AEDV 2023 Highlights

Con el patrocinio de:

ucb

BERLÍN

11-14 OCTUBRE

Iniciativa científica de:

AEDV 1909

ACADEMIA ESPAÑOLA DE DERMATOLOGÍA Y VENEREOLOGÍA

AEDV2023 Highlights

Novedades en

DERMATOPATOLOGÍA

José Mª Camino Salvador

Hospital Universitario de Guadalajara

Jose.caminosalvador@Gmail.com

josecamino96

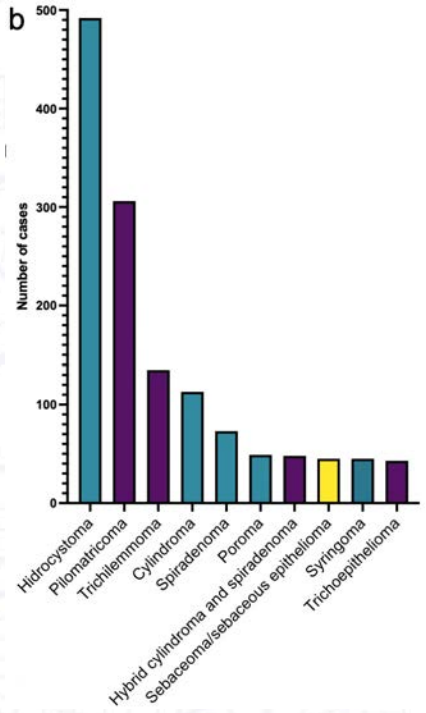
NO TENGO CONFLICTOS DE INTERÉS

BERLIN

11-14 OCTUBRE

TUMORES ANEXIALES

N = 1615 casos (1,5%).



Br J Dermatol. 2022 Jan;186(1):167-173. doi: 10.1111/bjd.20701. Epub 2021 Oct 21.

A 5-year retrospective review of skin adnexal tumours received at a tertiary dermatopathology service: implications for linked genetic diagnoses

S Cook [1], D Bajwa [2], L Hollestein [3 4], A Husain [1], N Rajan [2 5]

- 97% of cases benign, median age 55 yrs, M:F – 0.77:1
- 10% of cases paediatric (all benign)
- 80% of cases H&N, 35% periocular
- 90% of paediatric cases pilomatricoma
- most common adult tumor hydrocystoma

Commentary | Free to Read

Skin adnexal tumours in a tertiary dermatopathology service

Z. Marušić, E. Calonje

First published: 15 November 2021 | https://doi.org/10.1111/bjd.20816

- prebiopsy diagnosis correct in only 28%

Review > Histopathology. 2022 Jan;80(1):166-183. doi: 10.1111/his.14441.

Molecular pathology of skin adnexal tumours

Jaclyn M Plotzke [1], David J Adams [2], Paul W Harms [1 3 4]

Table 1. Syndromic associations with cutaneous adnexal tumors

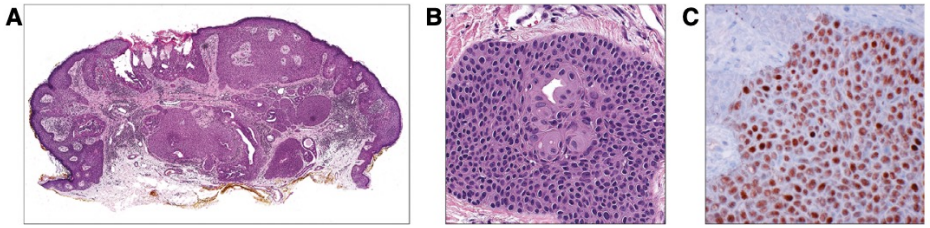
Syndrome	Gene function	Cutaneous tumors	Extracutaneous neoplasms	Other findings	Estimated prevalence
Bazex–Dupré–Christol (*ACTRT1*)	Ciliary function	BCC, less frequently trichoepitheliomas	N/A	Follicular atrophoderma, hypotrichosis, hypohidrosis, milia, facial hyperpigmentation, hair shaft anomalies	<1/1 000 000
Birt-Hogg-Dube (*FLCN*)	Inhibition of mTOR pathway	Fibrofolliculoma/ trichodiscoma, acrochordon	Pulmonary cysts, renal tumors (most commonly oncocytoma or renal cell carcinoma)	Spontaneous pneumothorax	Unknown
CYLD cutaneous syndrome/Brooke-Spiegler (*CYLD*)	Deubiquitinase (NF-κB inhibition)	Trichoepithelioma, spiradenoma, cylindroma, spiradenocylindroma with rare malignant transformation	Membranous basal cell adenoma (salivary gland)	N/A	<1/100 000
Clouston (*GJB6, GJB2*)	Connexins	Syringofibroadenoma	N/A	Palmoplantar keratoderma, hypotrichosis, nail dystrophy	Unknown
Cowden (PTEN)	Inhibition of PI3K signaling	Trichilemmoma, multiple hamartomatous lesions	High risk for breast, thyroid, and endometrial carcinoma	Acral keratoses, oral papillomas	1/200 000
Familial Pilomatricoma (*PLCD1*)	Phospholipase C (Protein kinase C, MAPK)	Multiple pilomatricomas	N/A	N/A	Unknown
FAP (*APC*)	Inhibition of Wnt/ β-catenin signaling	Multiple pilomatricomas, epidermoid cysts, cutaneous fibromas, lipoomas	Osteomas, colorectal adenomas, desmoid tumors, adrenal adenomas, nasopharyngeal angiofibroma Increased risk for colon, thyroid, hepatobiliary, and CNS malignancies	N/A	(~1/8000)
Muir-Torre (MMR genes: *MLH1, MSH2,* and *MSH6*)	DNA mismatch repair	Sebaceous adenoma, sebaceoma, sebaceous carcinoma, keratoacanthoma	Colonic adenocarcinoma (most common), genitourinary, breast, and hematologic malignancies	N/A	1/300
Malta syndrome/ Nicolau-Balus (*MYH9*, possible)	Myosin heavy chain	Syringoma, microcystic adnexal carcinoma-like lesions	N/A	Atrophoderma vermiculata, milia	Unknown
NBCCS (*PTCH1*)	Inhibition of Hedgehog signaling	Numerous BCCs; basaloid follicular hamartomas (infrequent)	Odontogenic keratocysts, CNS tumors, ovarian cysts	Palmoplantar pits, skeletal anomalies, coarse facial features, hypertelorism, macrocephaly	~1/31000
Schimmelpenning–Feuerstein–Mims (postzygotic *HRAS/KRAS*)	RAS-MAPK signaling	Nevus sebaceus	CNS, ocular, and skeletal anomalies	N/A	Unknown
Schöpf-Schulz-Passarge (*WNT10A*)	Wnt/β-catenin signaling	Syringofibroadenoma, eyelid apocrine hidrocystoma	N/A	Palmoplantar keratoderma, telangiectasia, dental anomalies, onychodystrophy, hypotrichosis	<1/1 000 000
Steatocystoma multiplex (*KRT17*)	Keratin	Steatocystomas, eruptive vellus hair cysts	N/A	Pachyonychia congenita	Unknown

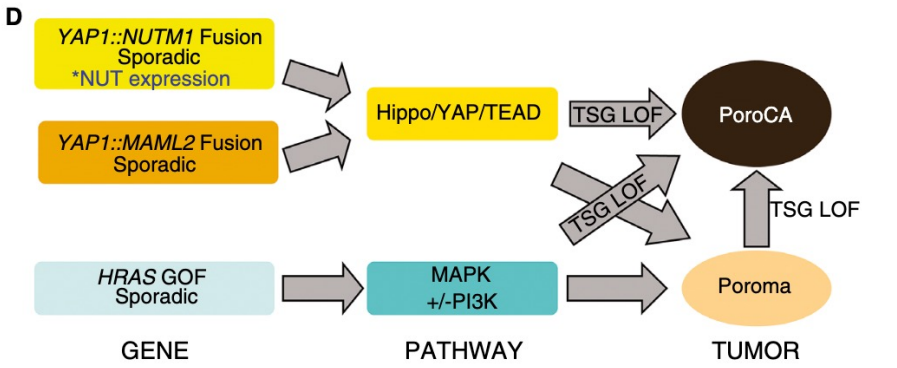
BCC, basal cell carcinoma; FAP, familial adenomatous polyposis; MMR, mismatch repair; NBCCS, nevoid basal cell carcinoma syndrome.

Review > Ann Dermatol Venereol. 2023 Sep;150(3):202-207.
doi: 10.1016/j.annder.2023.03.003. Epub 2023 Jun 1.

Genetics of adnexal tumors: An update

T Kervarrec [1], P Sohier [2], D Pissaloux [3], A de la Fouchardiere [3], B Cribier [4], M Battistella [5], N Macagno [6]





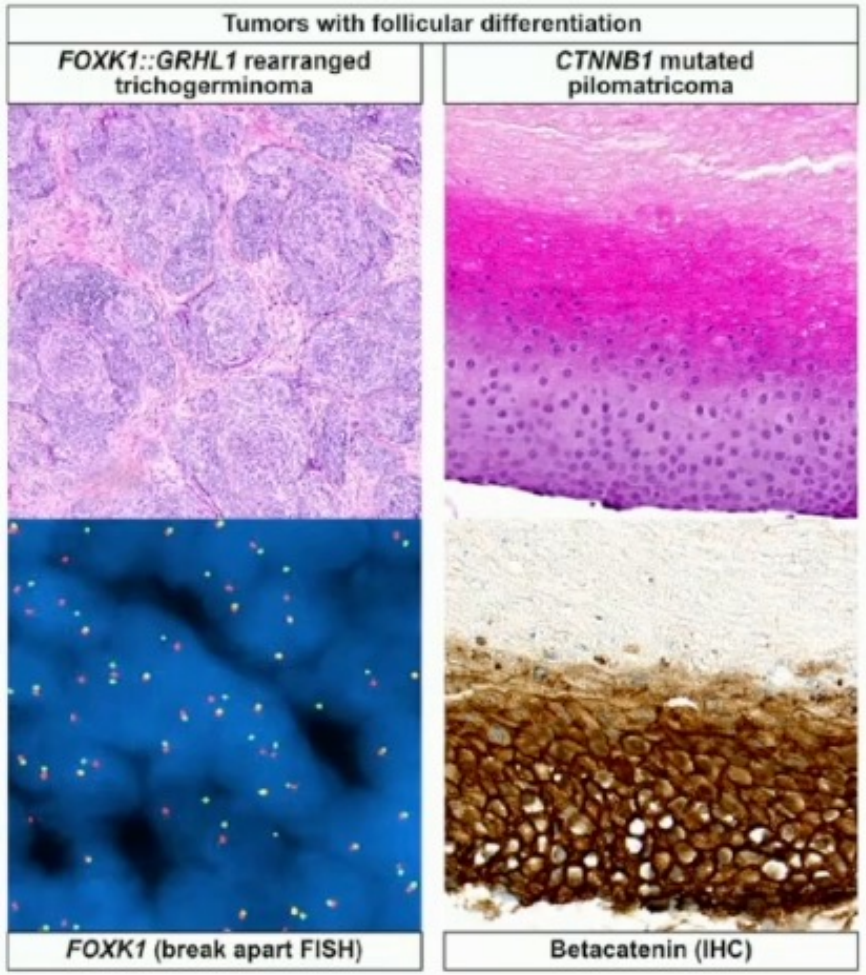
Review > Histopathology. 2022 Jan;80(1):166-183. doi: 10.1111/his.14441.

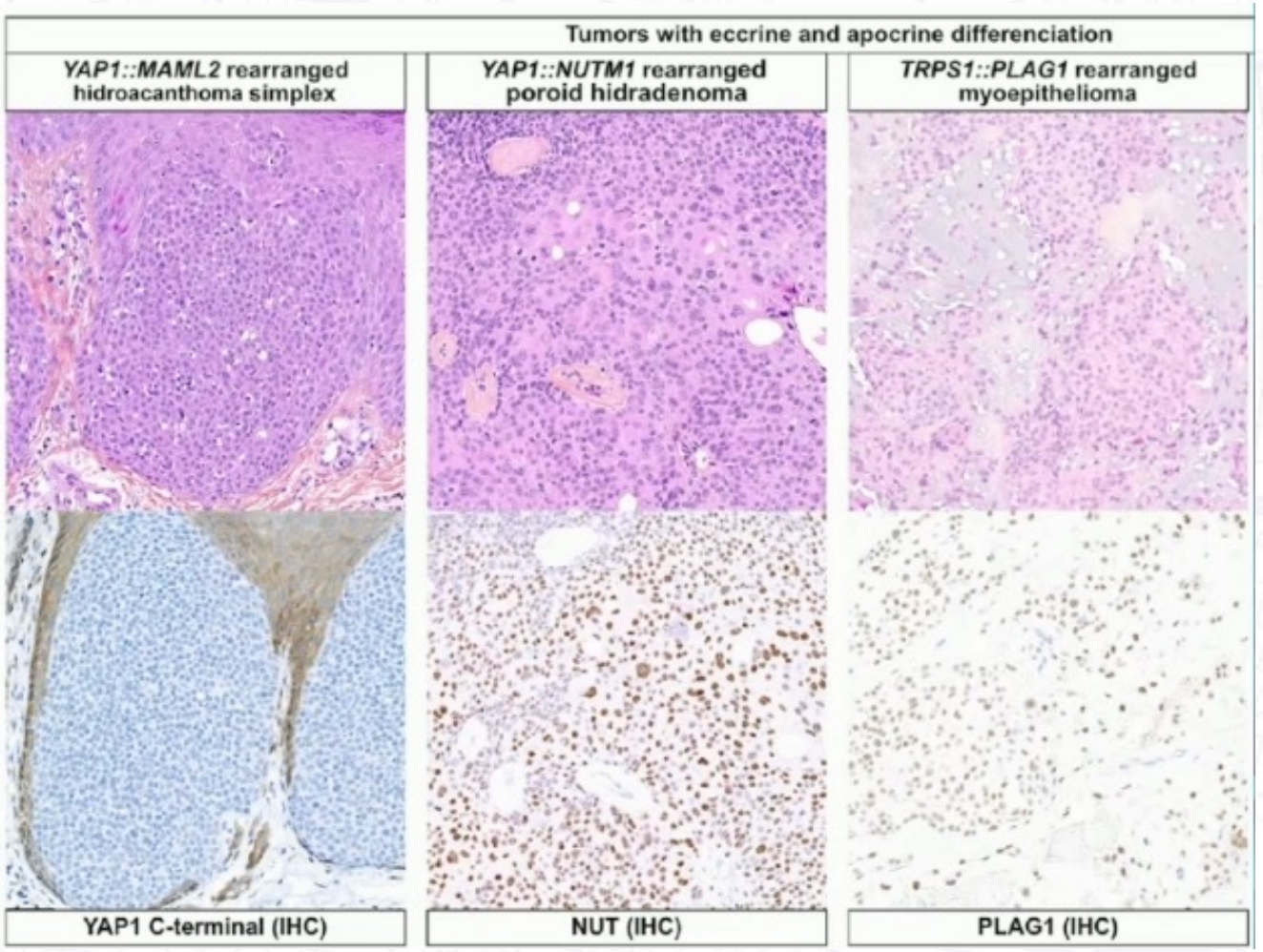
Molecular pathology of skin adnexal tumours

Jaclyn M Plotzke [1], David J Adams [2], Paul W Harms [1 3 4]

Table 1
Main recurrent genetic alterations in adnexal tumors. Int: virus integration, MMRd: MMR deficiency, mut: mutation, fus: fusion gene.

Follicular tumors	Genetic alteration	IHC marker
Trichilemmoma	*PTEN (mut)* (Cowden) *HRAS (mut)* (sebaceous nevus)	PTEN -
Trichoblastoma	*CYLD (mut)* (Brooke-Spiegler)	-
Adamantinoid trichoblastoma	*EGFR (mut)*	-
Trichogerminoma	*GRHL (fus)*	CK20
Pilomatricoma and other matrical tumors	*CTNNB1 (mut)*	β-catenin (nuclear)
Sebaceous tumors	**Genetic alteration**	**IHC marker**
Adenoma, sebaceoma and carcinoma	MMRd	MSH2, MSH6 MLH1, PMS2
Carcinoma	HPV *(int)*, *RB1(mut)*	Rb
Sweat gland tumors	**Genetic alteration**	**IHC marker**
Poroma and porocarcinoma	*YAP::MAML2, YAP1::NUTM1*	YAP1 (Cter) NUT
Poroid hidradenoma	*YAP1::NUTM1*	YAP1 (Cter) NUT
Spiradenoma	*ALPK1 (mut)*	-
Cylindroma	*CYLD (mut)*	-
Hidradenoma	*CRTC1/3::MAML2*	-
Tubular adenoma	*BRAF (mut)*	BRAF p. V600E
Syringocystadenoma papilliferum	*BRAF (mut)*	
Mixed tumors	*PLAG1 (fus)*	PLAG
Myoepithelioma	*PLAG1 (fus)*	PLAG
Adenoid cystic carcinoma	*MYB (fus), MYBL1 (fus)*	-
Digital papillary adenocarcinoma	HPV42 (Int)	-
Secretory carcinoma	*ETV6::NTRK3*	Pan TRK
NUT adnexal carcinoma	*BRD3::NUTM1, NSD3::NUTM1, BRD3::NUTM2B*	NUT


Tumors with follicular differentiation
FOXK1::GRHL1 rearranged trichogerminoma
CTNNB1 mutated pilomatricoma
FOXK1 (break apart FISH)
Betacatenin (IHC)


Tumors with eccrine and apocrine differenciation
YAP1::MAML2 rearranged hidroacanthoma simplex
YAP1::NUTM1 rearranged poroid hidradenoma
TRPS1::PLAG1 rearranged myoepithelioma
YAP1 C-terminal (IHC)
NUT (IHC)
PLAG1 (IHC)

CPD • Clinicopathological case

CED
Clinical and Experimental Dermatology

CPD

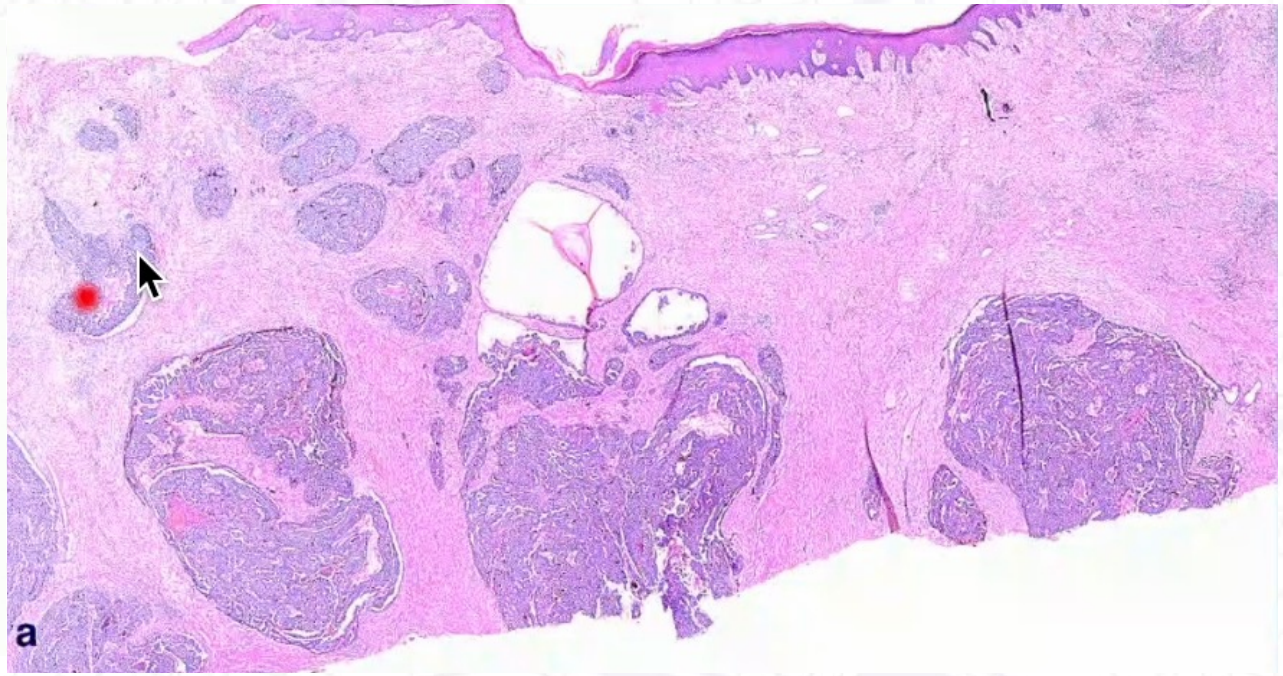
A rare and aggressive cutaneous thumb tumour

I. Svagelj,[1] J. Zarubica-Mavsar,[2] L. Labinac,[2] D. Herceg[3] and Z. Marusic[4]

[1]Department of Pathology and Cytology, General County Hospital Vinkovci, Vinkovci, Croatia; [2]Department of Pathology, General Hospital Pula, Pula, Croatia; [3]Oncology Clinic; and [4]Clinical Department of Pathology and Cytology, University Hospital Centre Zagreb, Zagreb, Croatia

doi: 10.1111/ced.14779

- 53 year old male
- ulcerated thumb nodule
- several months duration
- enlarged axillary lymph node



¿SCC?

