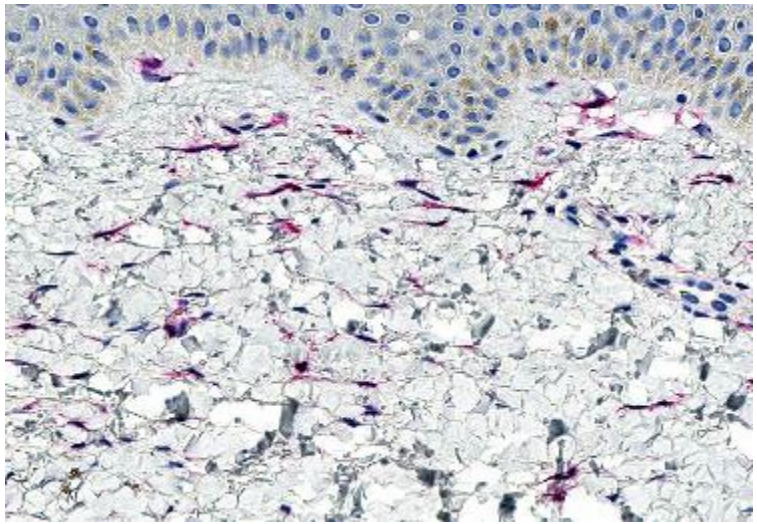




B25-88506

CD68





FXIIIa

Multinucleated angiohistiocytoma

First description: Wilson-Jones 1989

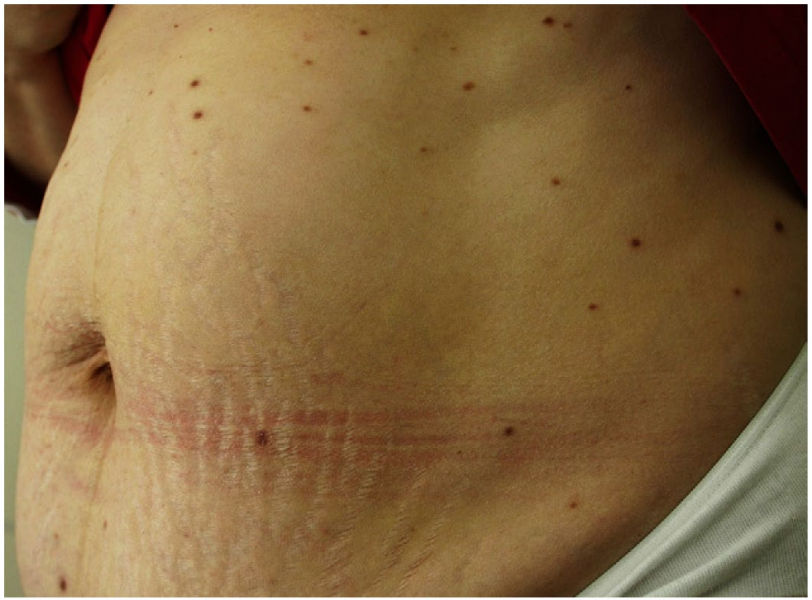
Average age of onset was 50.1 years,
mostly in women (80%)

Acral sites (30%), esp. dorsum of the hands,
and face (29%)

Frew JW Am J Dermatopath 2015

Eruptive forms are rare

Leclerc S et al. Ann Dermatol Venerol 2005
Villanueva CA et al. Actas Dermosifiliogr 2012



from: Fig. 1 Villanueva CA, et al. Actas Dermosifiliogr 2012

Multinucleated angiohistiocytoma

Histology

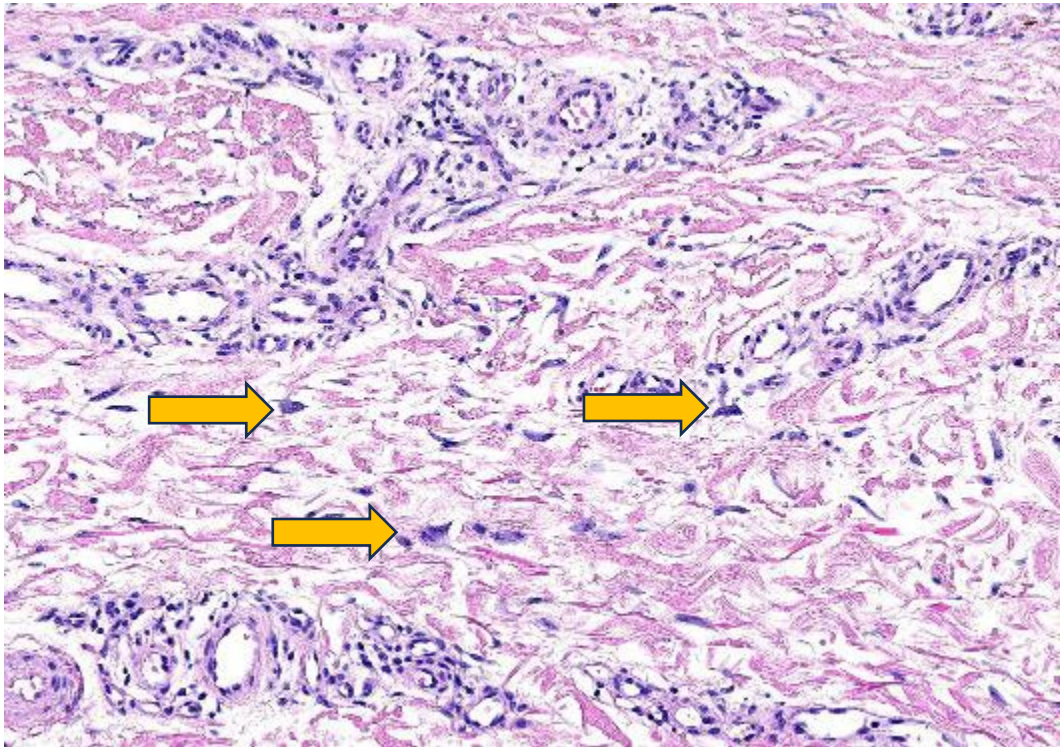
Small vessel proliferation

Bizarre basophilic multinucleated cells

Mild dermal fibrosis

Sparse lympho-histiocytic infiltrate

Dermal fibrosis associated with increased number of mast cells



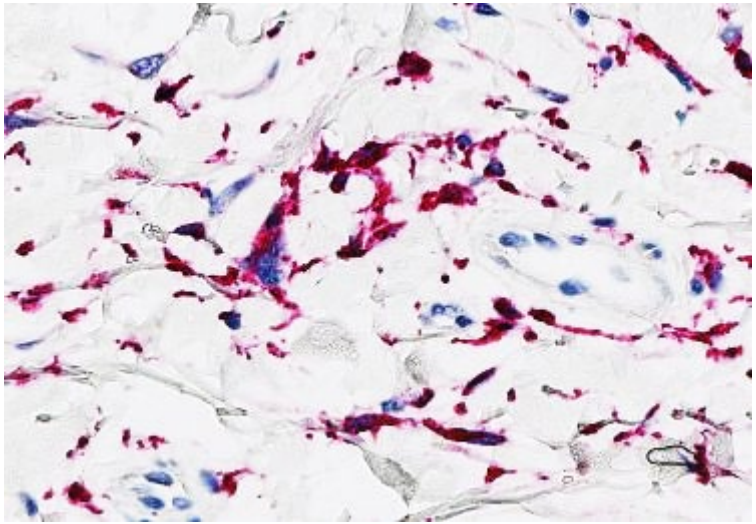
Multinucleated angiohistiocytoma

Bizarre basophilic multinucleated cells: Vimentin positive, Factor XIIIa and CD68 variable or absent.
Endothelial and Langerhans/myogenic markers absent.

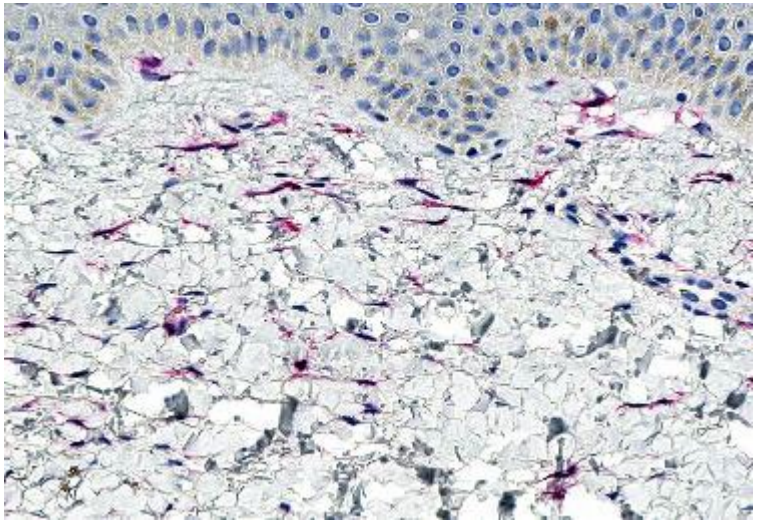
Requena L et al. J Cutan Pathol 2002
Jaconelli L et al. Dermatol Online 2009

Estrogen receptor alpha overexpression

Cesinaro AM et al. Am J Dermatopathol. 2010



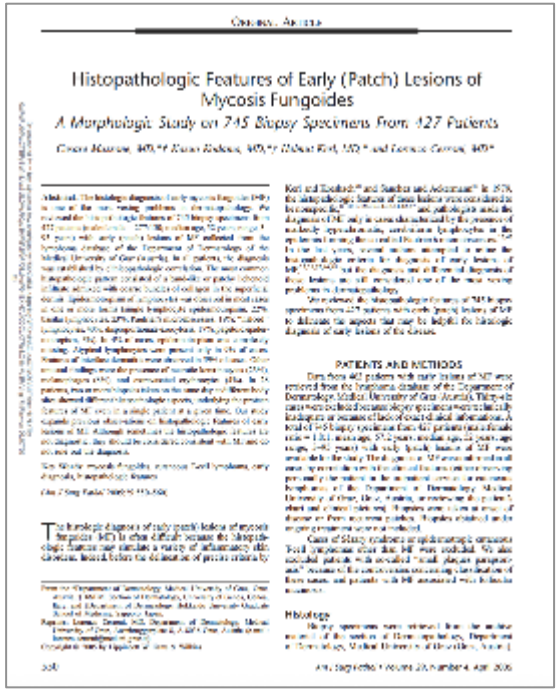
CD68



FXIIIa

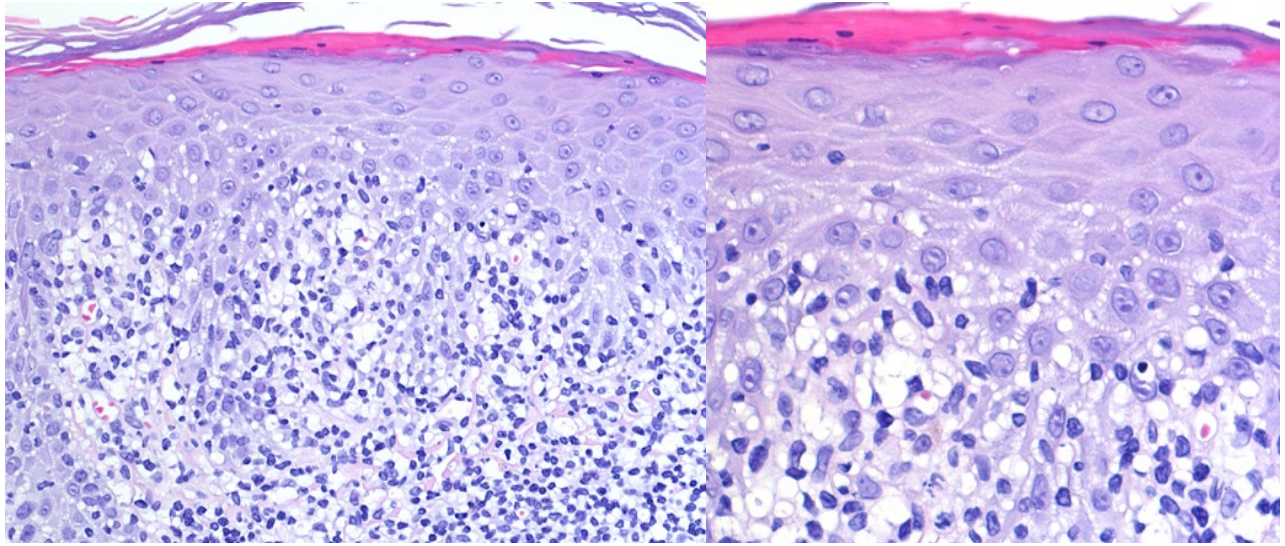
Europe, Jupiter moon, ridges and cracks with layered ice and snow
Kees Veenebos, 2006

MF (patch stage) - lichenoid pattern



Histopathologic Features of Early (Patch) Lesions of Mycosis Fungoides

A Morphologic Study on 745 Biopsy Specimens From 427 Patients

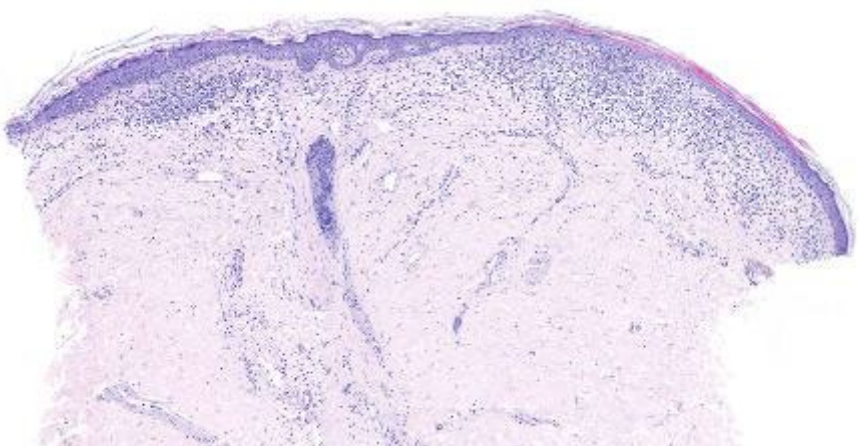


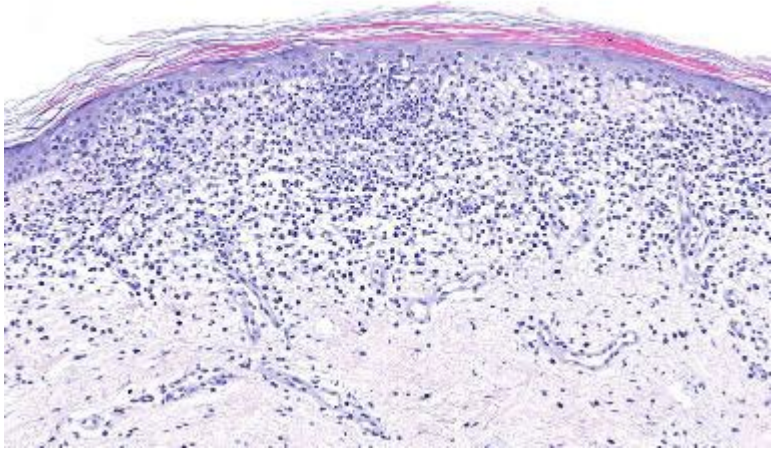
Interface changes with vacuolization in 59% of cases.

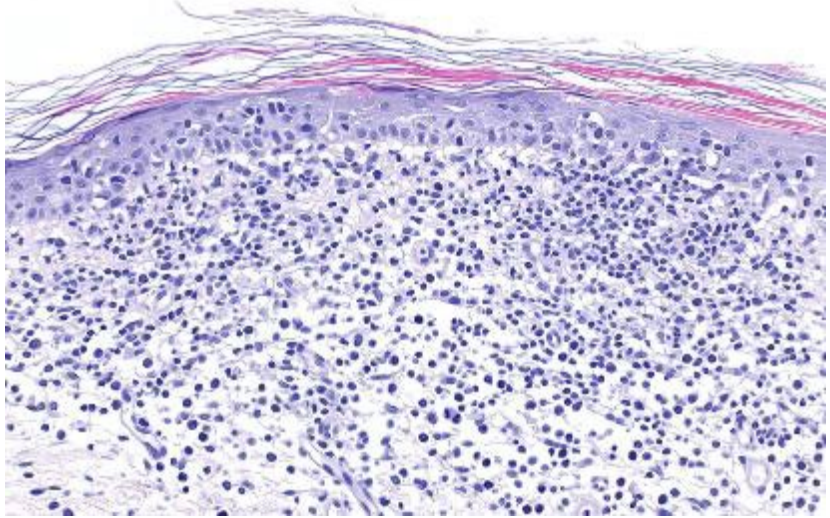
Necrotic keratinocytes (23%)

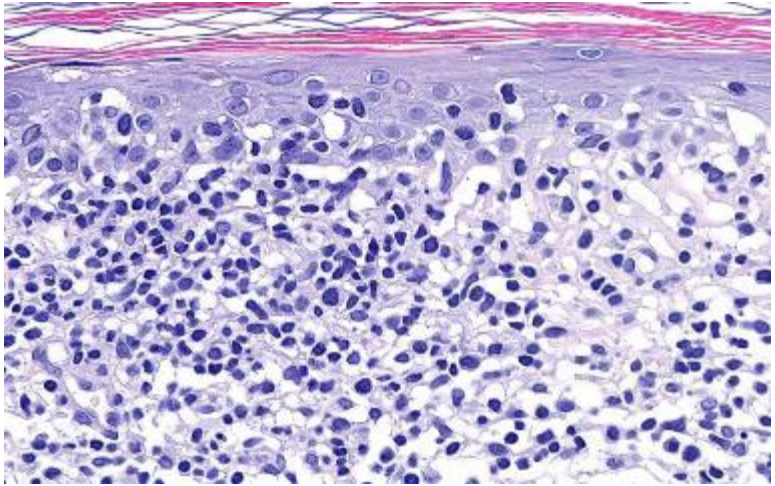
Massone et al. Am J Surg Pathol 2005

67-year-old woman with poikilodermatous patches on the trunk since 40 years.







