Histopathology of Common Nail Lesions

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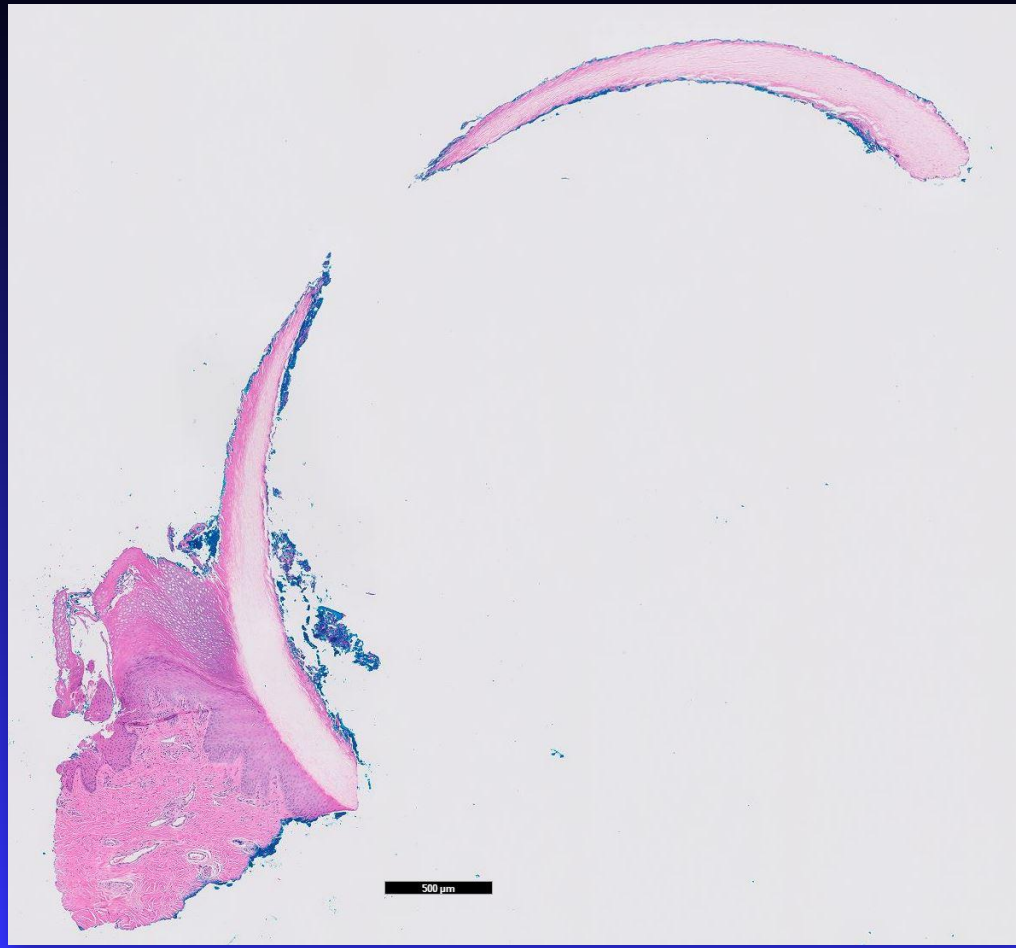
CTA PATHOLOGY
CURTIS THOMPSON, MD & ASSOCIATES
SKIN, HAIR AND NAIL EXPERTS

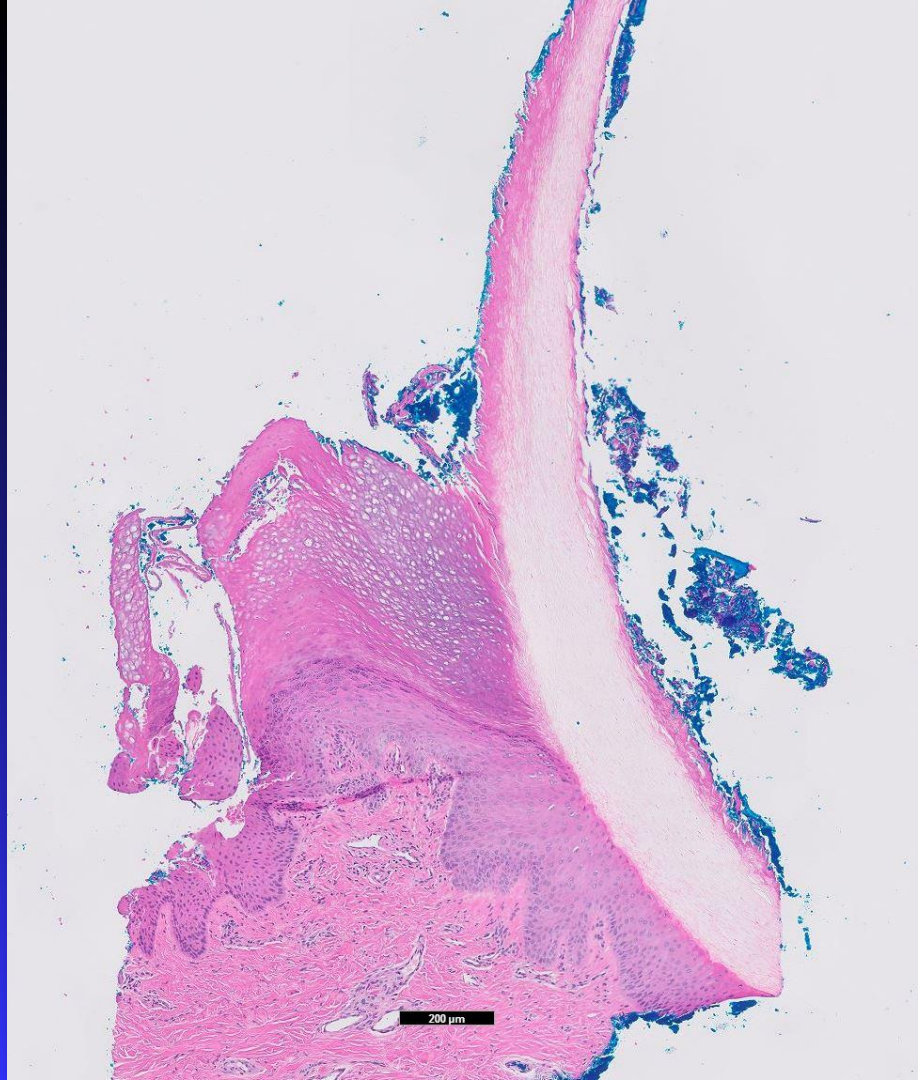
- Right index finger of 8 y/o male

8 y/o index

8 y/o index



8 y/o index



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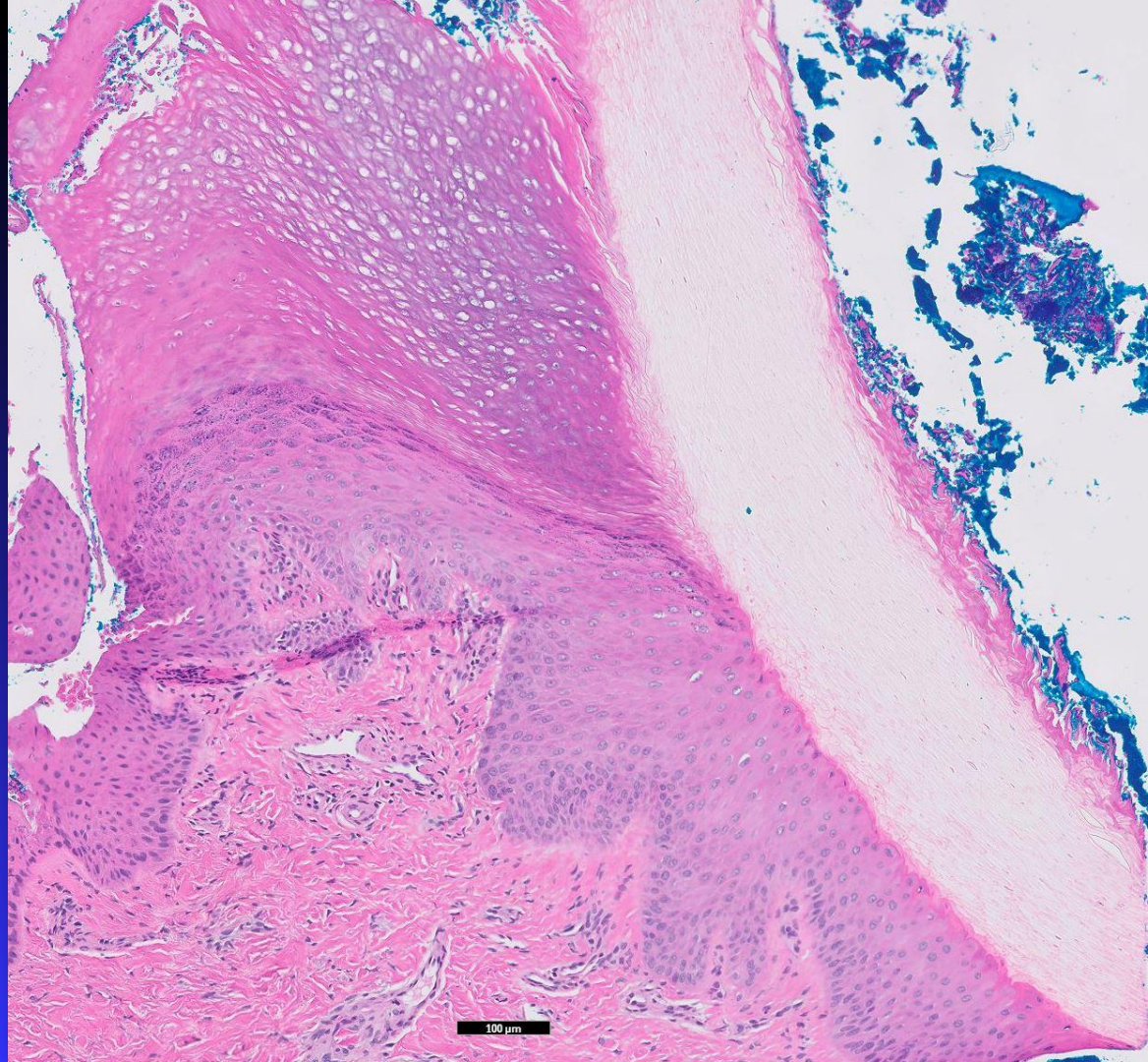

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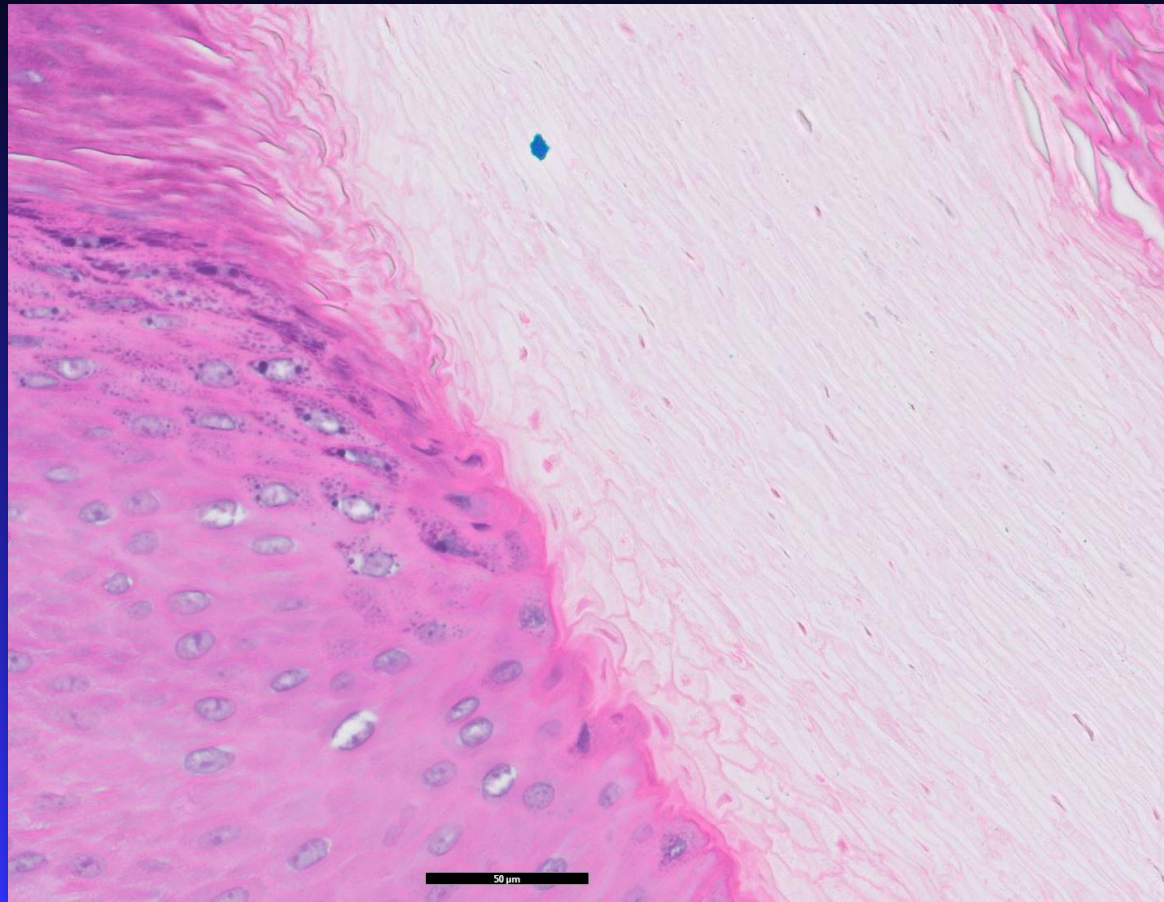