CPD • Clinicopathological case

CED
Clinical and Experimental Dermatology

CPD

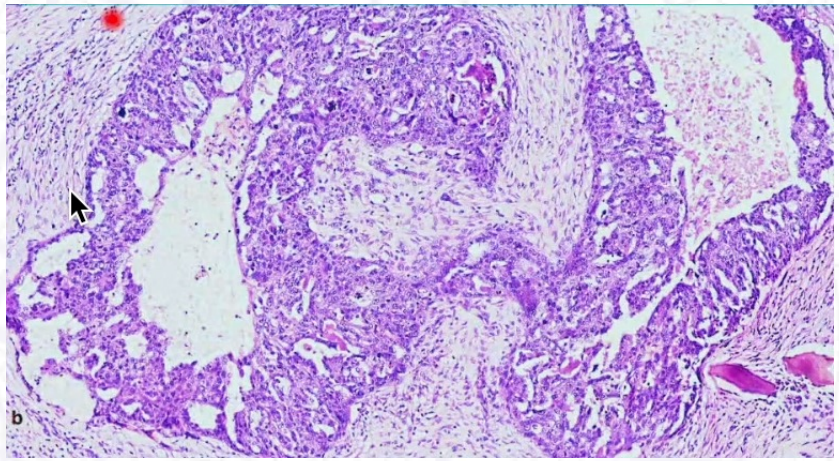
A rare and aggressive cutaneous thumb tumour

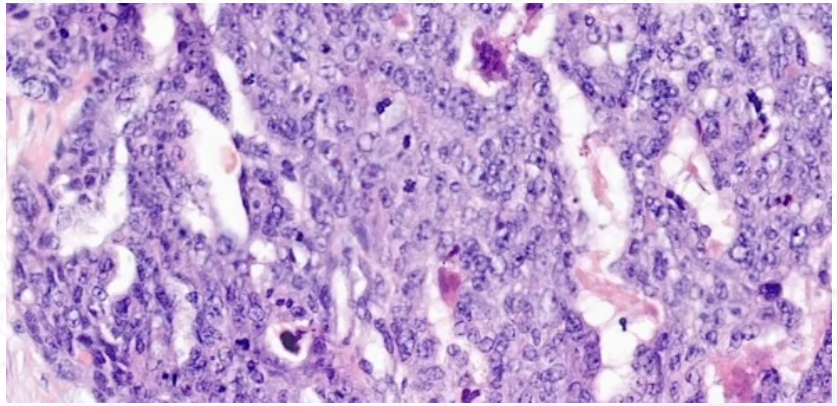
I. Švagelj,[1] J. Zarubica-Mavsar,[2] L. Labinac,[2] D. Herceg[3] and Z. Marusić[4]

[1]Department of Pathology and Cytology, General County Hospital Vinkovci, Vinkovci, Croatia; [2]Department of Pathology, General Hospital Pula, Pula, Croatia; [3]Oncology Clinic; and [4]Clinical Department of Pathology and Cytology, University Hospital Centre Zagreb, Zagreb, Croatia

doi: 10.1111/ced.14779

- 53 year old male
- ulcerated thumb nodule
- several months duration
- enlarged axillary lymph node





CPD • Clinicopathological case

CED
Clinical and Experimental Dermatology

CPD

A rare and aggressive cutaneous thumb tumour

I. Svagelj,[1] J. Zarubica-Mavsar,[2] L. Labinac,[2] D. Herceg[3] and Z. Marusic[4]

[1]Department of Pathology and Cytology, General County Hospital Vinkovci, Vinkovci, Croatia; [2]Department of Pathology, General Hospital Pula, Pula, Croatia; [3]Oncology Clinic; and [4]Clinical Department of Pathology and Cytology, University Hospital Centre Zagreb, Zagreb, Croatia

doi: 10.1111/ced.14779

- 53 year old male
- ulcerated thumb nodule
- several months duration
- enlarged axillary lymph node

PATIENT

DISEASE Skin adnexal carcinoma
NAME 3403046599, HR
DATE OF BIRTH 15 November 1967
SEX Male
MEDICAL RECORD # Not given

PHYSICIAN

ORDERING PHYSICIAN Herceg, Davorin
MEDICAL FACILITY UHC Zagreb
ADDITIONAL RECIPIENT None
MEDICAL FACILITY ID 501122
PATHOLOGIST Marusic, Zlatko

SPECIMEN

SPECIMEN SITE Skin
SPECIMEN ID 22142 /19 (3
SPECIMEN TYPE Block
DATE OF COLLECTION 20 October 2019
SPECIMEN RECEIVED 14 November 2019

Genomic Signatures

Microsatellite status - MS-Stable
Tumor Mutational Burden - TMB-Low (1 Muts/Mb)

Gene Alterations

For a complete list of the genes assayed, please refer to the Appendix.

FLT1 D1081N

0 Therapies approved in the EU
0 Therapies with Lack of Response
0 Clinical Trials

VEGF-R

GENOMIC SIGNATURES	ACTIONABILITY
Microsatellite status - MS-Stable	**No therapies or clinical trials.** see Genomic Signatures section
Tumor Mutational Burden - TMB-Low (1 Muts/Mb)	**No therapies or clinical trials.** see Genomic Signatures section

No therapies or clinical trials are associated with the Gene Alterations for this sample.

AdenoCa. papilar digital

- Raro.
- Crecimiento rápido. Agresivo.
- N+ y Mt viscerales (hasta > 20 años).
- Localización **acral** (dedos, varones) y **genital** (vulva, mujeres).

Aggressive digital papillary adenocarcinoma: A clinicopathological study of 19 cases

Noëlle Weingertner, MD,[a] Anne Gressel, JD,[a] Maxime Battistella, MD, PhD,[c,d] and Bernard Cribier, MD, PhD[b]
Strasbourg and Paris, France

Cutaneous Digital Papillary Adenocarcinoma
A Clinicopathologic Study of 31 Cases of a Rare Neoplasm With New Observations

Ravi Suchak, MBChB, MSc, MRCP, MRCGP, Dip RCPath, Wei-Lien Wang, MD,† Victor G. Prieto, MD, PhD,† Doina Ivan, MD,† Alexander J. Lazar, MD, PhD,† Thomas Brenn, MD, PhD, FRCPath,‡§ and Eduardo Calonje, MD, Dip RCPath**

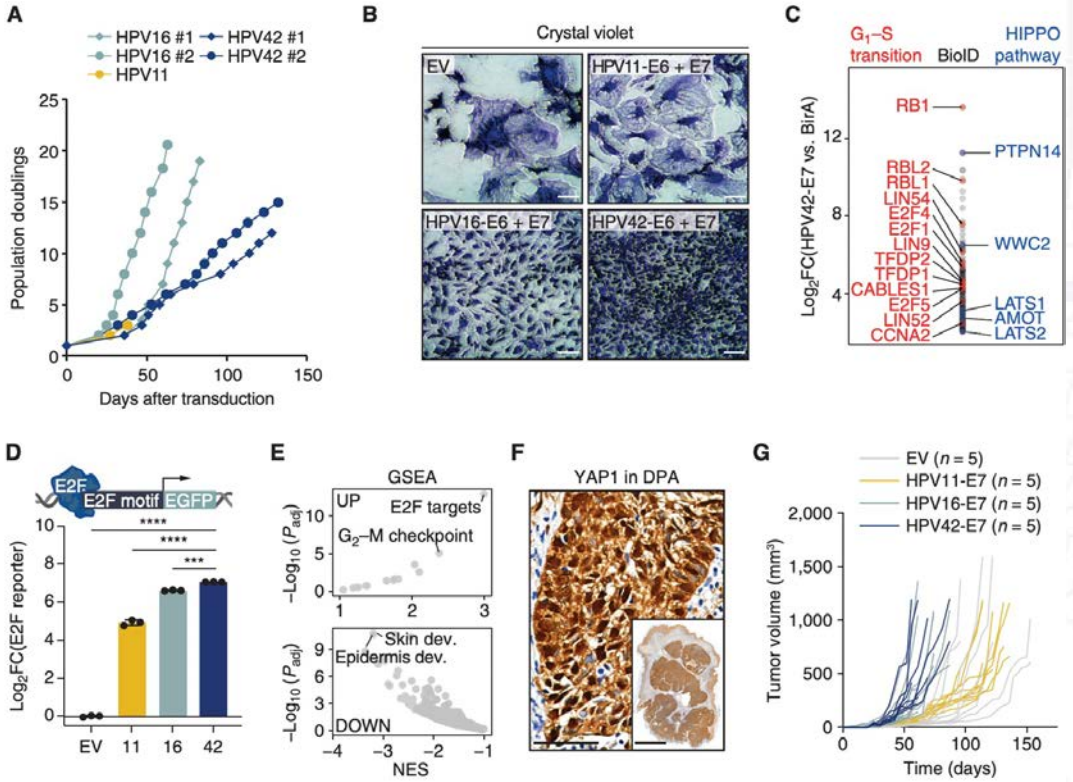
> Am J Surg Pathol. 2023 Oct 1;47(10):1077-1084. doi: 10.1097/PAS.0000000000002096. Epub 2023 Jul 31.

Digital Papillary Adenocarcinoma in Nonacral Skin: Clinicopathologic and Genetic Characterization of 5 Cases

Thibault Kervarrec [1 2 3], Sandrine Imbeaud [4], David Veyer [4 5], Helene Pere [4 5], Julien Puech [4], Agnes Pekár-Lukacs [6 7], Dorota Markiewicz [7], Michael Coutts [8], Anne Tallet [9], Christine Collin [9], Patricia Berthon [2], Ignacio G Bravo [10], Alice Seris [3 11], Thomas Jouary [3 11], Nicolas Macagno [12 13], Antoine Touzé [2], Bernard Cribier [14], Maxime Battistella [15], Eduardo Calonje [7]

RESEARCH BRIEF

Human Papillomavirus 42 Drives Digital Papillary Adenocarcinoma and Elicits a Germ Cell–like Program Conserved in HPV-Positive Cancers



AdenoCa. papilar digital

- Raro.
- Crecimiento rápido. Agresivo.
- N+ y Mt viscerales (hasta > 20 años).
- Localización **acral** (dedos, varones) y **genital** (vulva, mujeres).

Aggressive digital papillary adenocarcinoma: A clinicopathological study of 19 cases

Noëlle Weingertner, MD,[a] Anne Gressel, JD,[a] Maxime Battistella, MD, PhD,[c,d] and Bernard Cribier, MD, PhD[b]
Strasbourg and Paris, France

Cutaneous Digital Papillary Adenocarcinoma
A Clinicopathologic Study of 31 Cases of a Rare Neoplasm With New Observations

Ravi Suchak, MBChB, MSc, MRCP, MRCGP, Dip RCPath, Wei-Lien Wang, MD,† Victor G. Prieto, MD, PhD,† Doina Ivan, MD,† Alexander J. Lazar, MD, PhD,† Thomas Brenn, MD, PhD, FRCPath,‡§ and Eduardo Calonje, MD, Dip RCPath**

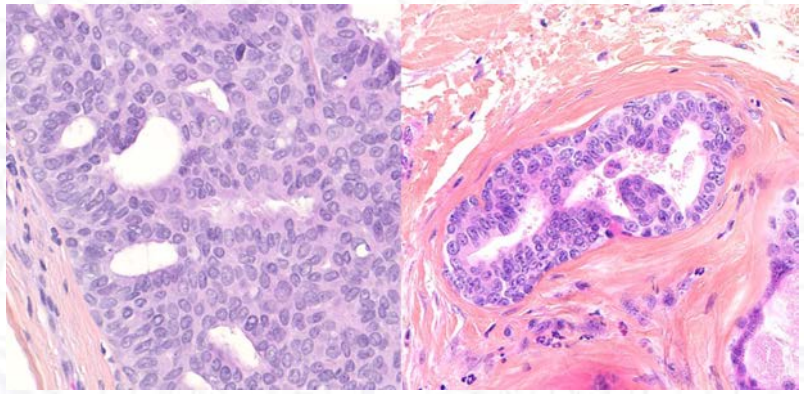
> Am J Surg Pathol. 2023 Oct 1;47(10):1077-1084. doi: 10.1097/PAS.0000000000002096. Epub 2023 Jul 31.

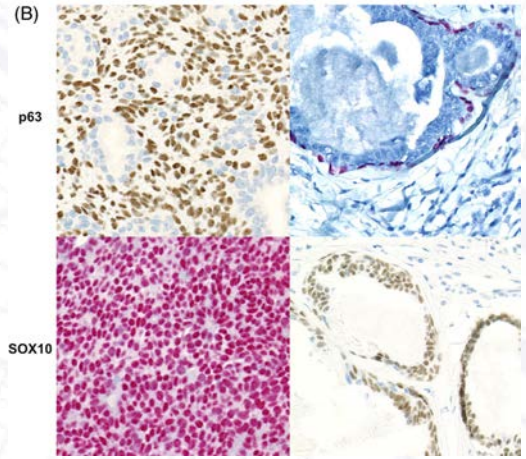
Digital Papillary Adenocarcinoma in Nonacral Skin: Clinicopathologic and Genetic Characterization of 5 Cases

Thibault Kervarrec [1 2 3], Sandrine Imbeaud [4], David Veyer [4 5], Helene Pere [4 5], Julien Puech [4], Agnes Pekár-Lukacs [6 7], Dorota Markiewicz [7], Michael Coutts [8], Anne Tallet [9], Christine Collin [9], Patricia Berthon [2], Ignacio G Bravo [10], Alice Seris [3 11], Thomas Jouary [3 11], Nicolas Macagno [12 13], Antoine Touzé [2], Bernard Cribier [14], Maxime Battistella [15], Eduardo Calonje [7]

Pitfall

Adenoma tubular





AdenoCa. papilar digital

- Raro.
- Crecimiento rápido. Agresivo.
- N+ y Mt viscerales (hasta > 20 años).
- Localización **acral** (dedos, varones) y **genital** (vulva, mujeres).

Aggressive digital papillary adenocarcinoma: A clinicopathological study of 19 cases

Noëlle Weingertner, MD,[a] Anne Gressel, JD,[a] Maxime Battistella, MD, PhD,[c,d] and Bernard Cribier, MD, PhD[b]
Strasbourg and Paris, France

Cutaneous Digital Papillary Adenocarcinoma
A Clinicopathologic Study of 31 Cases of a Rare Neoplasm With New Observations

Ravi Suchak, MBChB, MSc, MRCP, MRCGP, Dip RCPath, Wei-Lien Wang, MD,† Victor G. Prieto, MD, PhD,† Doina Ivan, MD,† Alexander J. Lazar, MD, PhD,† Thomas Brenn, MD, PhD, FRCPath,‡§ and Eduardo Calonje, MD, Dip RCPath**

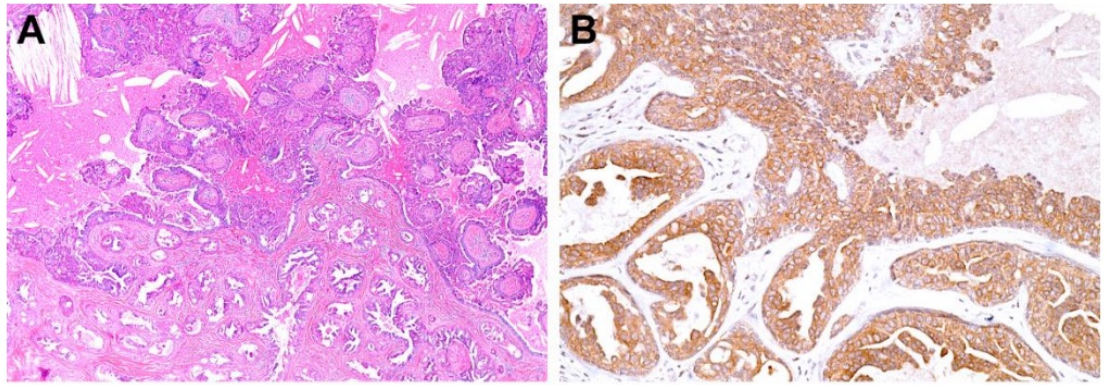
> Am J Surg Pathol. 2023 Oct 1;47(10):1077-1084. doi: 10.1097/PAS.0000000000002096. Epub 2023 Jul 31.

Digital Papillary Adenocarcinoma in Nonacral Skin: Clinicopathologic and Genetic Characterization of 5 Cases

Thibault Kervarrec [1 2 3], Sandrine Imbeaud [4], David Veyer [4 5], Helene Pere [4 5], Julien Puech [4], Agnes Pekár-Lukacs [6 7], Dorota Markiewicz [7], Michael Coutts [8], Anne Tallet [9], Christine Collin [9], Patricia Berthon [2], Ignacio G Bravo [10], Alice Seris [3 11], Thomas Jouary [3 11], Nicolas Macagno [12 13], Antoine Touzé [2], Bernard Cribier [14], Maxime Battistella [15], Eduardo Calonje [7]

Pitfall Adenoma tubular

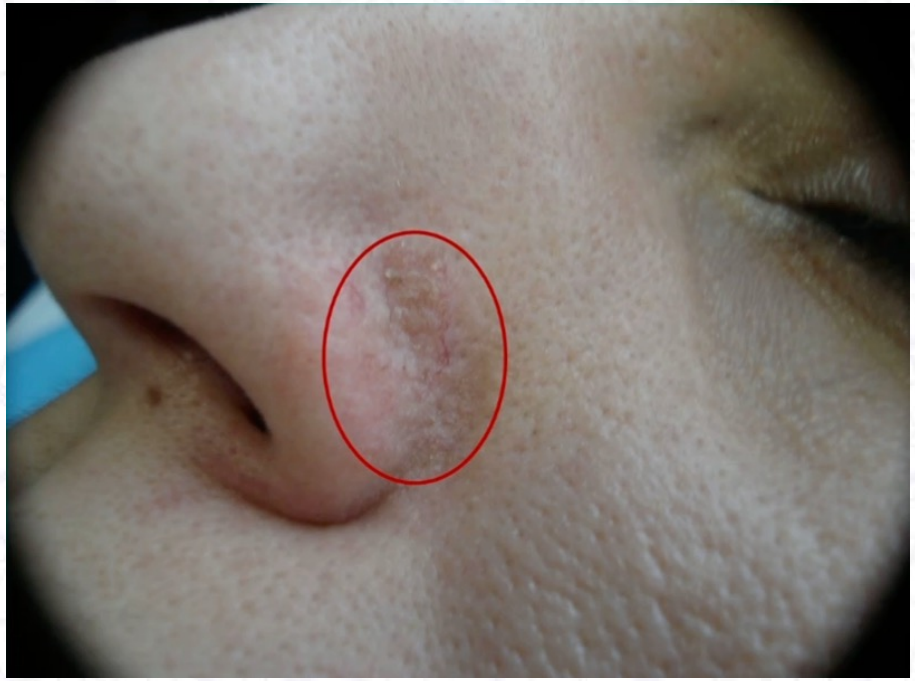
BRAF V600E

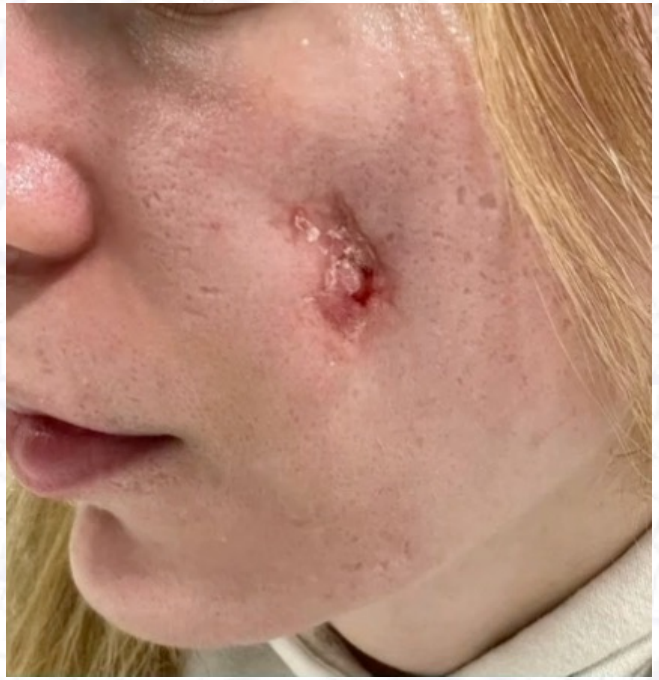


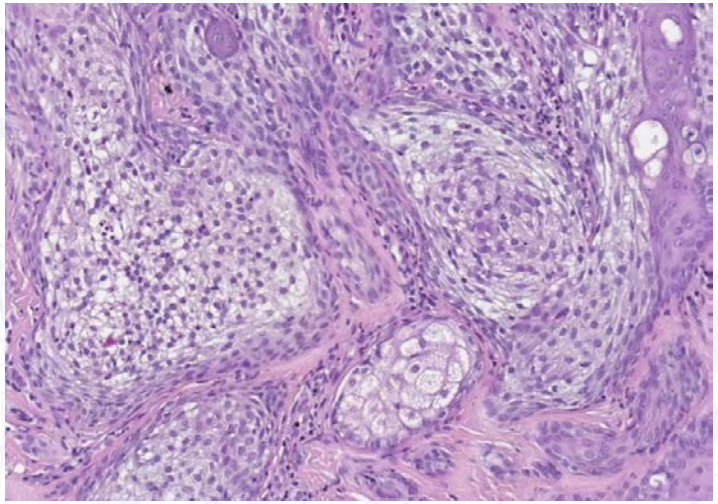
Comment > J Cutan Pathol. 2023 Jun;50(6):577-579. doi: 10.1111/cup.14430. Epub 2023 Apr 14.