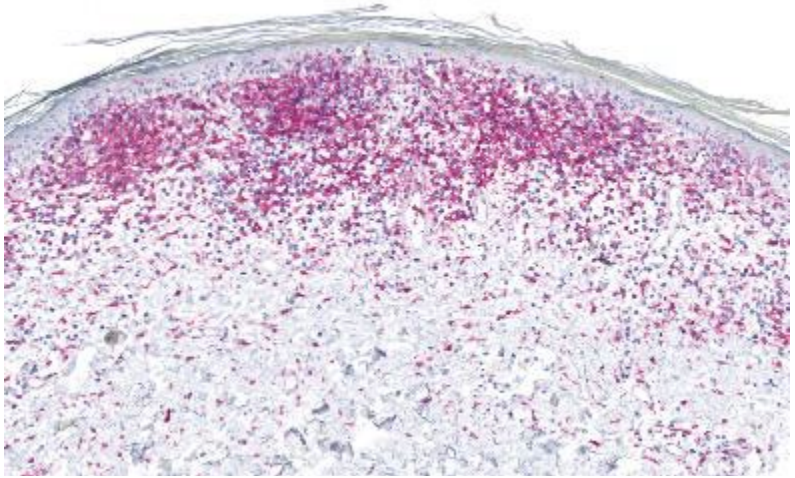


CD3+ CD4+ CD8- CD30-

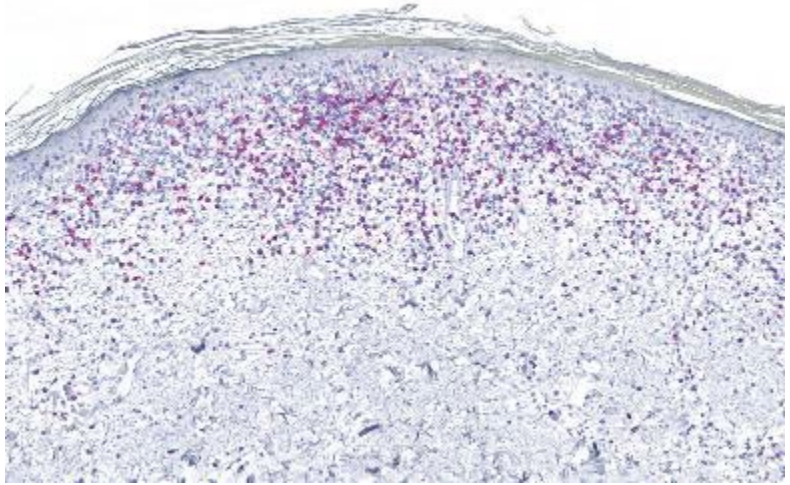
T-cell receptor gamma genes

monoclonal rearrangement

Identical in two biopsies



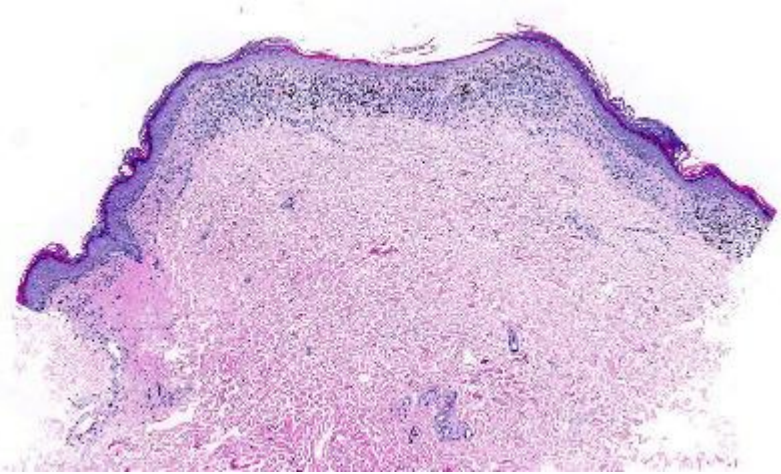
CD4

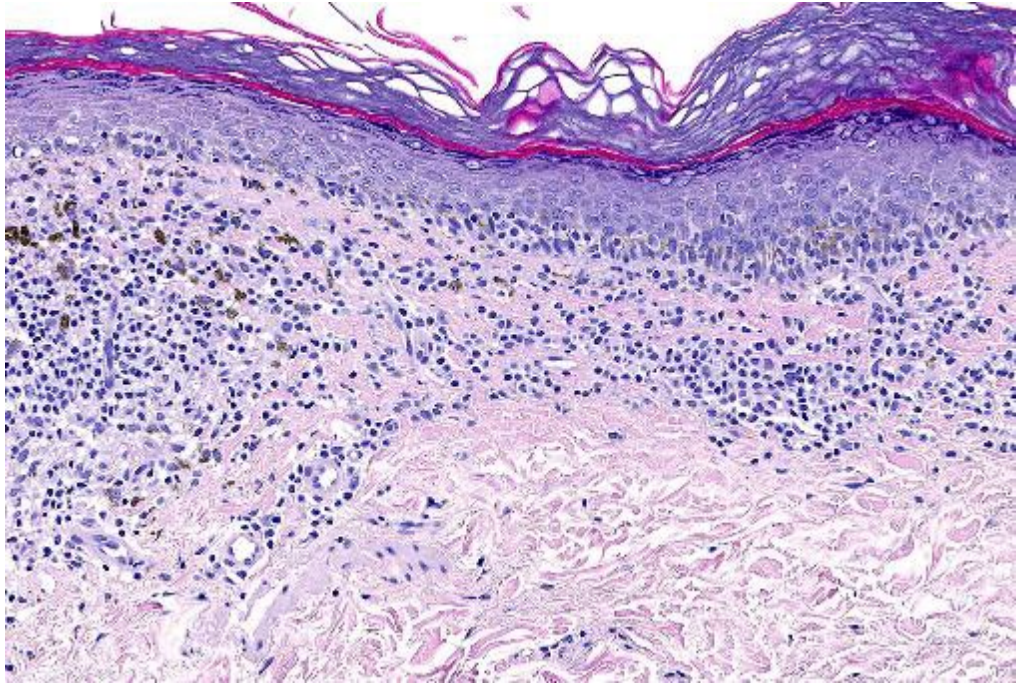


CD8

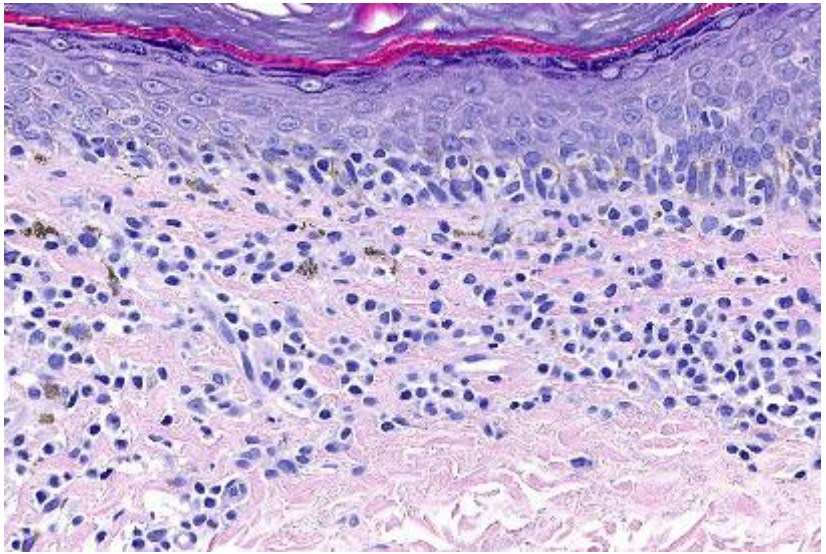
5o-year-old man with hyperpigmented papules and patches on the trunk and extremities since several years

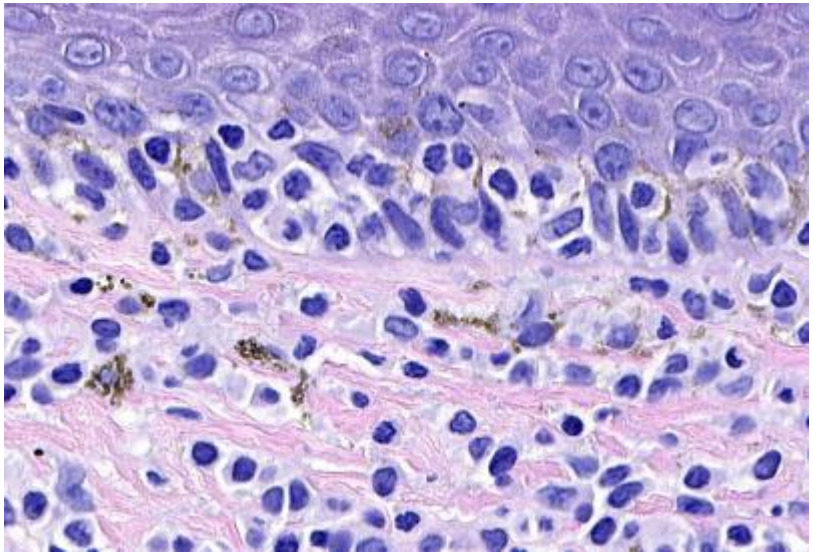
Consultation case: Dr. P-I. Stinga, Viollier, Lausanne

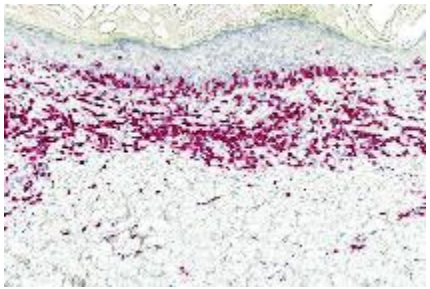




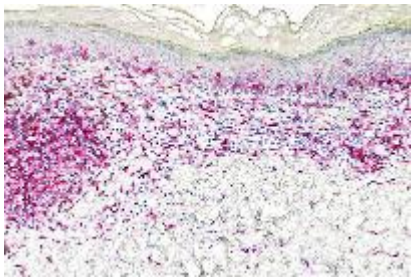
K25-192



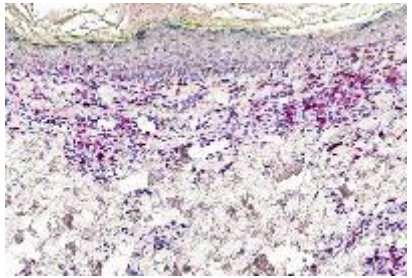




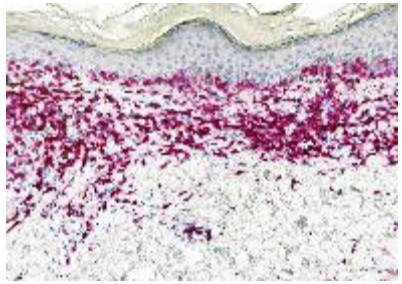
CD3



CD4

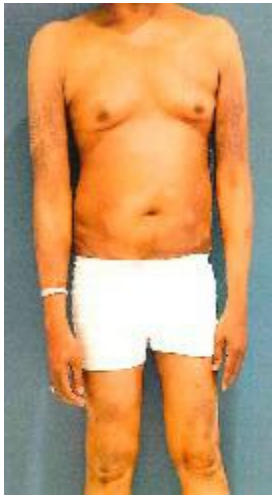
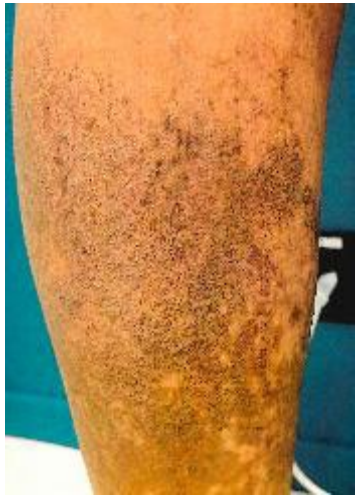
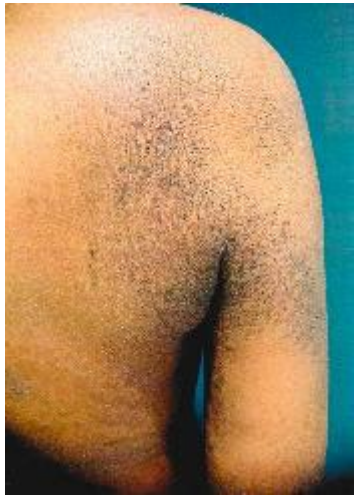


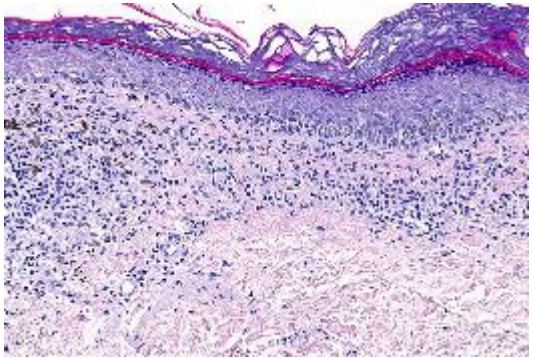
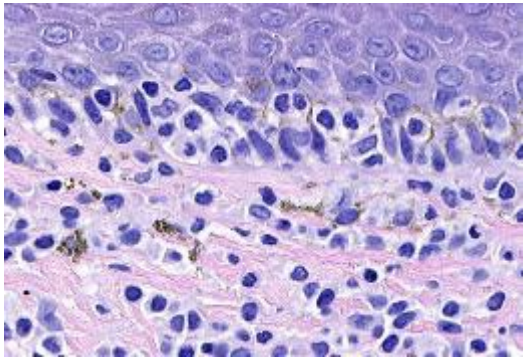
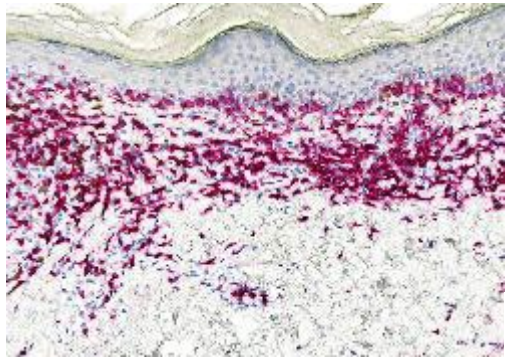
CD7



CD8

Mycosis fungoides simulating lichen planus

CD8

Hyperpigmented Mycosis Fungoides Masquerading as Longstanding Lichen Planus Pigmentosus: A Diagnostic Pitfall

Almut Böer-Auer, MD, Christian Jones, MD,† Jordan Jepson, PA,† and Masoud Azgari, MD‡*

Background: Mycosis fungoides (MF) is a rare primary cutaneous T-cell lymphoma, accounting for 50%–60% of all cutaneous T-cell lymphoma cases. It has a prevalence of approximately 5–6 cases per 1 million people annually and a higher incidence in dark-skinned populations.

Case presentation: We report a case of hyperpigmented MF in a 72-year-old dark-skinned man with a 5-year history of progressive, widespread poikilodermatous patches and thin plaques on the back and bilateral legs. The patient had been treated for lichen planus pigmentosus for 5 years without significant response to therapy.

Assessment: Multiple biopsies revealed a band-like lymphoid infiltrate in the dermis, accompanied by intraepidermal lymphocytes, some of which had larger hyperchromatic nuclei. CD4$^+$ T lymphocytes were predominant over CD8$^+$ T-positive cells located along the epidermis, dermoepidermal junction, and in the dermis.

Diagnosis: A diagnosis of hyperpigmented MF was made based on the clinical, histopathological, and immunohistochemical findings.

Conclusion: This case report highlights the importance of considering hyperpigmented MF as a differential diagnosis in patients with longstanding lichen planus pigmentosus, particularly when there is a lack of response to therapy.

Key Words: mycosis fungoides, hyperpigmentation, lichen planus pigmentosus, primary cutaneous T-cell lymphoma, CD4$^+$ T lymphocytes

(*Am J Dermatopathol* 2023;45:567–571)

From the *Dermatologikum Hamburg, Hamburg, Germany; †Golden State Dermatology, Turlock, CA; and ‡Fresno-Ie Pathology Medical Group, Modesto, CA.

Author contributions: The data are accessible to all the authors. C. Jones and J. Jepson visited and examined the patient and formulated the clinical differential diagnosis. M. Azgari reviewed the histological sections of the biopsies. M. Azgari wrote the manuscript. A. Böer-Auer edited the manuscript, enhanced its quality, and performed a PCR gene rearrangement study on the specimen. All the authors have read and approved the final manuscript.

The authors disclose no conflicts of interest.

Ethics Statement: Consent was obtained from the patient to use clinical photographs for publication.

Correspondence: Masoud Azgari, MD, Department of Pathology and Laboratory Medicine, Fresno-Ie Pathology Medical Group, 4341 North Star Way, Modesto, CA 95356-8718 (e-mail: mazgari@pmg.com).