Onychoheterotopia (Ectopic Nail)

- Nail is growth of nail unit tissue outside the usual anatomic area.
- Japan and India
- Congenital from syndromes such as Pierre-Robin Syndrome and Congenital Palmar Nail Syndrome.
- Trauma or chronic repetitive injury.

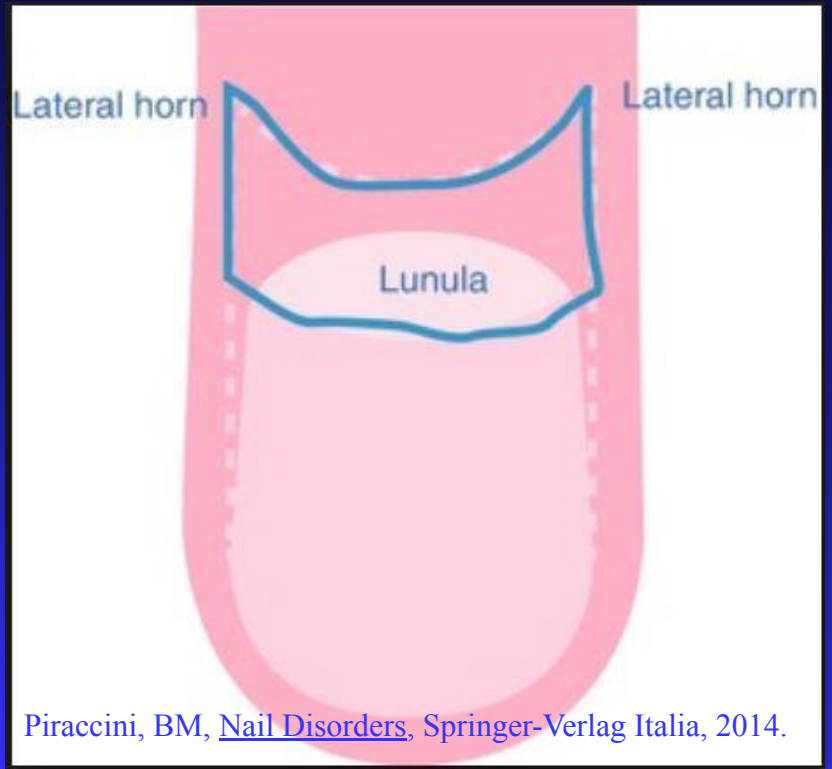
Onychoheterotopia (Ectopic Nail)

- Dorsal aspect of the hand.
- Osseous defects if matrix close to bone.

Traumatic ectopic nail

Çalişkan et al. Ann Dermatol, http://dx.doi.org/10.5021.ad.2014.26.6.768,

Matrix horns



Piraccini, BM, Nail Disorders, Springer-Verlag Italia, 2014.

Onychoheterotopia (Ectopic Nail)

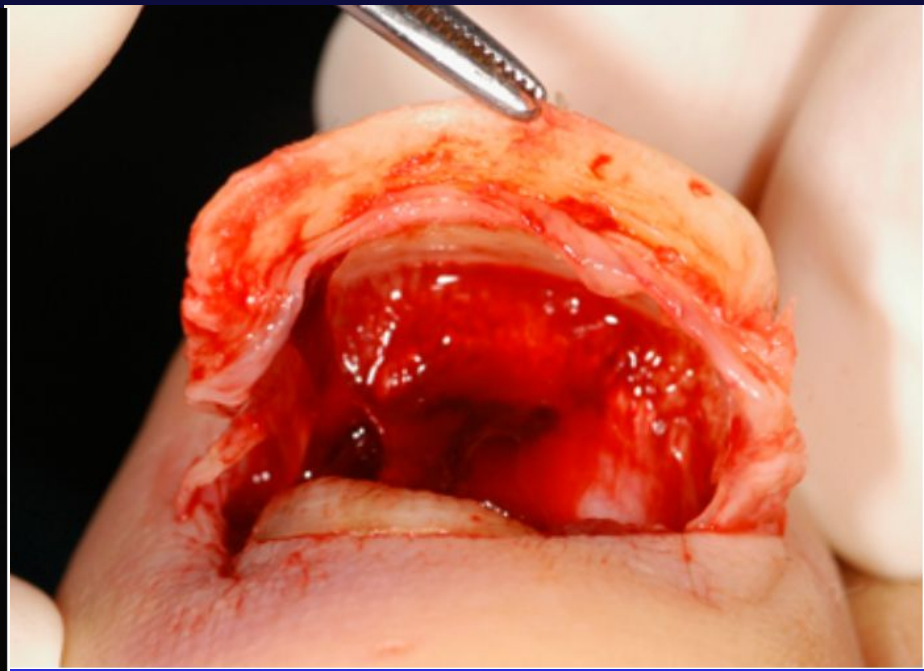
- All component of nail unit
  - Matrix
  - Place
  - Bed
  - Nail Fold

Onychoheterotopia (Ectopic Nail)

- Differential:
  - Retronychia
  - Squamous cell carcinoma in-situ (HPV)
  - Residual nail unit after incomplete excision

Retronychia

Retronychia



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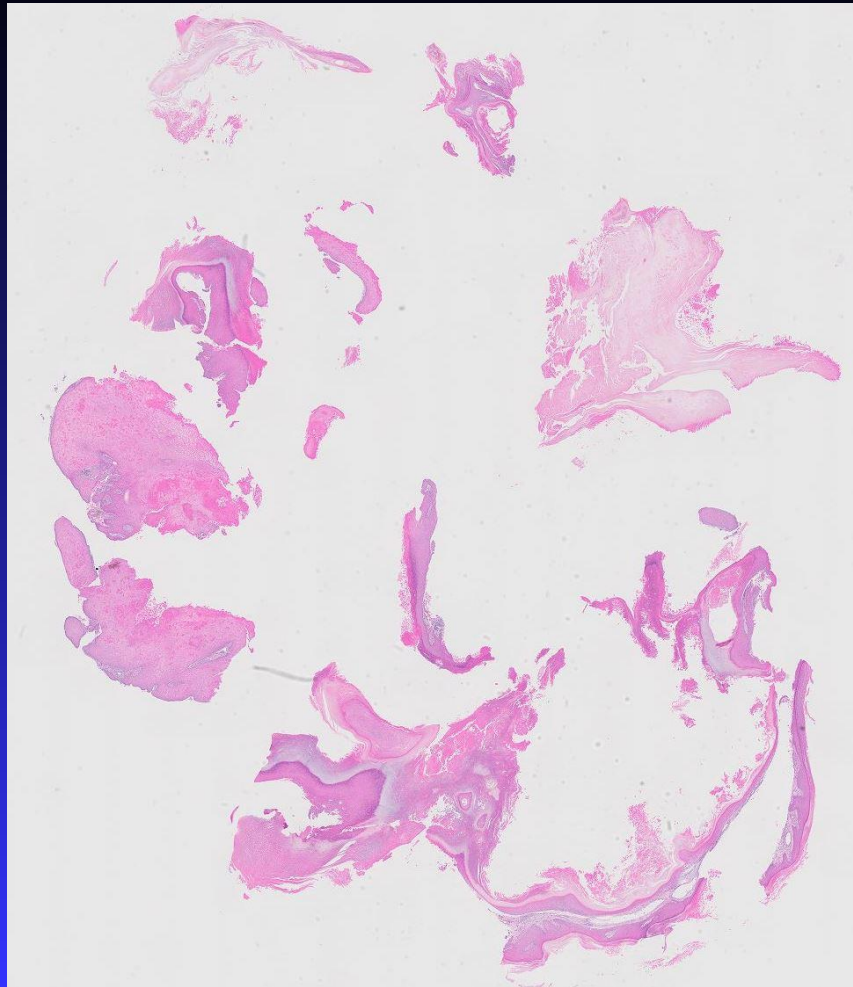
SKIN, HAIR AND NAIL EXPERTS

Baumgartner M and Haneke E. Retronychia: Diagnosis and Treatment. Dermatologic Surgery 36(10):1610-4, 2010