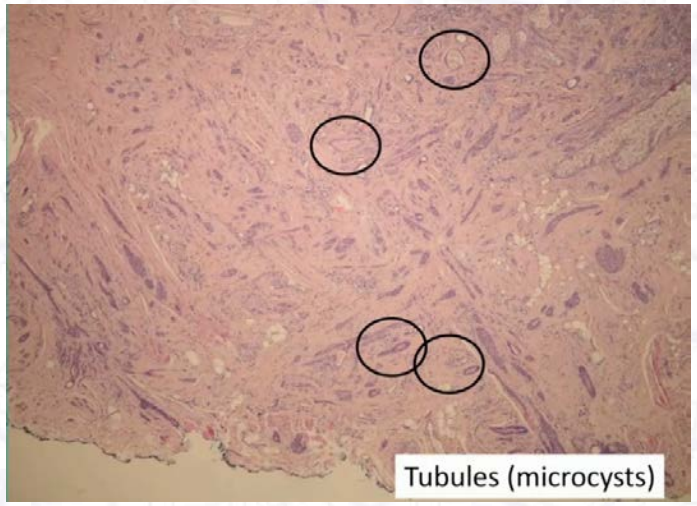
Acral BRAF-mutated tubular adenoma should be distinguished from HPV42-related digital papillary adenocarcinoma

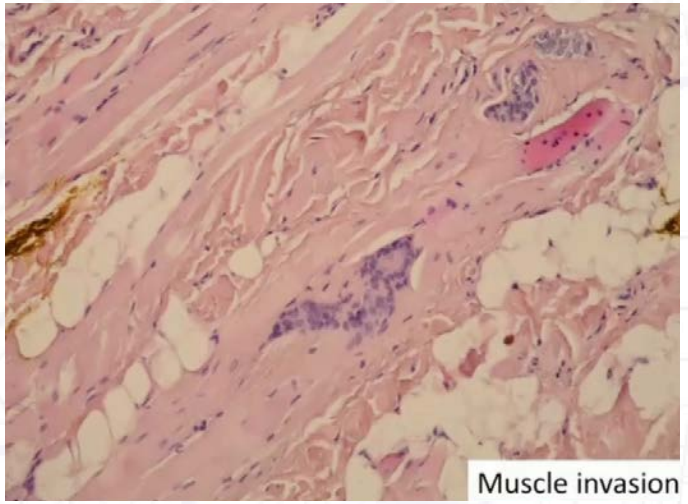
Thibault Kervarrec [1 2 3], Klaus J Busam [4]

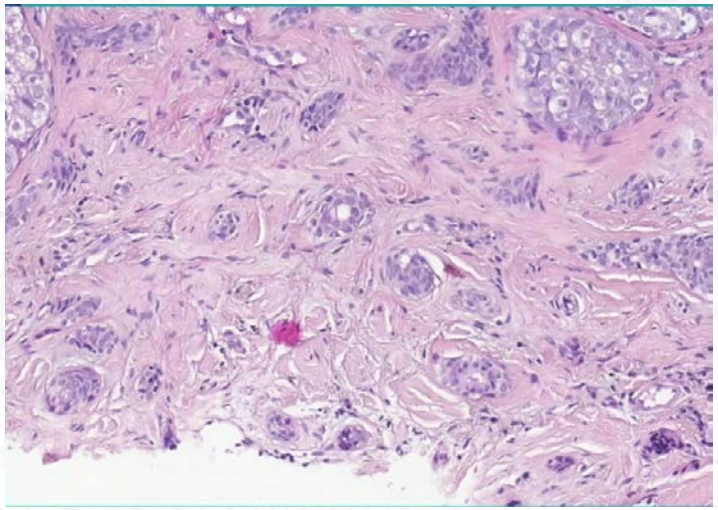


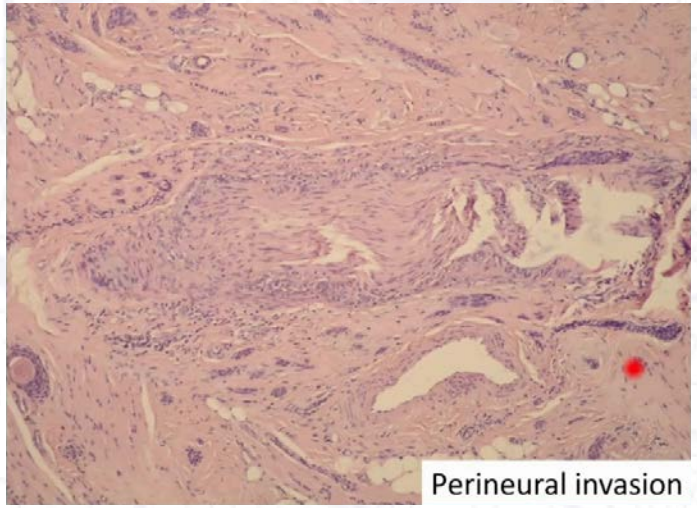


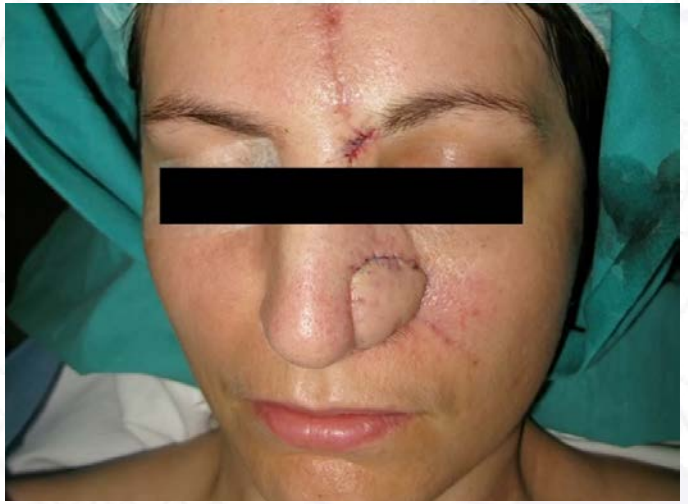



Tubules (microcysts)


Muscle invasion

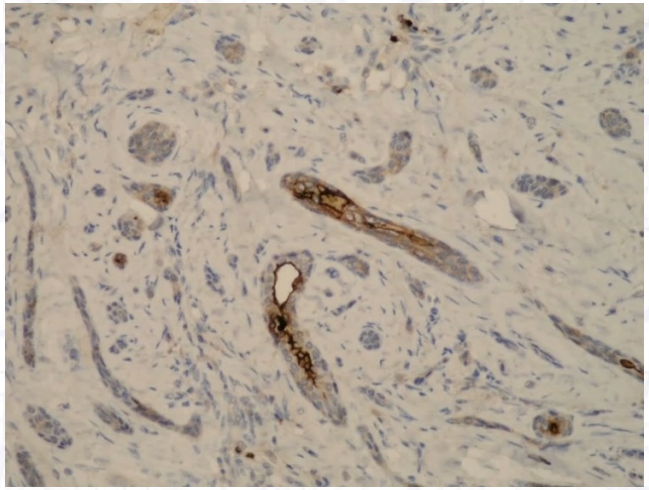



Perineural invasion



Carcinoma anexial microquístico

- Predominio en mujeres
- Localmente agresivo. 40-60% recidivas. Rara metástasis
- Mohs?



CEA

EA CONGRESS DV

Modern Pathology (2020) 33:1092–1103
https://doi.org/10.1038/s41379-019-0424-4

USCAP

ARTICLE

Next-generation sequencing implicates oncogenic roles for p53 and JAK/STAT signaling in microcystic adnexal carcinomas

May P. Chan[1,2,3] · Komal R. Plouffe[1,4,5] · Chia-Jen Liu[1,4] · Nallasivam Palanisamy[6] · Shannon Carskadon[6] · Lili Zhao[7] · Rosalynn M. Nazarian[8] · Alison B. Durham[2,3] · Timothy M. Johnson[2,3] · Aleodor A. Andea[1,2,3] · Rajiv M. Patel[1,2,3] · Lori Lowe[1,2,3] · Douglas R. Fullen[1,2,3] · Noah A. Brown[1] · Scott A. Tomlins[1,3,4,5] · Aaron M. Udager[1,4] · Paul W. Harms[1,2,3,4]

Yu et al. World Journal of Surgical Oncology (2022) 20:142
https://doi.org/10.1186/s12957-022-02601-6

World Journal of Surgical Oncology

RESEARCH Open Access

Four calcium signaling pathway-related genes were upregulated in microcystic adnexal carcinoma: transcriptome analysis and immunohistochemical validation

Shuaixia Yu[1,2†], Yang Wang[3†], Baijie Tang[1,2], Xiang Liu[1,2], Linhong Song[1,2], Gang Xu[1,2], Hong Zhu[1,2*] and Huajun Sun[1,2*]

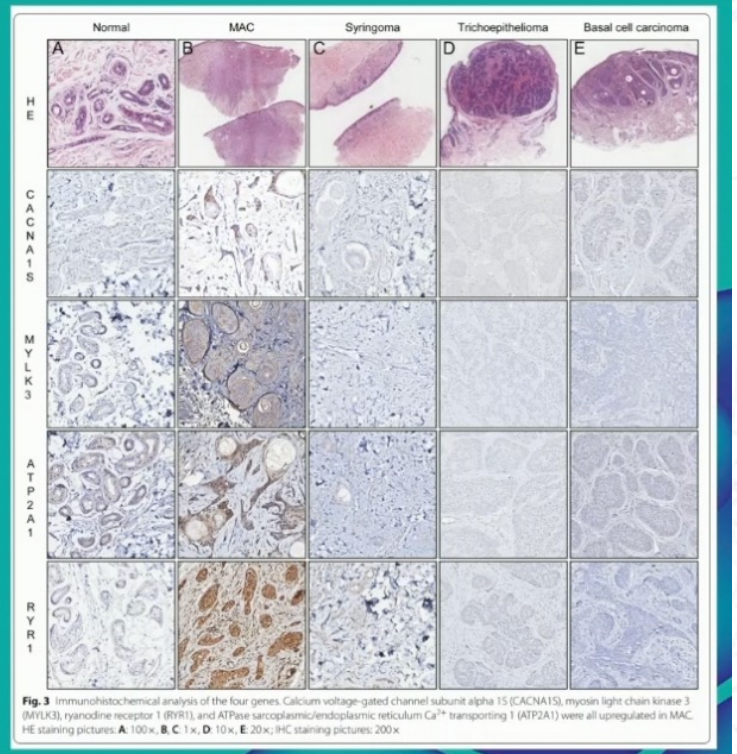


Fig. 3 Immunohistochemical analysis of the four genes. Calcium voltage-gated channel subunit alpha 1S (CACNA1S), myosin light chain kinase 3 (MYLK3), ryanodine receptor 1 (RYR1), and ATPase sarcoplasmic/endoplasmic reticulum Ca^{2+} transporting 1 (ATP2A1) were all upregulated in MAC. HE staining pictures: **A**: 100×, **B**, **C**: 1×, **D**: 10×, **E**: 20×; IHC staining pictures: 200×

Improved Margin Control of Microcystic Adnexal Carcinoma After Mohs Micrographic Surgery Compared With Wide Local Excision

Sharmitha Yerneni [1], Fadi Murad [2], Chrysalyne D Schmults [2], Emily S Ruiz [2]

MELANOMA

Table 1. The Melanocytic Pathology Assessment Tool and Hierarchy for Diagnosis Version 2.0

Class	Risk of tumor progression	Probability of progression, No. per population	Treatment recommendation	Examples[a]
0	NA	NA	Consider repeat biopsy	Nondiagnostic or unsatisfactory
I: low grade	Very low risk for continued proliferation and progression to invasive melanoma	1 in 10 000 to 1 in 100 000	No further treatment[b]	Common acquired nevi, no atypia
				Congenital nevi, no atypia
				Atypical and dysplastic nevi, low-grade atypia[c]
				Common blue nevi
II: high grade	Low risk for progression to invasive melanoma	1 in 100 to 1 in 1000	Re-excision with margins <1 cm[b]	Atypical and dysplastic nevi, high-grade atypia[c]
				Spitz nevi, tumors or melanocytomas, and atypical variants
				Cellular blue nevi or melanocytomas and atypical variants
				Plexiform or deep penetrating nevi or melanocytomas
				Lentigo maligna
				Melanoma in situ
III: melanoma pT1a	Relatively low risk for local and regional metastasis	1 in 10 to 1 in 100	Follow national guidelines (eg, wide excision with 1 cm margins)[b]	Melanoma AJCC stage pT1a, <0.8 mm Breslow thickness
				Melanoma pT1a lr (low risk)[d]
				Melanoma pT1a[e]
IV: melanoma ≥pT1b	Moderate to increased risk for regional or distant metastasis	1 in 2 to 1 in 10	Follow national guidelines (eg, wide excision with 1-2 cm margins[b] and consideration of sentinel lymph node staging and other therapies)	Melanoma AJCC stage pT1b or greater, ≥0.8 mm Breslow thickness

Consensus Statement | Pathology and Laboratory Medicine

Revision of the Melanocytic Pathology Assessment Tool and Hierarchy for Diagnosis Classification Schema for Melanocytic Lesions

A Consensus Statement

Classification of unusual histologic variants of cutaneous melanoma

(Rongioletti F, Smoller BR. J Cutan Pathol 2005;32)

1. Silhoutte
2. Stromal changes
3. Cytologic features
4. Combined patterns (silhoutte+cytology)

Franco Rongioletti
Melanoma

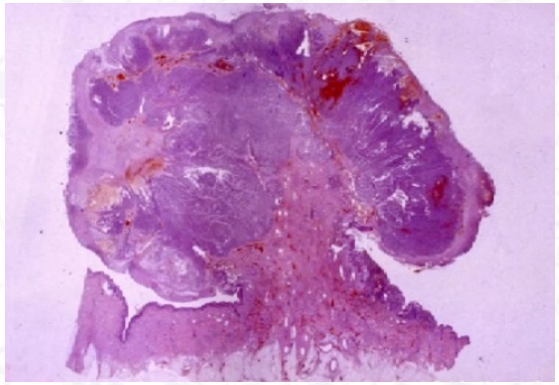
MM polipoideo

- Variante de MM nodular.
- Peor pronóstico.
- **↑ ulceración y Breslow.**

› Melanoma Res. 2023 Jun 1;33(3):257-261. doi: 10.1097/CMR.0000000000000886. Epub 2023 Mar 2.

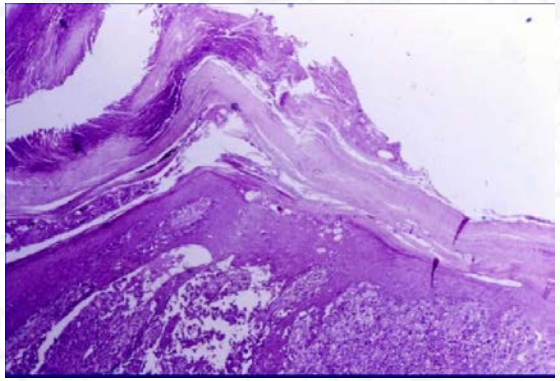
Prognosis of polypoid melanoma: a comparative study with non-polypoid melanomas

Velma Y Jasso-Sosa [1], Leonardo S Lino-Silva [1], Marín G Escobar-Jiménez [1], Joab R Galván-Bustillos [1], Dorian Y García-Ortega [2], Rosa A Salcedo-Hernández [2], César Zepeda-Najar [3], Pedro Frías-Fernández [4]



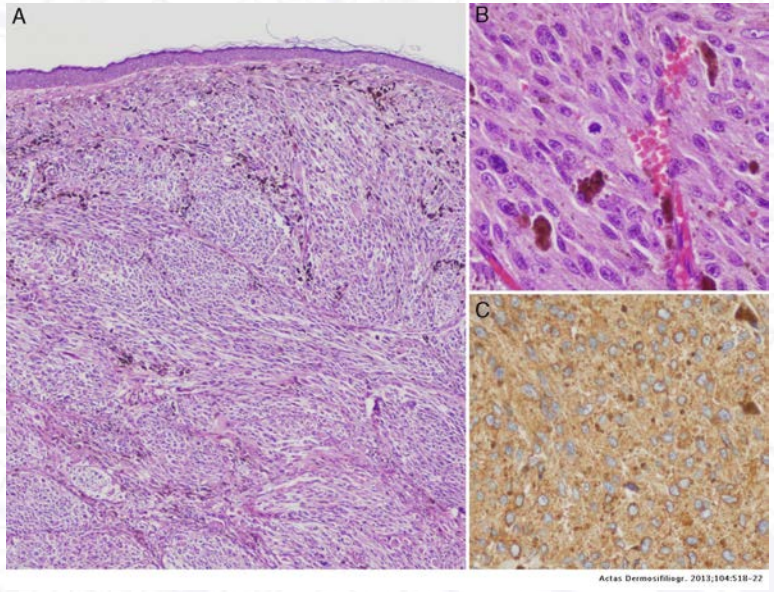
MM verrucoso (QS-like)

- *Misdiagnosis* 50%.
- Pronóstico = MM convencional.
- Acantosis y papilomatosis.
- Crecimiento pagetoide.
- ¿Breslow sobrestimado?



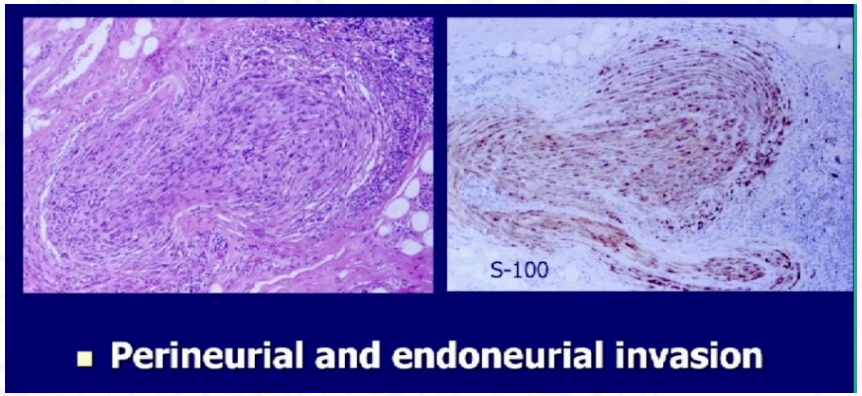
MM dérmico primario

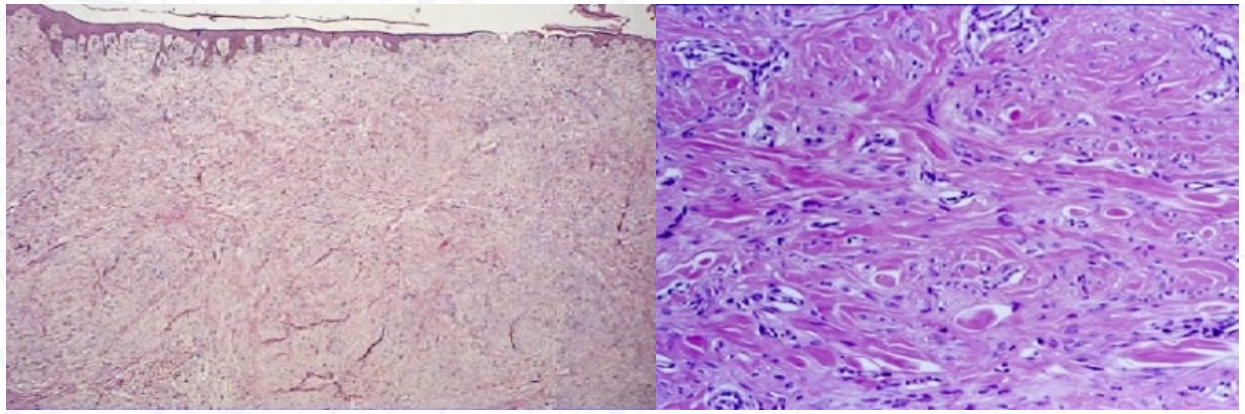
- *Challenging.*
- NO metástasis en estudio de extensión.
- Supervivencia prolongada.
- Mejor pronóstico que MM convencional de = espesor.



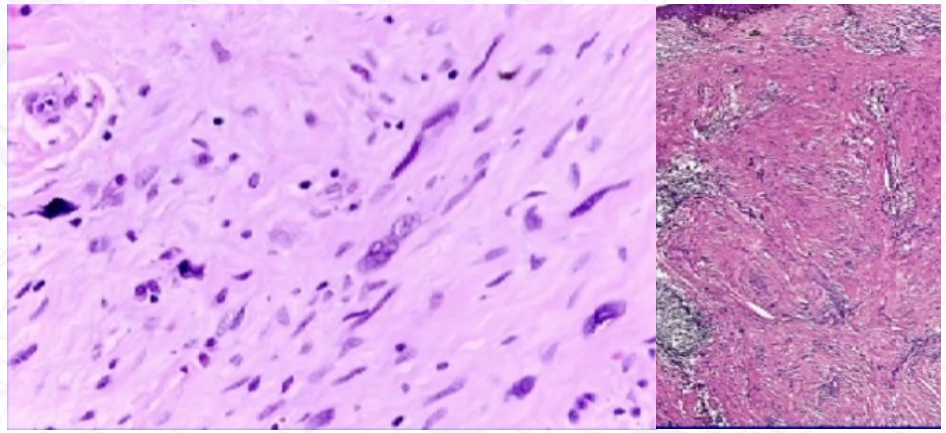
MM desmoplásico (neurotrópico)

- 4% del total.
- Fotoexposición (asociado a **LM**).
- ↑ carga mutacional (**NF-1**). NO BRAF, NRAS o c-kit.
- Recurrencia local. Rara metástasis.
- ¿Mejor pronóstico? NO si neurotropismo.





Pitfall: cicatriz, DF



Fibroblast-like atypical cells

Limphoid infiltrates

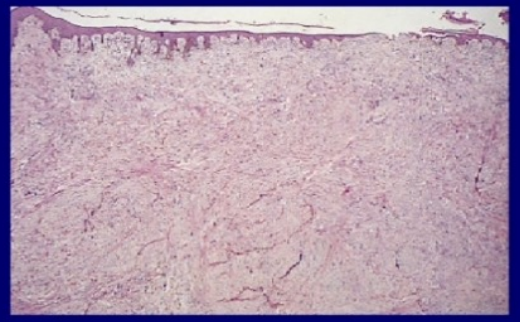
IHQ: **S-100 y SOX-10 +.** HMB-45 y Melan A -

¿Evitar BSGC en MM desmoplásico puro?