INTRODUCTION

Mycosis fungoides (MF) is an epidermotropic primary cutaneous small-to-medium sized T-cell lymphoma. MF represents the most common subtype of cutaneous T-cell lymphomas and accounts for 50%–60% of all primary cutaneous lymphomas.[1] MF presents with various clinical, histological, and phenotypic types, each with different prognostic implications.[2–4] In addition to the classic clinical presentation of patches and plaques, there are distinct pathological variants of MF, including folliculotropic MF, pagetoid reticulosis, granulomatous MF, and granulomatous slack skin, among others.[5] Several reports have recently documented hyperpigmented patches or plaques as the predominant or sole manifestation in the African American population.[6–8] Dark-skinned individuals are more likely to develop hyperpigmented MF, which is characterized by an interface change with usually predominant CD8$^+$ T cells.[6]

Several conditions need to be considered when diagnosing circumscribed hyperpigmented patches, such as post-inflammatory hyperpigmentation, erythema dyschromicum perstans, cutaneous amyloidosis, atrophoderma of Pasini-Pierini, idiopathic eruptive macular hyperpigmentation, drug eruptions, contact dermatitis, and lichen planus pigmentosus.[9–12] In addition, a few variants of MF, such as pigmented purpura-like MF, poikilodermia vasculare atrophicans, and hyperpigmented MF, may present with similar symptoms, but can usually be distinguished based on specific histopathologic features.[13]

Like classic MF, hyperpigmented MF is treated based on the clinical stages. Prompt aggressive therapy does not seem to improve survival. In addition to phototherapy, treatment options include topical corticosteroids, topical chemotherapy, interferon alpha-2a, and oral retinoids. Combination therapies have become increasingly popular over the past decade. Advanced stages of the disease with nodal and visceral involvement are typically treated with systemic chemotherapy or immunologic agents.[13]

CASE PRESENTATION

A 72-year-old male patient with a 5-year history of progressive, widespread poikilodermatous patches and thin plaques on the upper back and both legs (Figs. 1A, B). The patient was initially presented clinically with a nonresponsive lichen planus pigmentosus to medical therapy. Histopathologic sections obtained from early hyperpigmented patches demonstrated a relatively band-like lymphocytic infiltrate obscuring dermoepidermal junction in a few foci

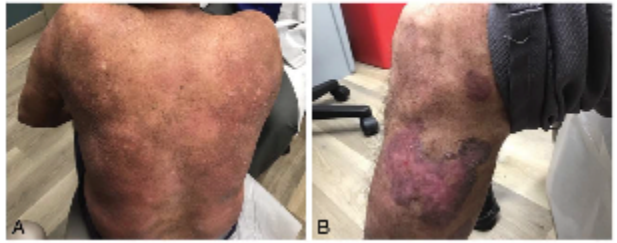
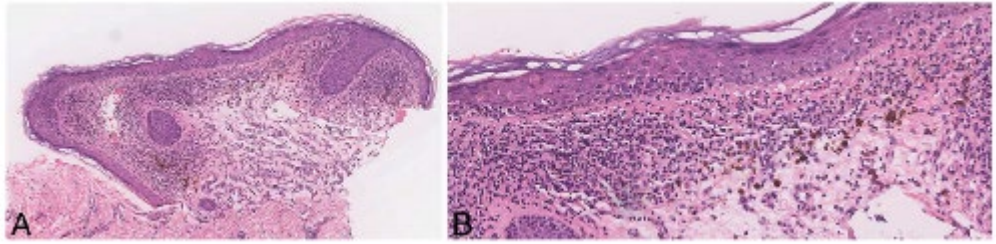
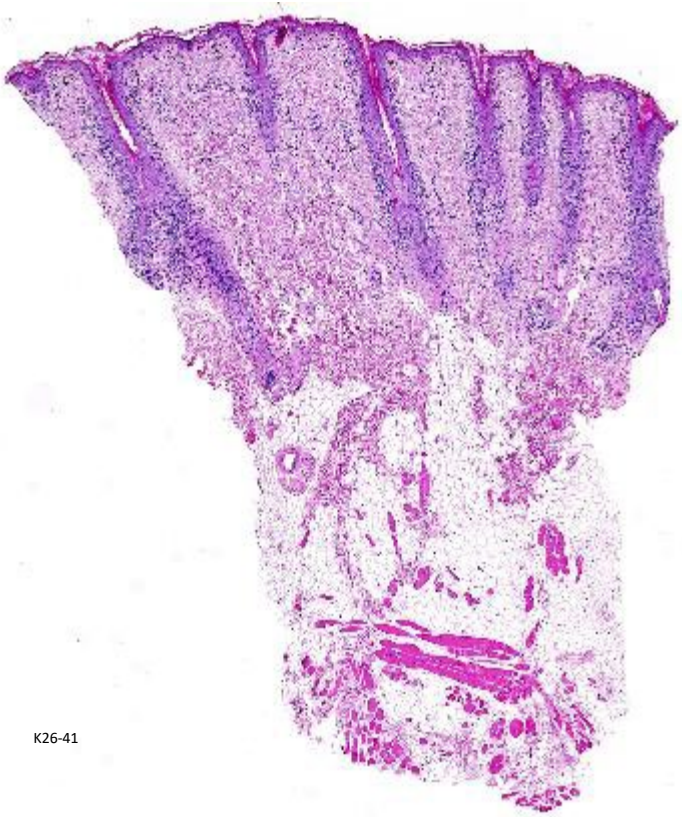


FIGURE 1. A–B, Pigmented erythematous patches and plaques with mild scaling on the back and legs. Lesions have ill-defined borders with asymmetric distribution (A) or are violaceous in various stages with slight scale (B) are characteristic. Circles indicate the biopsy site.

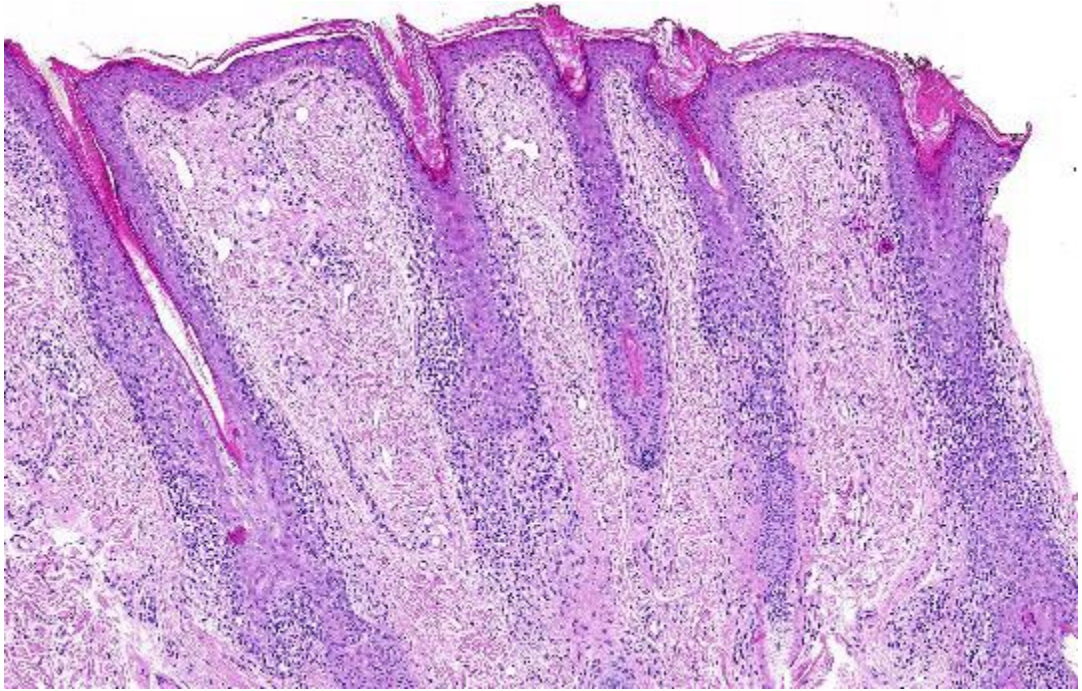


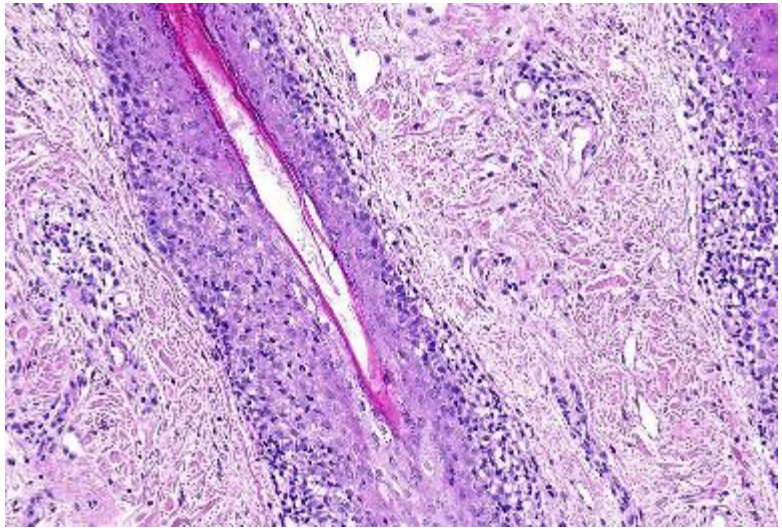
56-year-old woman with hyperpigmented lesions on the face

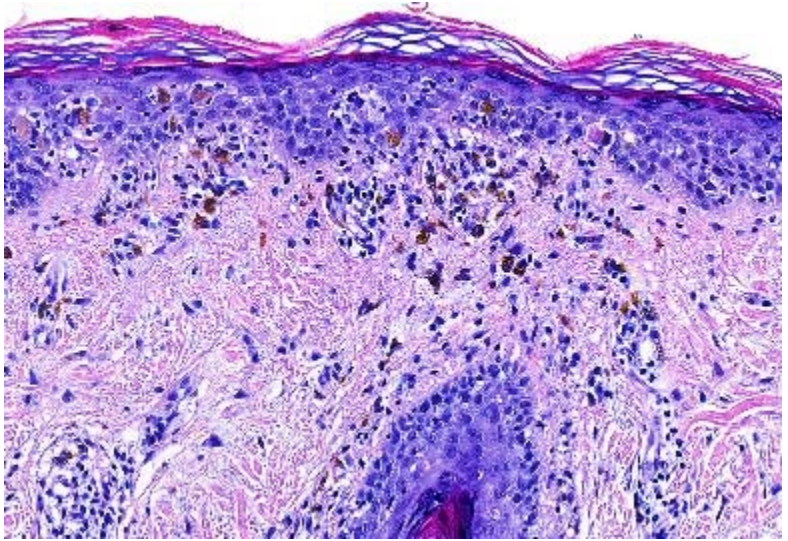
Consultation case: Dr. V. Schweinsberg, PD Dr. Iliana Tantcheva-Poor, Univ. Cologne, Germany