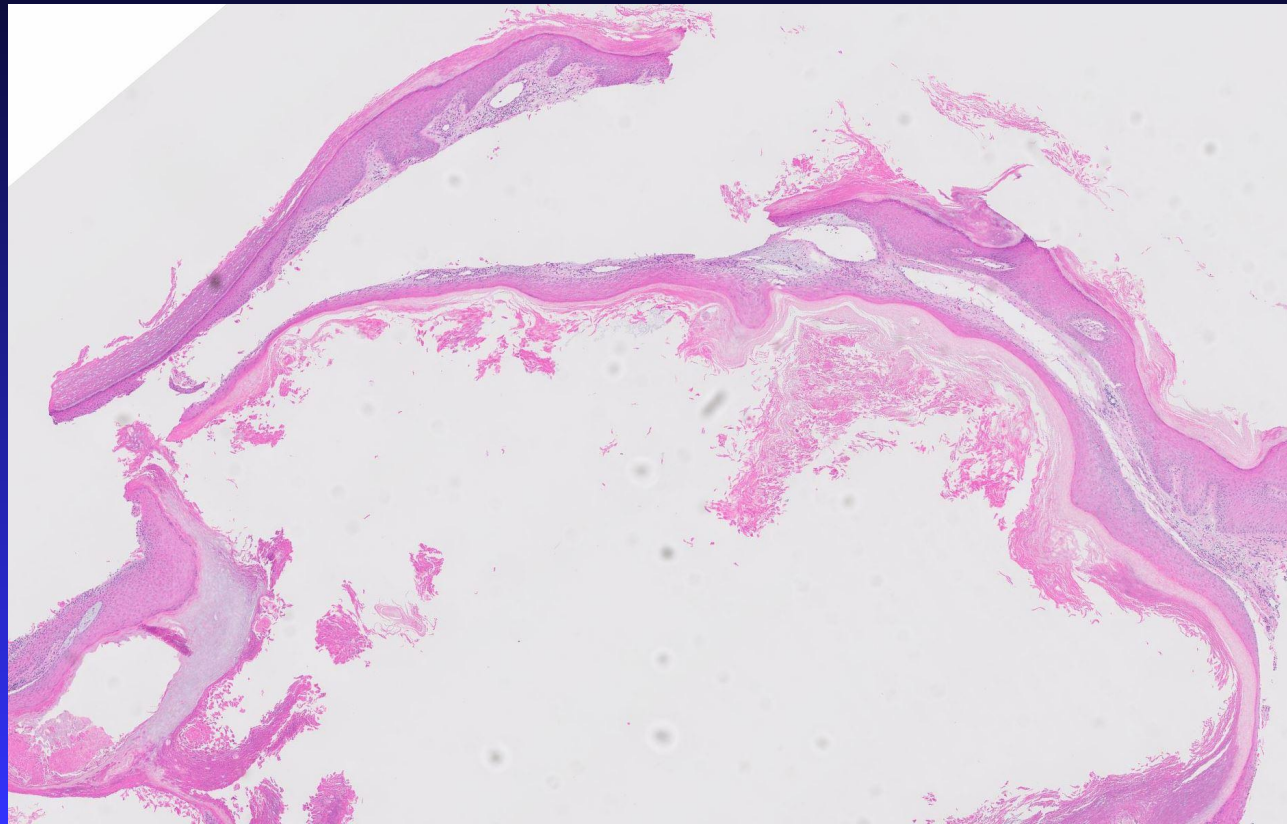
64 y/o male

- Rapidly enlarging nodule in fingernail for one month
- All other nails normal

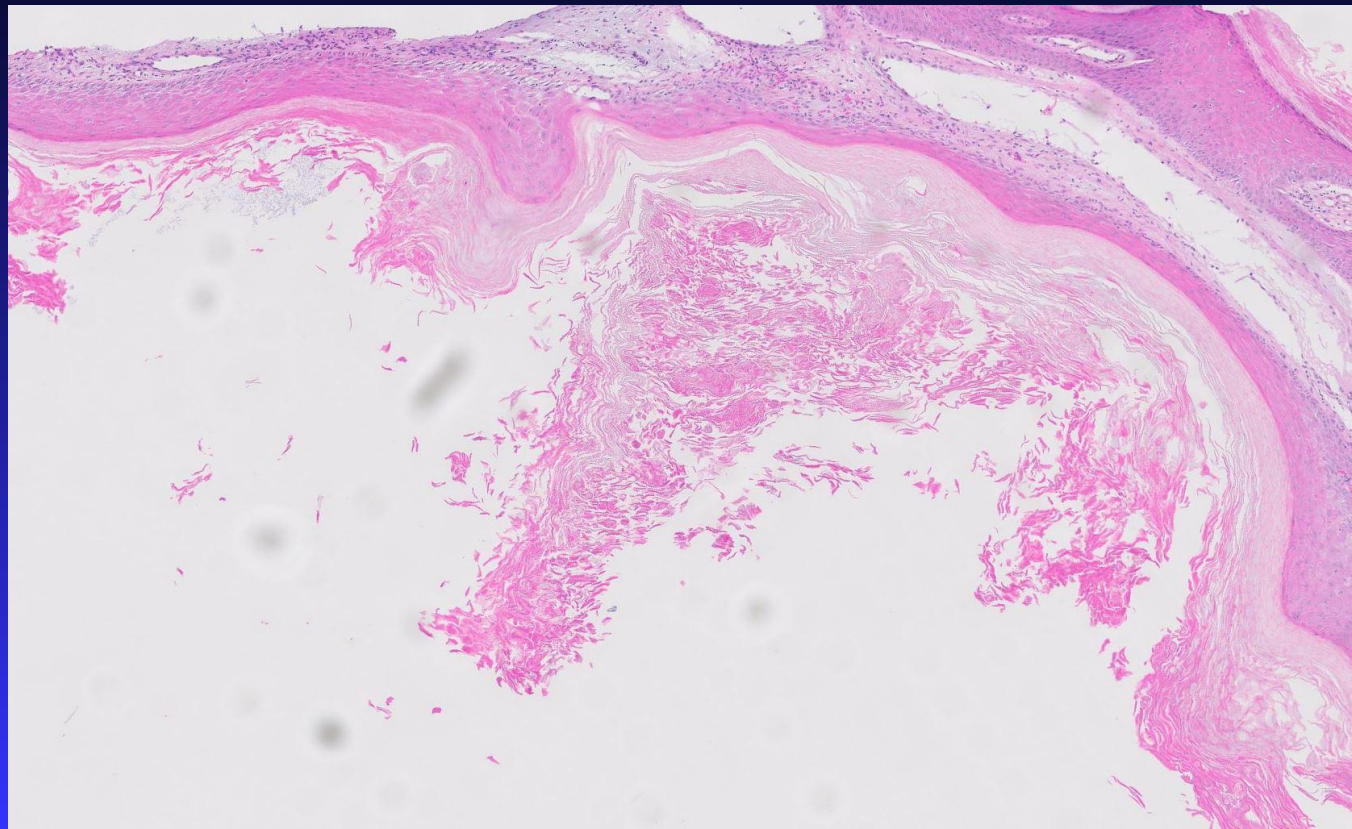
64 y/o male



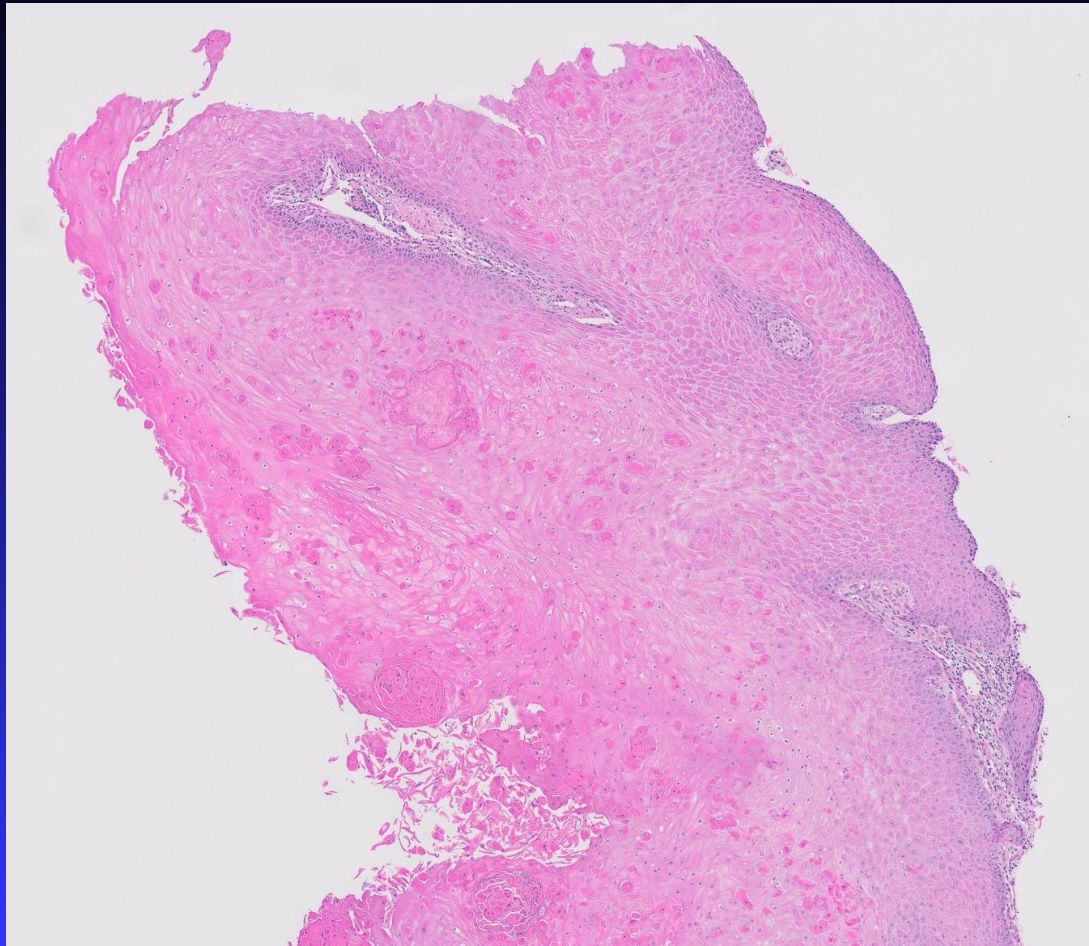
64 y/o male



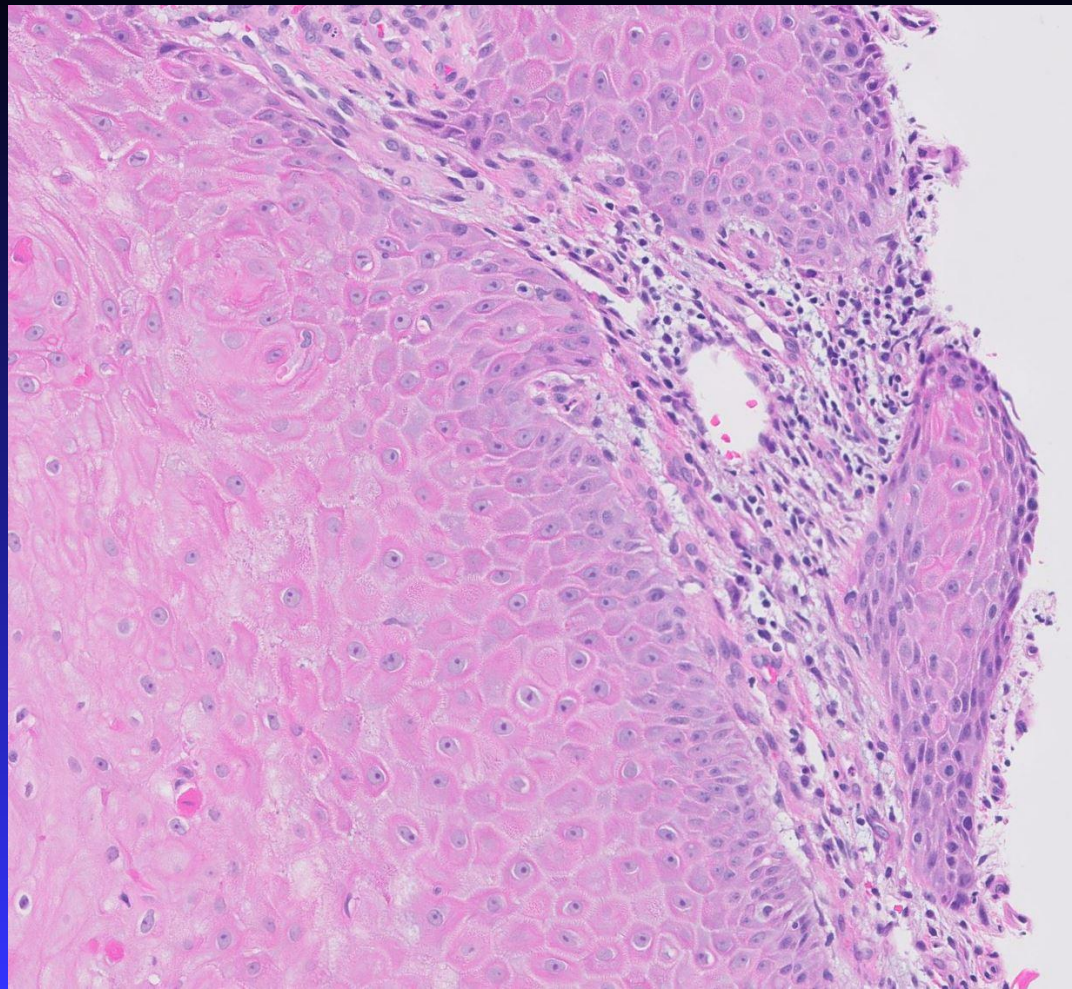
64 y/o male



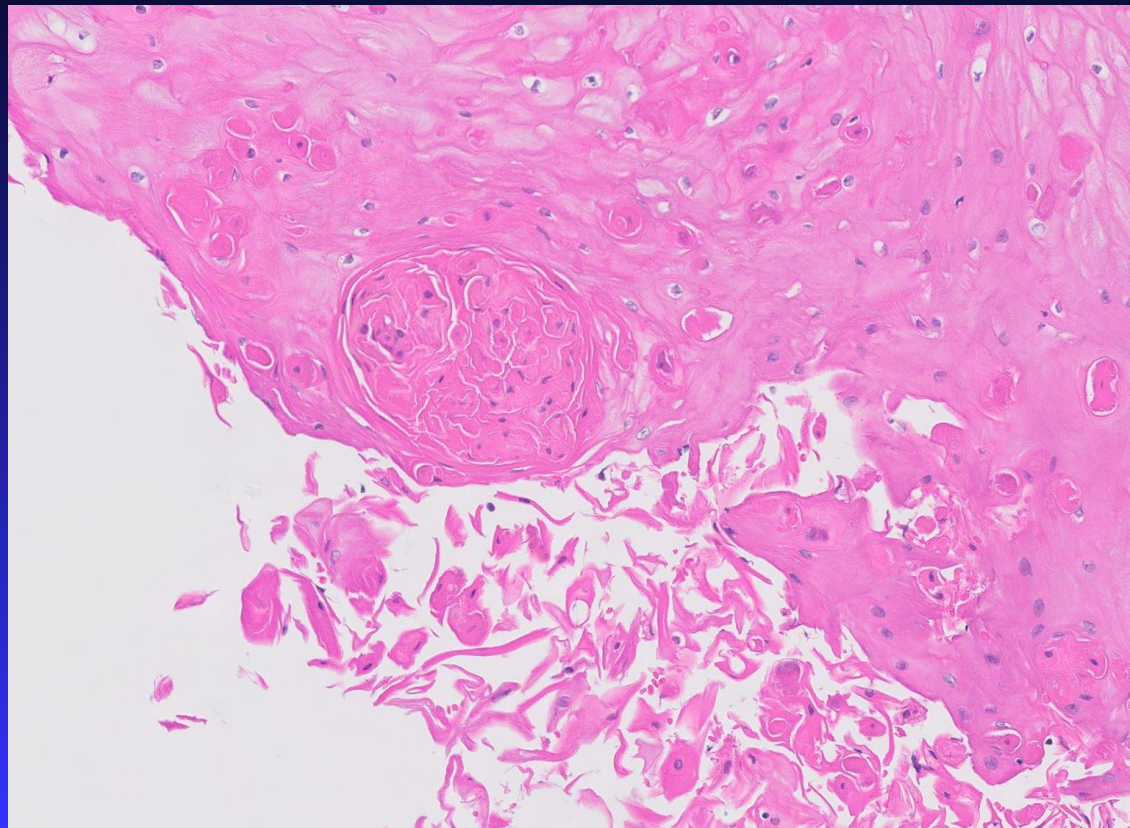
64 y/o male



64 y/o male



64 y/o male



Subungual Keratoacanthoma

- Similar to the keratoacanthoma-type of SCC elsewhere
- Destroys bone and does not regress but otherwise not aggressive
- Biopsy is curative

Subungual Keratoacanthoma

- Initial biopsy is curative
  - Calling squamous cell carcinoma often leads to an unnecessary amputation

Subungual Keratoacanthoma

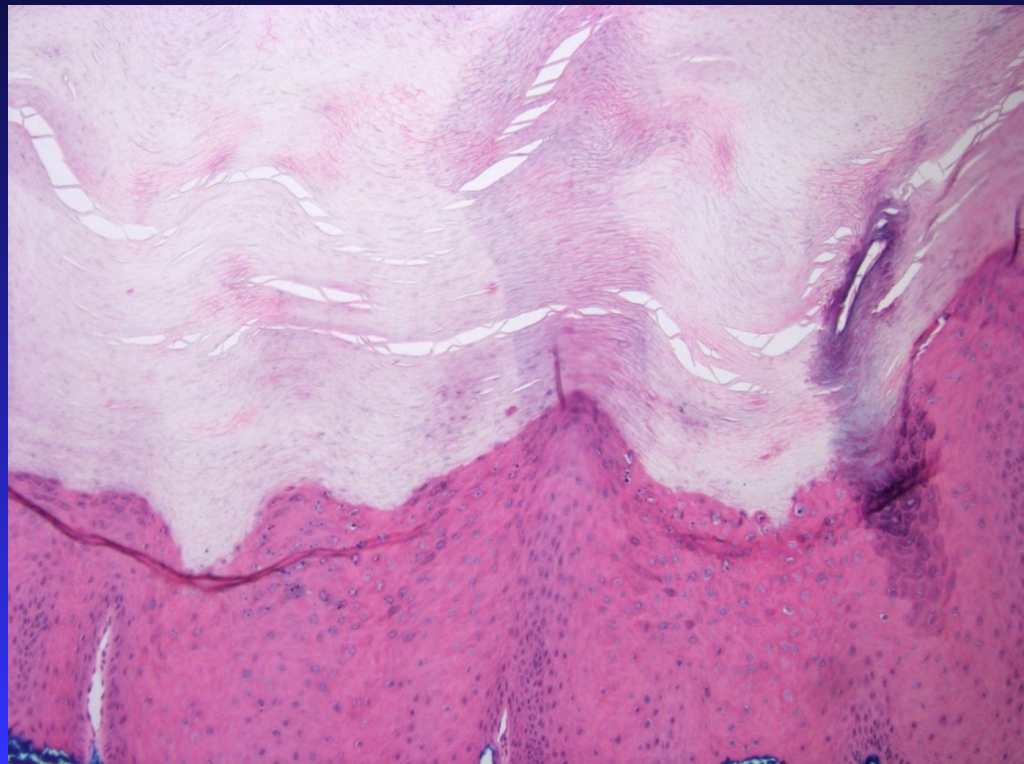
- Crateriform squamous proliferation with abundant keratin and parakeratotic foci
- Minimal keratinocytic atypia
- Variable mixed inflammatory cell infiltrate with intraepithelial neutrophils and surrounding lymphocytes, plasma cells and sometimes eosinophils

Subungual Keratoacanthoma

- Differential
  - **Verruca**
  - **Squamous cell carcinoma of the nail unit**