Two subtypes of desmoplastic melanoma

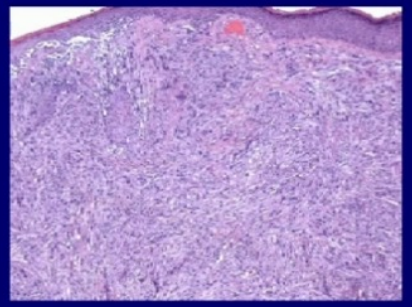
1. Pure

desmoplasia is prominent troughout the full tumor (90%)



2. Myxed

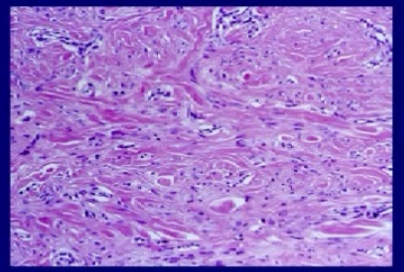
fibrosis is present in only part of an otherwise non desmoplastic melanoma



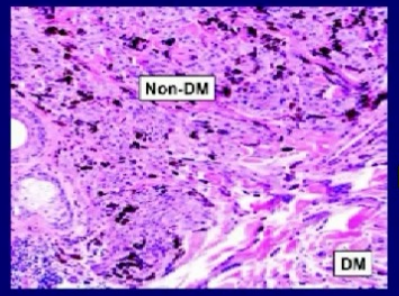
Clinical utility of classifying desmoplastic melanoma into pure and mixed subtypes

(McClain et al. J Cutan Pathol 2009;36; Hawkins et al. Ann Surg Oncol 2005;12)

- **Pure subtypes** are unlikely to disseminate to regional lymphnodes and are associated with a more favorable outcome than **mixed subtype** or **conventional type**
- Sentinel node biopsy restricted to only mixed type?
- Different mutation pattern of the 2 variants



Pure



Mixed

Tumores spitzoides

Front Oncol. 2022; 12: 889223.
Published online 2022 Jun 7. doi: 10.3389/fonc.2022.889223
PMCID: PMC9209745
PMID: 35747831

The Spectrum of Spitz Melanocytic Lesions: From Morphologic Diagnosis to Molecular Classification

Tiffany W. Cheng, [1] Madeline C. Ahern, [1] and Alessio Giubellino[1,2,*]

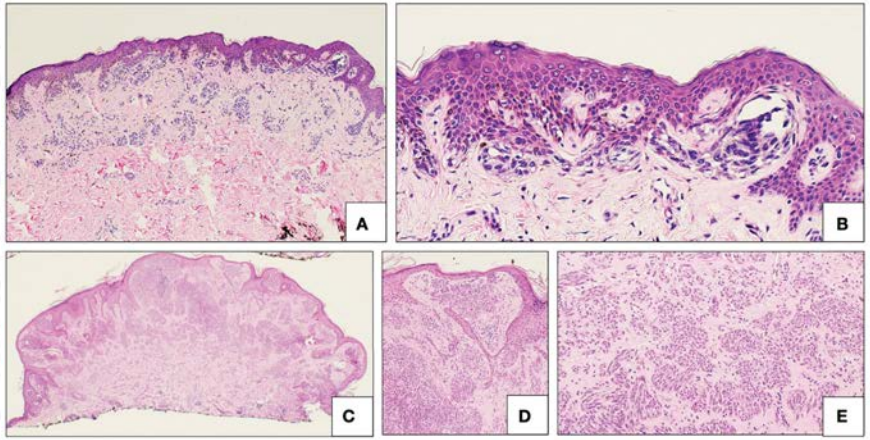
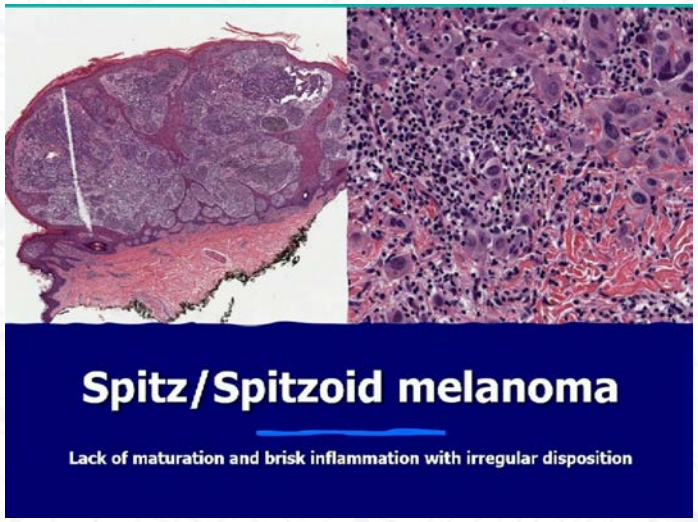


TABLE 2 | Histopathologic Characteristics of Spitz Tumors.

Histopathologic Feature	Spitz Nevus	Atypical Spitz Tumor	Spitzoid Melanoma
Size	<6 mm	6-10 mm	>1 cm
Symmetry	Symmetrical	May be symmetrical or asymmetrical	Asymmetrical
Circumscription	Well-Circumscribed	May be well or poorly circumscribed	Poorly circumscribed
Ulceration	Rare	More common than SN	Present
Epithelioid/ Spindle Cell Morphology	Can be primarily epithelioid, primary spindle cell, or combination of both; well-organized into dermo-epidermal nests found in banana bunch configuration	Can be primarily epithelioid, primarily spindle cell, or combination of both; greater epithelioid predominance in AST with MAP3K8 alterations; greater nuclear plemorphism and higher nuclear:cytoplasmic ratio	Can be primarily epithelioid, primarily spindle cell, or combination of both; irregular nesting patterns; fascicular melanocyte nests in SM with ALK fusions; dermal rosette structures in SM with NRTK1 fusions; greater epithelioid predominance in SM with MAP3K8 fusions
Pagetoid Spread	Uncommon, typically focal if present	If present, typicaly peripheral, involves upper epidermis	Present, may be extensive
Kamino Bodies	Present, located at periphery of melanocyte nests	Infrequent, may be smaller in size	Rare
Melanocyte Maturation	Commonly Present	May be absent	Absent
Mitotic Activity	Low, 0-2/mm^2	Moderate, 2-6/mm^2	High, >6/mm^2
Lymphocytic Inflammatory Infiltrate	Commonly present	May be present	May be present
Multinucleation	May be present, typically found in SN of primarily epithelioid origin	May be present in AST with MAP3K8 alterations	Rare
Epidermal Hyperplasia	Present	Rare	Rare

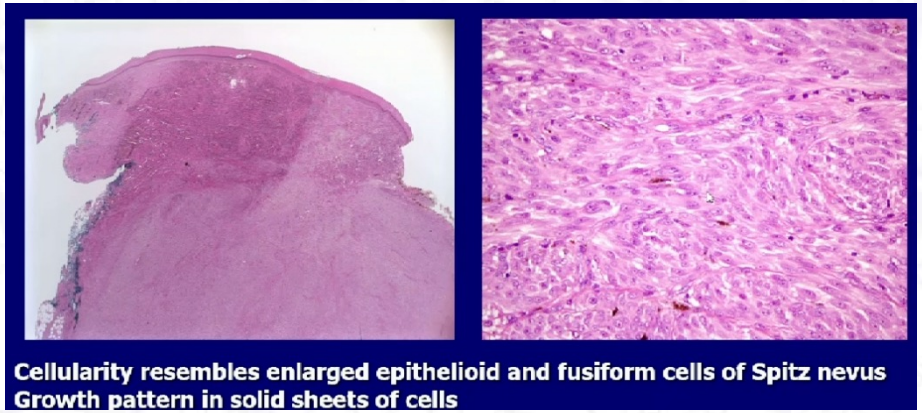
Melanoma de Spitz (tumor de Spitz maligno)

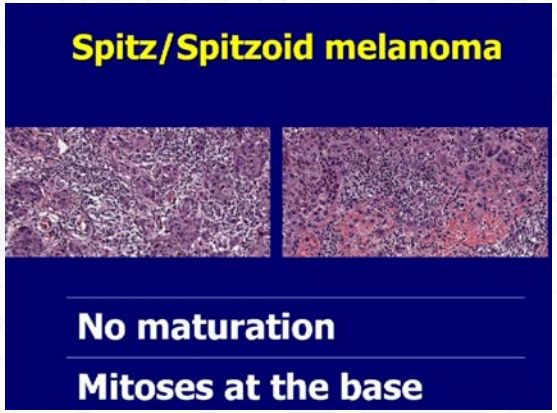
- Espectro de tumores de Spitz (**H-RAS**, fusiones de ROS1, ALK)
- Jóvenes.
- Buen pronóstico.

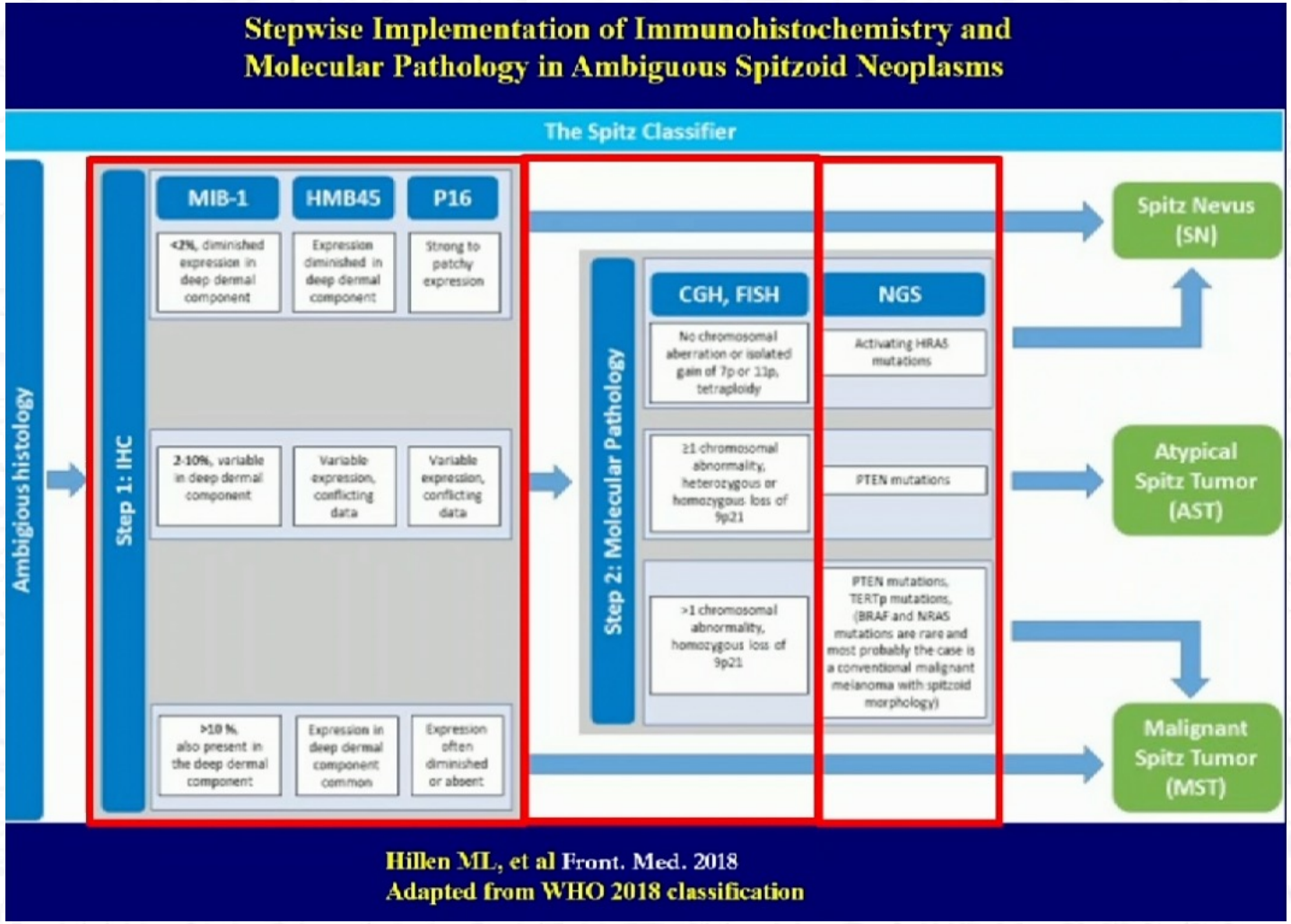


Melanoma spitzoide

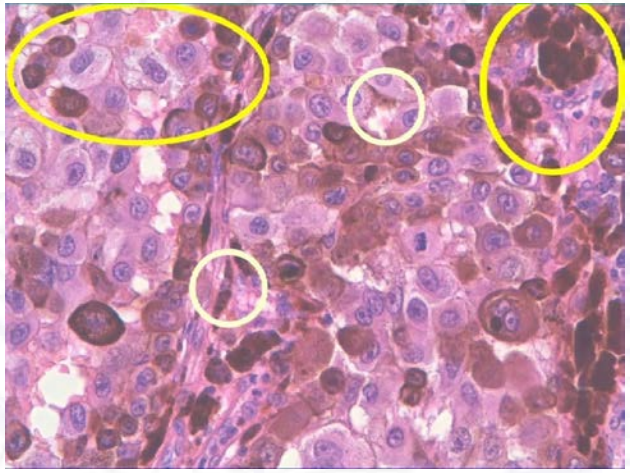
- ¿Melanoma convencional? (**BRAF**)
- Adultos.
- Peor pronóstico.






Stepwise Implementation of Immunohistochemistry and Molecular Pathology in Ambiguous Spitzoid Neoplasms
The Spitz Classifier
Ambigious histology
Step 1: IHC
MIB-1
HMB45
P16
<2%, diminished expression in deep dermal component
Expression diminished in deep dermal component
Strong to patchy expression
2-10%, variable in deep dermal component
Variable expression, conflicting data
Variable expression, conflicting data
>10 %, also present in the deep dermal component
Expression in deep dermal component common
Expression often diminished or absent
Step 2: Molecular Pathology
CGH, FISH
NGS
No chromosomal aberration or isolated gain of 7p or 11p, tetraploidy
Activating HRAS mutations
≥1 chromosomal abnormality, heterozygous or homozygous loss of 9p21
PTEN mutations
>1 chromosomal abnormality, homozygous loss of 9p21
PTEN mutations, TERTp mutations, (BRAF and NRAS mutations are rare and most probably the case is a conventional malignant melanoma with spitzoid morphology)
Spitz Nevus (SN)
Atypical Spitz Tumor (AST)
Malignant Spitz Tumor (MST)
Hillen ML, et al Front. Med. 2018
Adapted from WHO 2018 classification

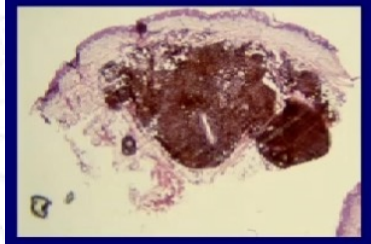
Melanoma *animal-type*



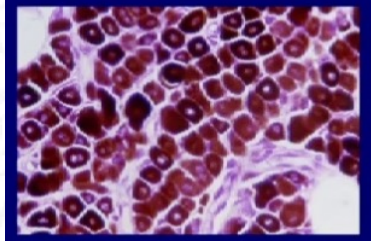
- The combination of striking melanin deposition and relatively indolent behavior has led to abandon the denomination animal-type melanoma in favor of the umbrella term "Pigmented epithelioid melanocytoma"

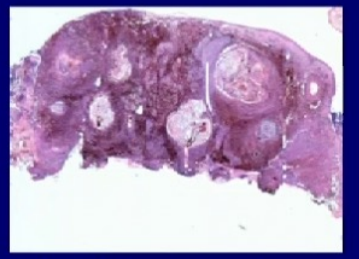
Pigmented epithelioid melanocytoma

Zembowicz A, Carney JA, Mihm MC.Am J Surg Pathol. 2004;28
Mandal et al Am J Surg Pathol. 2009;33

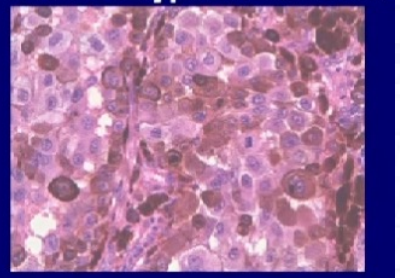


Epithelioid blue nevus





Animal-type melanoma



- A clinical-anatomic "entity" said to include both benign and malignant pigmented melanocytic neoplasms ranging from epitheliod blue nevus to animal-type melanoma characterized by an indolent course
- PRKAR!A-inactivated melanocytoma

PRKAR1A y PRKCA

Melanocytic Tumour of Uncertain Malignant Potential (MelTUMP)
A Diagnostic Dilemma in Children

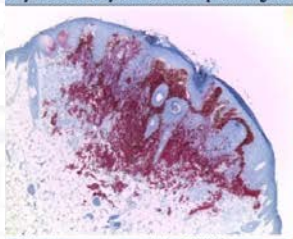
Main author: Diana-Maria CETINĂ[1]
Co-author: Ioana BILIUȚĂ[1]
Scientific coordinators: Elena Diana OLTEANU[2], Annamaria FÜLOP[2], Prof. Dr. Gabriela Adriana FILIP[1], Rodica Voichița COSNAROVICI[2]
Affiliated: *University of Medicine and Pharmacy "Iuliu Haţieganu" Cluj-Napoca, Romania[1], Oncology Institute "Prof. Dr Ion Chiricuță" Cluj-Napoca, Romania[2]*

INTRODUCTION

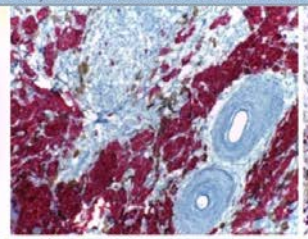
The histopathological diagnosis and classification of melanocytic skin tumors is a practical challenge in paediatric dermatopathology. Sometimes, the major difficulty is distinguishing between benign and malignant microscopical patterns of a skin lesion. These borderline entities are defined as melanocytic tumours of uncertain malignant potential (MelTUMPs) [1].

CASE REPORT

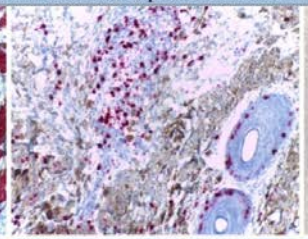
We are reporting the case of a 12-year-old female patient was referred to a local hospital for a pigmentary skin lesion which recently increased in size. The infracentimetric lesion was located on the upper posterior trunk. Due to dermoscopy findings, the lesion was excised, and the initial histopathological diagnosis was nodular melanoma. Reevaluation of the lesion performed in our oncology center raised the suspicion of a MelTUMP. Microscopy described a proliferation of large rather fusiform melanocytes arranged in confluent jonctional nests in a hyperplastic epidermis with lymphocytic infiltrate. Minimal cell atypia and no mitotic activity were identified. HMB45 was positive and Ki67 index was 20%. Due to the diagnosis difference, other opinions were requested from experts. Two more versions were obtained: compound blue nevus (CBN) and pigmented epithelioid melanocytoma (PEM) with loss of PRKAR1A expression. The RNA-Sequencing revealed PRKAR1A-ACTB fusion. As ultrasonography (US) revealed bilateral axillary polyadenopathies, sentinel lymph node biopsy (SNB) was performed, showing subcapsular metastases with the same histopathological pattern. PET scan revealed no FDG up-take after the excision. The patient underwent 3 mg/kg Nivolumab immunotherapy every 4 weeks for 1 year. The follow-up including blood tests, US check and CT scan once in 6 months did not show the recurrence. The patient has been free of disease for 2 years.



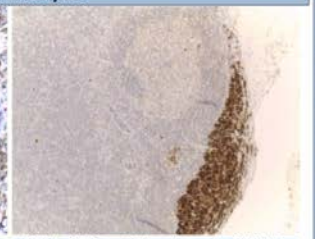
Obj. 4x: tegument containing HMB45+ melanocytes after depigmentation; symmetrical architecture; epidermal hyperplasia



Obj. 20x: HMB45+ melanocytes after depigmentation



Obj. 20x: melanocytes with abundant pigment; Ki67=5-10% of the lymphocytes nuclei one month after biopsy



Obj. 4x:SNB containing subcapsular HMB45+ melanocytic proliferation

DISCUSSIONS

The studies show that MelTUMPs are not a well-defined histological entity. They have a predilection for paediatric population. The diagnosis is made by IHC, but the markers used are frequently nonspecific. In a considerable number of cases they metastasize locally and systemically only in exceptional cases [2]. Local excision should be performed in all cases, but studies show that positive SNB has low importance in diagnosis and prognosis [1]. PEM represents one of the most common MelTUMP [1] although it is recognized by the WHO 2018 guideline as an individual lesion. It is ruled out of MelTUMP category by PRKCA fusion [2], but inactivation of PRKAR1A is also highly correlated with the diagnosis of PEM [3]. The PRKAR1A-ACTB fusion remains yet undescribed in literature, but sustains the diagnosis. As a class characteristic of MelTUMPs, recurrence and mortality rates are low [4]. This condition should be treated as a high grade melanocytoma, according to WHO [2].

CONCLUSION

Here we report a case of a rare paediatric skin tumour. Owning their frequency and non-specific diagnostic methods, individual entities of MelTUMPs are challenging to be differentiated from one another. The positive diagnosis is often made after being examined by experienced dermatopathologists, as was the case of our patient.

REFERENCES

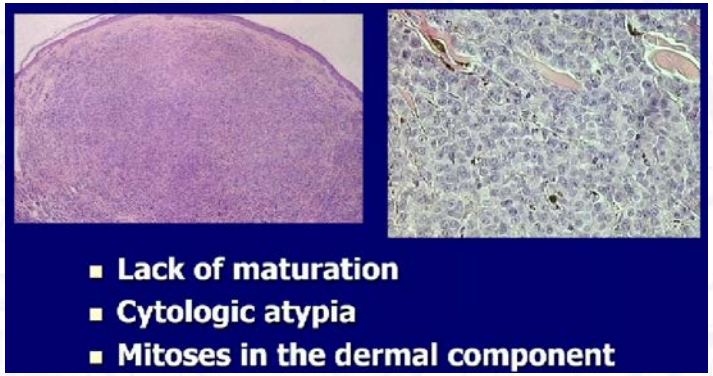
1. Varey AHR, Williams GJ, Lo SN, Taing CY, Maurichi A, Santinami M, Scolyer RA, Thompson JF. Clinical management of melanocytic tumours of uncertain malignant potential (MelTUMPs), including melanocytomas: A systematic review and meta-analysis. J Eur Acad Dermatol Venereol. 2023 May;37(5):859-870.
2. Ferrara G, Argenziano G. The WHO 2018 Classification of Cutaneous Melanocytic Neoplasms: Suggestions From Routine Practice. Front Oncol. 2021 Jul 2;11:675296.
3. Cohen JN, Joseph NM, North JP, Onodera C, Zembowicz A, LeBoit PE. Genomic Analysis of Pigmented Epithelioid Melanocytomas Reveals Recurrent Alterations in PRKAR1A, and PRKCA Genes. Am J Surg Pathol. 2017 Oct;41(10):1333-1346
4. Ensslin CJ, Hibler BP, Lee EH, Nehal KS, Busam KJ, Rossi AM. Atypical Melanocytic Proliferations: A Review of the Literature. Dermatol Surg. 2018 Feb;44(2):159-174

EADV CONGRESS

- Adenopatías axilares bilaterales.
- Nivolumab.
- TLD: 2 años.