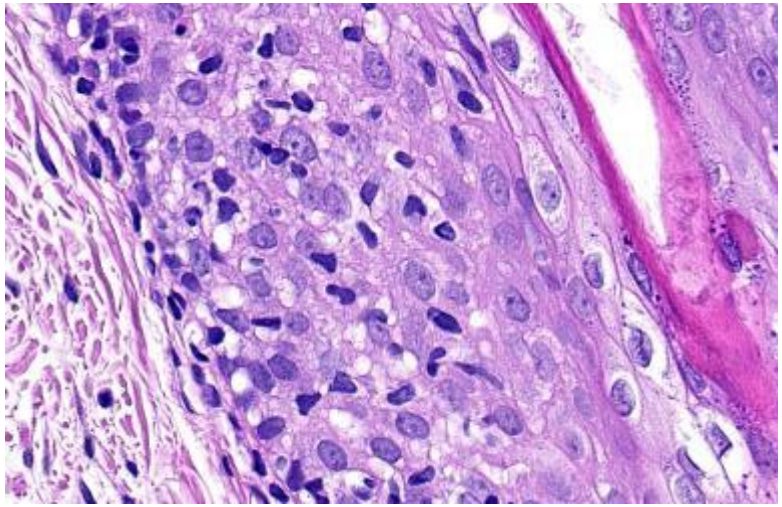


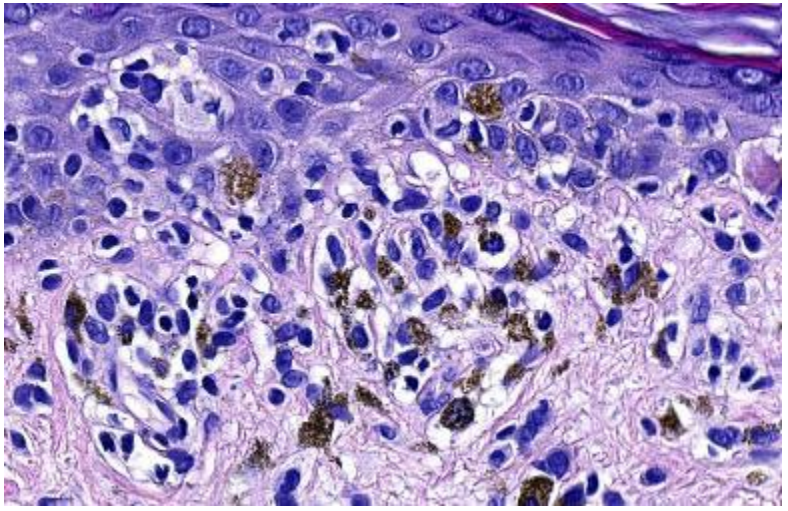
K26-41

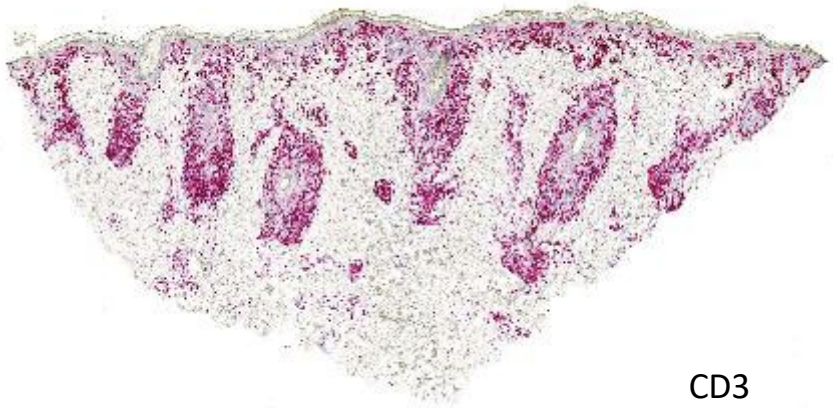




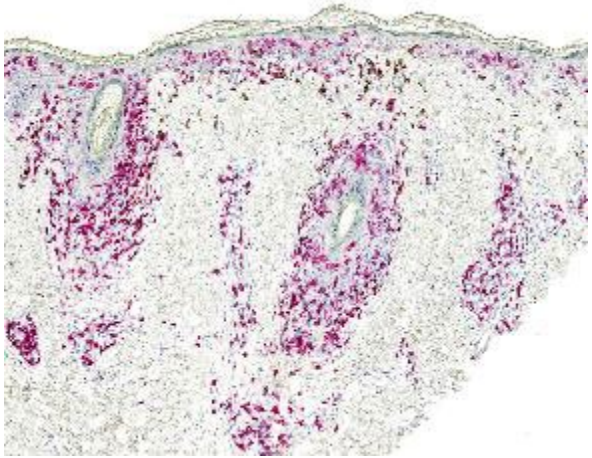




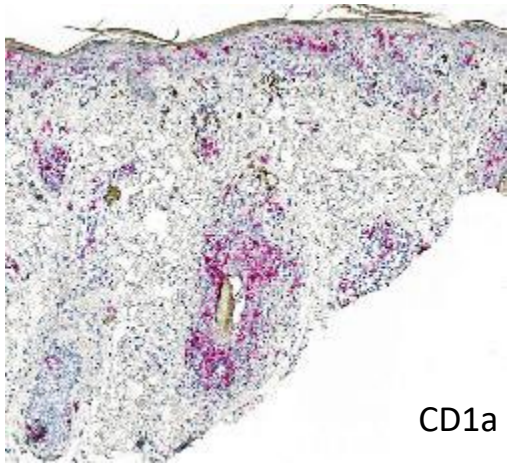




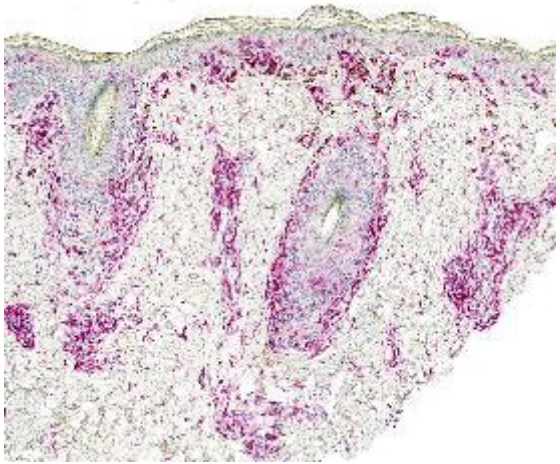
CD3



CD8

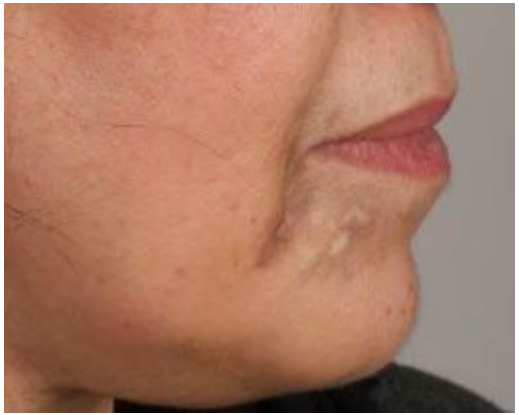


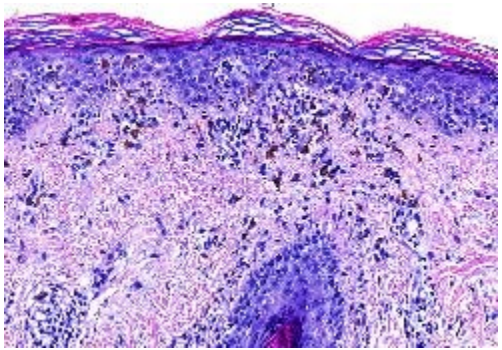
CD1a

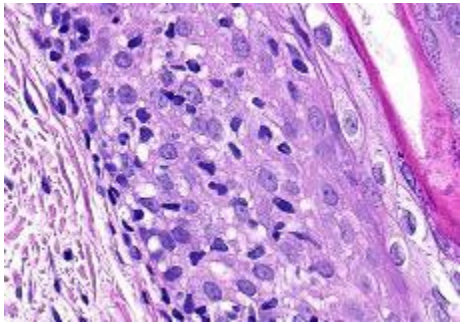


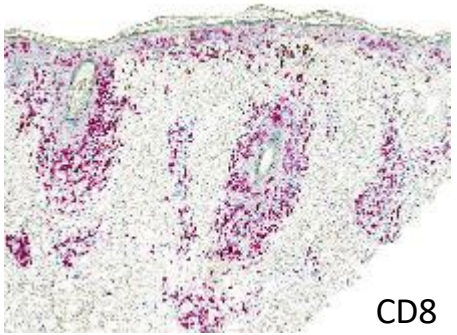
CD4

Lichen ruber follicularis et actinicus simulating folliculotropic MF



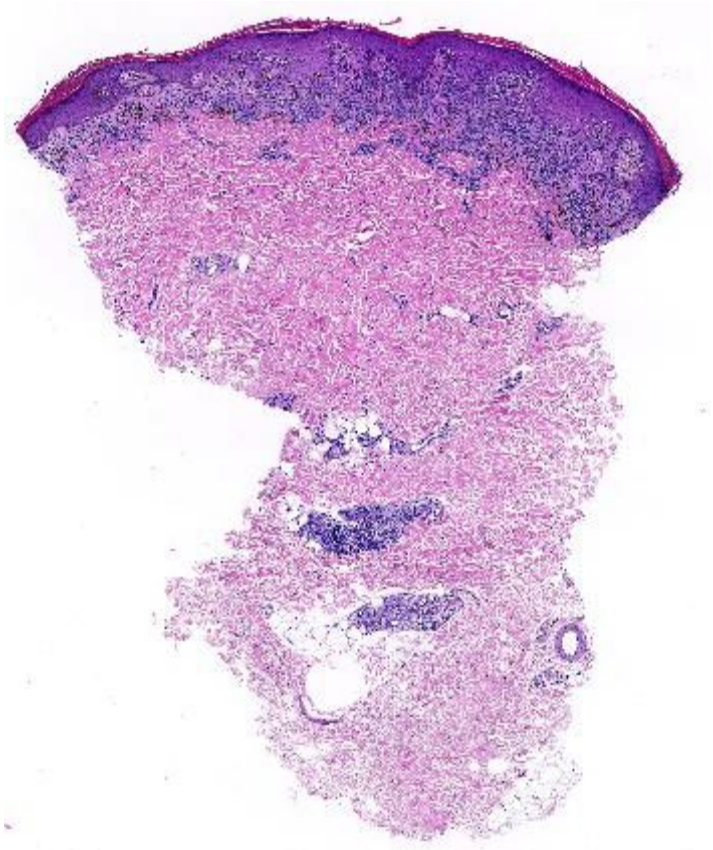


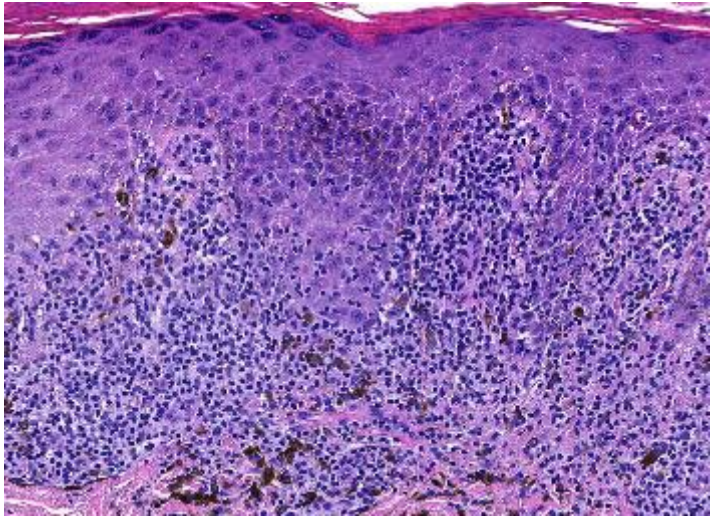


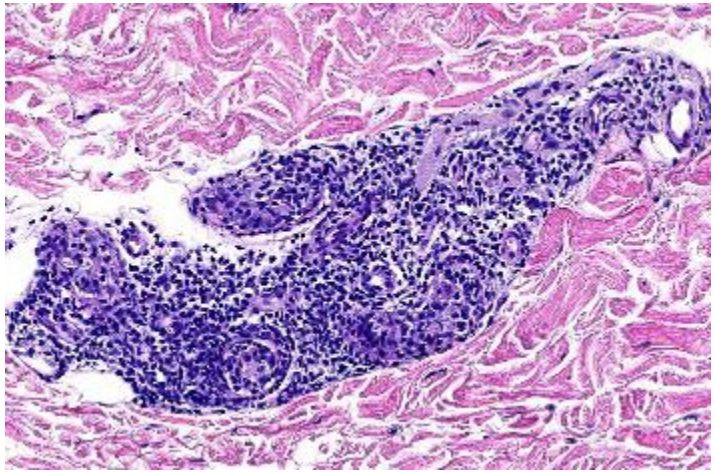


Clinical images: Dr. V. Schweinsberg, PD Dr, Iliana Tantcheva-Poor, Univ. Cologne

42-year-old man with papular lesions on the knee







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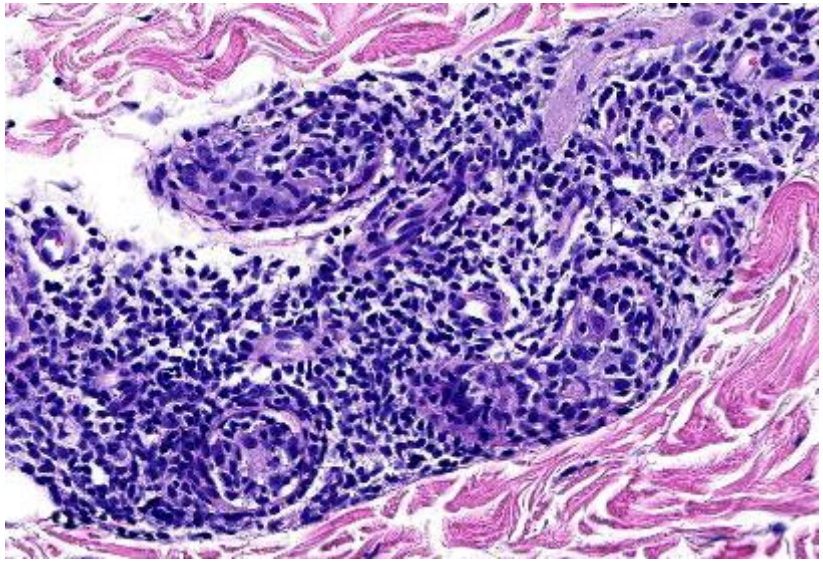

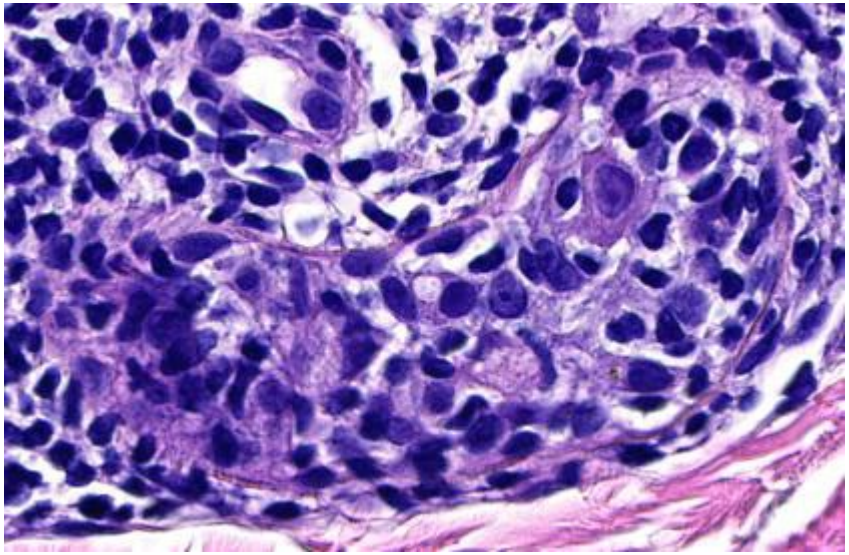

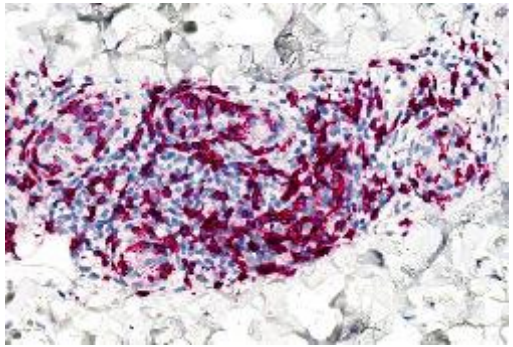

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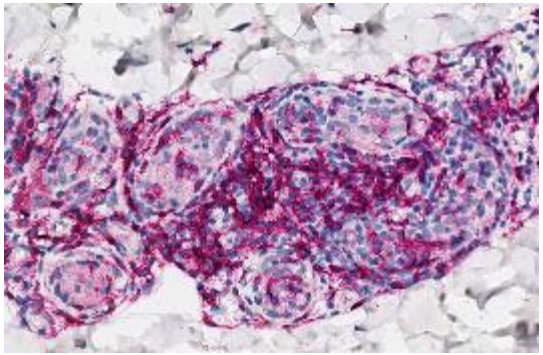

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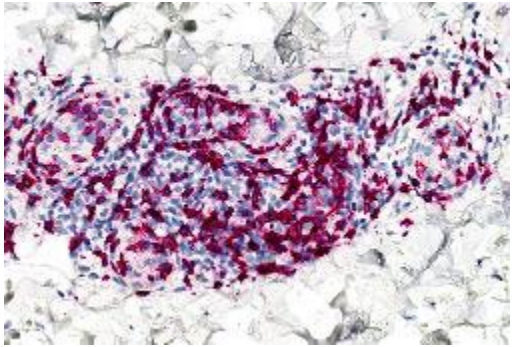

CD8

Journal of Cutaneous Pathology

Lichen striatus with syringotropism and hyperplasia of eccrine gland cells: a rare phenomenon that should not be confused with syringotropic mycosis fungoides

Lei Wang, Fengming Chen, Yu Liu, Tianwen Gao and Gang Wang

Department of Dermatology, Xijing Hospital, Fourth Military Medical University, Xi'an, China

Background: Syringotropism is characterized by lymphocyte infiltration in the eccrine gland and is usually associated with various degrees of hyperplasia of eccrine gland cells. This phenomenon has been reported in rare cases of mycosis fungoides, which are also called as syringotropic mycosis fungoides.

Methods: We studied seven cases of lichen striatus associated with syringotropism and hyperplasia of eccrine gland cells, diagnosed at our dermatology department in the past 5 years. The hematoxylin and eosin stained slides from these cases were analyzed, and immunohistochemical and T-cell receptor gene rearrangement studies were performed.

Results: Of the seven cases, two showed prominent and five showed subtle syringotropism and hyperplasia of eccrine gland cells. Immunohistochemical study showed mixed infiltration by T-cells and B-cells around the eccrine glands. The T-cells were composed of CD4 and CD8-positive cells. T-cell receptor gene rearrangement study showed negative results in all the cases.

Conclusion: Syringotropism and hyperplasia of eccrine gland cells is a rare phenomenon in lichen striatus. Dermatopathologists should be aware of this to avoid misdiagnosis as syringotropic mycosis fungoides.

Keywords: eccrine gland, hyperplasia, lichen striatus, mycosis fungoides, syringotropism

Wang L, Chen F, Liu Y, Gao T, Wang G. Lichen striatus with syringotropism and hyperplasia of eccrine gland cells: a rare phenomenon that should not be confused with syringotropic mycosis fungoides. J Cutan Pathol 2016; 43: 927–931. © 2016 John Wiley & Sons A/S. Published by John Wiley & Sons Ltd

Gang Wang, MD, PhD, and Tianwen Gao, MD, PhD, Department of Dermatology, Xijing Hospital, Fourth Military Medical University

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Extraordinary Case Report

Syringotropic Lichen Planus: A Potential Histopathologic Mimicker of Syringotropic Mycosis Fungoides

Mauro Mazzeo, MD,† Andrea Saggini, MD,*† Tea Rocco, MD,† Maria Concetta Fargnoli, MD,† and Lorenzo Cerroni, MD**

Abstract: Perieccrine inflammation may be observed in several different dermatoses, but true permeation of the secretory coil by lymphocytes (lymphocytic syringotropism) is a rather uncommon finding, usually observed in mycosis fungoides (MF—syringotropic MF). Rare cases of syringotropic lichen striatus and lymphocytic autoimmune hidradenitis showing a similar pattern have been described as well. We describe an exceptional case of lichen planus (LP) characterized by marked lymphocytic syringotropism with focal hyperplasia of the eccrine epithelium. Histopathology was characterized by the combination of features of conventional LP, prominent permeation of the secretory portion of the eccrine glands by reactive lymphocytes, and focal involvement of a hair follicle. Syringotropic LP may be regarded as a histologic mimicker of syringotropic MF, thus representing a potential diagnostic pitfall.

Key Words: lichen planus, syringotropic lichen planus, adnexotropic lichen planus, adnexotropic mycosis fungoides, syringotropic mycosis fungoides, syringometaplasia

(*Am J Dermatopathol* 2019;41:e51–e53)

chemotherapeutic agents), or of palmoplantar eccrine hidradenitis,[3] an idiopathic condition with a predilection for children. Histologic evidence of lymphocytic syringotropism has been conventionally linked to mycosis fungoides (MF syringotropic MF),[4-6] although rare cases of syringotropism in lichen striatus (LS)[7] or lymphocytic autoimmune hidradenitis[8] have been described as well.

We report an unusual case of lichen planus (LP) with prominent involvement of the eccrine glands.

CASE REPORT

A 35-year-old woman with a history of surgically treated fibrocystic breast disease presented with itchy, lichenoid, erythematous-brownish papules, and flat, small plaques present for the past 8 months. The lesions were located on the upper and lower extremities as well as on the trunk, also involving the right breast at the site of the previous surgical intervention (Fig. 1). Systemic antihistaminic treatment could not control the pruritic symptoms, and local steroid treatment had resulted in only minimal improvement.

Three biopsies were obtained from the right breast (2 specimens) and the right lower arm, respectively. The biopsy from the

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PATHOLOGICA 2026;118:53-55
DOI: 10.32074/1591-951X-1996

Letter to the Editor

"Adnexotropic" lichen striatus: a potential histological mimicker of mycosis fungoides

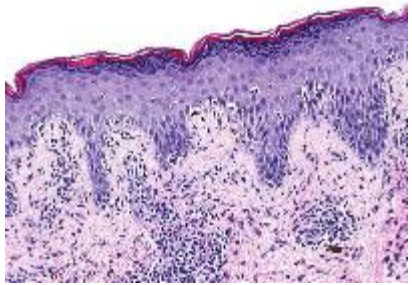
Francesco Fortarezza[1], Francesca Caroppo[2,3], Alessandra Maneghel[4], Francesco Tinti[5], Cesare Tiengo[5], Anna Belloni Fortina[3], Francesco Zulian[4], Angelo Paolo Dei Tos[1,5]

[1] Surgical Pathology and Cytopathology Unit, Department of Integrated Diagnostics, University Hospital of Padua, Padua, Italy; [2] Pediatric Dermatology Regional Center, Department of Women and Children's Health, University of Padua, Padua, Italy; [3] Department of Medicine (DIMED), Dermatology Unit, University of Padua, Padua, Italy; [4] Pediatric Rheumatology Unit, Department for Women and Children's Health, University Hospital of Padua, Padua, Italy; [5] Plastic and Reconstructive Surgery Unit, Department of Neurosciences, University of Padua

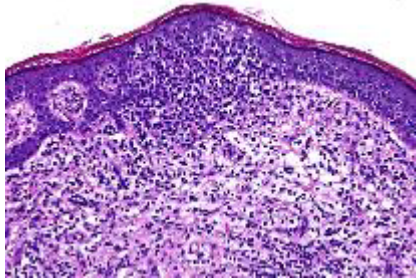
B22-48431

MF and interface dermatitis

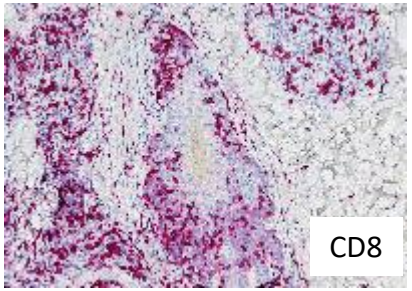
Drug reaction



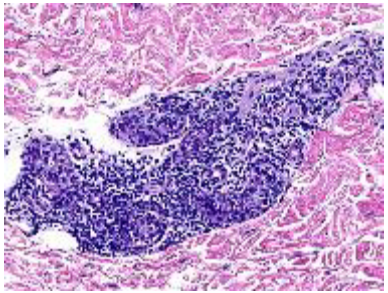
L. sclerosus



LR follicularis



L. striatus



Predominance of CD8 positive lymphocytes
Clonality does not allow distinction
Nuclear atypia - pitfall

dermatopathology

MDPI

Review

Adnexotropic Variants of the Interface Dermatitides: A Review

Carla Stephan [1,†], Ossama Abbas [1,†] and Jag Bhawan [2,*]

[1] Department of Dermatology, American University of Beirut, Beirut 11-0236, Lebanon; cs63@aub.edu.lb (C.S.); ossamaabbas2003@yahoo.com (O.A.)
[2] Department of Dermatology, Boston University School of Medicine, Boston, MA 02118-2413, USA
* Correspondence: jbhawan@bu.edu
† These authors contributed equally to this work.

Abstract: The interface dermatitides encompass a vast array of cutaneous entities which, at times, may present with particular clinical variants with adnexal predilection. Similarly, hair follicle and eccrine gland involvement of some of these entities has been observed on histopathology. This review aims to describe the various adnexotropic presentations of the interface dermatitides. Recognizing that the adnexa can be a frequent site of involvement of these conditions may aid dermatopathologists in making the correct diagnosis and avoid misinterpreting adnexotropism for other conditions such as the great imitator, mycosis fungoides.

Keywords: interface dermatitis; folliculotropic; syringotropic; lichen planus; lichen sclerosus; graft versus host disease; lichen nitidus; lichen striatus; mycosis fungoides

check for updates

Citation: Stephan, C.; Abbas, O.; Bhawan, J. Adnexotropic Variants of the Interface Dermatitides: A Review. *Dermatopathology* **2021**, *8*, 125–146. https://doi.org/10.3390/dermatopathology8010015

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1. Introduction

Interface dermatitis is a pathological pattern characterized by the presence of basal cell vacuolization and apoptotic keratinocytes. A variety of dermatoses exhibit interface dermatitis on pathology including the lichenoid dermatoses, graft versus host disease, connective tissue diseases, and drug reactions, among others [1,2].