  - **Nail bed inclusions**
  - **Onycholemmal cyst**
  - **Subungual tumor of incontinentia pigmenti**

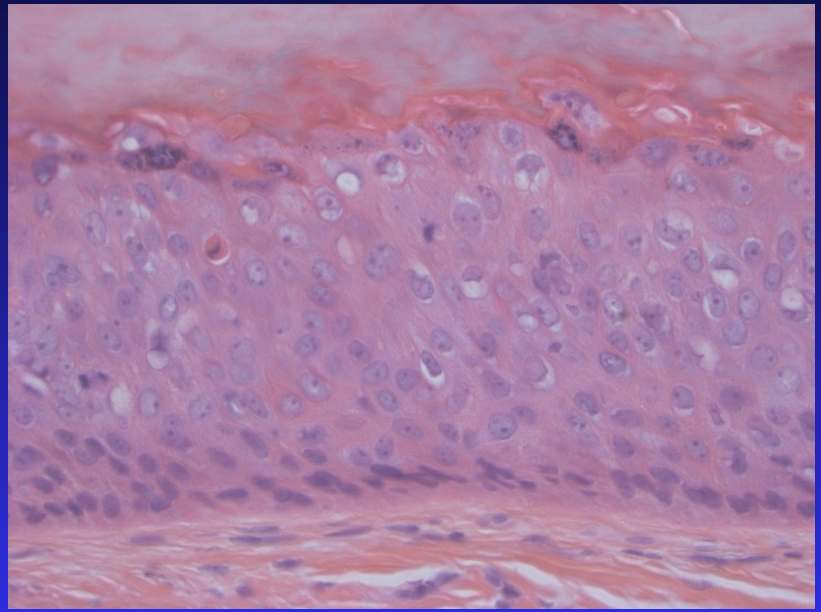
Subungual Keratoacanthoma Differential

- Verruca or SCC
- Both HPV



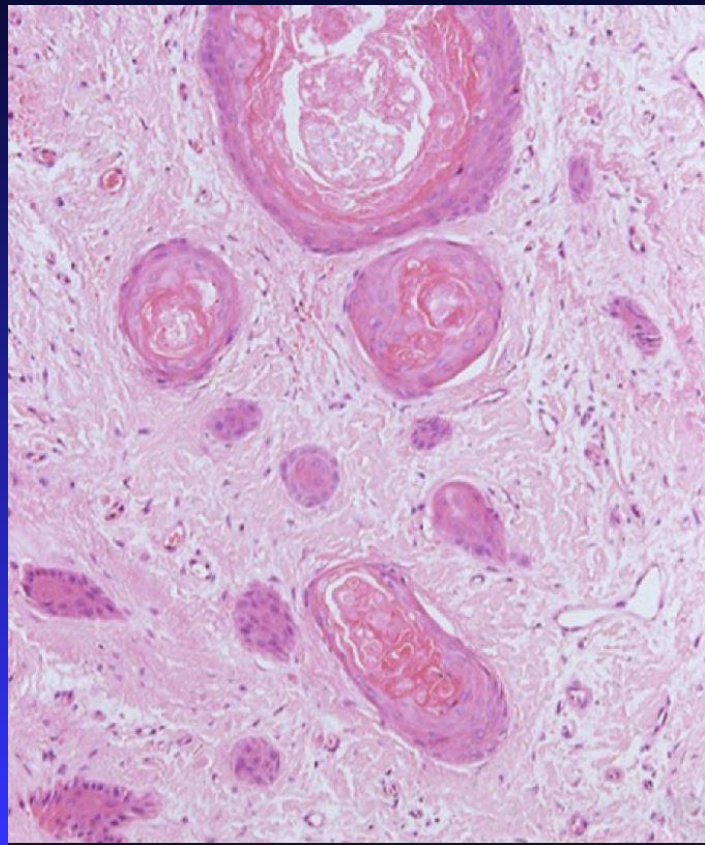
Subungual Keratoacanthoma Differential

- Verruca or SCC
  - Both HPV
  - Sampling important



Subungual Keratoacanthoma Differential

- Nail bed inclusions
- Onycholemmal cyst
  - Likely result of trauma



Martin B. Nail histopathology. Actas Dermosifiliogr 104:564-78, 2013

Subungual Keratoacanthoma Differential

- Subungual tumor of incontinentia pigmenti (IP)
  - Suspect if young female
  - May be first presentation of IP in mosaic cases

Donati et al. Eur J Dermatol 19:243-7, 2009

Confusing Nail Tumor Terminology

- Onychopapilloma
- Onychomatricoma
- Onychocytic matricoma
- Onycholemmal (cyst, horn tumor)

Onychopapilloma

Onychopapilloma

- Clinical
  - Logintudinal erythronychia (redness)
  - Distal nail split

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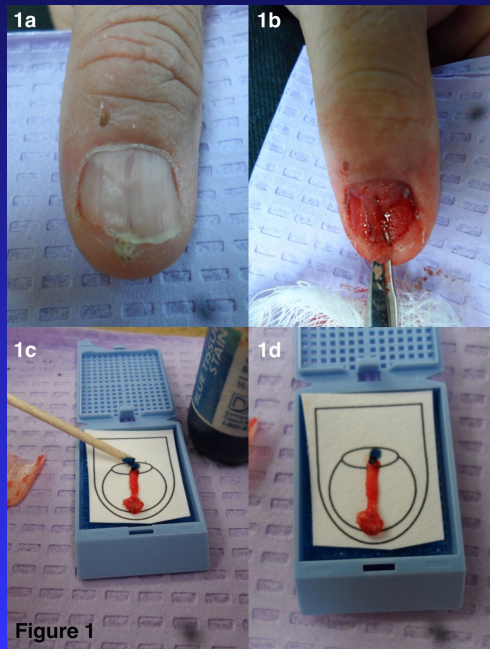
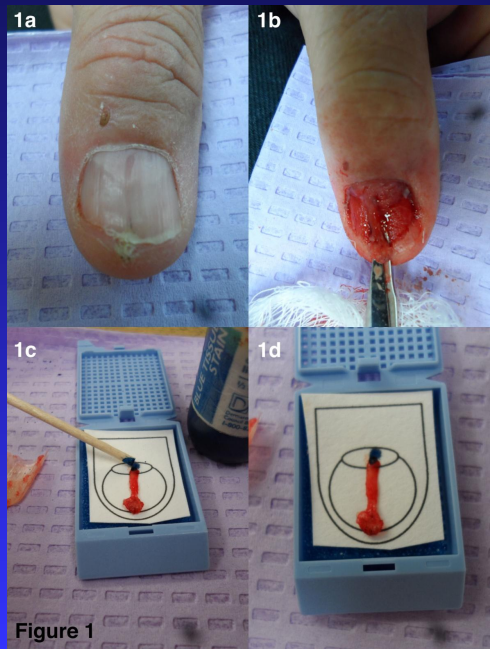