MM nevoide

- *Nightmare*.
- Variantes: 1) papilomatosa. 2) *dome-shaped*.
- **HMB-45 y KI-67 ↑** en dermis profunda.



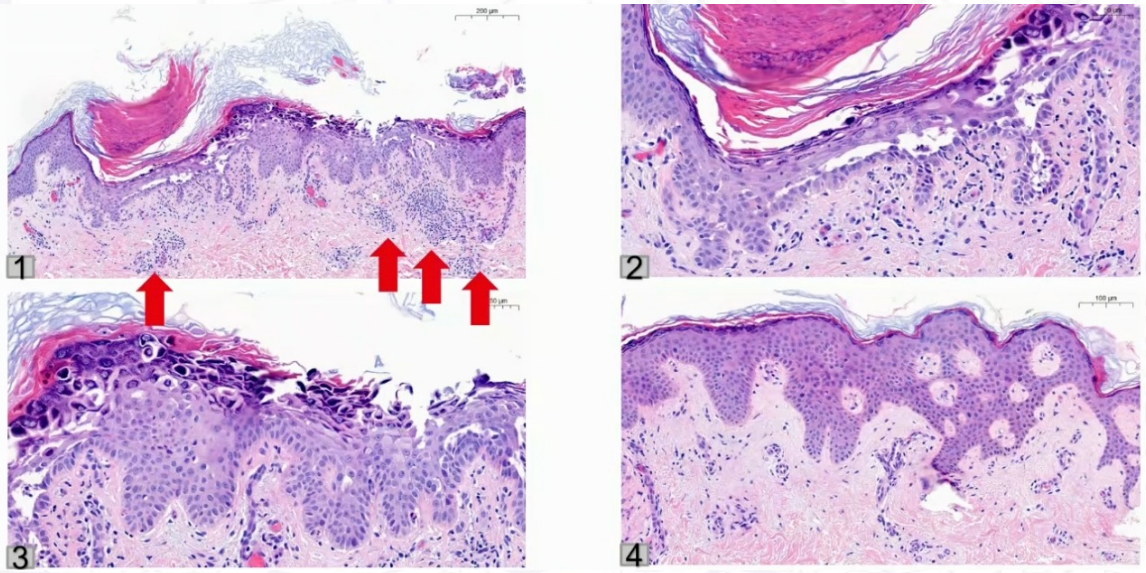
"A melanoma that I have diagnosed as nevus, and I wish I hadn't»
(McKee PH.Histopathology.2010)

The diagnosis of the day after
(Rongioletti)

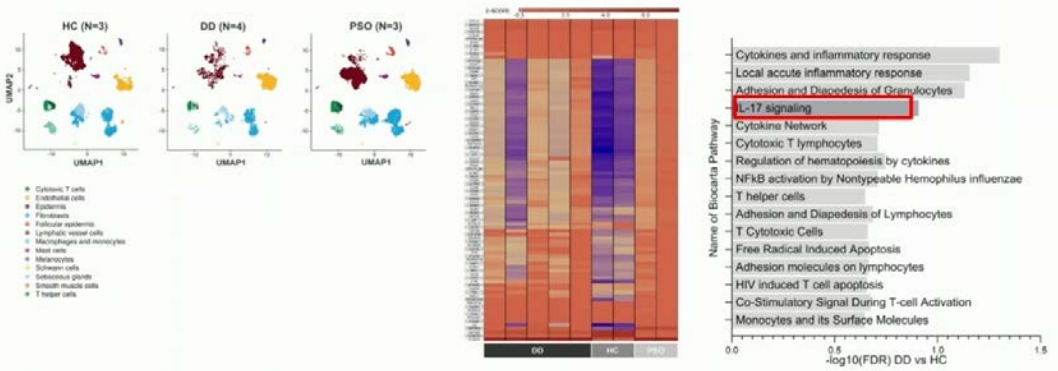
Take-home message:

- **Pure desmoplastic:** > recurrences < metastases?
- **Neurotropic**: less survival?
- **Balloon cell**: > skin metastases?
- **Spitz melanoma:** better prognosis in young individuals
- **Animal-type**: more indolent behaviour
- **Malignant blue nevus**: aggressive with late-onset metastases
- **Clear cell sarcoma**: aggressive with recurrences and metastases

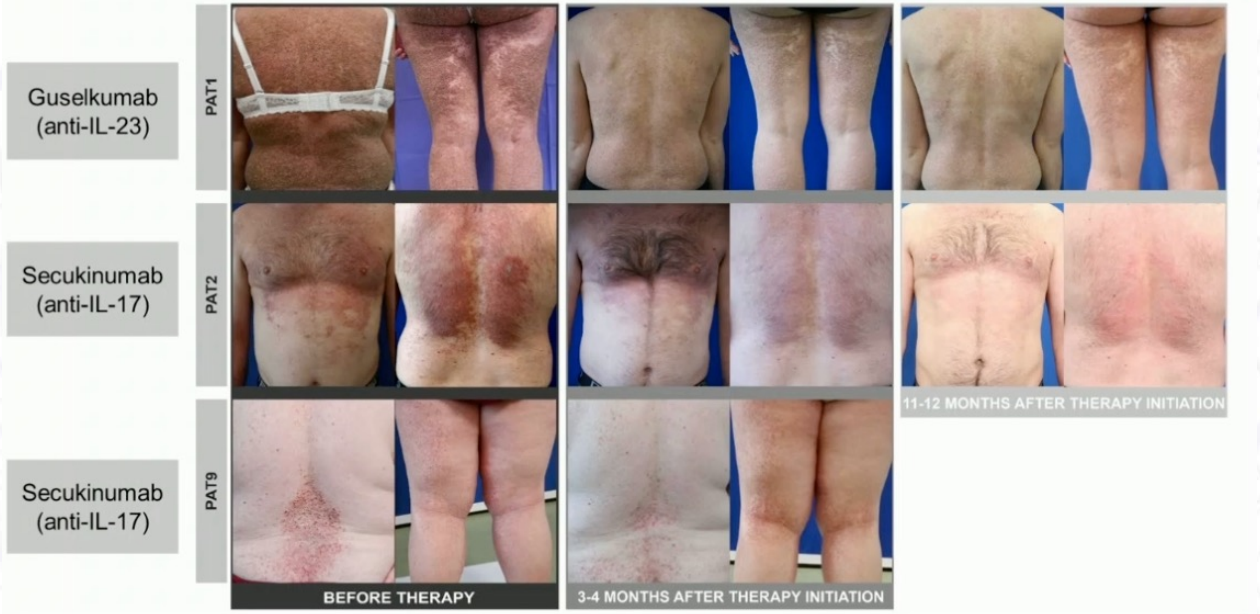
LINFOPROLIFERATIVOS



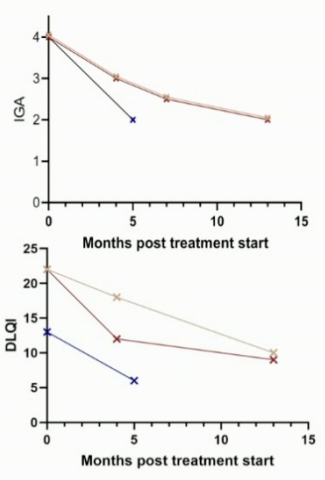
Increased expression of IL-17 signaling related genes in lesional DD skin

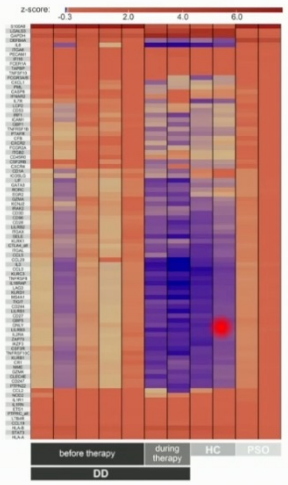


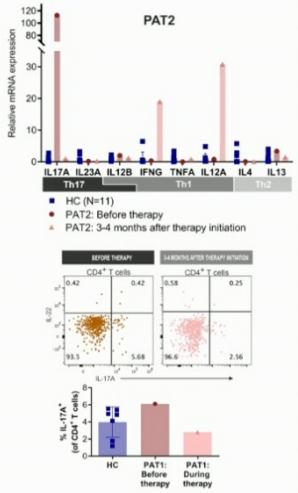
Successful therapy of DD patients with biologics



Successful therapy reduces Th17 cells in patients







Post-transplant lymphoproliferative disorder (PTLD)

Post-transplant lymphoproliferative disorder, monomorfic type, in a kidney transplant patient following administration of anti-influenza vaccine

Miriam Fernández-Parrado1, Jacques Alzoghby-Abi Chaker2, Angel Fernandez-Flores3, Daniel Nieto Rodríguez4, Elena Sendagorta4, Luis Miguel Valladares5 de 1 Hospital Universitario de Navarra, Dermatology, 2 Complejo Asistencial Universitario de León, Pathology, 3Hospital Universitario del Bierzo, Dermatopahology, Ponferrada, Spain, 4 Hospital Universitario La Paz, Dermatology, Madrid, Spain, 5 Complejo Asistencial Universitario de León, Dermatology, Leon, Spain

BERLIN EADV 11-14 OCTOBER 2023 EUROPEAN ACADEMY OF DERMATOLOGY AND VENEREOLOGY

INTRODUCTION

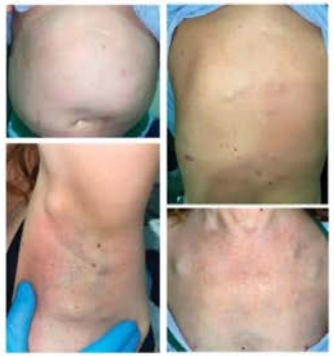
Post-transplant lymphoproliferative disorders (PTLD) are an uncommon complication in patients who have undergonesolidorgantransplantation. Their clinical presentation is often dramatic, leading to suspicion of aggressive lymphoma.

These proliferative disorders are unequally represented in the 2 main current classifications of lymphomas: the International Consensus Classification and the WHO classification (fifth Edition). In the International Consensus Classification, the group of immunodeficiency-associated lymphoproliferative disorders is recognized, which includes PTLD (including non destructive forms, polymorphic forms, monomorphic forms, and post-transplant Hodgkin lymphoma), and other iatrogenic lymphoproliferative disorders (Table1). In the fifth edition of the WHO classification, the monomorphic disorder is not reflected as such.

CLINICAL CASE

A 49-year-old woman came to the clinic with disseminated skin lesions that appeared day after receiving the influenza A. vaccine (1 doce of Vaxigrip) and increased in number for 2 weeks until the day of the consultation.The patient presented with B symptoms (with evening low-grade fever and nocturnal sweating).

On examination, multiple skin-colored nodules were observed, disseminated over the trunk, upper extremities, axillae, and the neck. The nodules were elastic, non erythematous, non pruritic, non painful, and not adherent to Deep planes. Ultrasound examination showed that they were subcutaneous nodules (Figure 1).



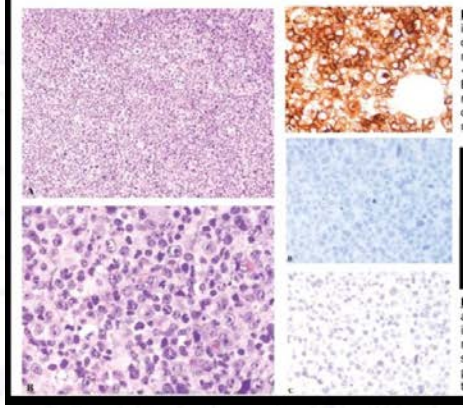


Figure 2. Histology A. Subcutaneous infiltration by a lymphoproliferative disorder composed of atypicall argeblastic monomorphic cells (Hematoxylin-eosin ·100).
B. At higher magnification, numerous mitoses and apoptotic cells were observed. Many cells had a prominent nucleolus (hematoxylin-eosin-400).

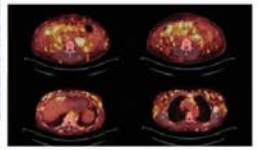


Figure 3. PET-scan showed numerous adenopathies with uptake at supra and infradiaphragmatic levels, subcutaneous nodules, involvement of the right submandibular gland, pancreas, both pleurae, bones, dorsal musculature, and the peritoneum.

DISCUSSION

We present a case of PTLD coinciding with an antiviral vaccine for influenza. This is a very rare event, and we are only a ware of occasional similar cases with the influenza vaccine and with the anti-COVID-19 vaccine in the recent pandemic. There is also a published case of paradoxical response to an influenza vaccine in which the patient (a 27 year-old man) developed an aggressive lymphoid proliferation of Tcells, EBER-negative, which started at the vaccine site, but extended and killed the patient. Unlike our case, the one described by these authors was CD30-negative.

Cases of pseudolymphomas and various lymphoid proliferations in the áreas of injection of the influenza vaccine are not uncommon.

In all these cases of local response, pseudolymphomatous reactions are confined to the cutaneous área of the vaccine injection site or to regional lymph nodes. In contrast, in our case, a disseminated lymphoproliferative disorder developed. Cutaneous involvement occurs in only 5% of PTLD cases, with most cases being monomorphic (more frequently B-cell than T-cell).

Cutaneous involvement occurs in only 5% of PTLD cases,

TABLE 1. Classification of Lymphoproliferative Disorders Associated With Immunodeficiency, According to the International Consensus Classification

Immunodeficiency-Associated Lymphoproliferative Disorders
Post-transplant lymphoproliferative disorders
Nondestructive post-transplant lymphoproliferative disorders
Plasmacytic hyperplasia post-transplant lymphoproliferative disorder
Infectious mononucleosis post-transplant lymphoproliferative disorder
Florid folicular hyperplasia post-transplant lymphoproliferative disorder
Polymorphic post-transplant lymphoproliferative disorder
Monomorphic post-transplant lymphoproliferative disorder
B-cell type
T-cell type
Classic Hodgkin lymphoma post-transplant lymphoproliferative disorder
Other iatrogenic immunodeficiency-associated lymphoproliferative disorders

with most cases being monomorphic (more frequently B-cell than T-cell).

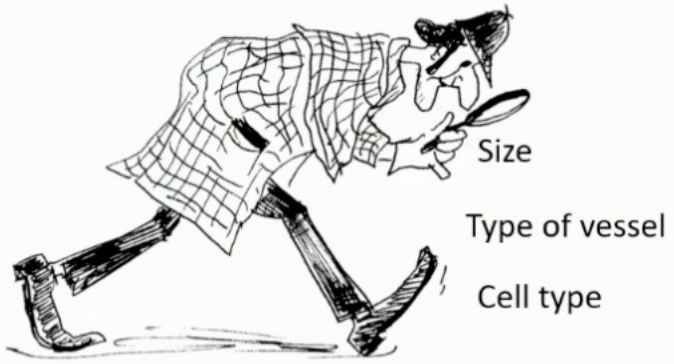
Patients with solid organ transplantation usually receive the influenza vaccine annually. It is worth asking why this type of lymphoid proliferation after influenza vaccination is not more common in these patients. One of the reasons is the low immunogenicity of the type of vaccine used. Thus, for example, the vaccine used in our case (Vaxigrip) is made of inactivated Split virus and is characterized by inducing a lower intensity and duration of immune response tan other types of vaccines.

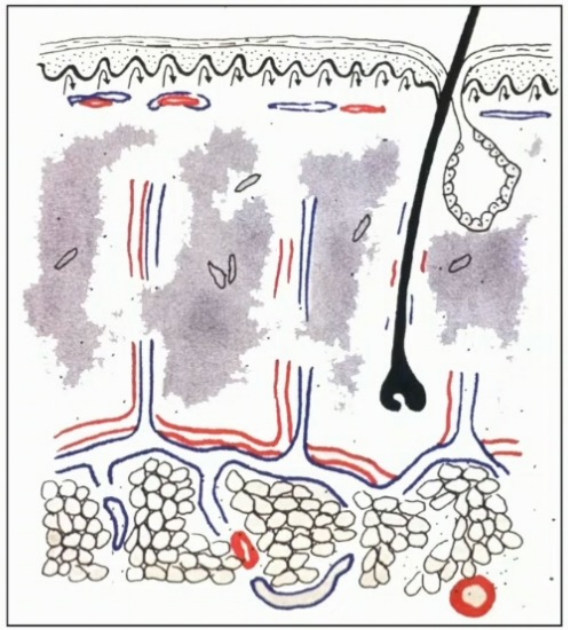
REFERENCES

VASCULITIS

Marc-Cyriel-Marie Haspeslagh
Vasculitis and vasculopathies

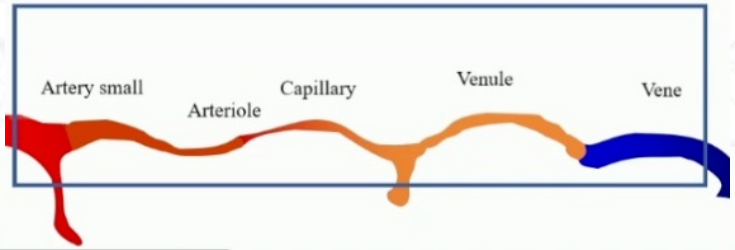
5-skin limited vasculitis or systemic vasculitis?





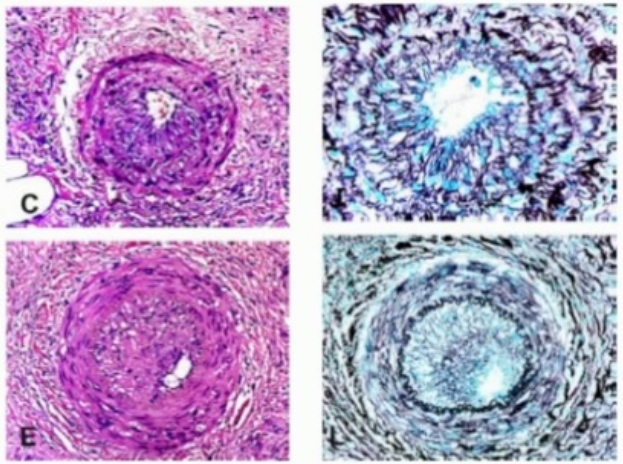
VASCULITIS SMALL VESSELS

VASCULITIS MEDIUM VESSELS



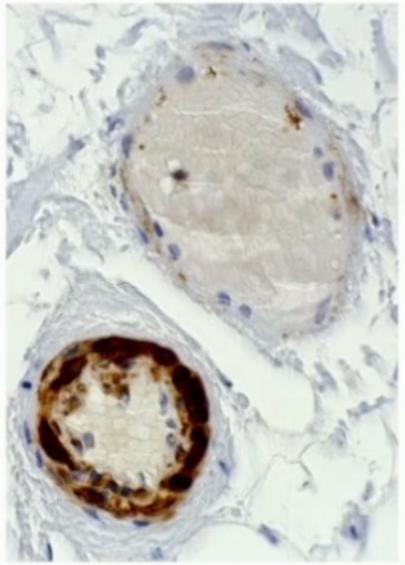
Vasculitis in erythema induratum of Bazin: A histopathologic study of 101 biopsy specimens from 86 patients

Sonia Segura, MD,[a] Ramón M. Pujol, MD,[a] Felicidade Trindade, MD,[b] and Luis Requena, MD[b]
Barcelona and Madrid, Spain



ELA - SMA

Arteriole and venule



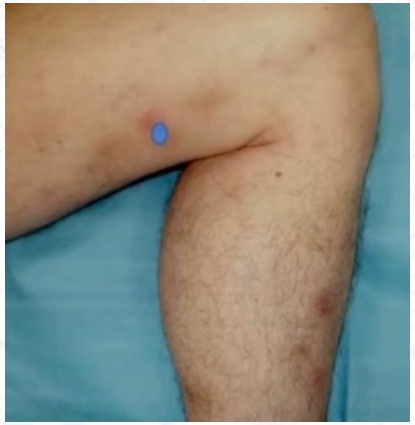
Lower leg veins of the connective tissue septa of the subcutis show a thicker and more compact muscular layer than the veins of the subcutis in other areas of the skin and they often are misinterpreted as arteries.[23]

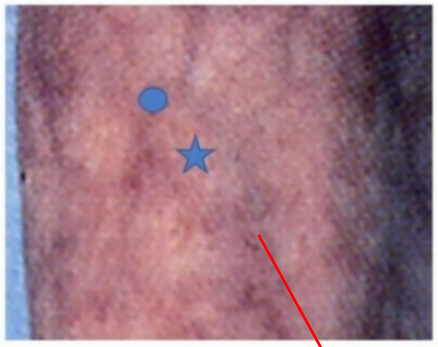
VASCULITIS DE MEDIANO VASO

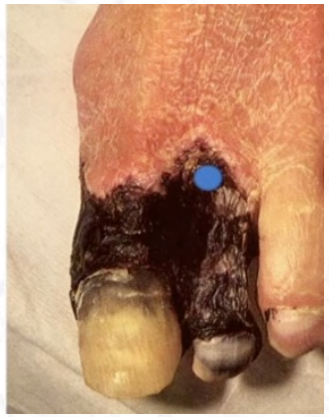
Nódulos

Livedo reticularis

Infarto/gangrena acral

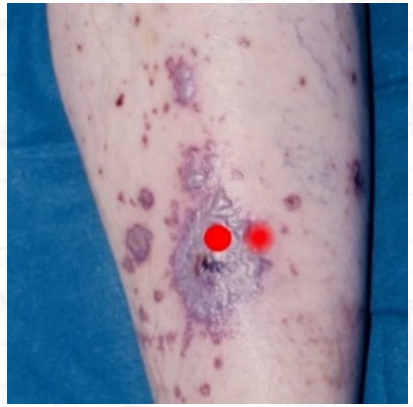


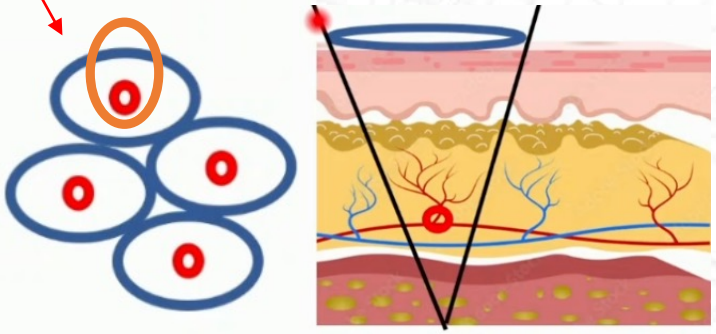




VASCULITIS DE PEQUEÑO VASO

Púrpura palpable





Vasculitis de pequeño vaso

- **VLC**:
  - Término histológico.
  - ¿Predictores de IgA -? Siempre IFD.