Several entities of interface dermatitides are known to have distinct rare adnexotropic variants whereby the inflammation involves the adnexa of the skin such as the hair follicle or the sweat gland. In lichen planus for example, follicular and syringotropic variants have been classically described.

Adnexal involvement can also at times be seen on histopathology of the interface dermatitides that do not have distinct adnexotropic variants. For example, adnexal inflammation can be seen in lichen striatus or in pityriasis lichenoides.

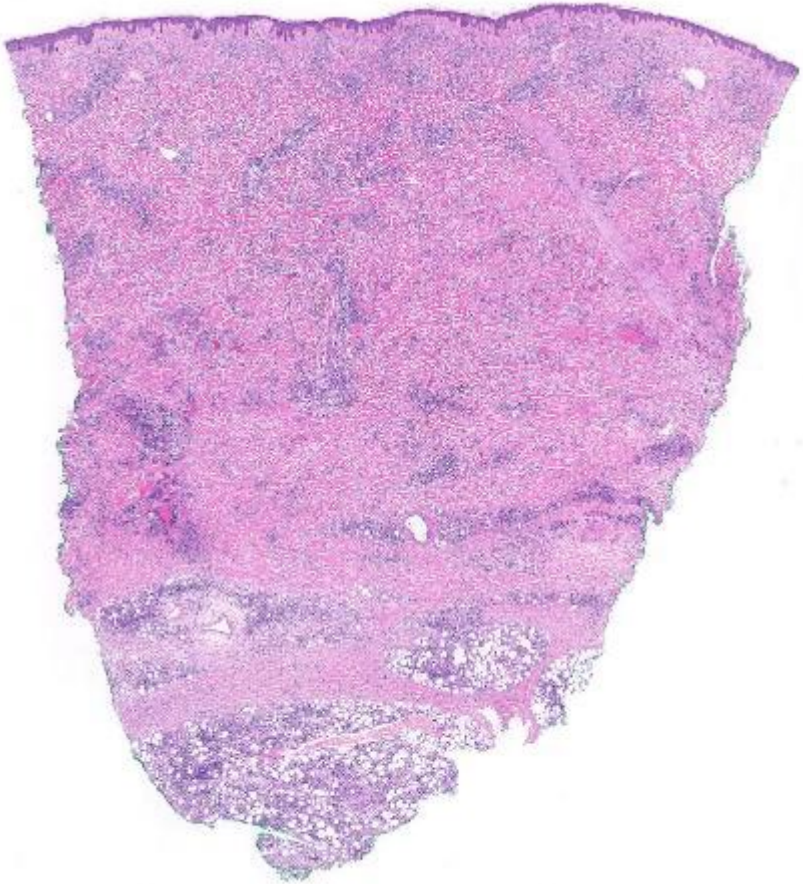
Recognizing the possibility of adnexal involvement in the interface dermatitides is important, as adnexal involvement is commonly seen in dermatopathological mimickers such as mycosis fungoides.

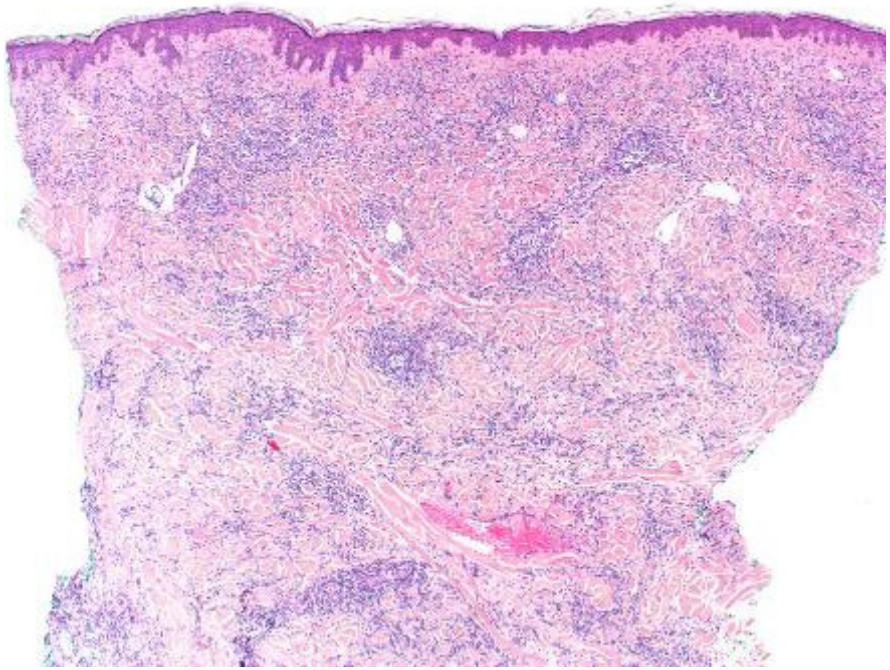
2. Interface Dermatitides with Distinct Clinicopathologic Adnexotropic Variants

2.1. Lichen Planus

Lichen planus is the prototype lichenoid dermatitis characterized by a self-limiting eruption of pruritic flat-topped violaceous papules. On histopathology, hyperkeratosis, hypergranulosis, irregular acanthosis, and subepidermal clefts known as Max Joseph spaces are observed. A band-like lymphohistiocytic infiltrate is also seen. Distinct adnexotropic variants of lichen planus have been described (Figure 1). These include follicular lichen planus, lichen planus follicularis tumidus, and syringotropic lichen planus.

73-year-old woman with indurated hyperpigmented plaque on the left breast.





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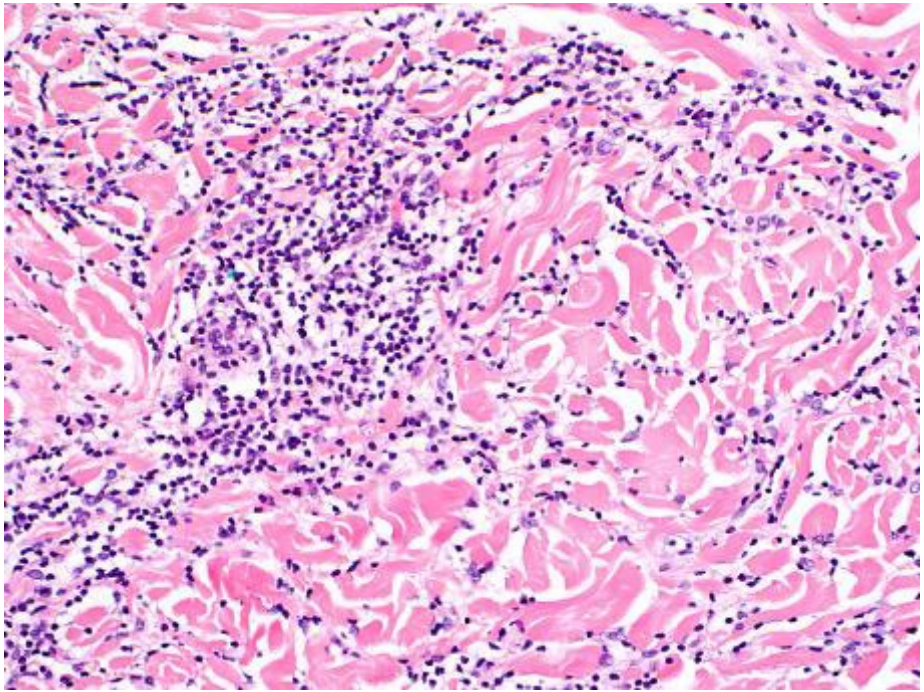
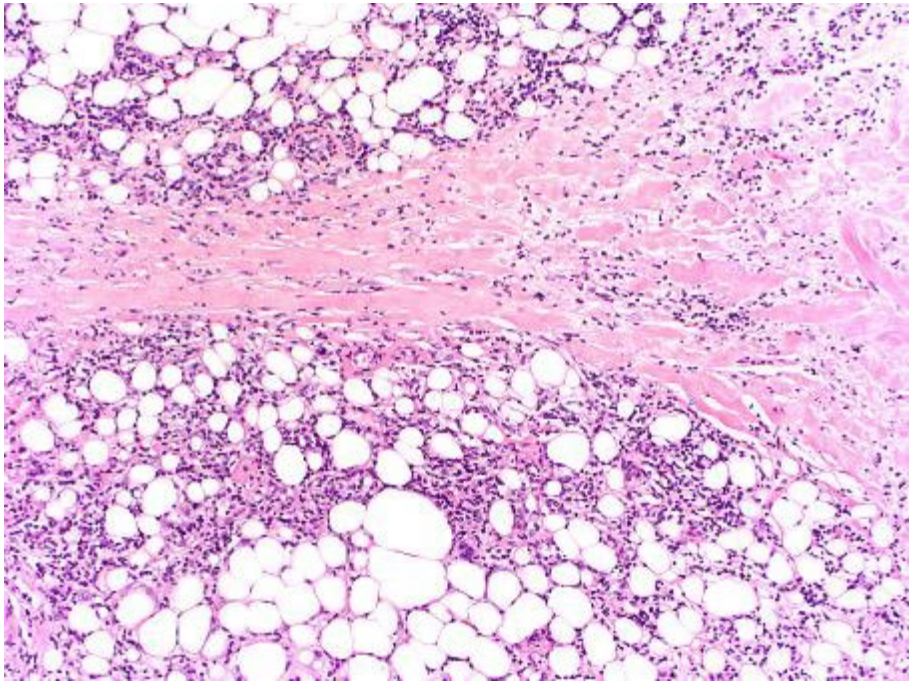
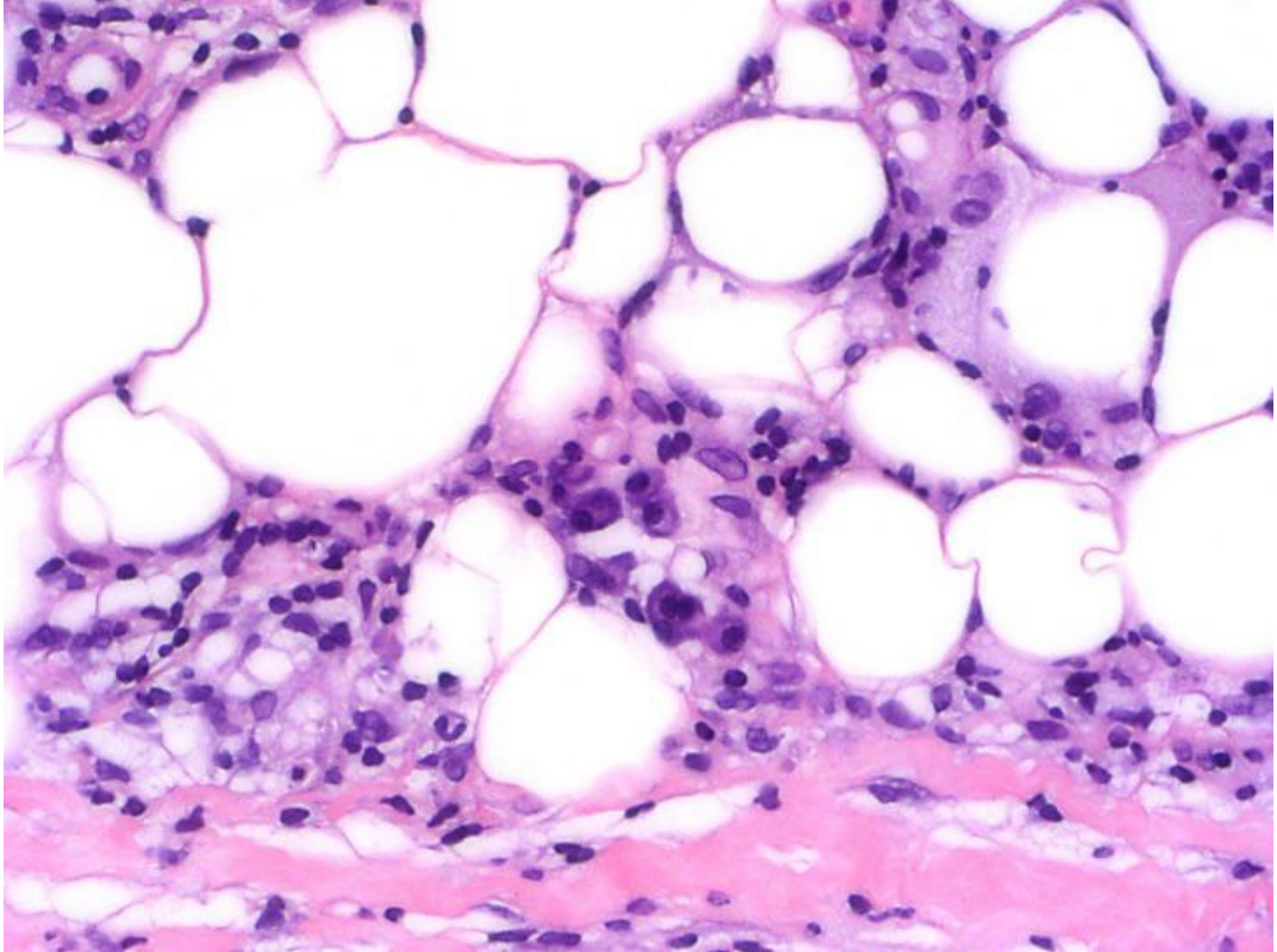
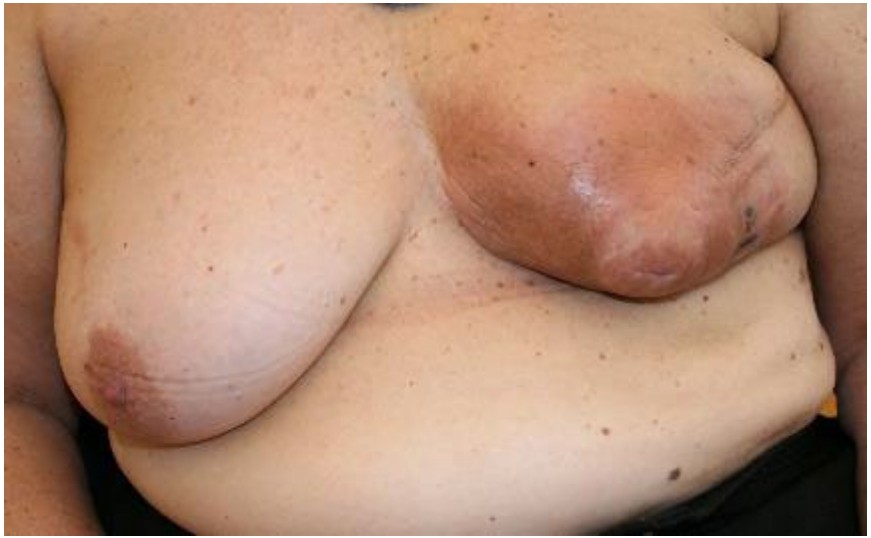
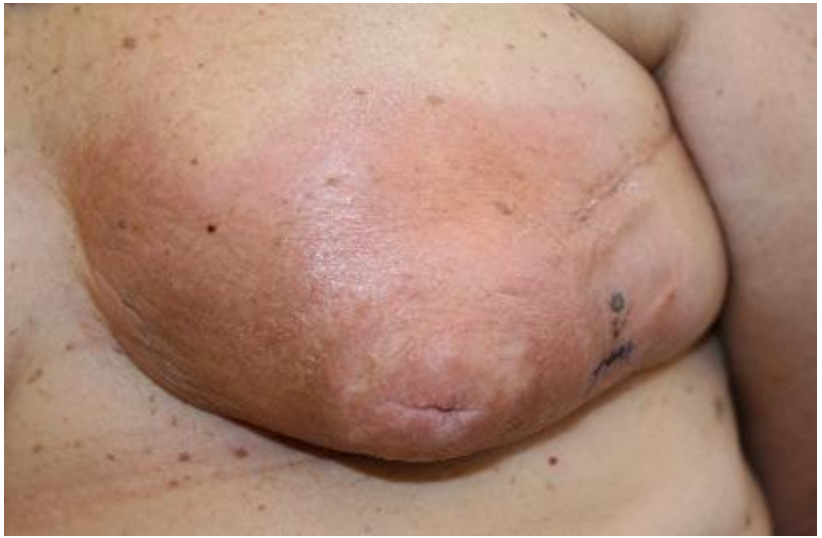




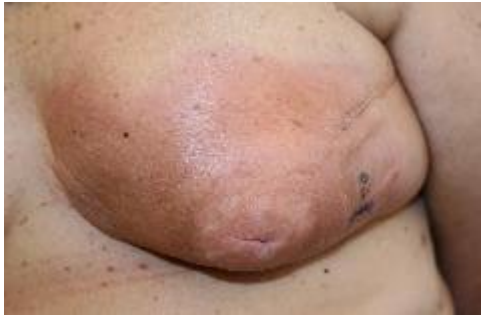


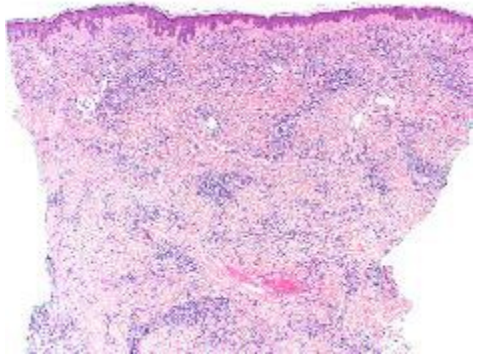
73-year-old woman with indurated hyperpigmented plaque on the left breast.
Radiotherapy for breast carcinoma(2006).