Figure 1

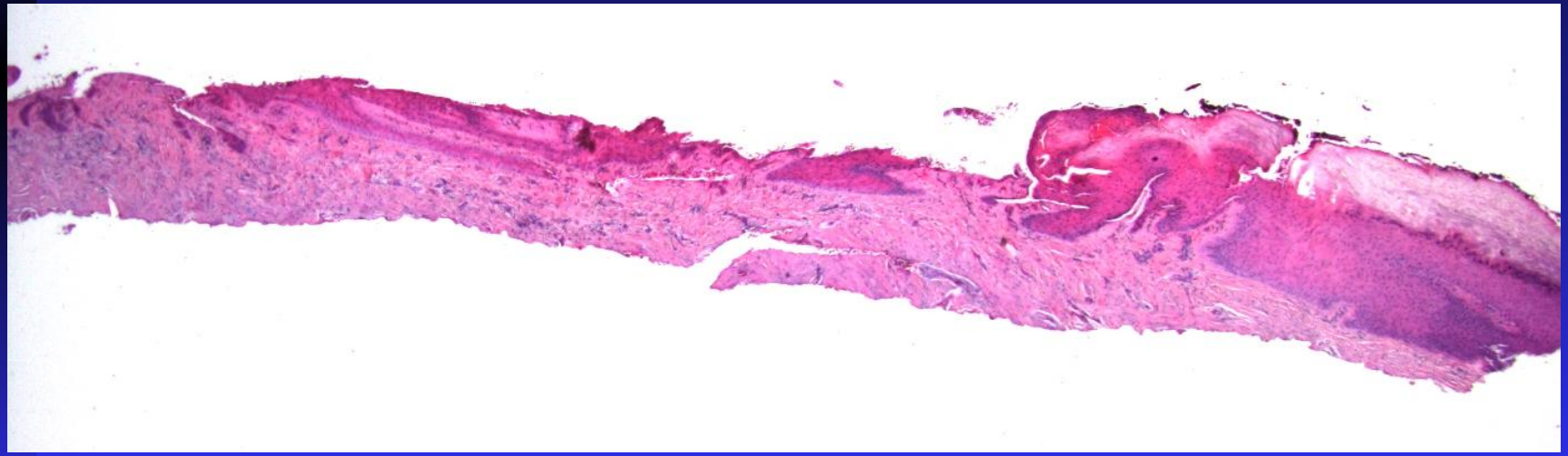
Onychopapilloma

- Clinical
  - Logintudinal erythronychia (redness)
  - Distal nail split

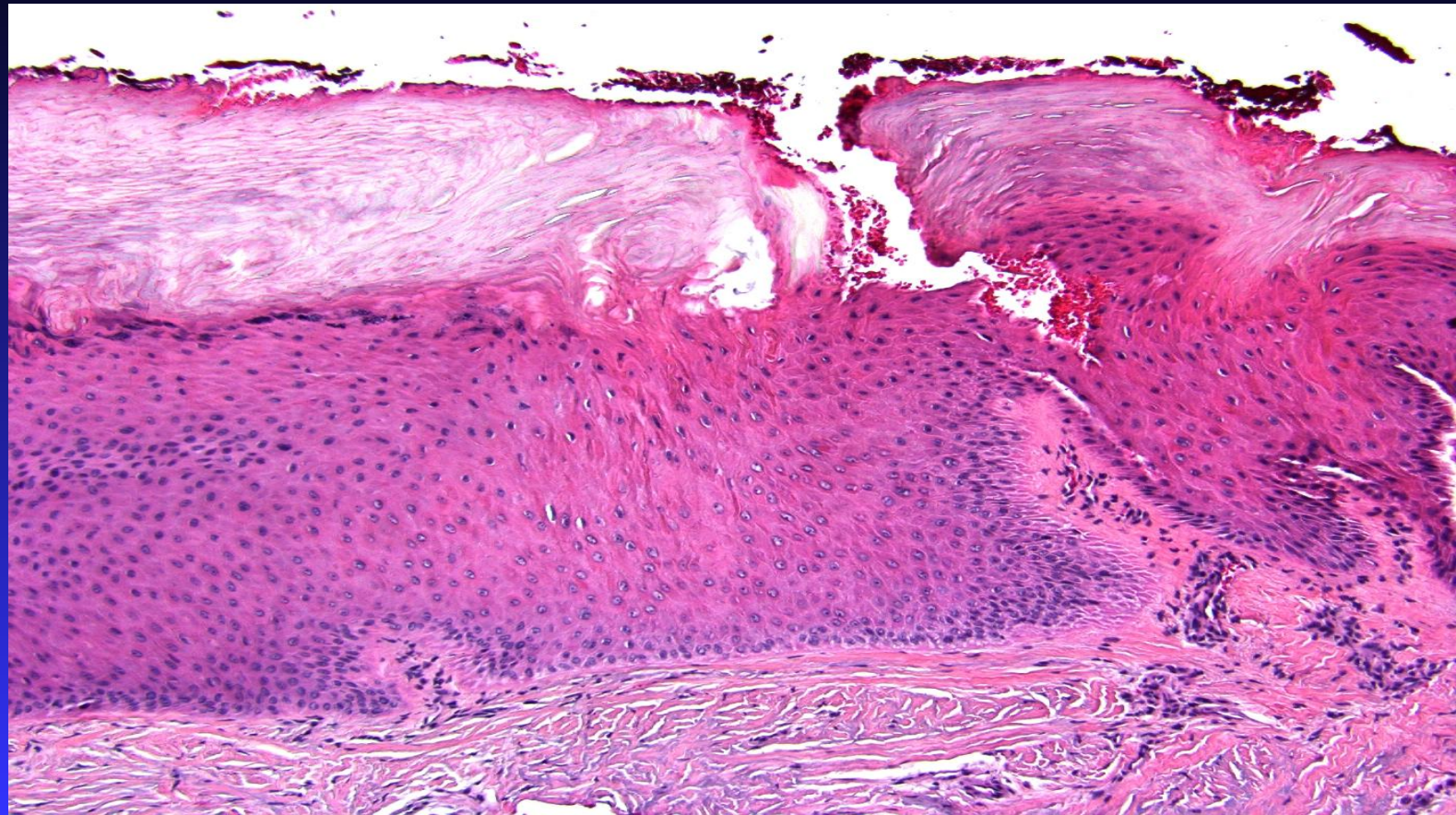
Embed proximal to distal



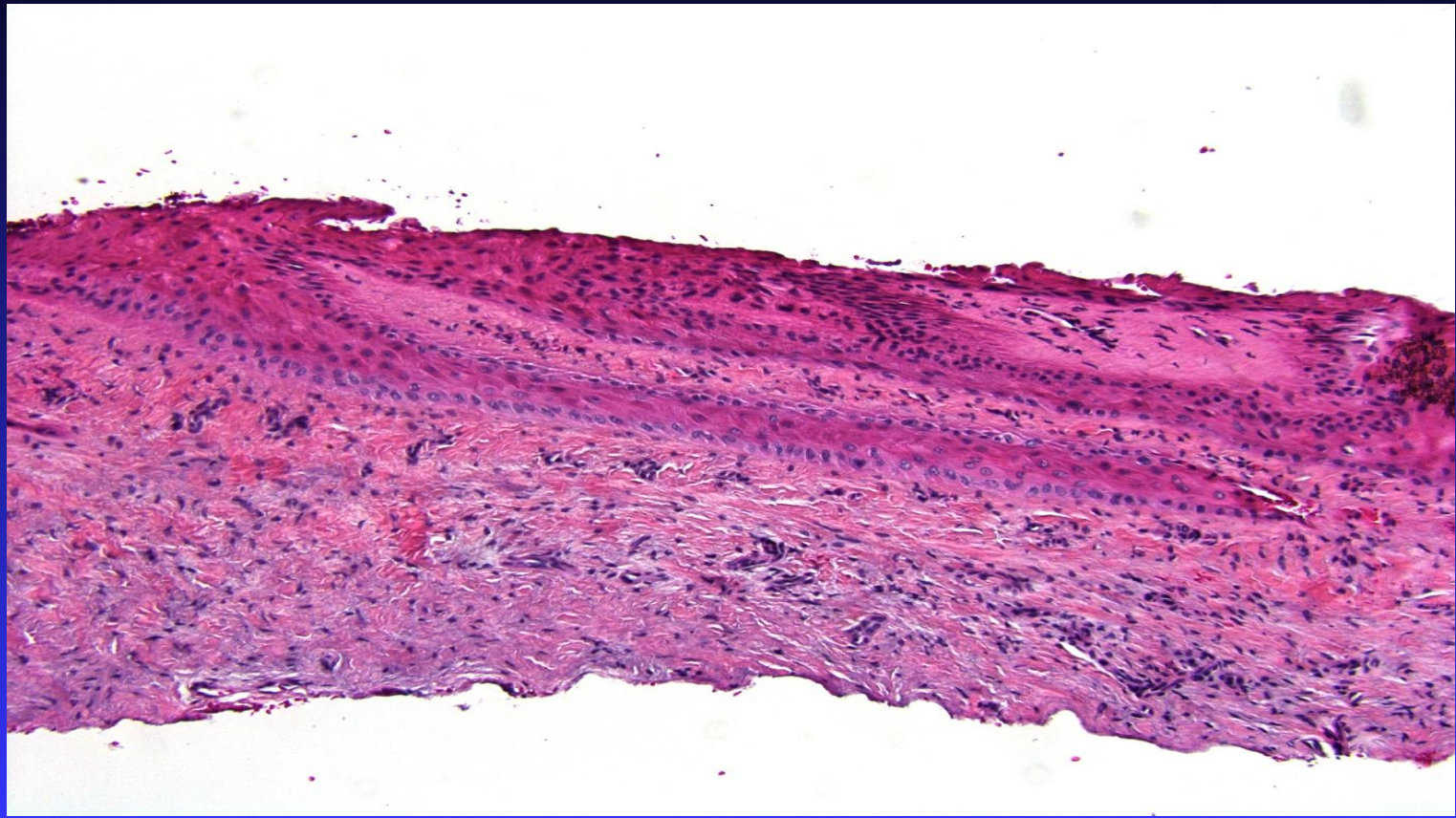
Onychopapilloma—Keratin Producing



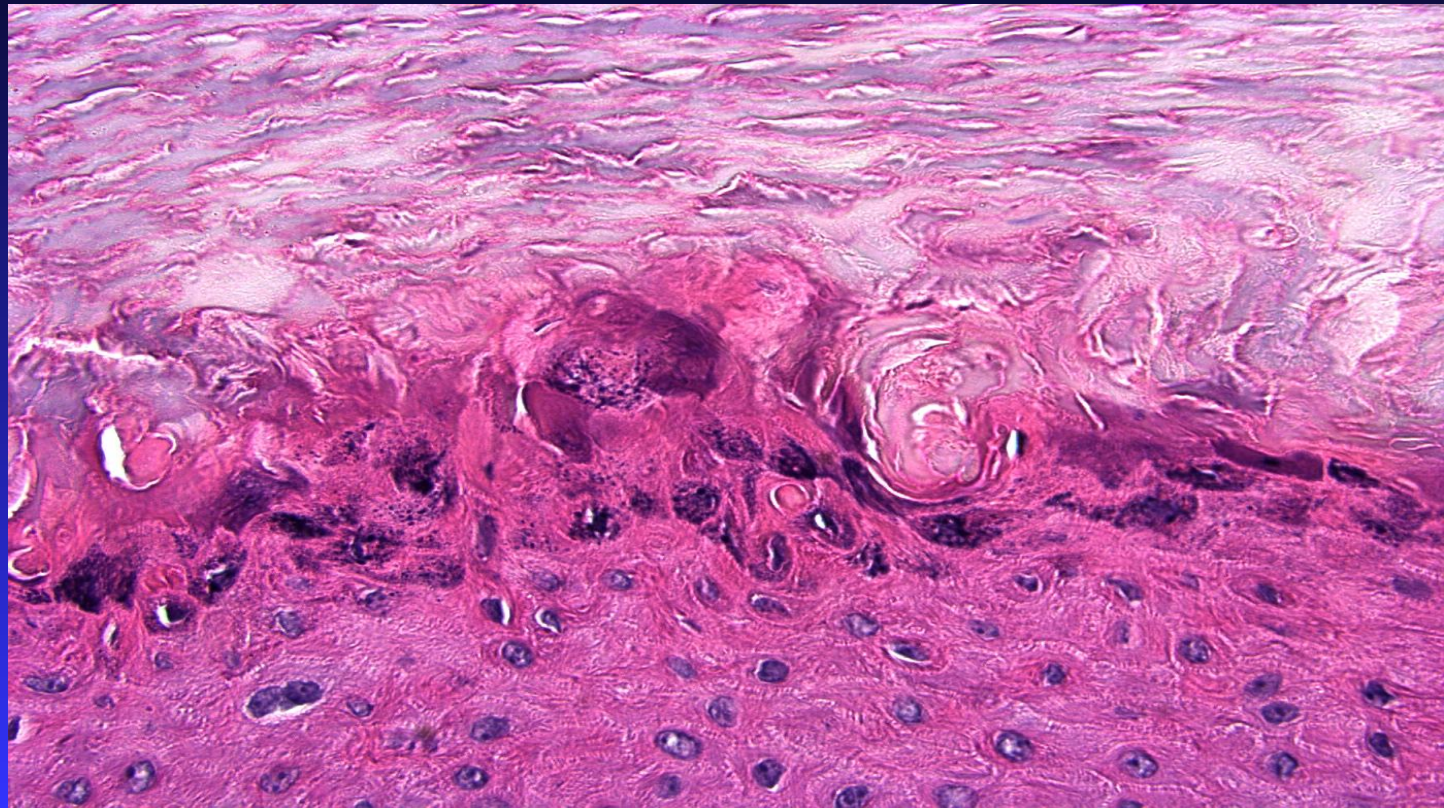
Onychopapilloma—Keratin Producing