J Cutan Pathol. 2023 Jul;50(7):681-686. doi: 10.1111/cup.14436. Epub 2023 May 7.

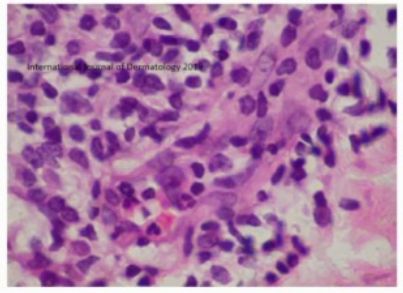
Histopathologic features predictive of perivascular deposition of IgA on direct immunofluorescence in cases of leukocytoclastic vasculitis: A retrospective study of 112 specimens

Fangyi Xie [1], Emma F Johnson [1 2], David A Wetter [1], Michael J Camilleri [1 2], Austin Todd [3], Julia S Lehman [1 2]

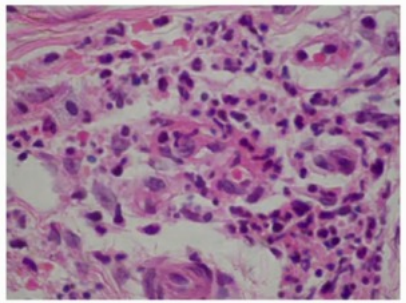
	IgA+ (n = 56)	IgA− (n = 56)	*p* Value
Epidermal spongiosis	14 (25.0%)	12 (21.4%)	0.654[a]
Dermal edema	38 (67.9%)	37 (66.1%)	0.841[a]
Subepidermal separation			0.697[a]
None	34 (60.7%)	38 (67.9%)	
Focal	4 (7.1%)	4 (7.1%)	
Extensive	18 (32.1%)	14 (25.0%)	
Neutrophils present	56 (100.0%)	56 (100%)	1.0[a]
Eosinophils present	31 (55.4%)	41 (73.2%)	**0.049**[a]
Mid and deep dermal inflammation—perivascular inflammation in lower 2/3 of the dermis	14 (25.0%)	28 (50.0%)	**0.006**[a]

Vasculitis de pequeño vaso

- VLC:
  - Término histológico.
  - ¿Predictores de IgA -? Siempre IFD.
- **Urticaria-vasculitis**:
  - **Normo-C'** (+frec): linfocítica.
  - **Hipo-C'** (anti-C1q): neutrofílica.



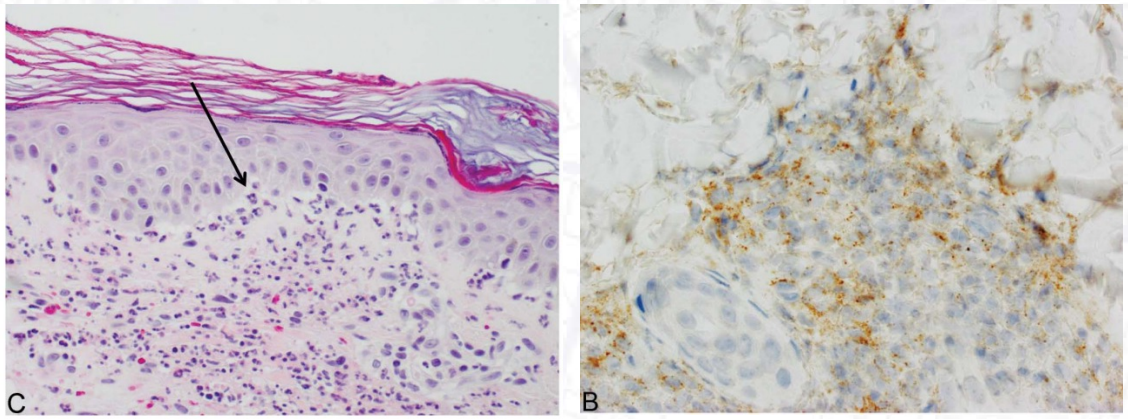
The lymphocyte predominant group constitutes the majority, are normocomplementemic



Neutrophilic predominant hypocomplementemic

› Am J Dermatopathol. 2020 Jun;42(6):399-406. doi: 10.1097/DAD.0000000000001501.

Dermal C4d Deposition and Neutrophil Alignment Along the Dermal–Epidermal Junction as a Diagnostic Adjunct for Hypocomplementemic Urticarial Vasculitis (Anti-C1q Vasculitis) and Underlying Systemic Disease



C4d

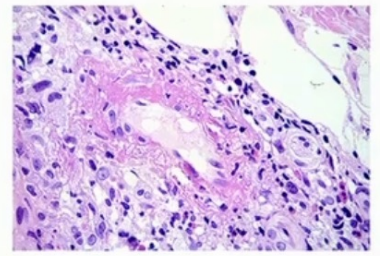
*_Pearl_: alineación de PMNs en MB y depósito de C4d en dermis

Vasculitis de pequeño vaso

- VLC:
  - Término histológico.
  - ¿Predictores de IgA -? Siempre IFD.
- Urticaria-vasculitis:
  - **Normo-C'** (+frec): linfocítica.
  - **Hipo-C'** (anti-C1q): neutrofílica.
- VVP IgM/IgG similar a Schönlein-Henoch.
- **MIS-C** en contexto de COVID.

MIS-C : multisystem inflammatory syndrome in children with COVID

Severely ill children few weeks after corona infection.
Dermatologic manifestations are one of the hallmark findings of MIS-C, affecting roughly 74% of children diagnosed with the condition.
A study by Young et al classifies skin lesions found in children with MIS-C into four categories: morbilliform lesions, reticulated lesions, scarlatiniform lesions, and urticarial lesions.



LCV

Corona vaccination

Small-vessel vascular injury was seen in two specimens, which were diagnosed as urticarial vasculitis and leukocytoclastic vasculitis (IgA), respectively.

Received: 12 October 2022 | Revised: 29 March 2023 | Accepted: 10 May 2023
DOI: 10.1002/jvc2.174

CASE REPORT

JEADV CLINICAL PRACTICE

Cutaneous leukocytoclastic vasculitis following COVID-19 vaccination

Manuel Zoppi[1] | Victoria Gandara[1] | Jorge Zoppi[2]

NOTES & COMMENTS

Response to Berry et al's "Cutaneous small-vessel vasculitis following single-dose Janssen Ad26.COV2.S vaccination"

Association of COVID-19 antigenicity with the development of antineutrophilic cytoplasmic antibody vasculitis

Jamie R. Felzer[1] | Delvise T. Fogwe[1] | Shaher Samrah[2] | Clement J. Michet Jr[3] | Ulrich Specks[4] | Misbah Baqir[4] | Aahd F. Kubbara[4]

Corona disease

Received: 5 September 2021 | Accepted: 13 October 2021
DOI: 10.1111/1346-8138.16211

CONSICE COMMUNICATION

JOURNAL OF DERMATOLOGY

Coronavirus disease 2019-associated immunoglobulin A vasculitis/Henoch–Schönlein purpura: A case report and review

Patrick M. Jedlowski | Mahdieh F. Jedlowski

SVV part of Multisystem inflammatory disease in children

Urticarial vasculitis

Lymphocytic vasculitis pernio type

CASE REPORT

A case of generalized Sweet syndrome with vasculitis triggered by recent COVID-19 vaccination

Neha Kinariwalla, MPhil,[a] Ashley O. London, MS,[a] Yssra S. Soliman, MD,[b] George W. Nicc
Sameera Husain, MD,[b] and Stephanie M. Gallitano, MD[b]
New York, New York

Vasculitis de mediano vaso

POLYARTERITIS NODOSA

SYSTEMIC PAN

medium vessels : skin infarction, livedo reticularis, intradermal nodules, and deep ulcers

purpura, hemorrhagic macules, erythema, and hemorrhagic blisters have also been reported

→ Subgroup of PAN with a component of small vessel vasculitis (with likely involvement of venules).

Only SKIN PAN

small arteries and arterioles in the panniculus and the dermal-subcutaneous junction

not directly involve veins

livedo reticularis, intradermal nodules, and ulcers

SVV

Déficit de ADA-2

The average age at presentation to the NIH was 15.95 years

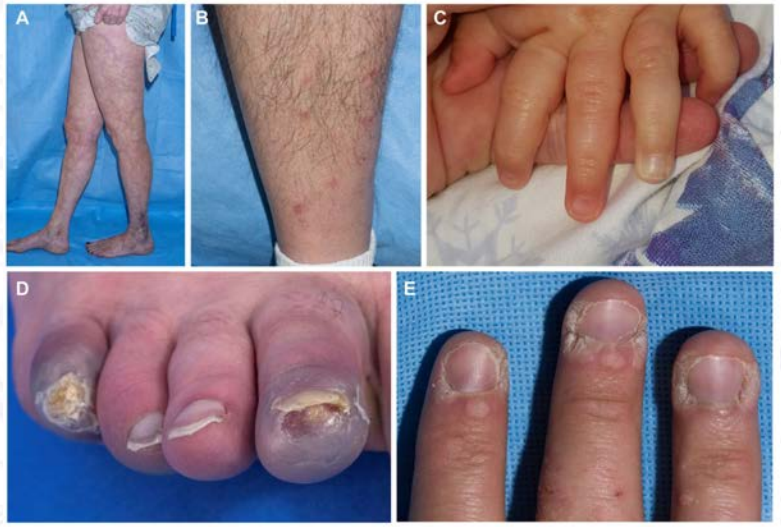
It mimics PAN, and has to be excluded escpecially in children with PAN or familial PAN cases.

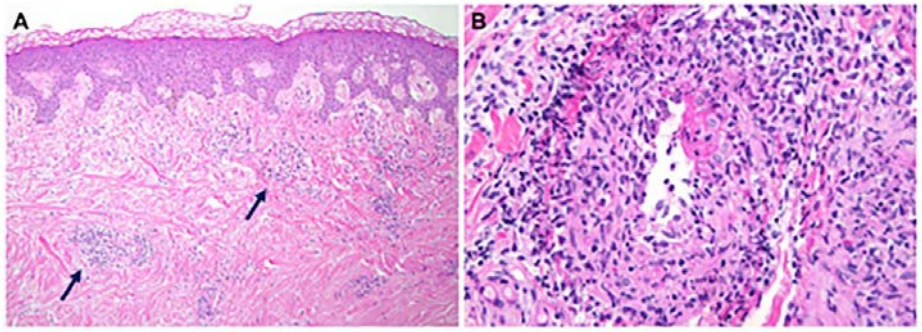
Phenotypic variability was noted among affected family members
Skin involvement in 90% and livedo racemose in 74%

Diagnosis : ADA2 enzyme activity or genetic mutation

Front Immunol. 2021; 12: 811473.
Published online 2022 Jan 10. doi: 10.3389/fimmu.2021.811473
PMCID: PMC8790931
PMID: 35095905

The Spectrum of the Deficiency of Adenosine Deaminase 2: An Observational Analysis of a 60 Patient Cohort

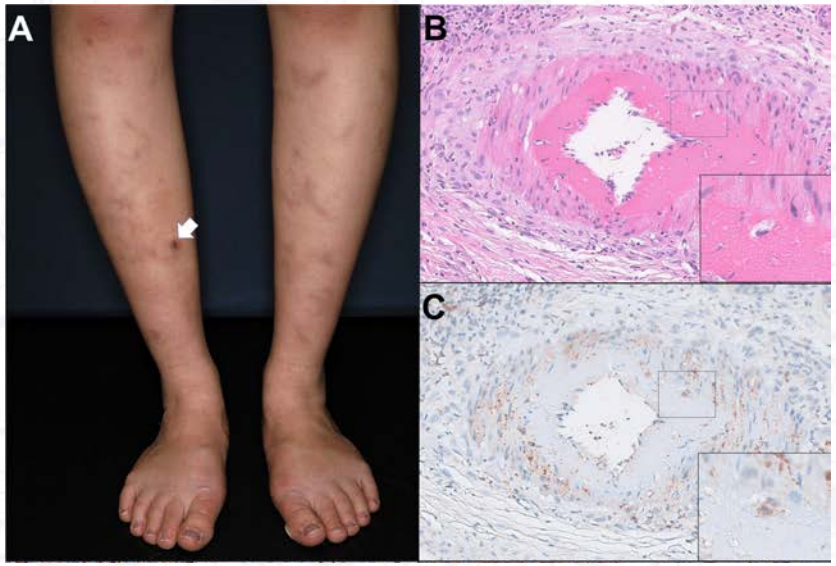




SVV

> Virchows Arch. 2023 Jun;482(6):1079-1083. doi: 10.1007/s00428-023-03531-8.
Epub 2023 Mar 24.

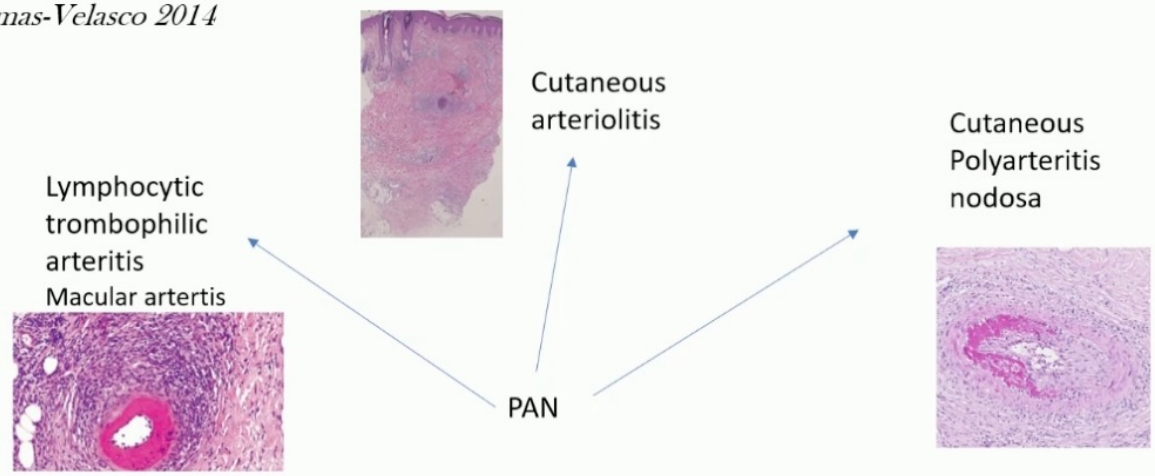
Cutaneous arteritis with intimal fibrin ring and immature myeloid cell infiltrate: lymphocytic thrombophilic arteritis or histiocytoid polyarteritis nodosa?



MPO+

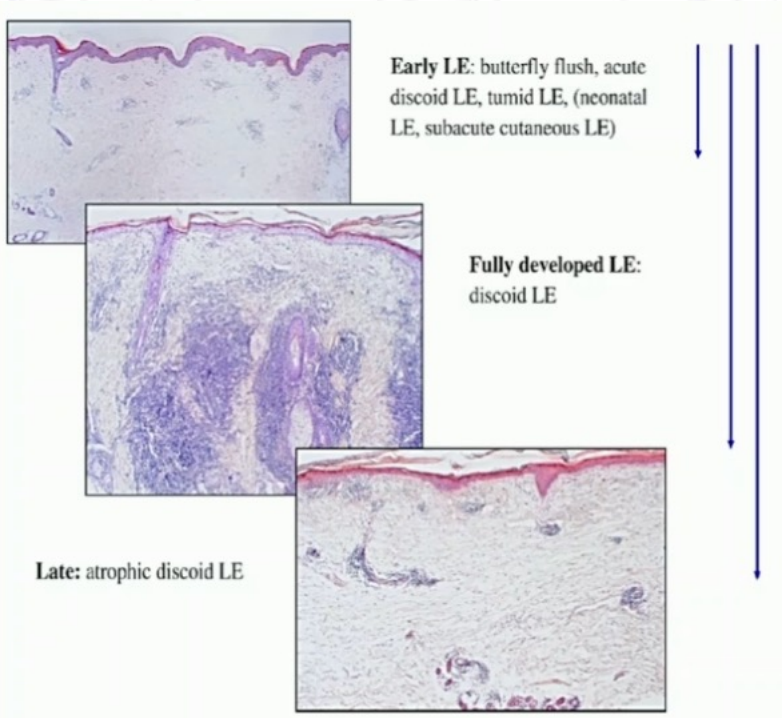
Clinicopathologic spectrum that exhibits increasing severity from (cutaneous arteriolitis), macular arteritis to lymphocytic thrombophilic arteritis and, cutaneous PAN

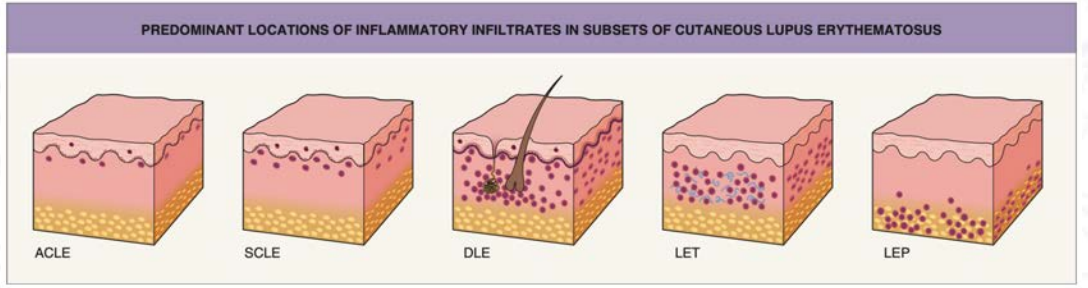
Mar Llamas-Velasco 2014



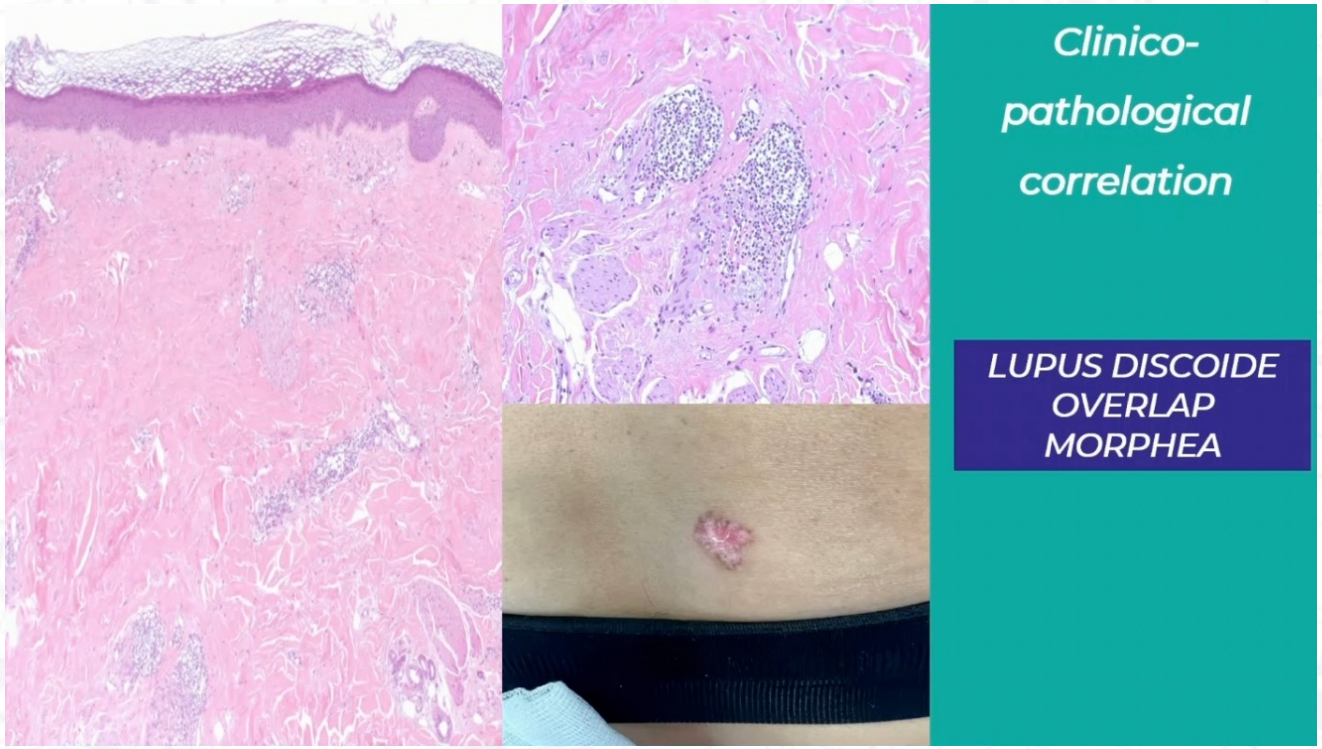
ENFERMEDADES DEL TEJIDO CONECTIVO

LUPUS





- LE has a **broad spectrum** of histological signs which are related to the stages of the lesions
- Clinical subsets of LE (except LE profundus) show an **extensive overlap** in their **histological features**
- Not every case of LE can be allocated to a subset because **transitions** from one type to another as well as **intermediate forms** occur
- For accurate subtyping a ***CLINICO-PATHOLOGICAL CORRELATION*** is thus indispensable.


Clinico-
pathological
correlation
LUPUS DISCOIDE
OVERLAP
MORPHEA

AP de LES

Review > Indian J Dermatol Venereol Leprol. 2021 Jan-Feb;87(1):3-13.
doi: 10.25259/IJDVL_638_19.