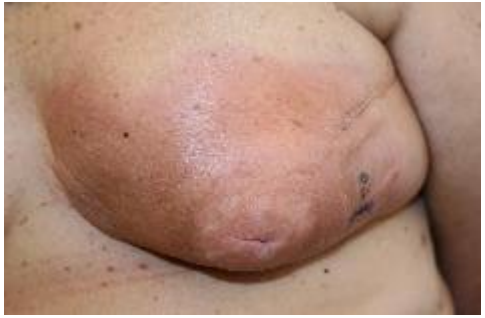
Differentialdiagnosis

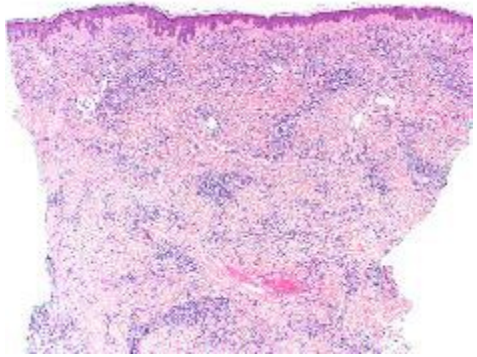




- Metastatic breast carcinoma (Carcinoma erysipeloides)
- Chronic radiation dermatitis
- Chemotherapy-induced sclerosis (bleomycin)
- Postradiation morphea
- Mastitis due to infectious process (erysipelas, fungal)
- Lichen sclerosus et atrophicus
- Lupus panniculitis

Differentialdiagnosis





- Metastatic breast carcinoma (Carcinoma erysipeloides)
- Chronic radiation dermatitis
- Chemotherapy-induced sclerosis (bleomycin)
- **Postradiation morphea**
- Mastitis due to infectious process (erysipelas, fungal)
- Lichen sclerosus et atrophicus
- Lupus panniculitis

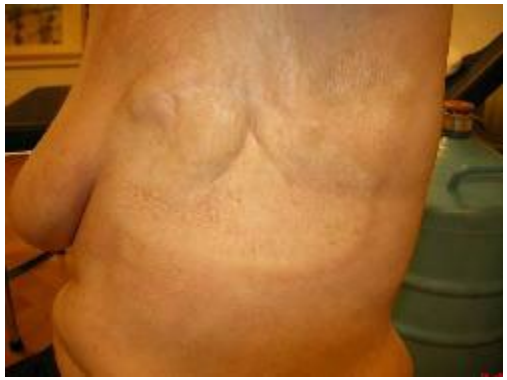
Postradiation morphea

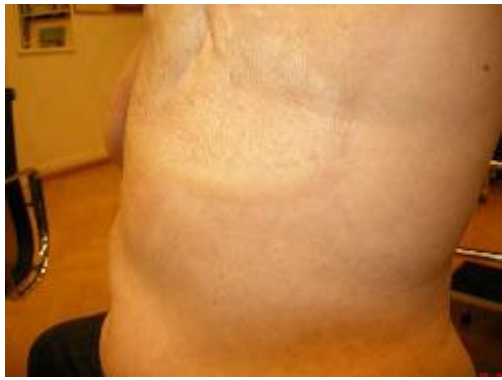
Approx. 40 patients reported in the literature

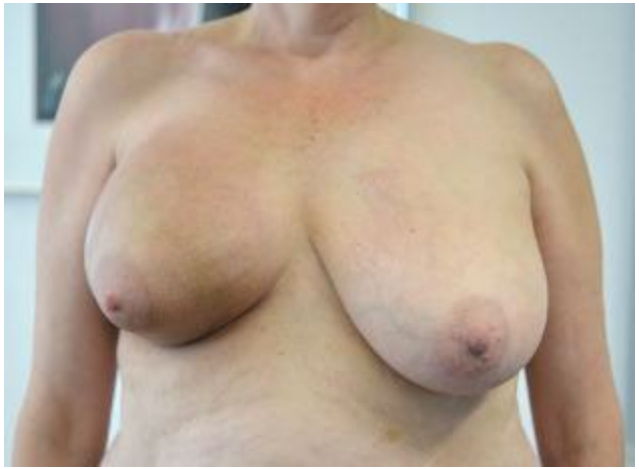
Estimated incidence = 1:500 patients undergoing RT

Indurated painful lesions involving the irradiated field, fewer than one-third of patients have plaques extending beyond the treatment area

Impredictable response to treatment





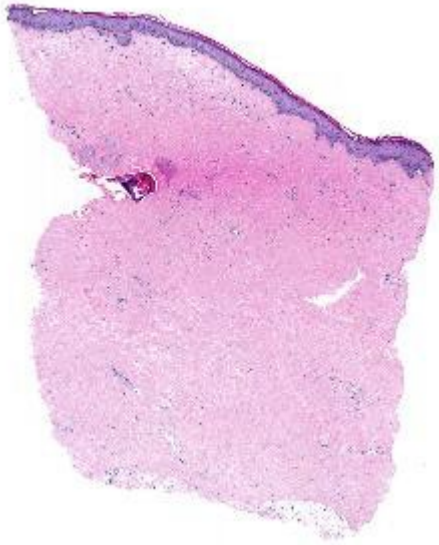


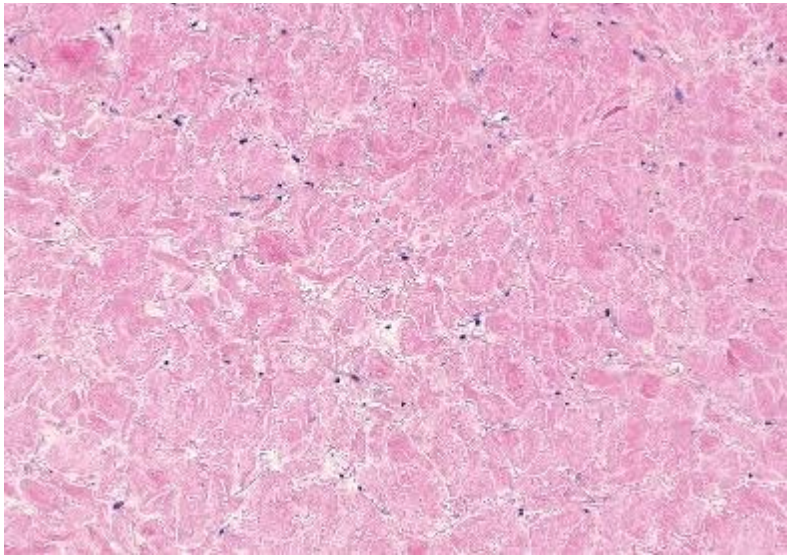
Morganroth et al. Am J Dermatopathol 2010
Peters et al. Am J Dermatopathol 2013

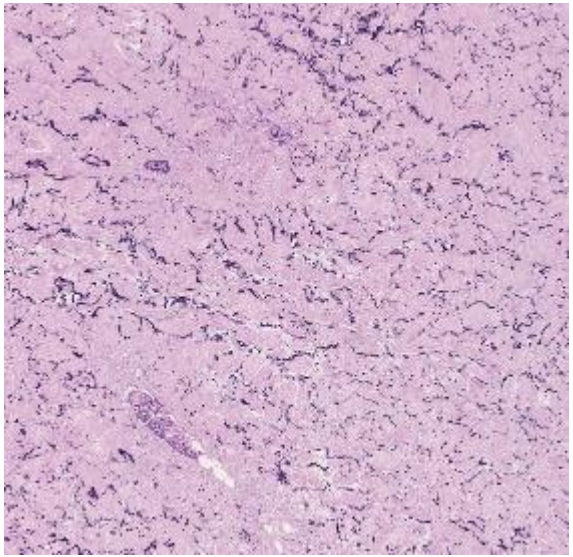
Postradiation morphea - Histology

Dermal sclerosis and/ or lichen sclerosus-like changes
Lymphocytic and plasmacellular infiltrate
Deep form with broadened subcutaneous septa and septal inflammation

Walsh et al. Hum Pathol 2008;39:1680–88
Peters et al. Am J Dermatopathol 2013;35:289–307







Elastica

Postradiation morphea

Late reaction (3y or more)

Abrupt onset in most cases with initial erythema and induration

Dermal sclerosis

Lichen sclerosus-like changes

Lymphocytic and plasmacellular infiltrate

Deep form with broadened subcutaneous septa, septal and lobular inflammation

Tx: Topical steroids as early as possible.

Radiation induced fibrosis

Early reaction within first 3 months

No initial erythema

Dermal sclerosis

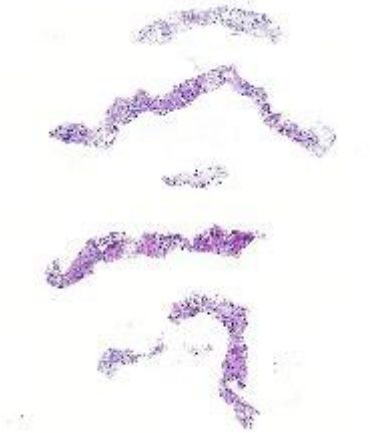
Lichen sclerosus-like changes

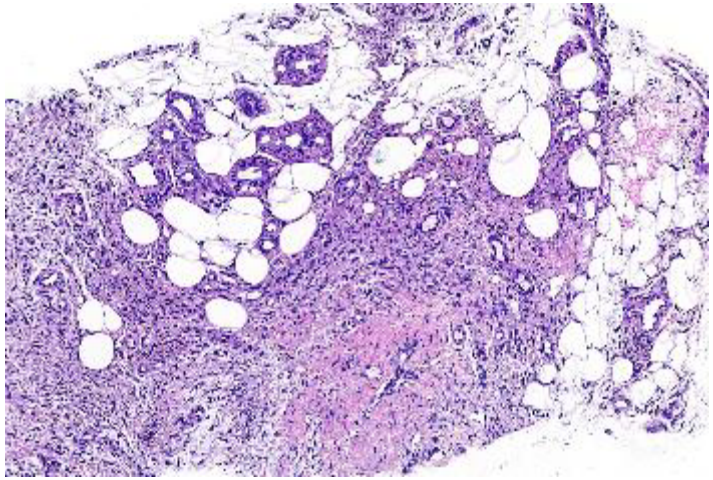
No dermal infiltrate

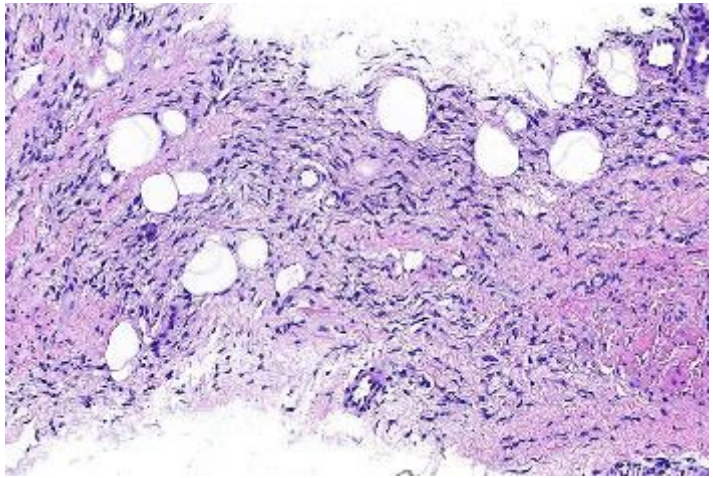
Post-RT sclerosing panniculitis: Lobular infiltrates

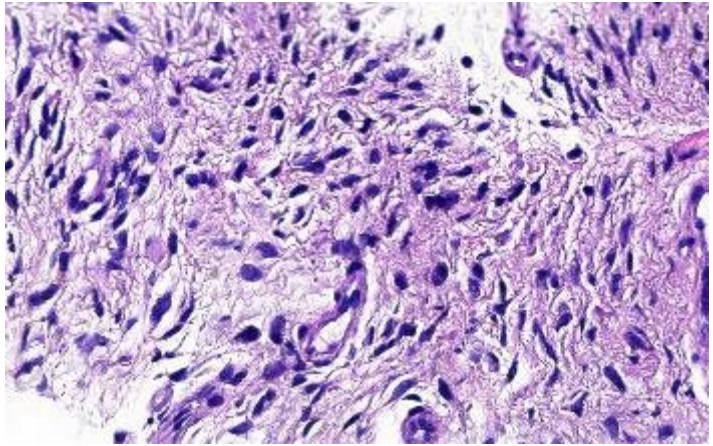
Liu Bolin

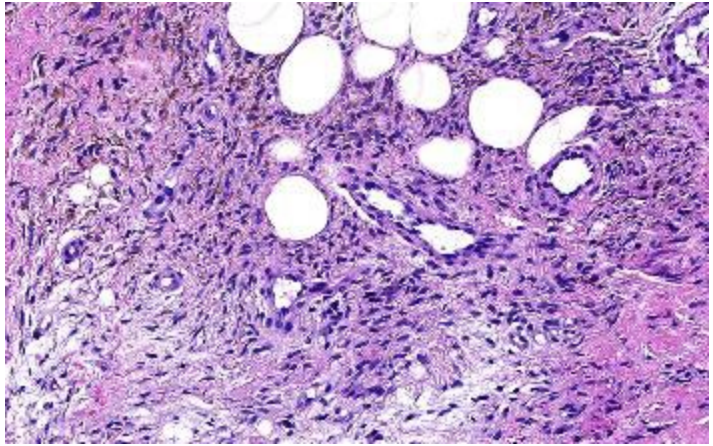
71-year-old man with swollen toe (Dig 2, right foot)











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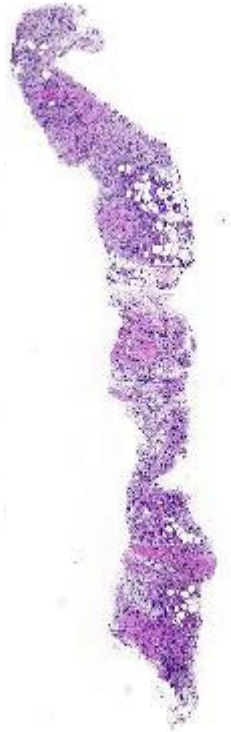
Negative: SMA CD68, S100, desmin, ERG



CD34

FXIIIa

Iron stain

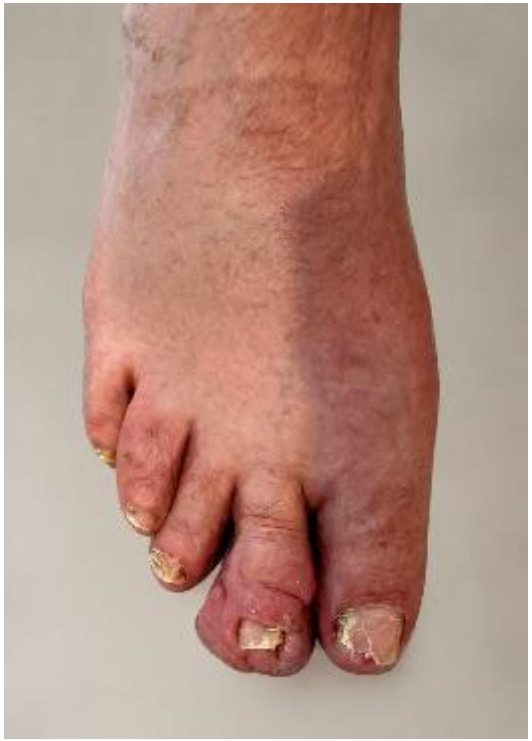
Differential diagnosis: acral CD34-positive spindle cell neoplasms

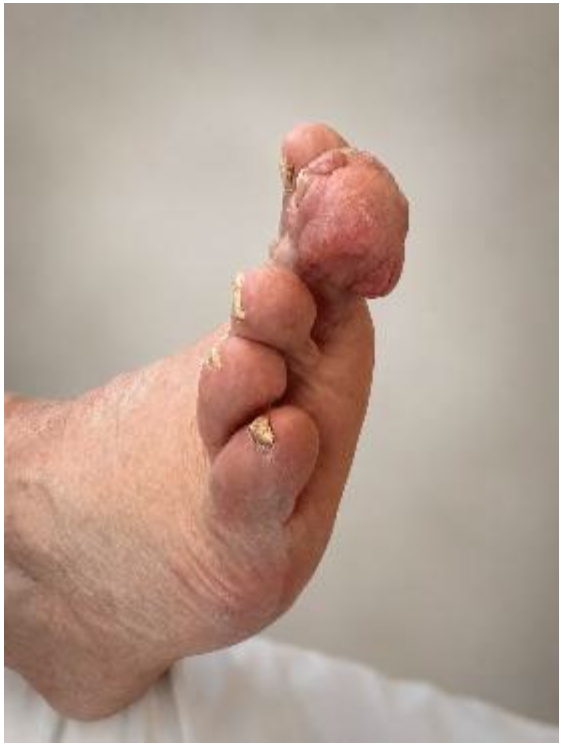
Entity	Clinical setting	Histology	Phenotype / molecular	Prognosis / behavior
HFLT	**Adult; F>M; foot / ankle / toe**	**Infiltrative fat-septal spindle cells; hemosiderin; fibrosis ± inflammation**	**CD34+; S100− in spindle cells; TGFBR3 / MGEA5 subset; overlaps PHAT–MIFS**	**Local recurrence risk; metastasis exceptional in classic lesions; beware hybrid / high-grade areas**
Superficial acral fibromyxoma	Middle-aged; M>F; peri-/subungual finger or toe	Bland spindle / stellate cells in myxocollagenous stroma; variable mast cells	CD34+, CD99+; EMA variable; S100, keratin, SMA usually−	Benign; recurrence mainly after incomplete excision
Cellular digital fibroma	Adults; slight M; small papule on digit / toe	Bland cellular dermal spindle-cell proliferation; collagenous; minimal myxoid change	Diffuse CD34+; EMA−, S100−, SMA−	Benign; no convincing metastatic potential; recurrence not typical
DFSP, acral variant	Adults; rare on hand / foot / digit	Monomorphic storiform spindle cells; honeycomb subcutis infiltration	Diffuse CD34+; factor XIIIa−; COL1A1::PDGFB	Locally aggressive; metastasis rare, higher with fibrosarcomatous transformation
MIFS	Adults; distal extremities incl. hands / feet	Myxoid + hyaline zones; mixed inflammation; virocyte / Reed–Sternberg-like cells	CD34 variable; TGFBR3 / MGEA5 or related rearrangements in subset	Frequent local recurrence; rare metastasis; high-grade variants behave worse
PHAT	Adults; slight F; lower limb, ankle / foot	Ectatic hyalinized vessels; pleomorphic spindle cells; hemosiderin	CD34+; S100−, keratin−; TGFBR3 / MGEA5 subset	Intermediate / locally recurrent; metastasis very rare or absent in classic PHAT
Superficial CD34+ fibroblastic tumor	Middle-aged adults; superficial extremity; acral rare	Pleomorphic spindle / epithelioid cells with glassy cytoplasm; low mitotic rate	Diffuse CD34+; keratin variable/focal; S100 / SOX10−	Borderline / low-grade; rare nodal metastasis; local recurrence uncommon
Spindle cell / pleomorphic lipoma	Older men; acral uncommon	Mature fat + bland spindle cells; ropey collagen ± floret cells	CD34+; Rb loss / 13q; MDM2 & CDK4−	Benign; excision usually curative
Distal epithelioid sarcoma	Young adults; M>F; finger / hand / foot / toe	Nodular epithelioid + spindle cells; granuloma-like necrosis possible	Keratin+, EMA+; CD34 ~50%; INI1 / SMARCB1 loss	Aggressive sarcoma; local recurrence, nodal and distant metastasis

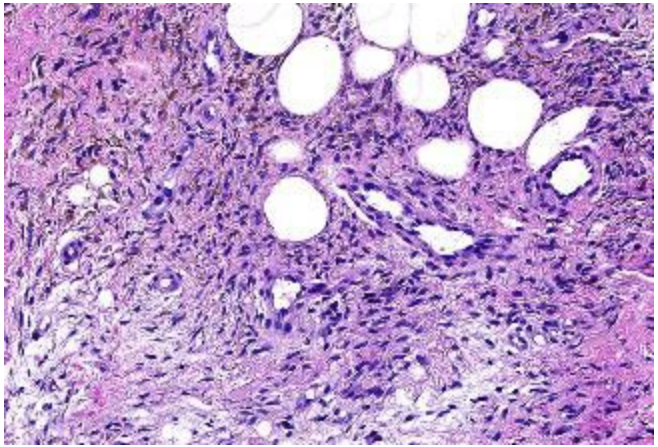
DFSP dermatofibrosarcoma protuberans; MIFS myxoinflammatory fibroblastic sarcoma; PHAT pleomorphic hyalinizing angiectatic tumor.