Onychopapilloma



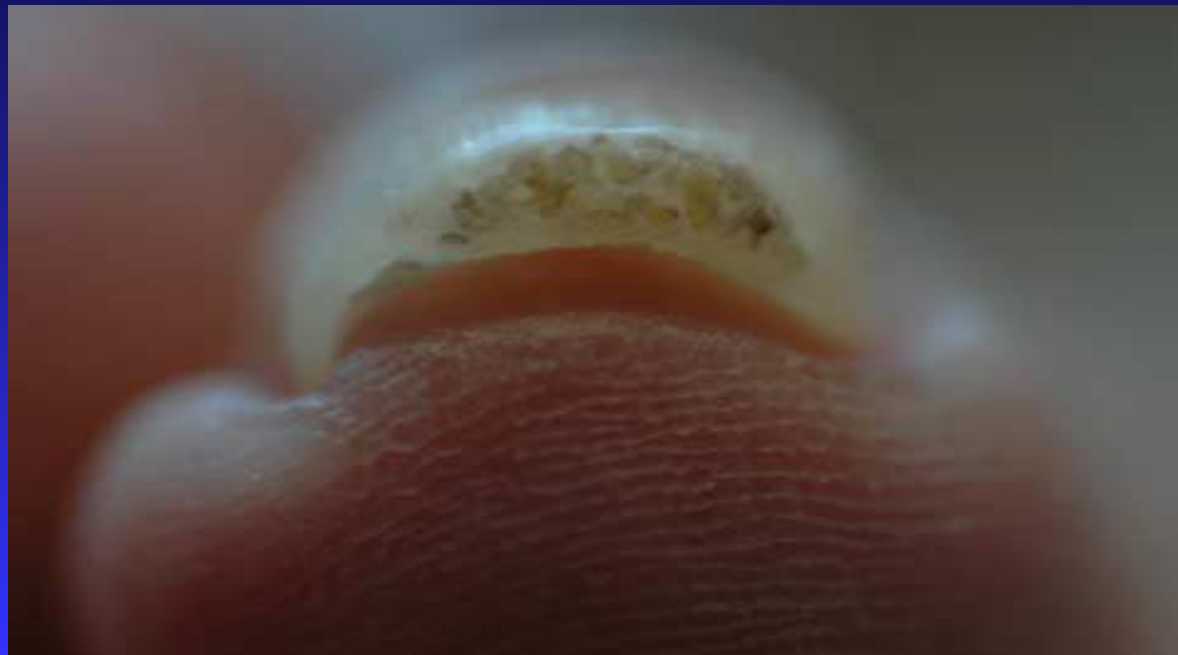
Onychopapilloma—Not a wart



Onycomatricoma

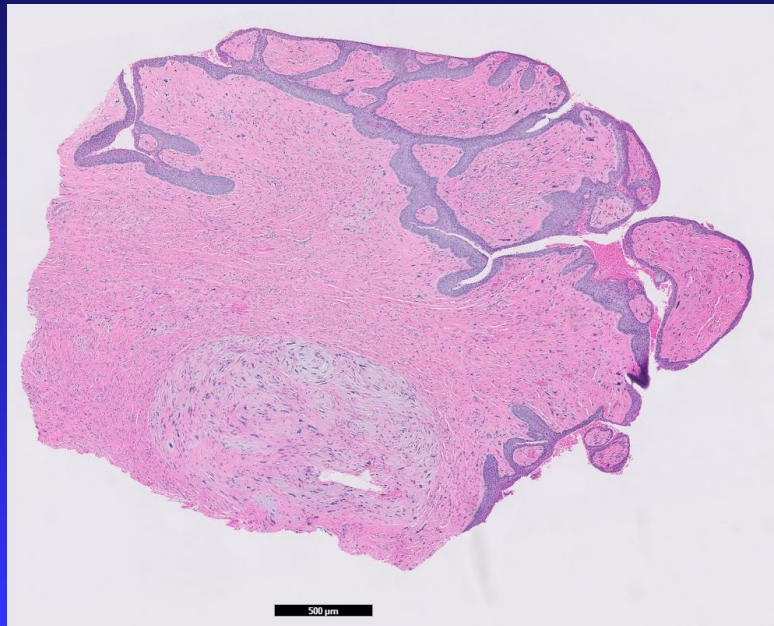
Onycomatricoma

- Examine nail for holes—Transverse sections of dystrophic nail

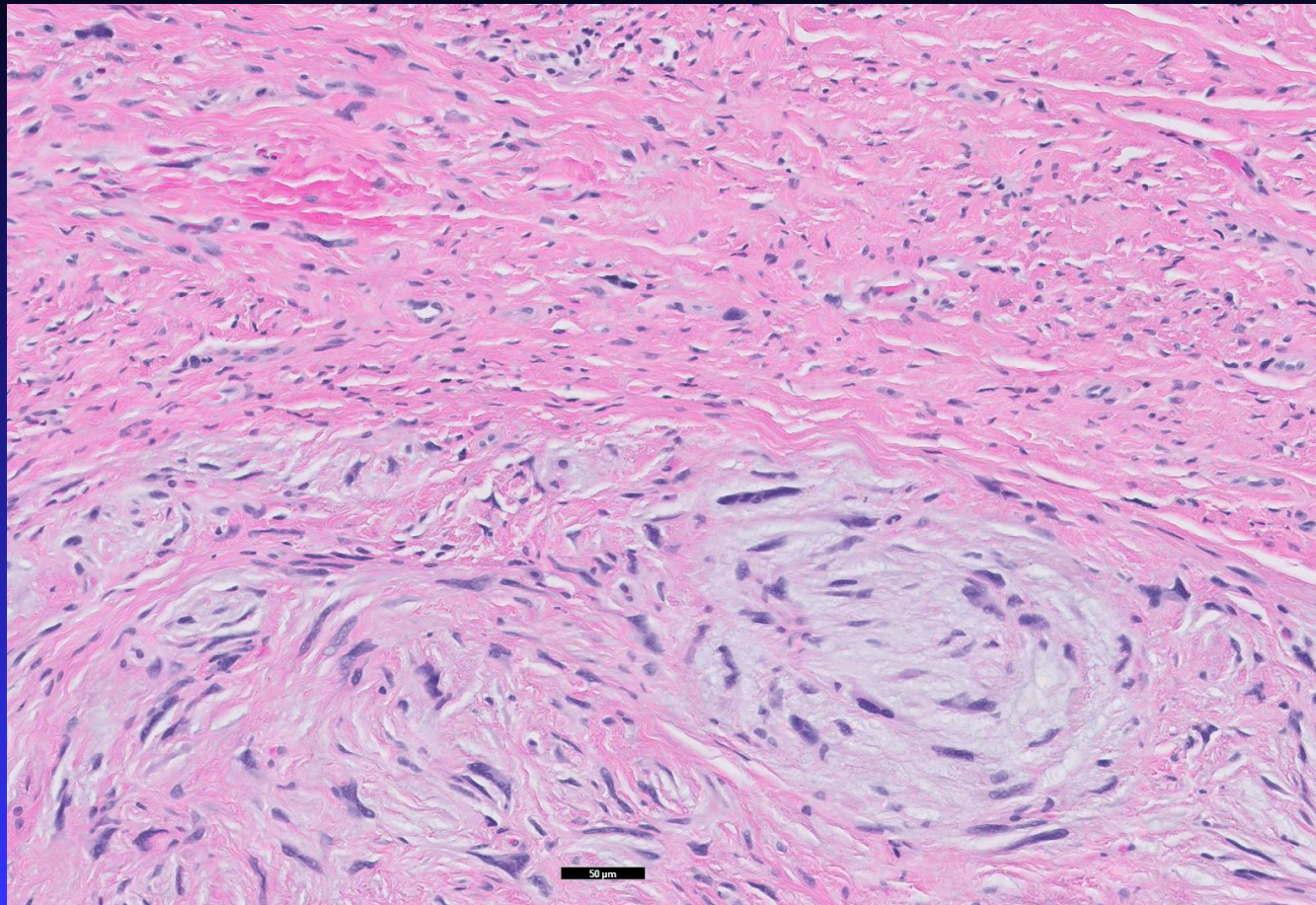


Onychomatricoma

- Two components
  - Epithelial (?reactive)
  - Dermal spindle
    - (May be myxoid)