Diagnostic utility of plasmacytoid dendritic cells in dermatopathology

Tara Bardawil [1], Samar Khalil [1], Mazen Kurban [1], Ossama Abbas [1]

CD123 +

- ✓ The *NUMBER* and *DISTRIBUTION of PDCs* differ markedly in inflammatory and neoplastic diseases
- ✓ *CD123+ PDC clusters* seem to be highly specific for *CLE*

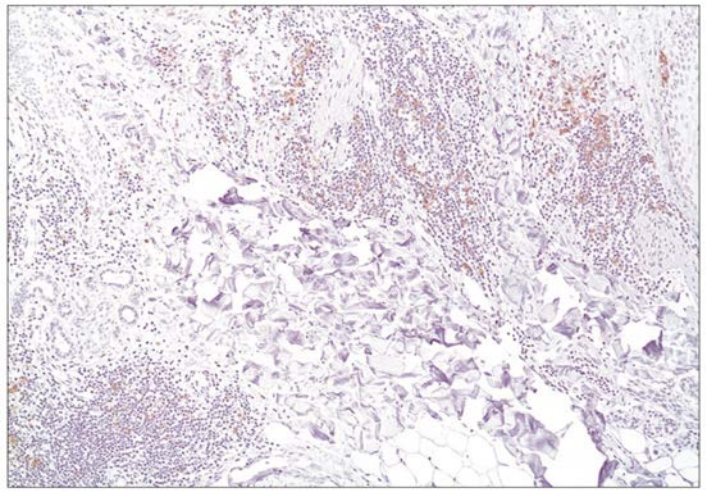


Figure 2d: Numerous plasmacytoid dendritic cells in clusters and as single cells, in superficial and deep locations in cutaneous lupus erythematosus (×100)

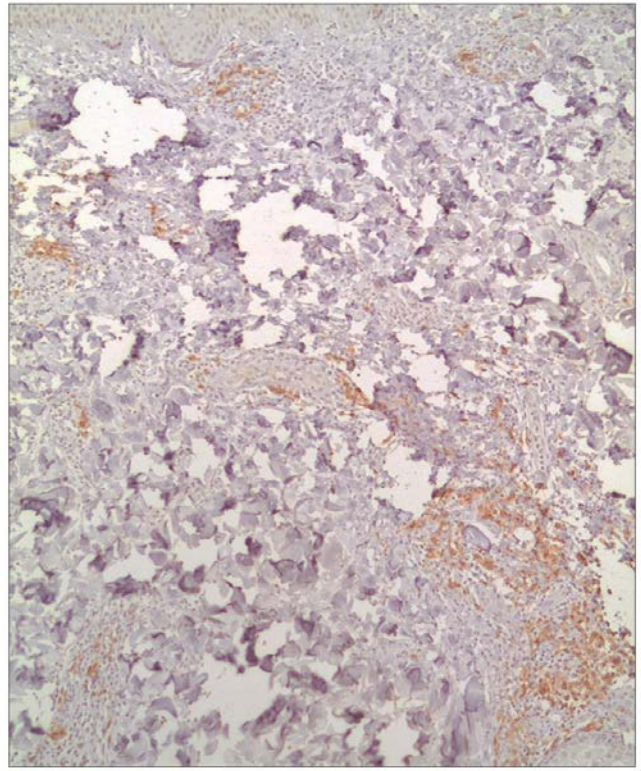


Figure 3d: Numerous plasmacytoid dendritic cells in clusters and as single cells, in superficial and deep locations in cutaneous lupus erythematosus (×100)

Otras entidades

- Morfea generalizada 2ª a radiodermitis
- Dermatomiositis paraneoplásica
- Nódulo reumatoide
  - ¿Colonización? por ***Leishmania major***.
  - Literatura = 2 casos previos.

Multicenter Study > J Am Acad Dermatol. 2019 Jul;81(1):260-262.
doi: 10.1016/j.jaad.2019.02.039. Epub 2019 Feb 21.

Radiation-induced morphea: Association with autoimmune comorbidities, severity, and response to therapy

Medicine (Baltimore). 2020 Aug 21; 99(34): e21733.
Published online 2020 Aug 21. doi: 10.1097/MD.0000000000021733
PMCID: PMC7447459
PMID: 32846794

Malignancy in dermatomyositis

A retrospective paired case–control study of 202 patients from Central China

Lili Chang, MM,[a,d] Lina Zhang, MD,[b] Haiquan Jia, MM,[c] Zhiyong Nie, MD,[a] and Lei Zhang, MD[d,*]

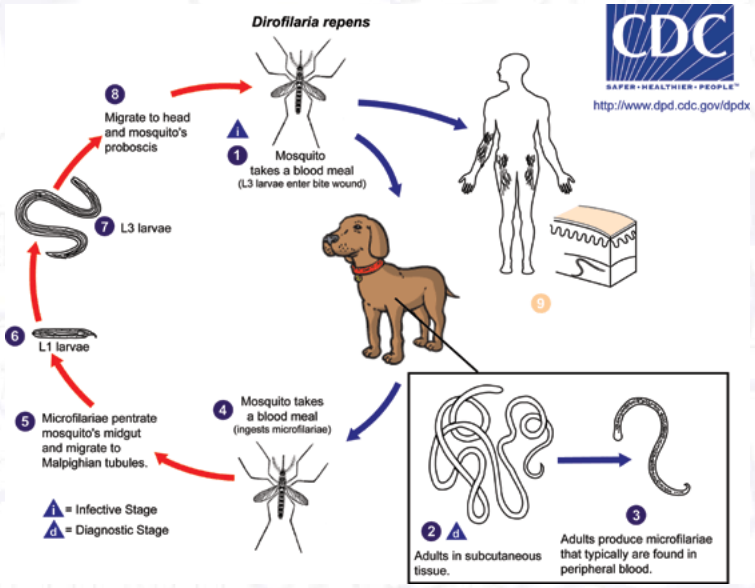
CPD

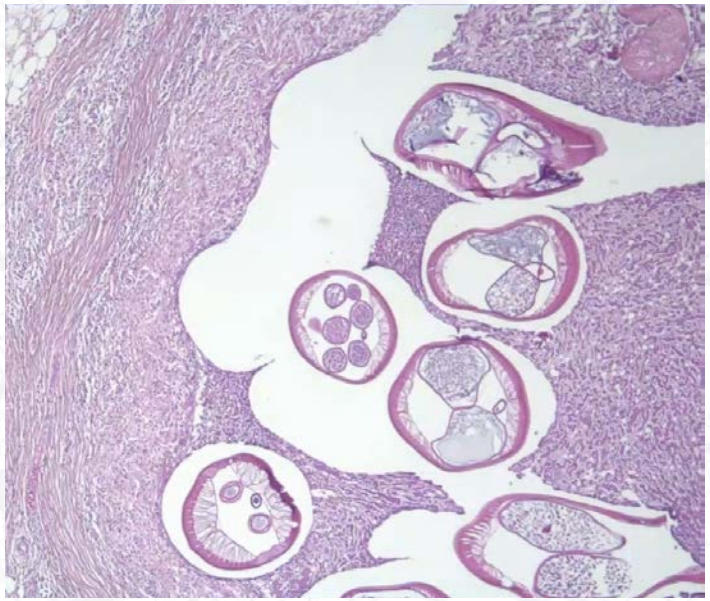
Cutaneous leishmaniasis mimicking dactylitis in a patient with rheumatoid arthritis treated with certolizumab

J. Herrerias-Moreno,[1] V. Expósito-Serrano,[1] E. Agut-Busquet,[1] M. Corbacho,[1] E. Sáez[2] and J. Luelmo[1]

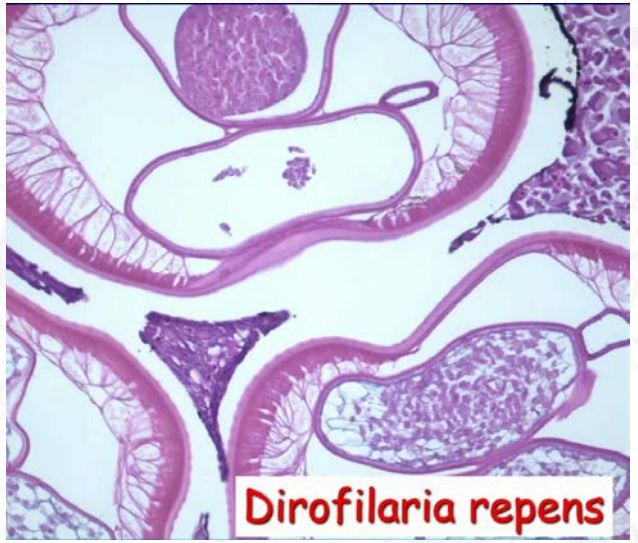
Departments of [1]Dermatology and [2]Anatomy–Pathology, Corporació Sanitària Parc Taulí, Hospital Universitari de Sabadell, Universitat Autónoma de Barcelona, Sabadell, Spain

INFECCIONES EMERGENTES





- Nódulo subcutáneo.
- Larva viva.
- Dx: AP.

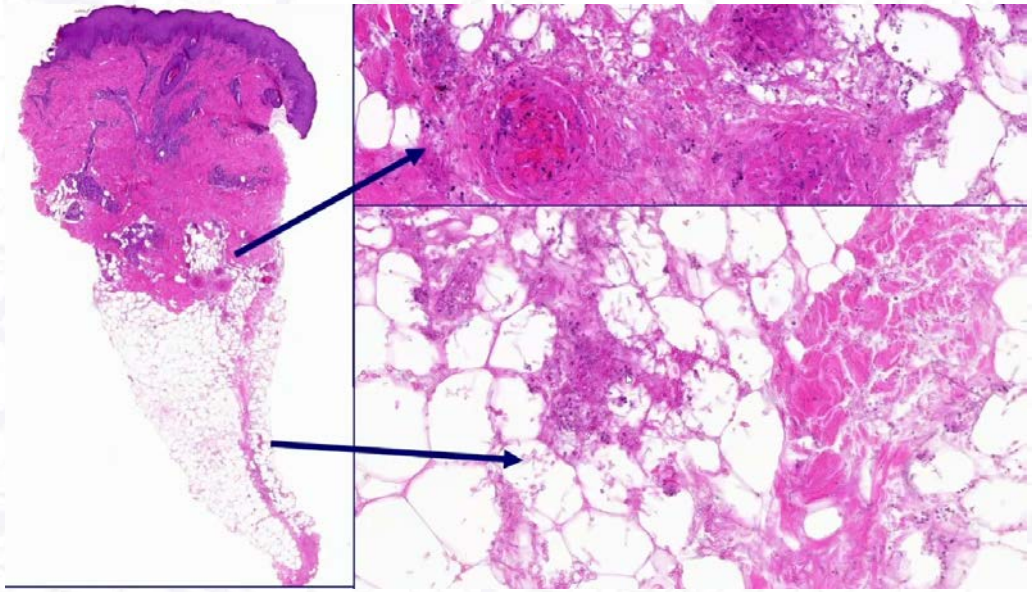


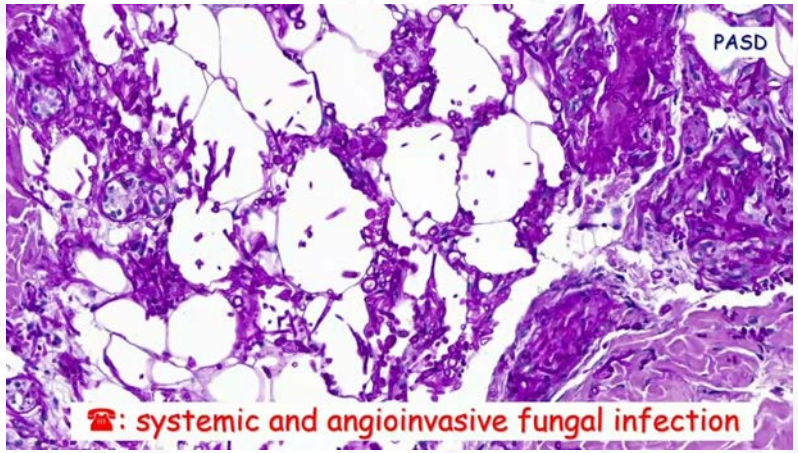
5 yo, AML relapse, QT 2 weeks prior, papules, fever

Angioinvasive fungal infections

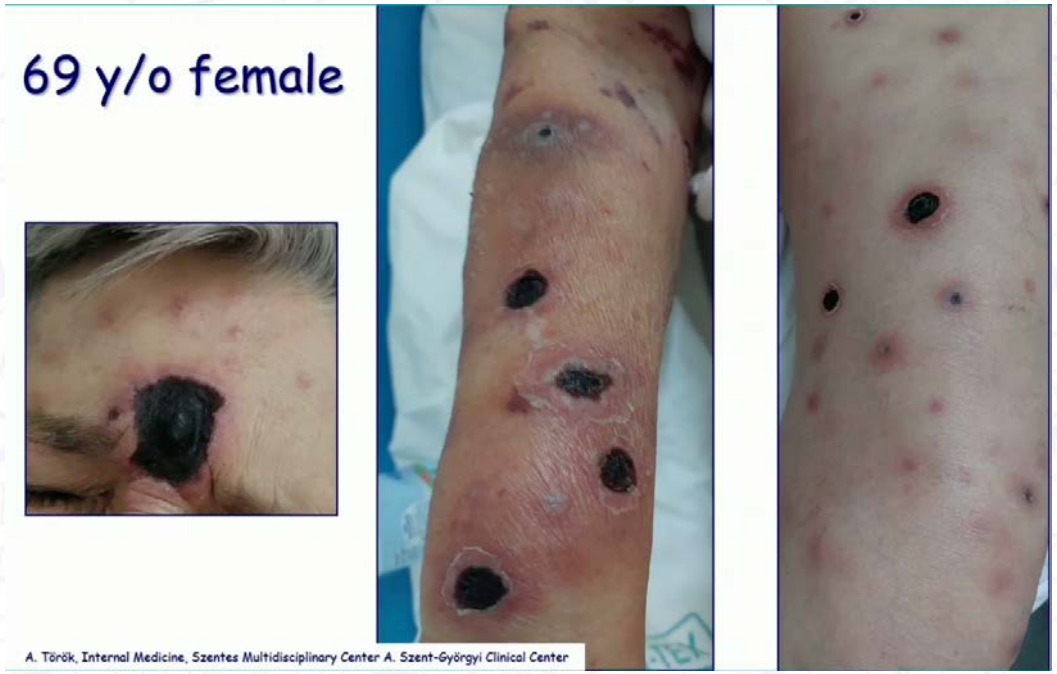
- immunosuppressed patients
- biopsies: fungal culture (multiple), histopathology
- fungal hyphae in the tissue and/or vessels, necrosis
- HE, PAS, GMS
- specification cannot be done on histopathology alone
- pitfalls in characterizing the exact fungus
- septate (Aspergillus, Fusarium, Scedosporium)
- nonseptate (class Mucormycetes: Rhizopus spp, Absidia spp, Mucor spp)
- first-line for Mucormycetes: liposomal amphotericin B
- first-line for Aspergillus: voriconazole

Arch Pathol Lab Med 2019

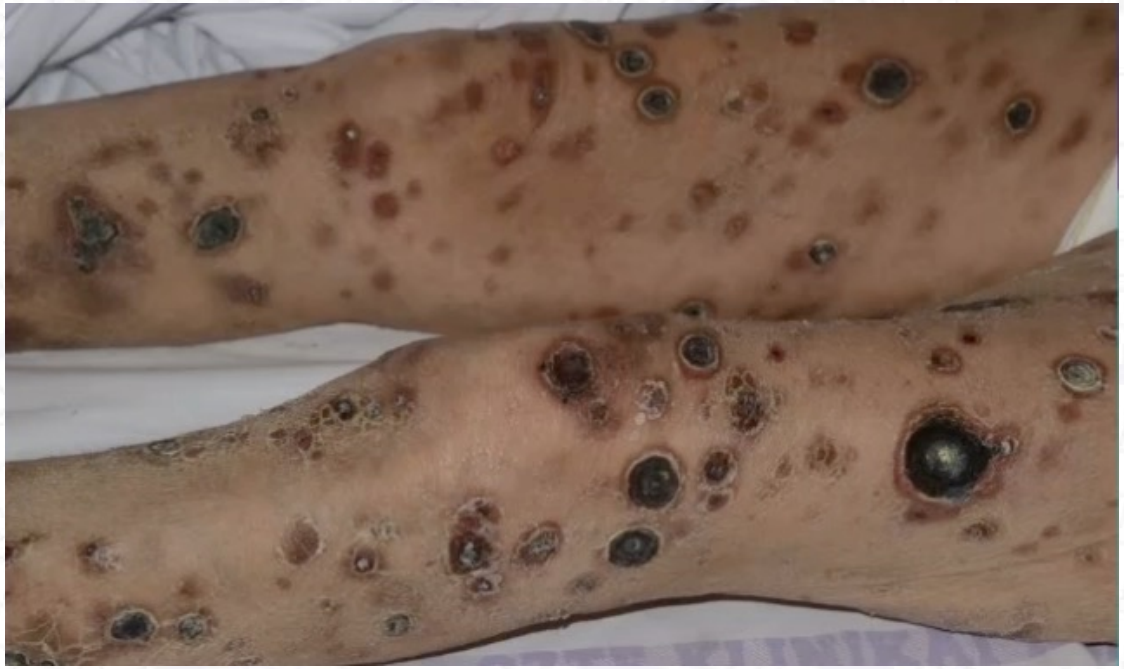




69 yo, Ibrutinib for B-CCL, Mieloma, Sjögren



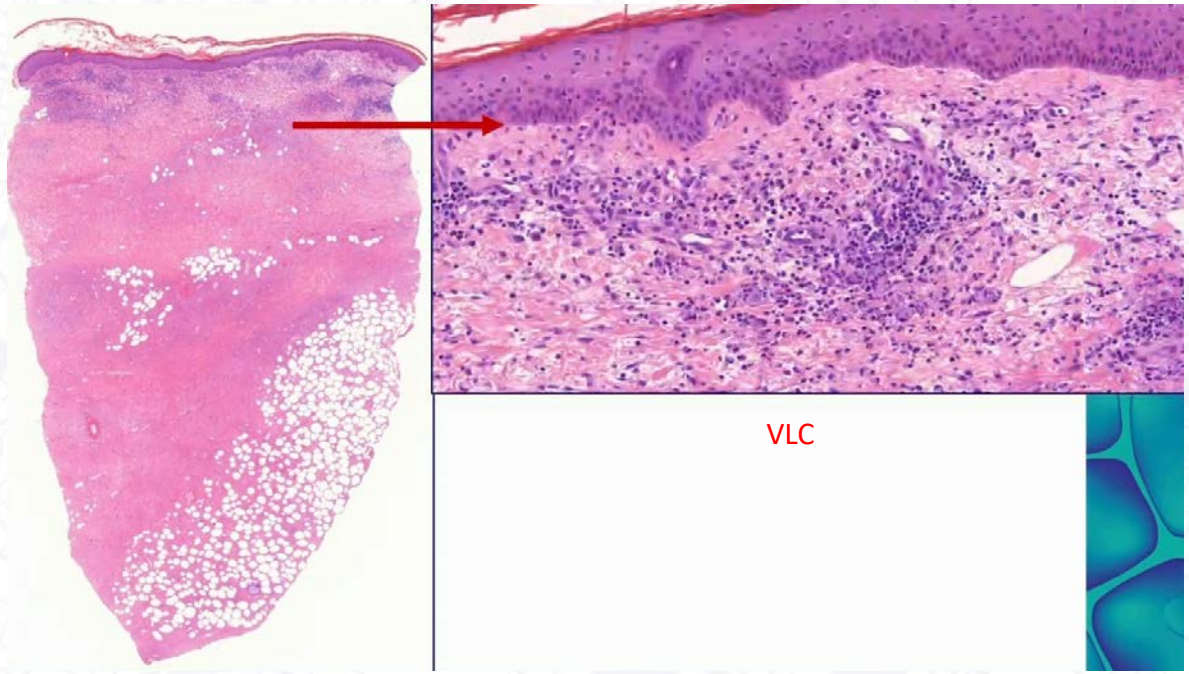
Conjuntivitis crónica previa



¿PLEVA?