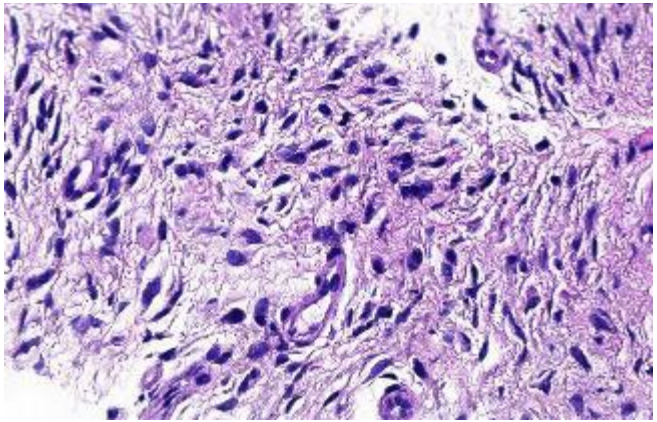
Selected refs: Antonescu 2011; Cohen 2020; Hao 2020; Crepaldi 2019; Zafar 2023; Regalbuto 2020.

71-year-old man with swollen toe (Dig II, right foot)









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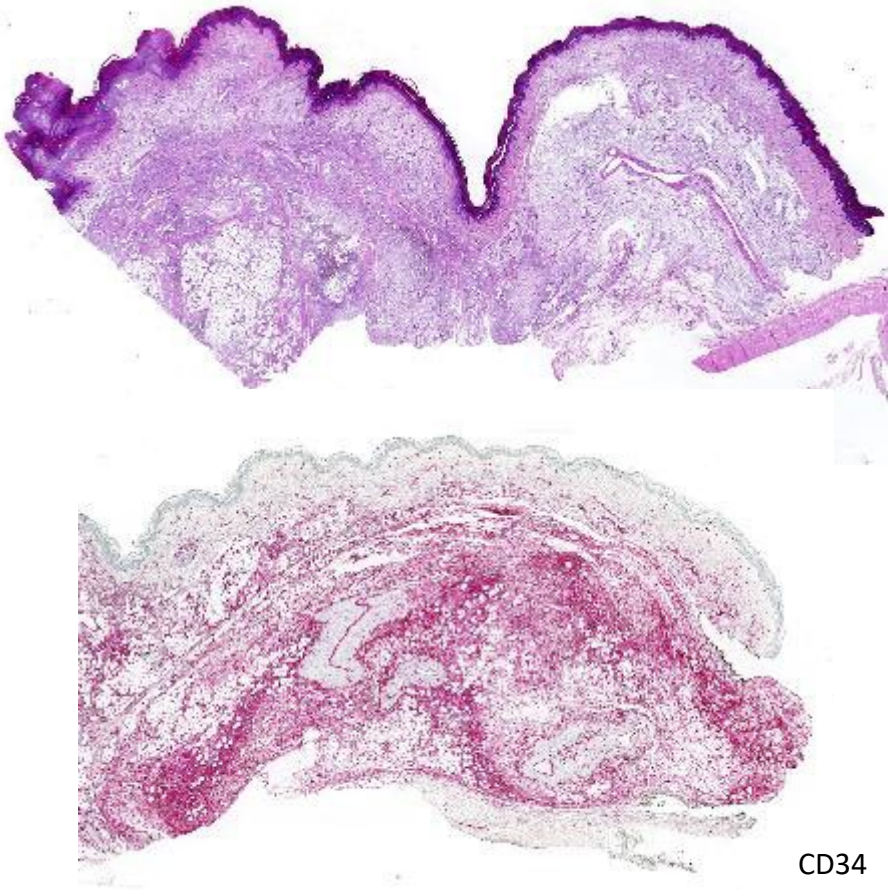
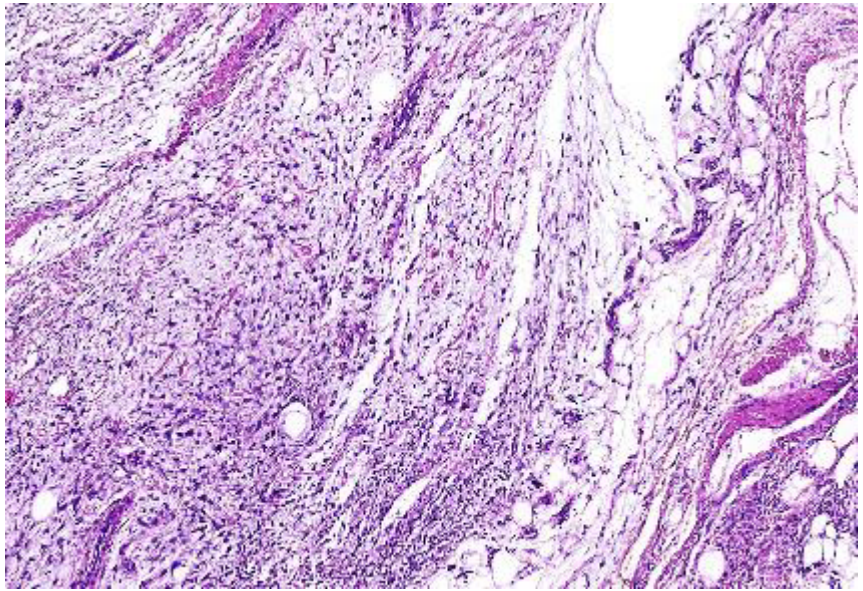


CD34



By courtesy: Dr. M. Kurrer, Pathologikum, Zürich; Consultation: Prof. B. Bode, Lucerne

Hemosiderotic fibrolipomatous tumor (HFLT)

Rare, locally aggressive neoplasm

Marshall-Taylor C et al. Mod Pathol 2000

Middle-aged adults , f > m

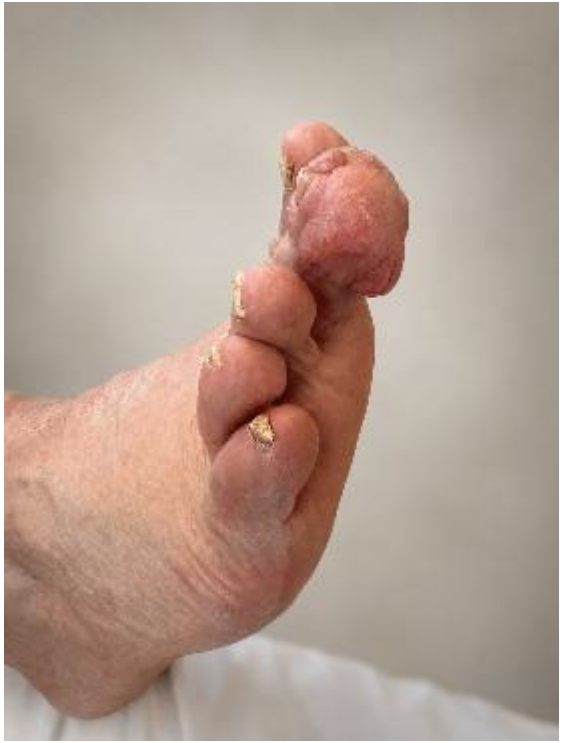
Distal extremities (foot and ankle)

Slow-growing, painless, ill-defined subcutaneous mass, often present for months to years

Usually superficial, rarely deep (bone)

Local recurrence (30-50%)
No metastasis

Progression to MIFS/high-grade sarcoma



Browne T and Fletcher C Histopathology 2006
Solomon D et al. Am J Surg Pathol 2013
Liegl-Atzwanger, B. Encyclopedia of Pathology. 2020

Hemosiderotic fibrolipomatous tumor - Histology

Unencapsulated, infiltrative lesion

Composite pattern:
Mature adipose tissue
spindle-cell proliferation
Irregular fascicles of bland fibroblastic
and myofibroblastic spindle cells.

No marked cytologic atypia

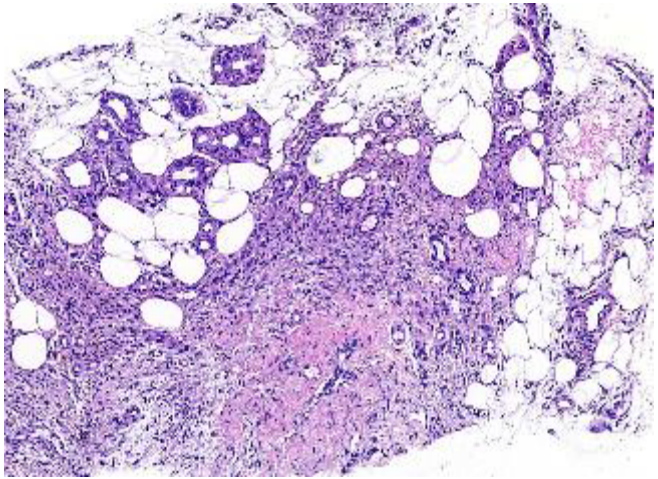
Multinucleated giant cells may be present

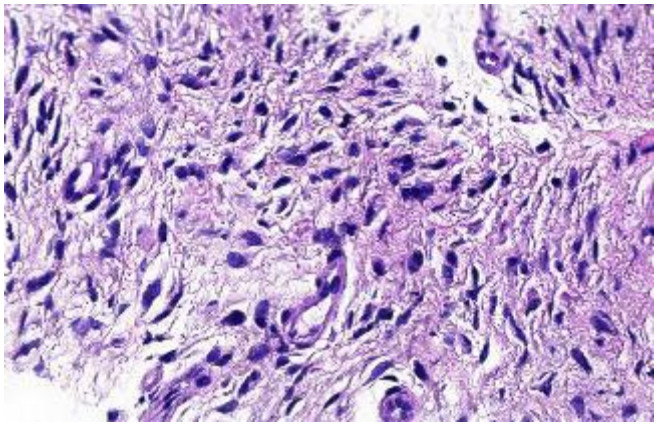
Abundant hemosiderin deposition

Inflammatory infiltrate

CD34 + (80%)
Negative: SMA CD68, S100, desmin.

Browne T and Fletcher C Histopathology 2006





CD34 Iron stain

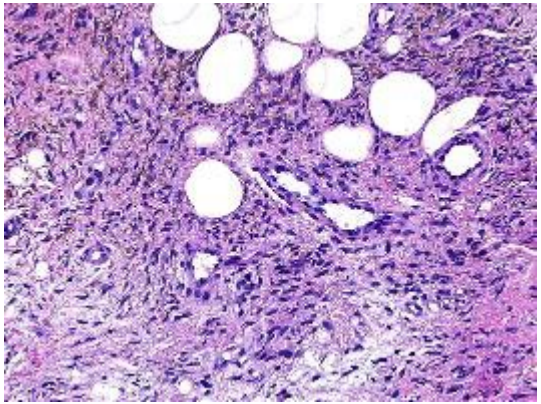
HFLT - Genetics

Recurrent t(1;10)(p22;q24) rearrangements involving TGFBR3 and MGEA5

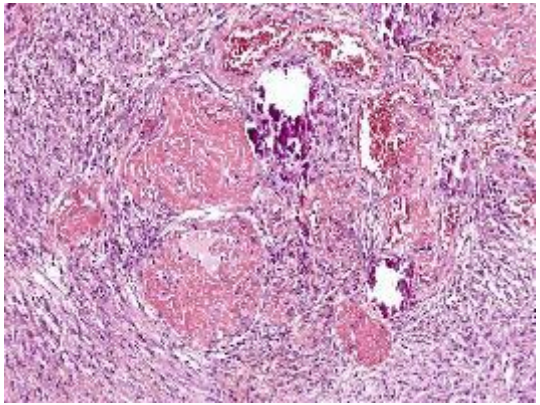
Morphologic and genetic overlap with pleomorphic hyalinizing angiectatic tumor (PHAT) and myxoinflammatory fibroblastic sarcoma (MIFS = progression of HFLT?)

Boland J and Folpe A Adv Anat Pathol 2027

HFLT

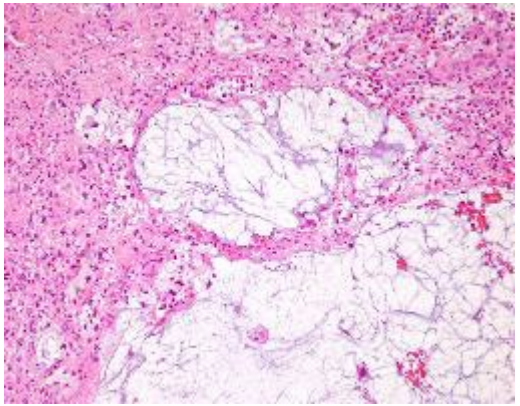


PHAT



From: Dr. Perret, Pathology Outlines

MIFS

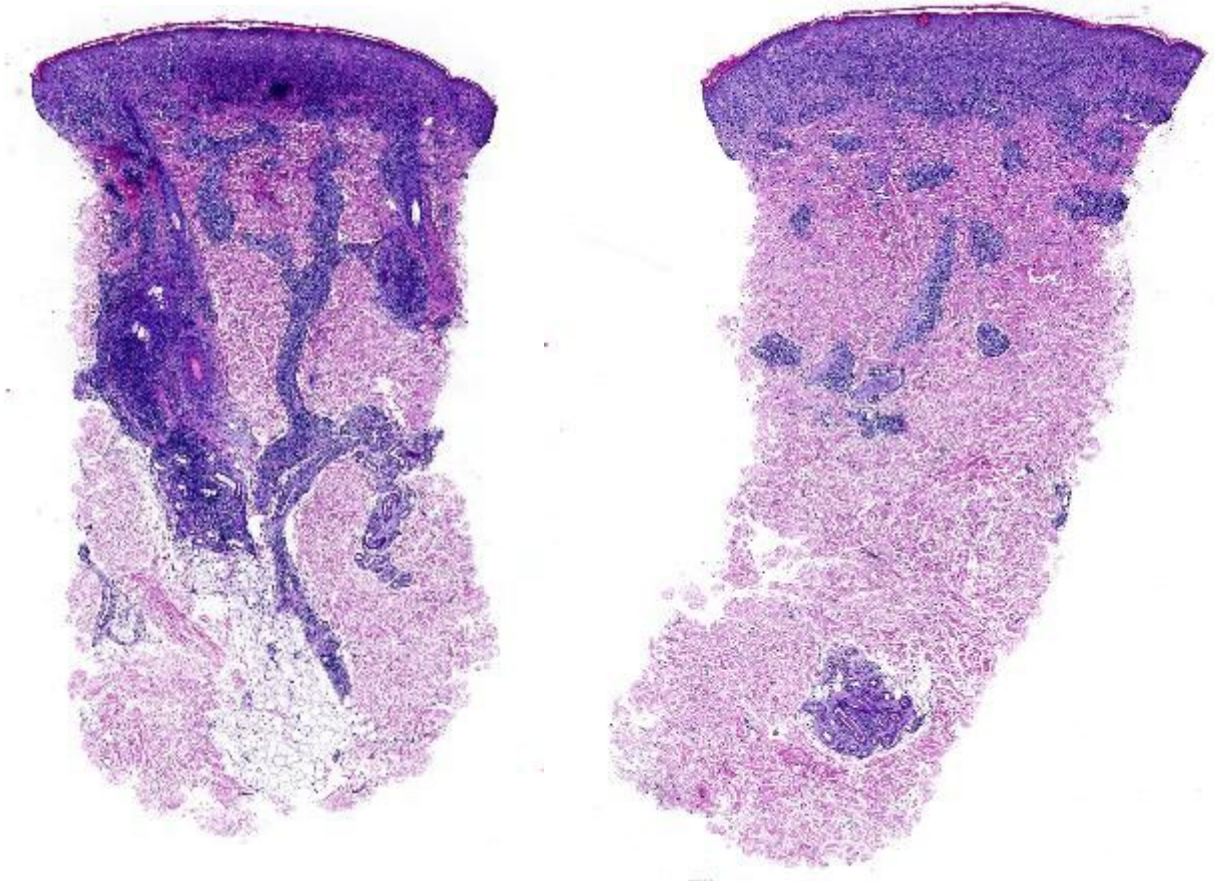


From: Dr. Suster, Pathology Outlines

Alchisme grossa

Bild: Javier Aznar Gonzalez de Rueda

29-year-old man with papular lesions on the trunk



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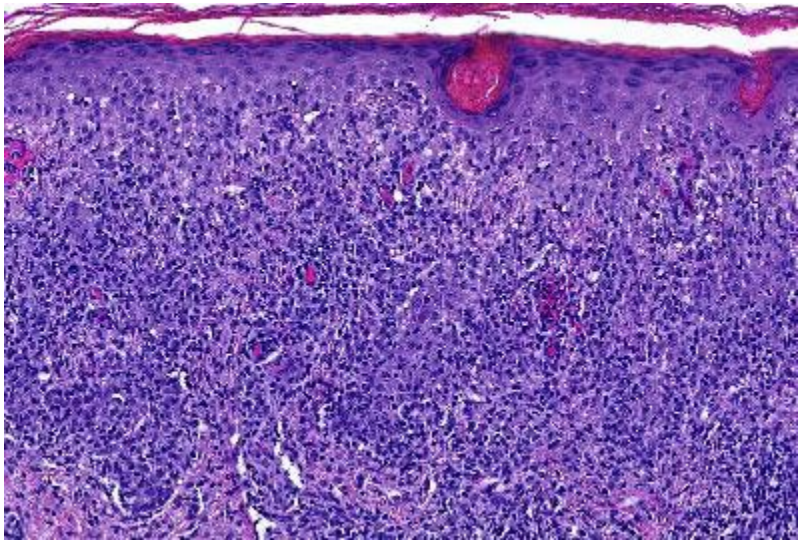