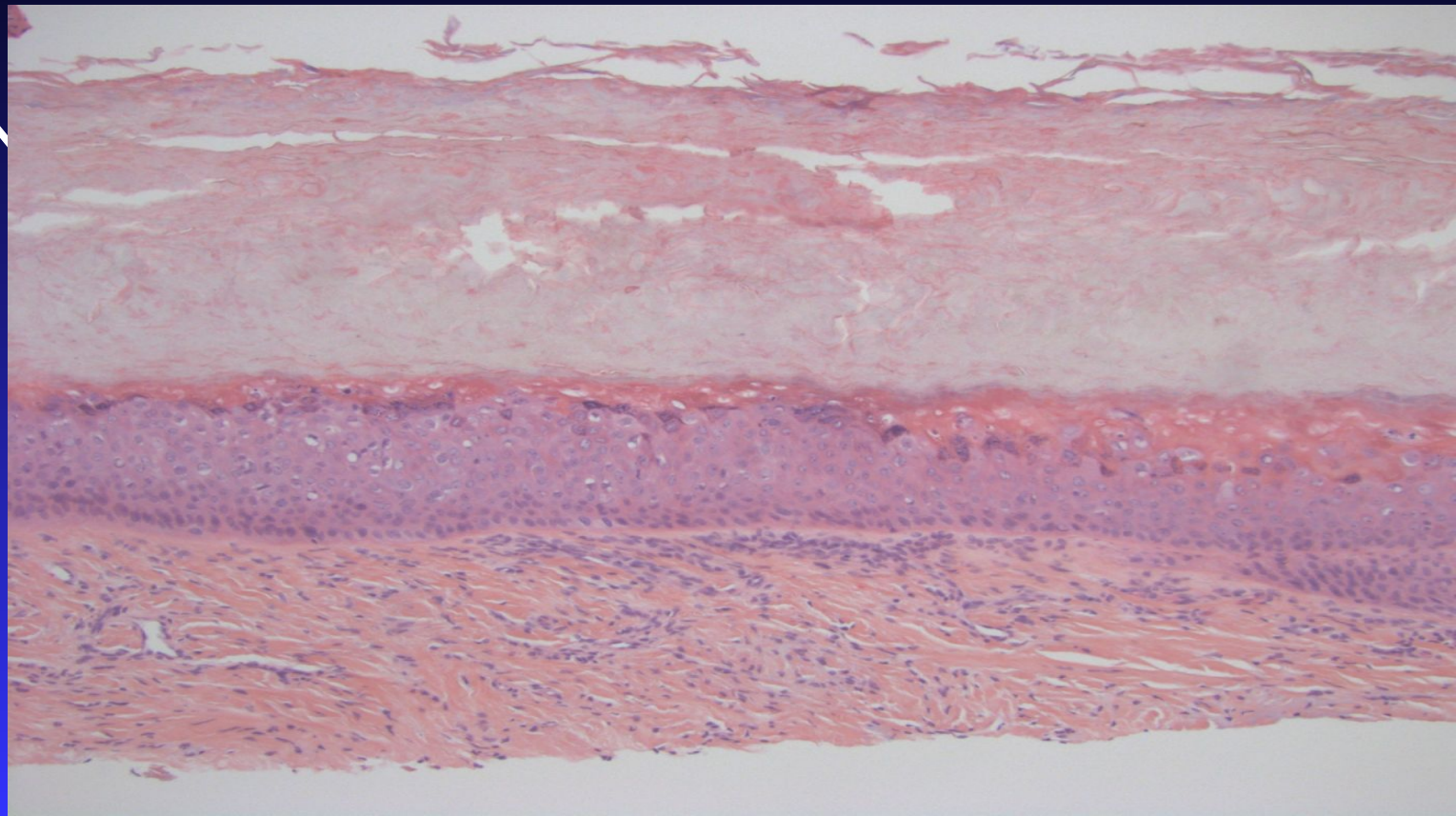
Onycomatricoma



Squamous cell carcinoma

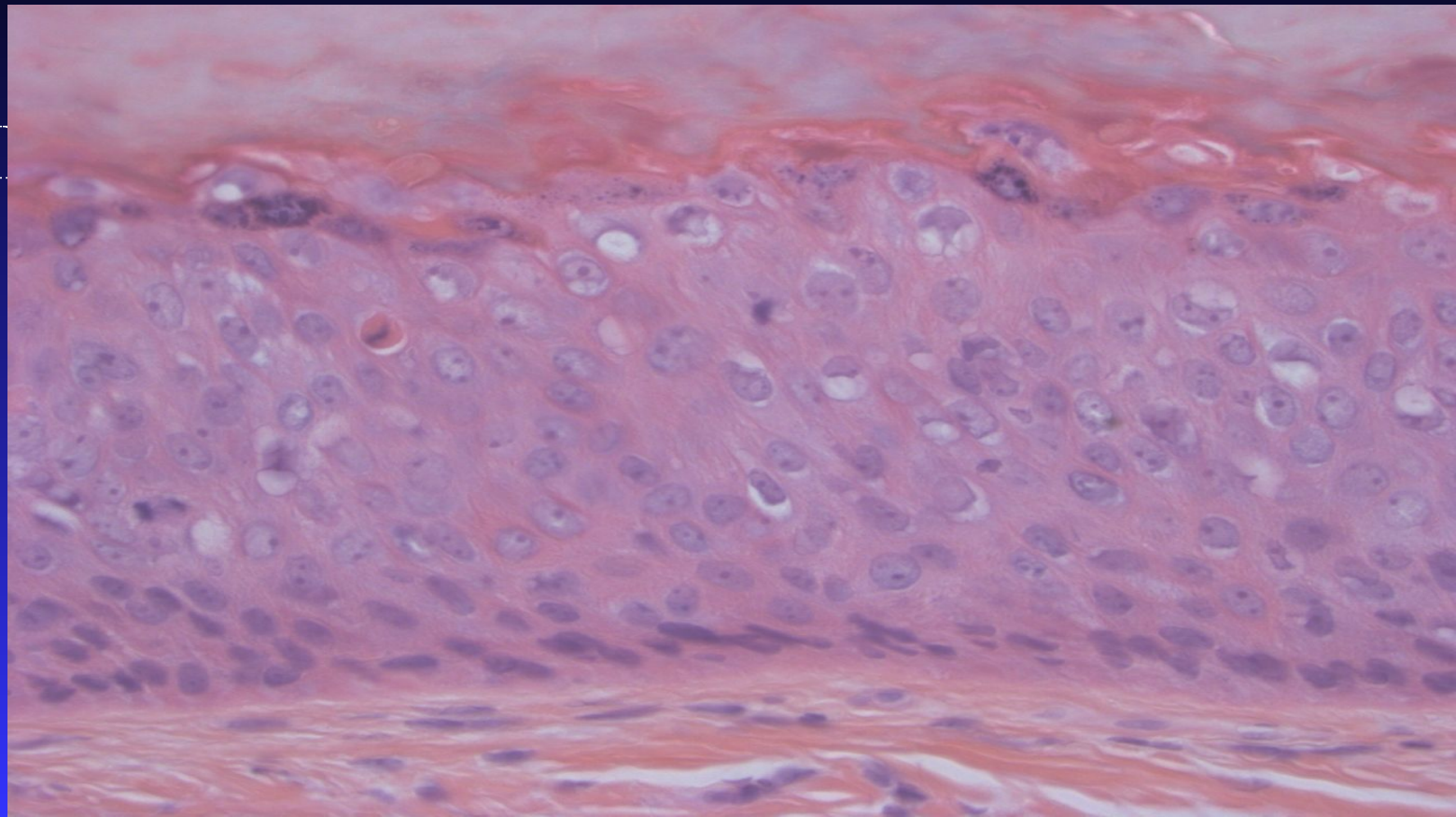
Squamous cell carcinoma

- IN



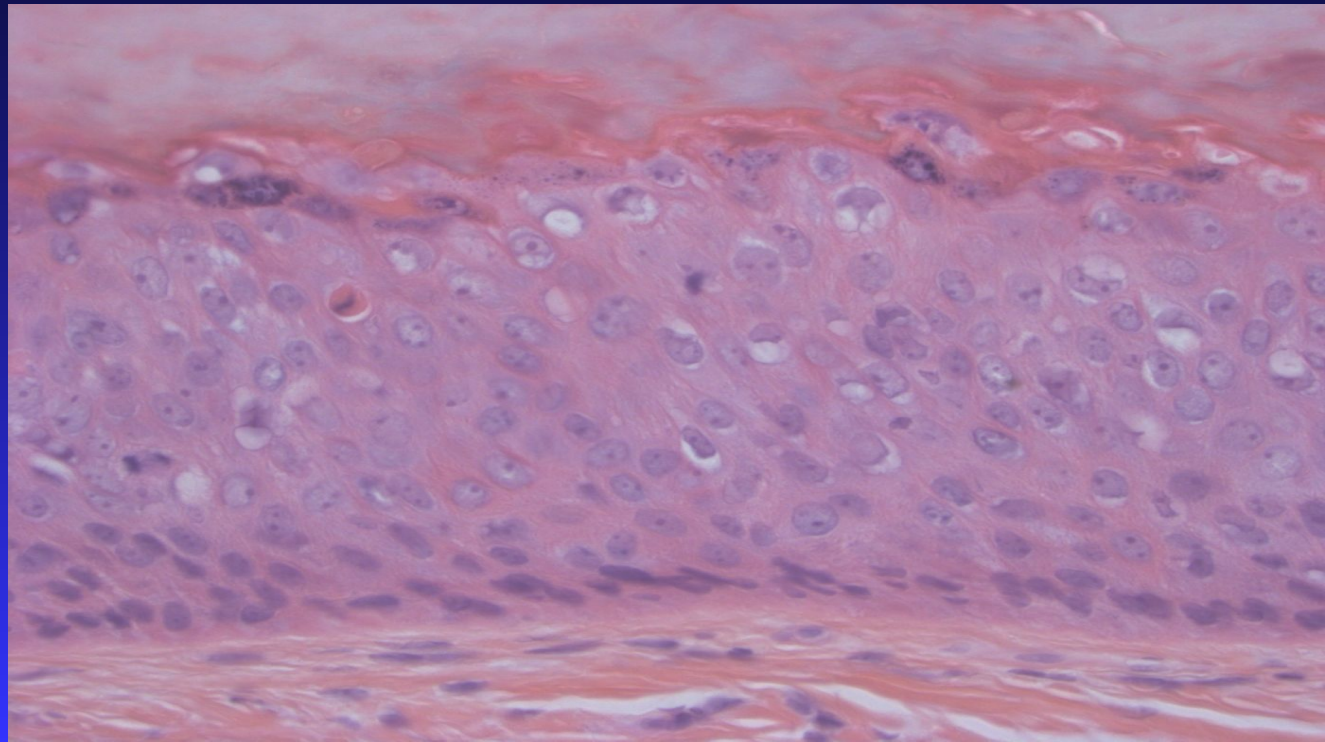
Squamous cell carcinoma in-situ

- I

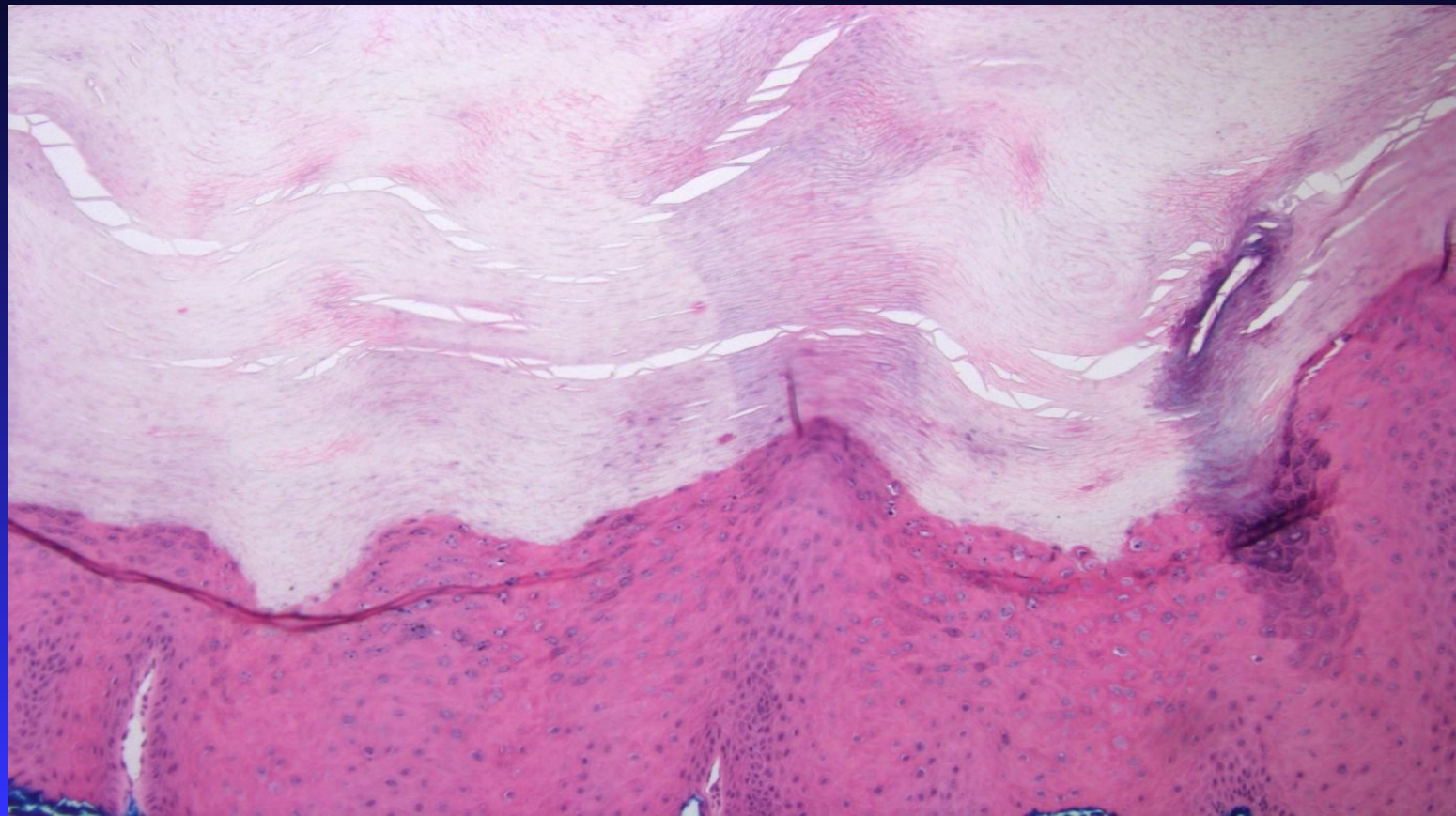


Squamous cell carcinoma in-situ

- Human Papillomavirus (HPV) features



SCC versus Wart/Verruca



SCC versus Wart/Verruca

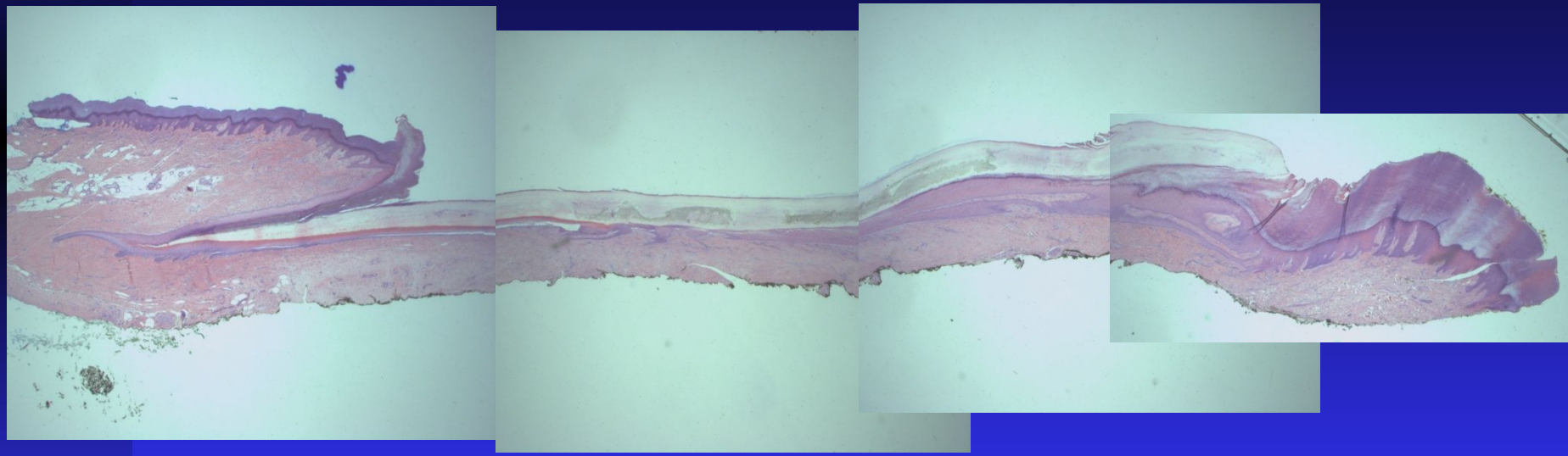
- Clinical correlation often necessary
  - Immunosuppression (esp HIV)
  - If it is destroying bone, it is not benign!
  - Sample more if suspicious

HPV In-situ Hybridization (ISH)

- HPV Subtypes—same as cervical SCC
  - Low risk--Verruca
  - High risk—Squamous cell carcinoma
  - Pan HPV test—Benign and malignant

Hutchinson's Sign

Hutchinson's Sign



Hutchinson's Sign

- J Am Acad Dermatol. 2001 Feb;44(2):305-7.
- **Two kinds of Hutchinson's sign, benign and malignant.**