69 yo, Ibrutinib for B-CCL, Mieloma, Sjögren

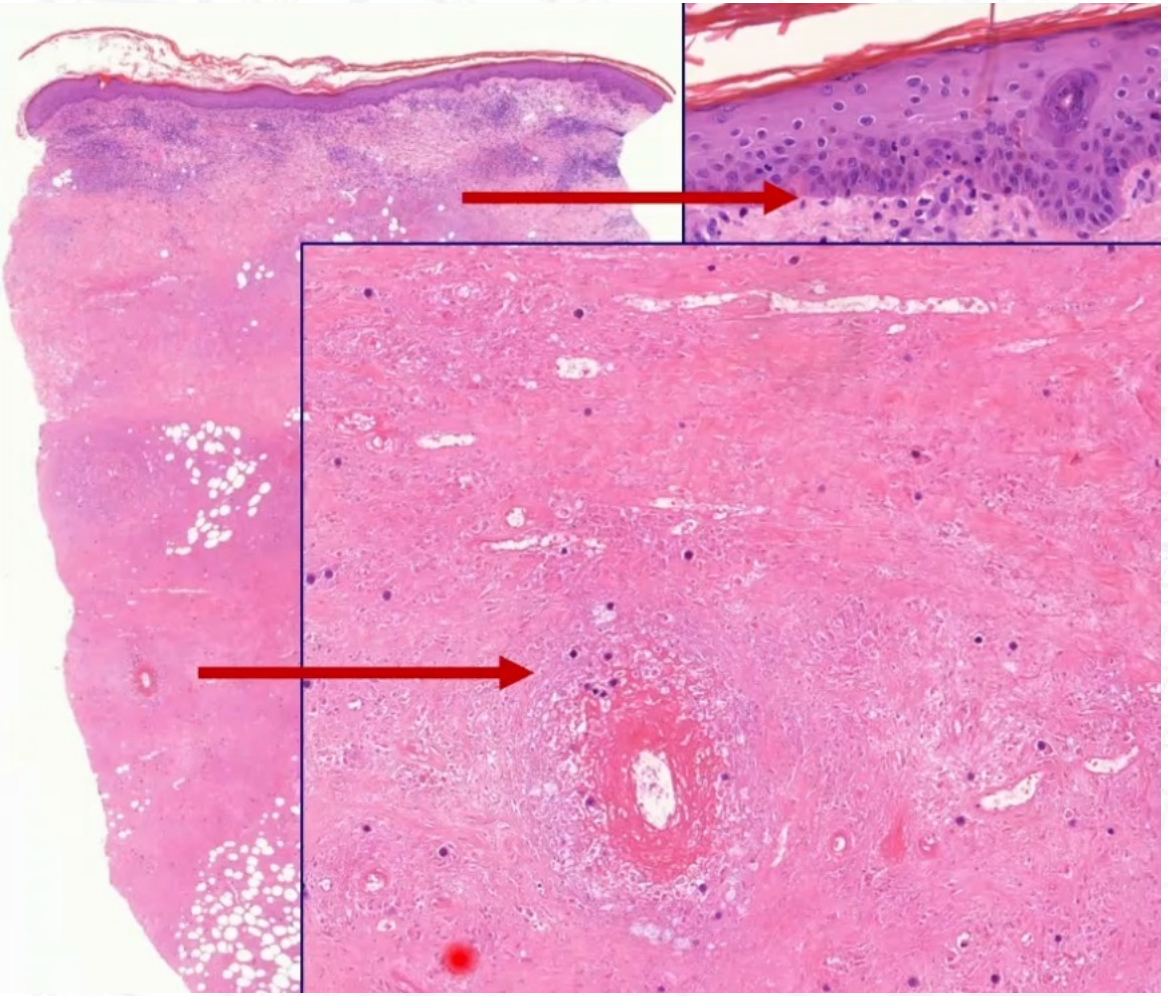
Conjuntivitis crónica previa



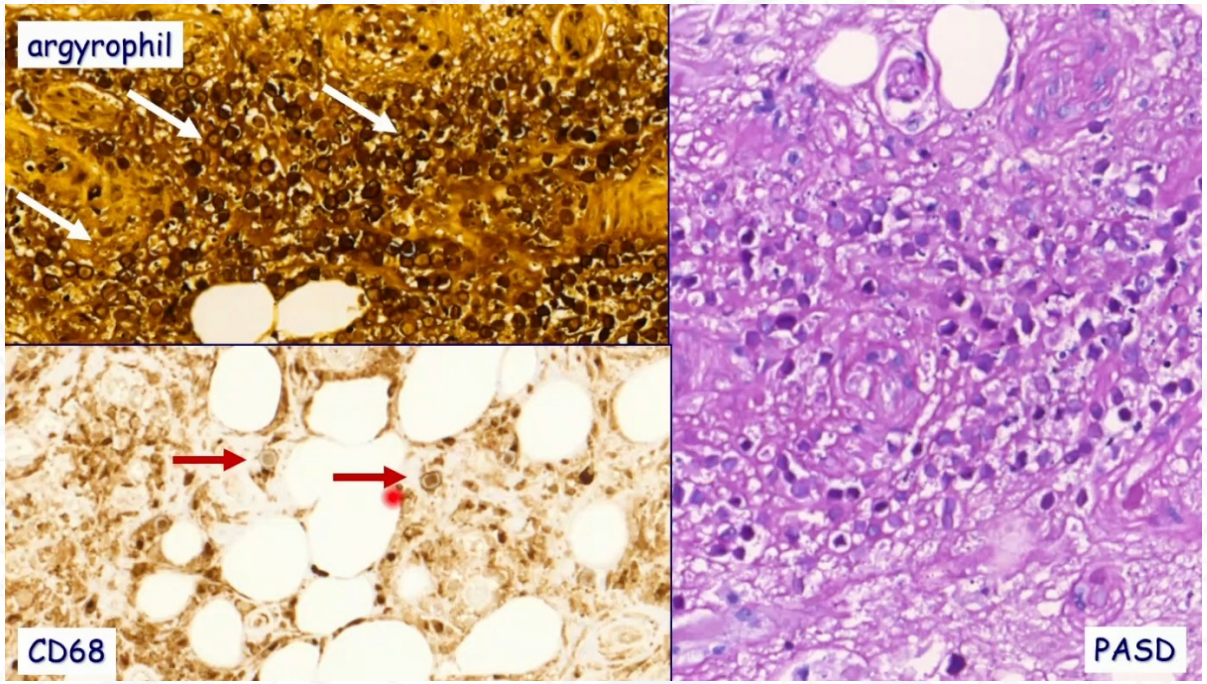
VLC

69 yo, Ibrutinib for B-CCL, Mieloma, Sjögren

Conjuntivitis crónica previa



69 yo, Ibrutinib for B-CCL, Mieloma, Sjögren



Conjuntivitis crónica previa

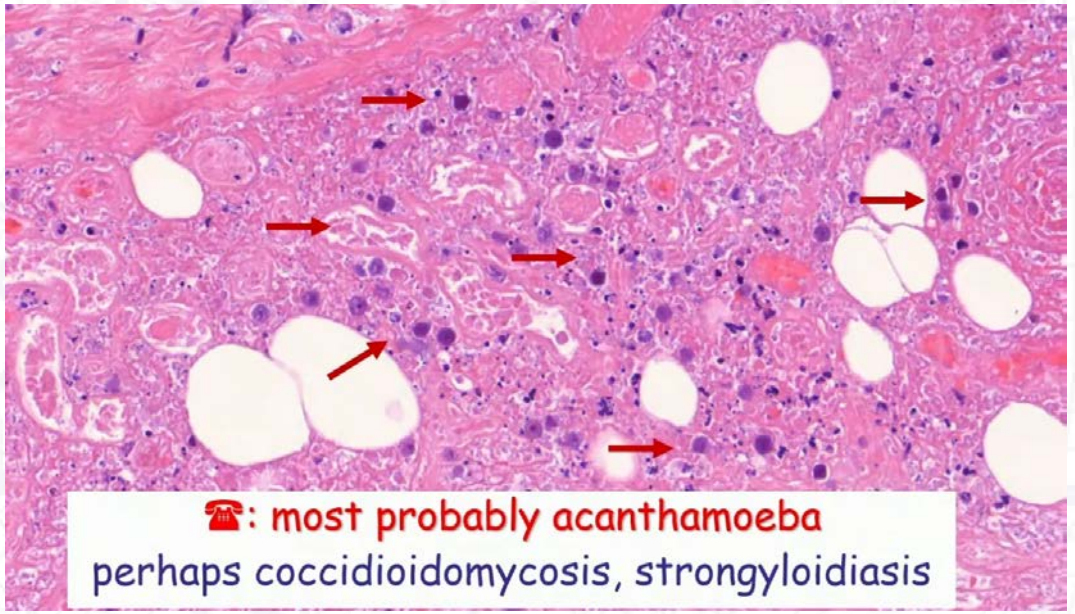
Diseases caused by Acanthamoeba

Cutaneous and mucosal infections
- multiple lesions
- nodular, ulcerated, necrotic, papulo-pustular

Keratitis
- rising incidence due to contact lens usage!!!

Central nervous system
- granulomatous amoebic encephalitis (GAE)
- primary amoebic meningoencephalitis (PAM)

Pneumonitis, rhinosinusitis

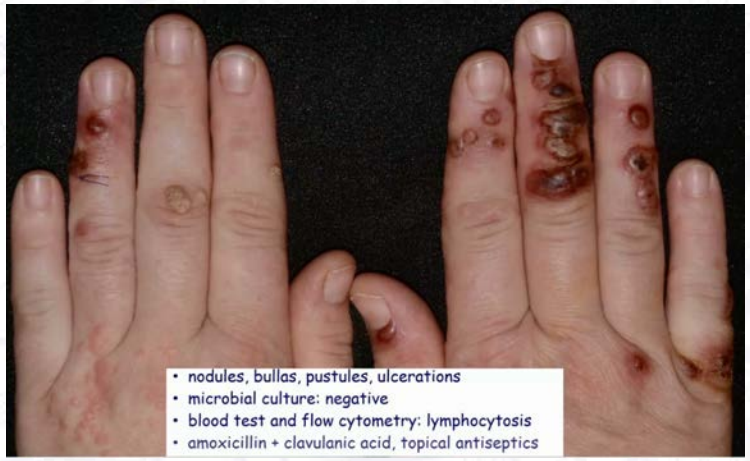


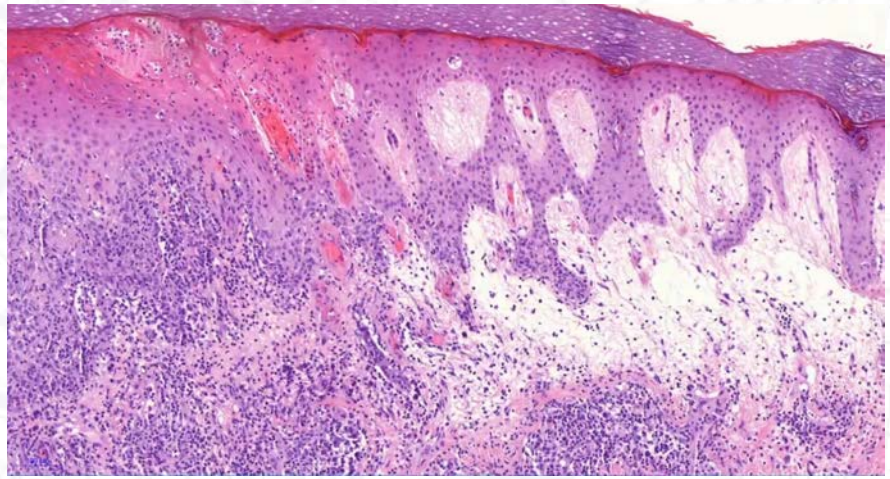
- early recognition, prompt diagnosis
- rapid initiation of treatment
- combination therapy: antibiotics, antifungals, antiprotozoal agents
- miltefosine, amphotericin B, metronidazole, ceftriaxone, rifampin, isoniazid, trimethoprim-sulfamethoxazole, fluconazole, flucytosine, azithromycin…

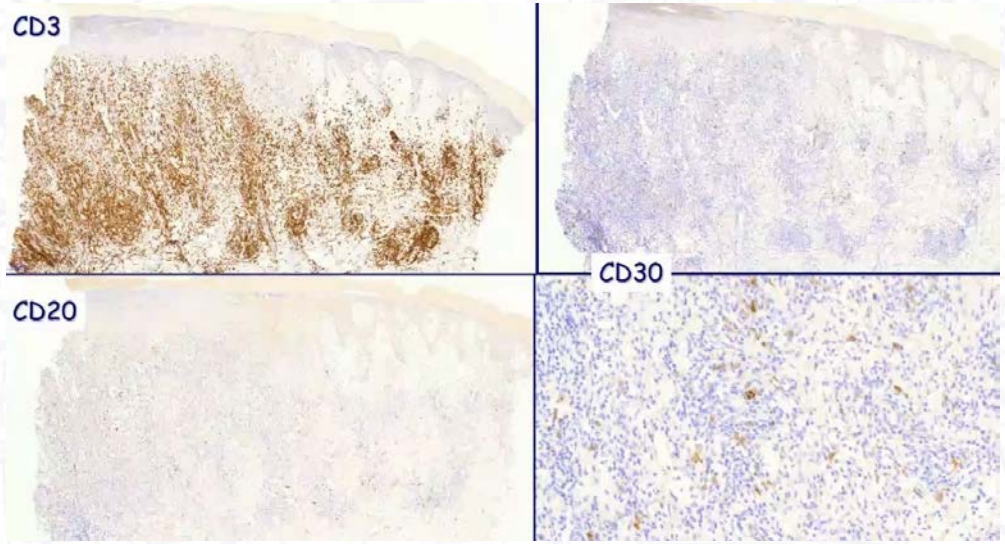
Acanthamoeba PCR: positive, sequencing: T4 genotype

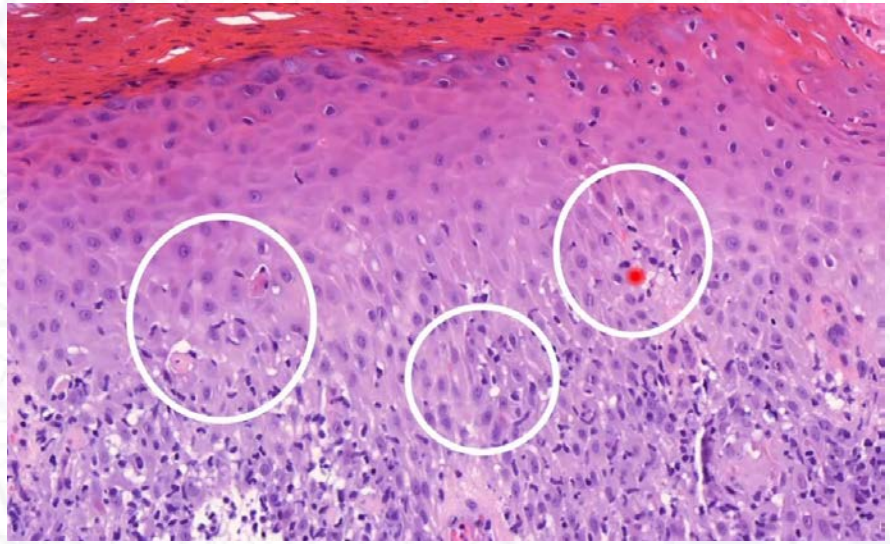
G. Terhes, Insitute of Clinical Microbiology

40 yo, cattle farm worker, bitten by calf 1 wk prior





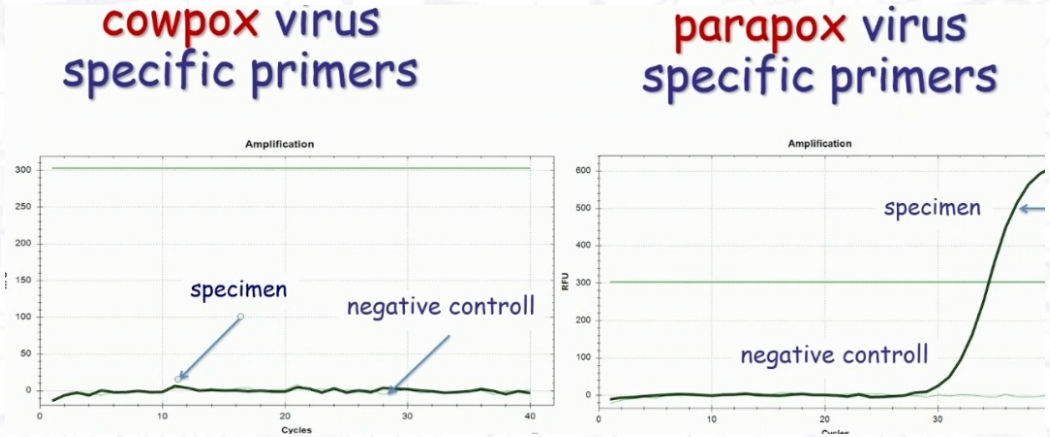


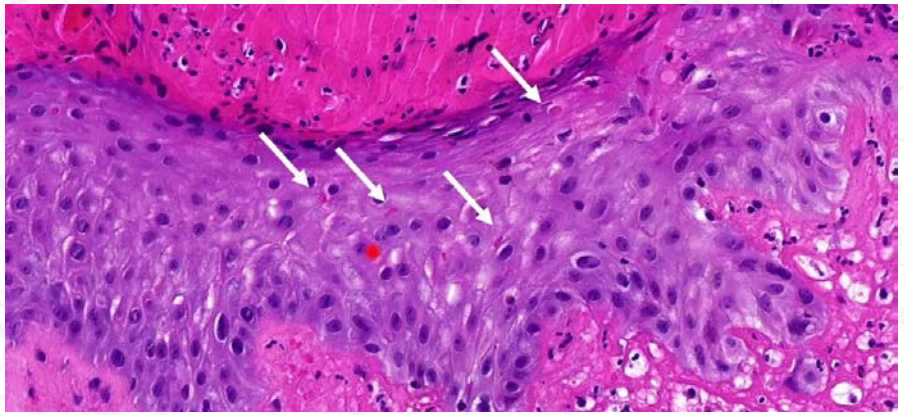


¿Papulosis linfomatoide?

	Milker's nodule	Orf / echtyma contagiosum	Cowpox
Virus	Paravaccinia / Pseudocowpox	Orf	Cowpox
Genus	Parapoxvirus	Parapoxvirus	Ortopox
Family	Poxviridae	Poxviridae	Poxviridae
Host	cattle	goats, sheep	cattle

- lymphomatoid papulosis!!! → emerged pathologically





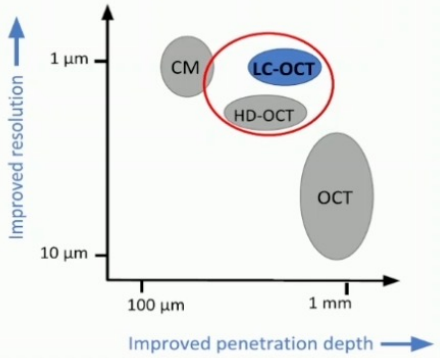
- treatment: spontaneous remission
- importance of protecting clothing

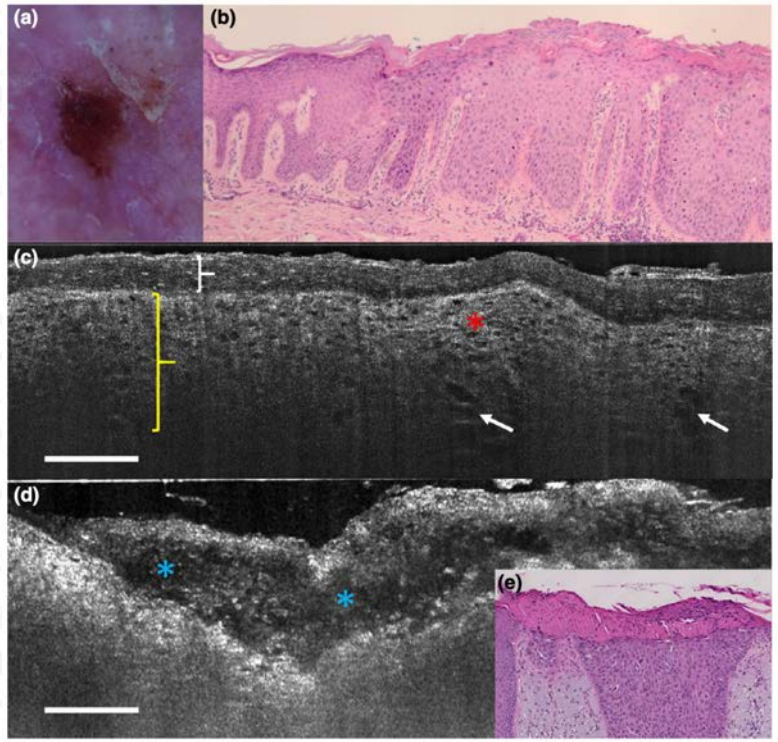
Original Article | Full Access

Morphological evaluation of melanocytic lesions with three-dimensional line-field confocal optical coherence tomography: correlation with histopathology and reflectance confocal microscopy. A pilot study

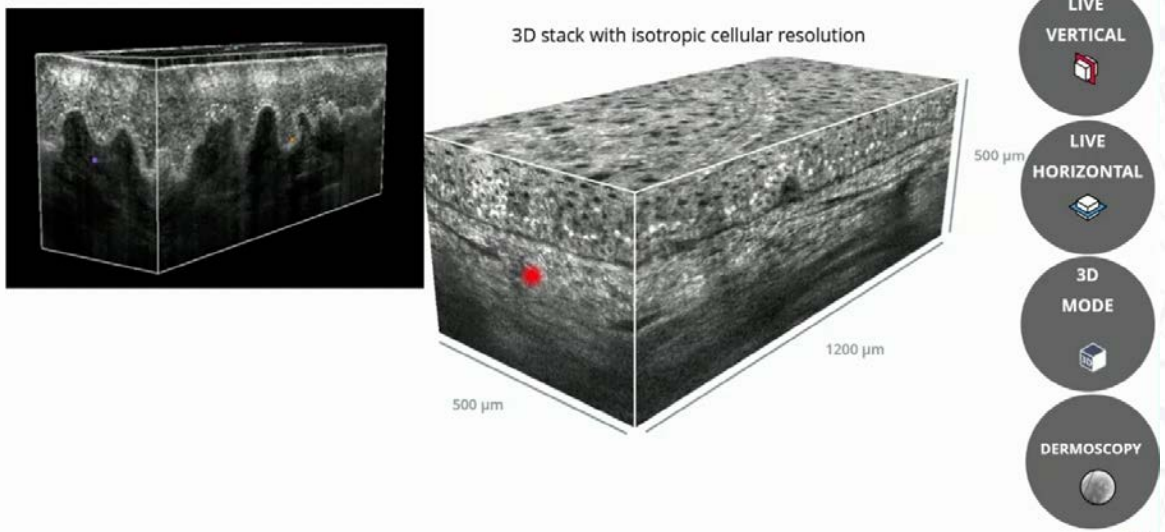
Javiera Perez-Anker, Susana Puig, Llucia Alos, Adriana García, Beatriz Alejo, Elisa Cinotti, Carmen Orte Cano, Linda Tognetti, Clement Lenoir, Jilliana Monnier ... See all authors

First published: 21 August 2022 | https://doi.org/10.1111/ced.15383 | Citations: 5





Line-Field Confocal Optical Coherence Tomography (LC-OCT)



Line-field confocal optical coherence tomography for actinic keratosis and squamous cell carcinoma: a descriptive study

E Cinotti [1], L Tognetti [1], A Cartocci [2], A Lamberti [1], S Gherbassi [1], C Orte Cano [3], C Lenoir [3], G Dejonckheere [3], G Diet [3], M Fontaine [3], M Miyamoto [3], J Perez-Anker [4 5], V Solmi [1], J Malvehy [4 5], V Del Marmol [3], J L Perrot [6], P Rubegni [1], M Suppa [3 7]

La Academia Española de Dermatología y Venereología expresa su agradecimiento al patrocinador UCB, por su especial apoyo y contribución con la actividad formativa Highlights 2023.

Con el patrocinio de:

ucb

MUCHAS GRACIAS

Iniciativa científica de:

AEDV 1909 ACADEMIA ESPAÑOLA DE DERMATOLOGÍA Y VENEREOLOGÍA