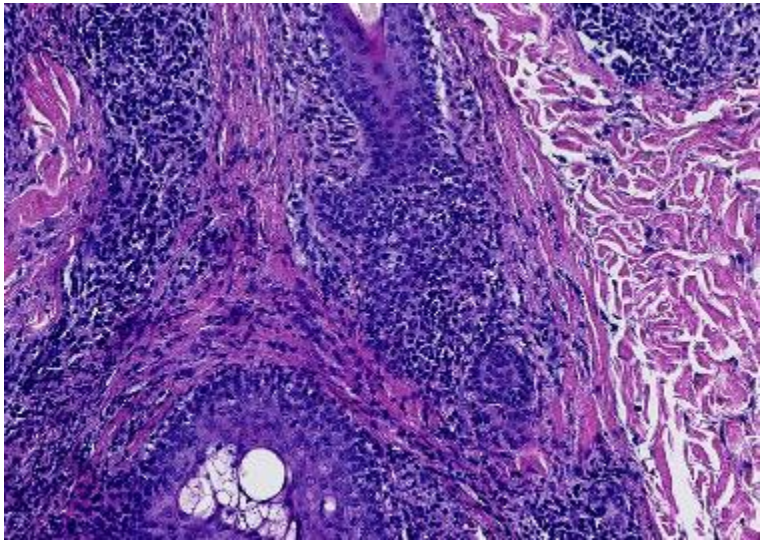

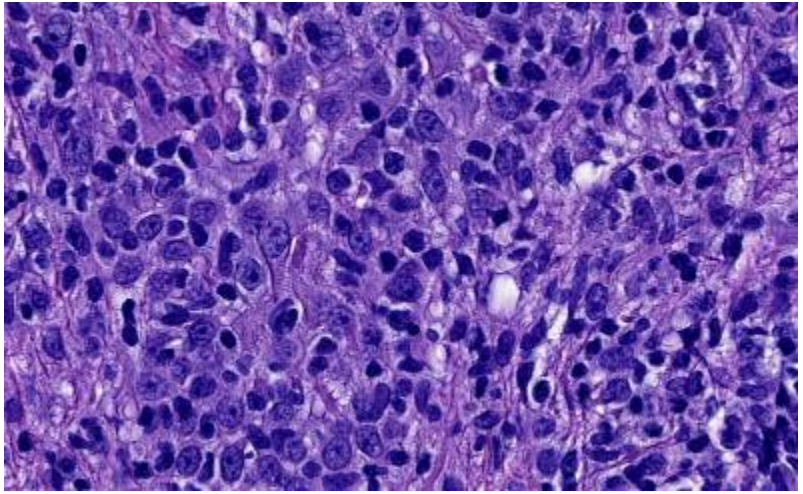

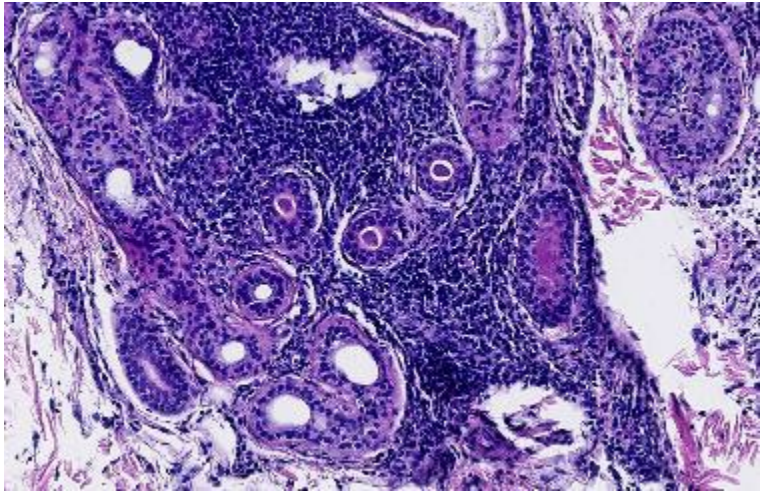

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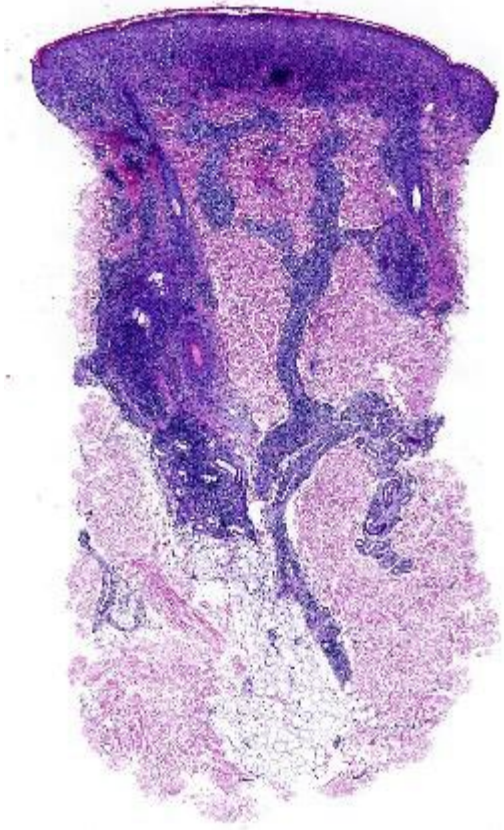
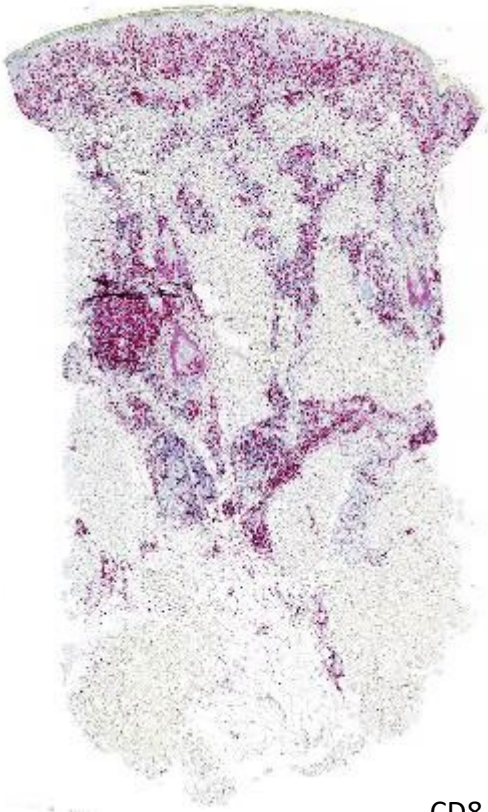
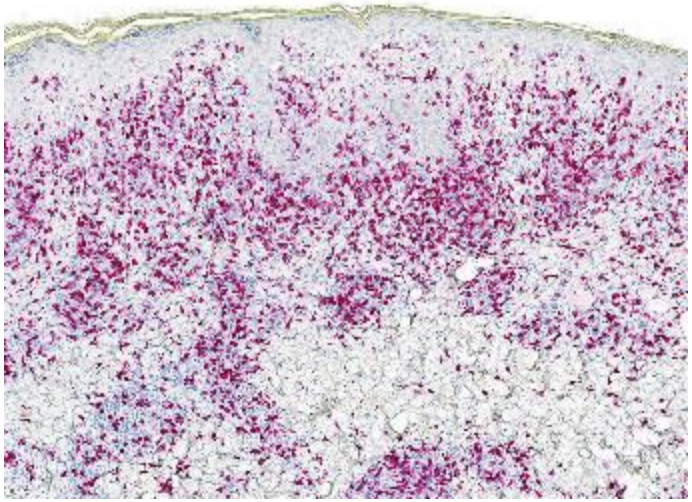
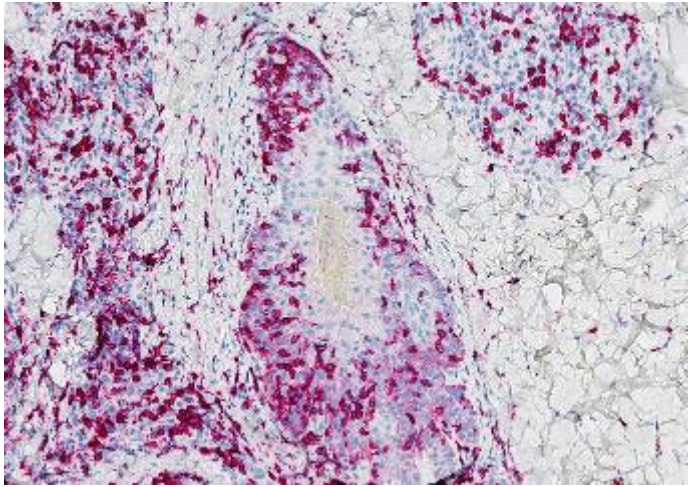




CD8





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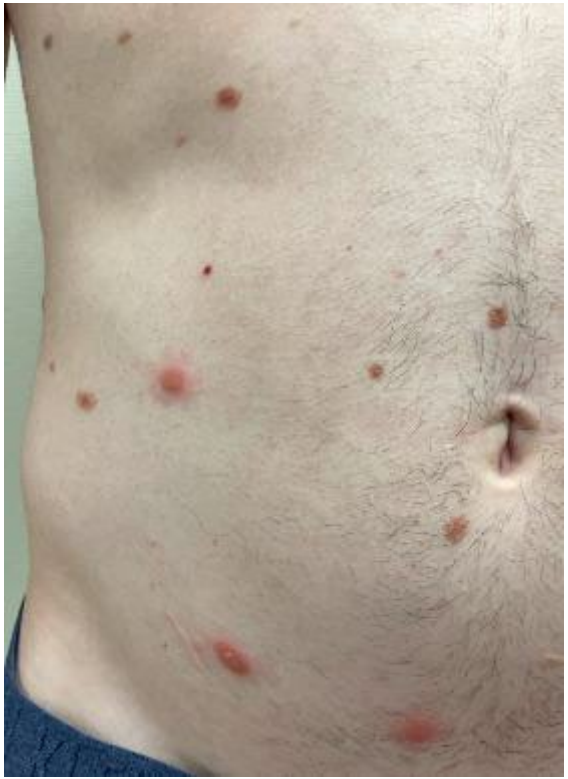
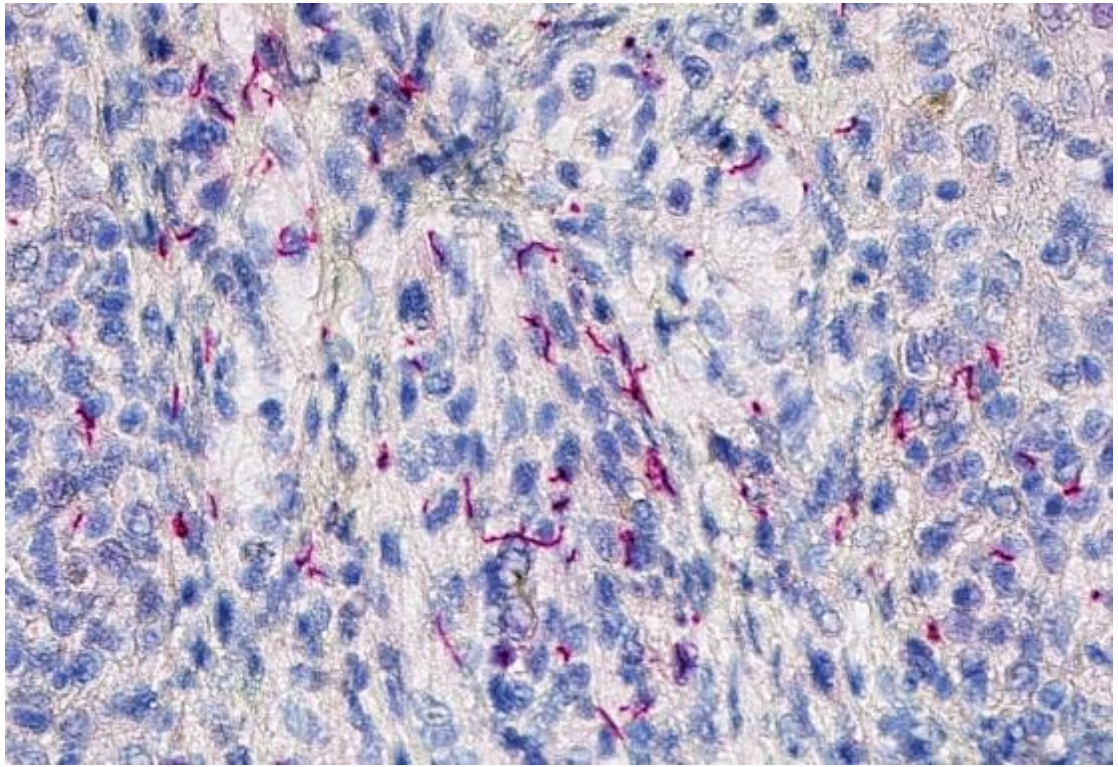
29-year-old man with papular lesions on the trunk





Trep. pall. IHC

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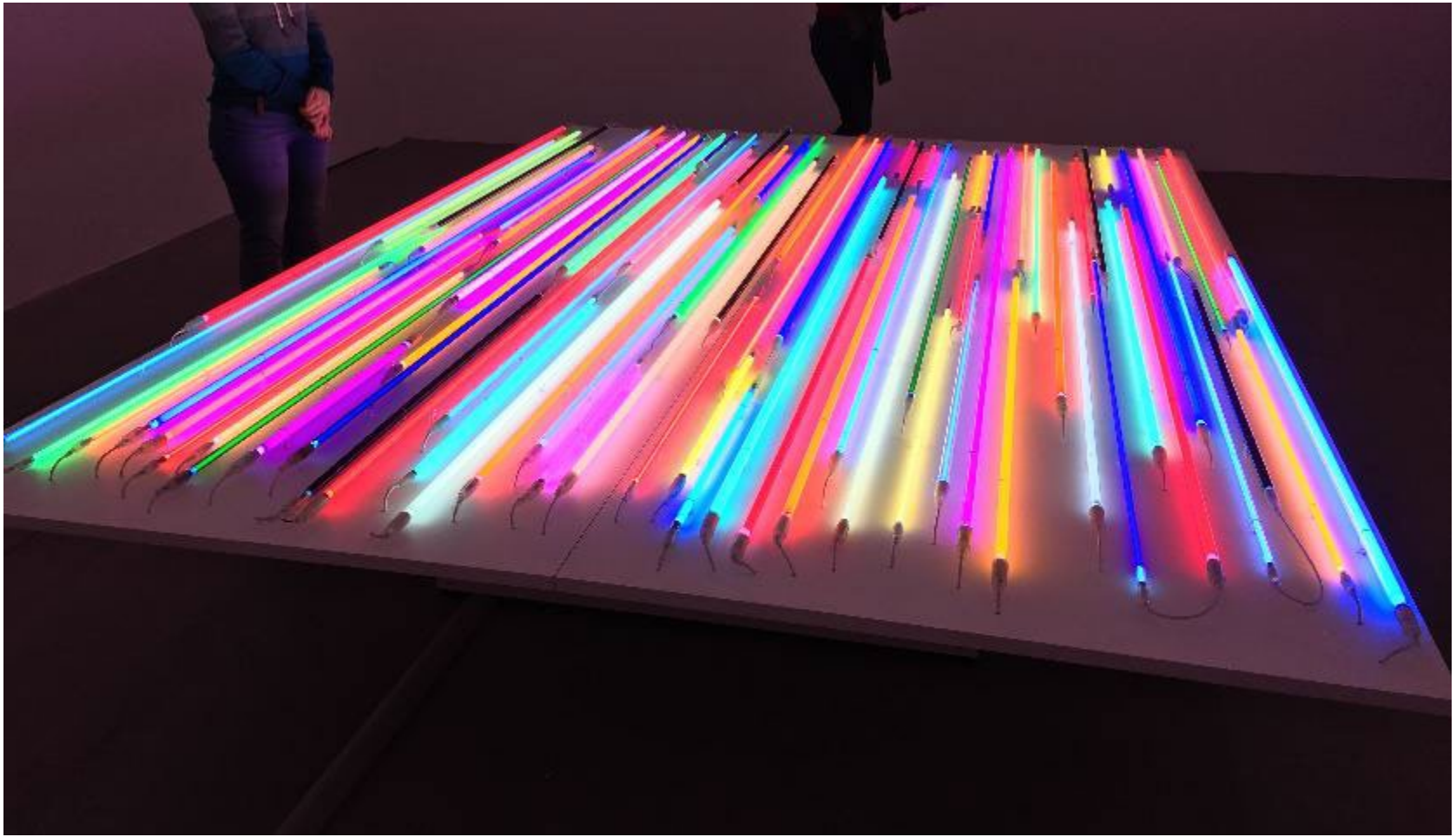

Christian Herdeg