- Kawabata YKawabata Y, Ohara KKawabata Y, Ohara K, Hino H, Tamaki K.
- Department of Dermatology, Faculty of Medicine, University of Tokyo, Japan. KAWABATA-der@h.u-tokyo.ac.jp
- We examined 6 subungual melanomas in situ and 18 melanocytic nevi and compared pigmentation of the nail plates and hyponychium with the use of a dermatoscope. Hutchinson's sign on the hyponychium was not always evidence of subungual melanoma because it can be seen in both diseases. However, there was a wide difference in their dermatoscopic features. We believe that observation of pigmentation on the hyponychium with the use of a dermatoscope contributes to the

Dr. Rich's Differential Diagnosis

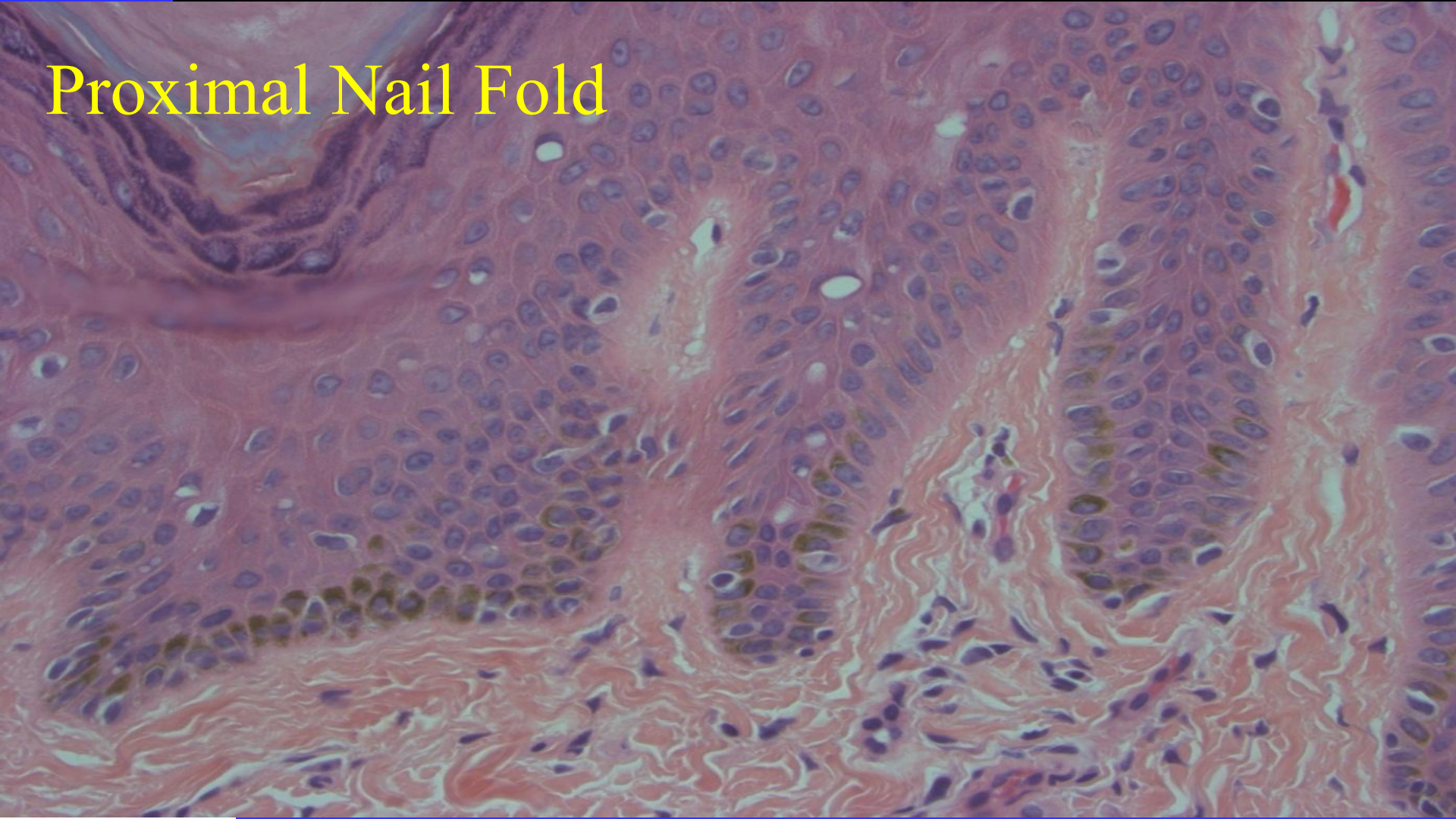
- Trauma pigment
- Nevus
- Lentigo
- R/O Melanoma

Biopsy

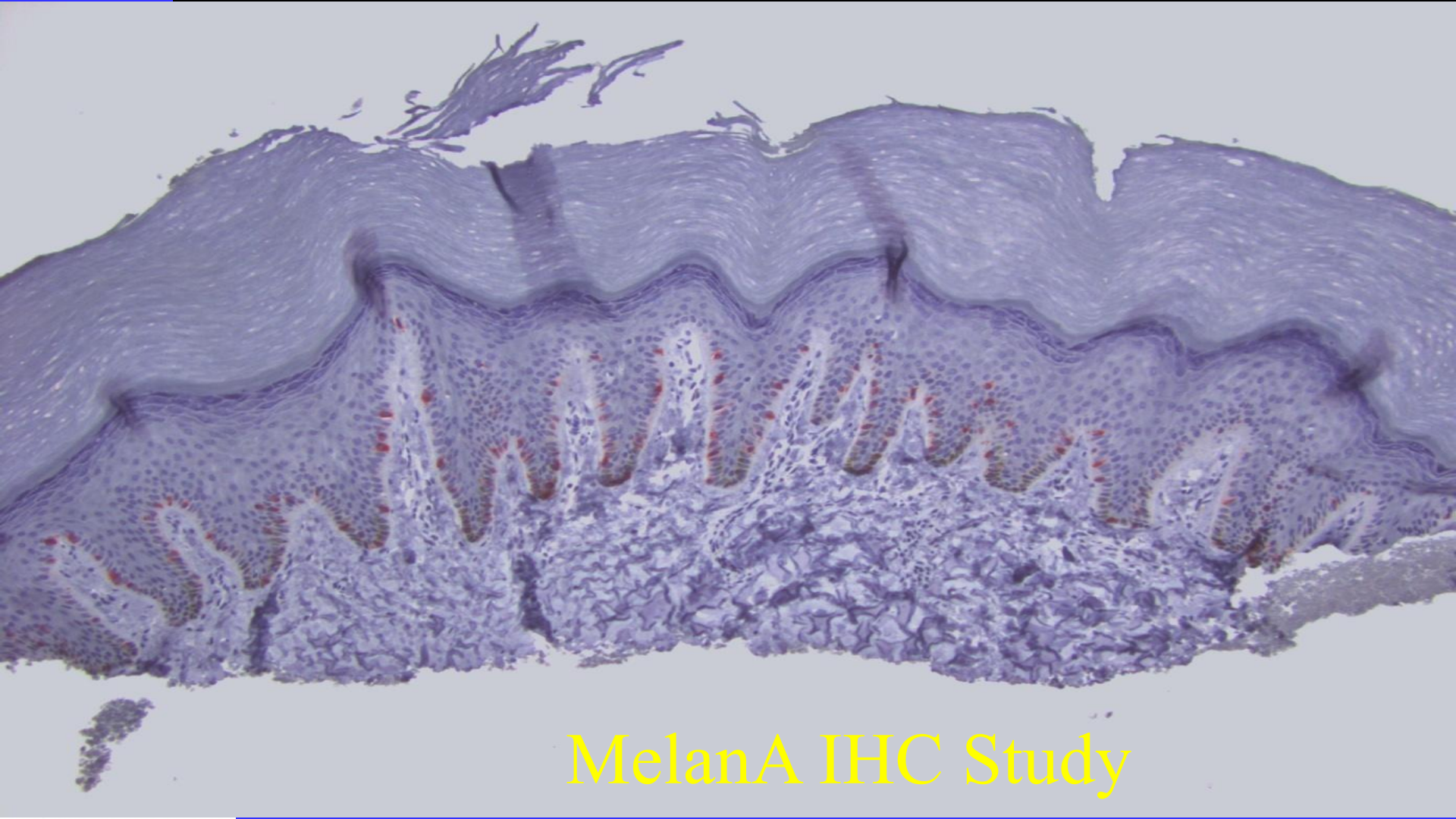
- Nail plate reflected and matrix sampled
- Proximal nail fold sampled

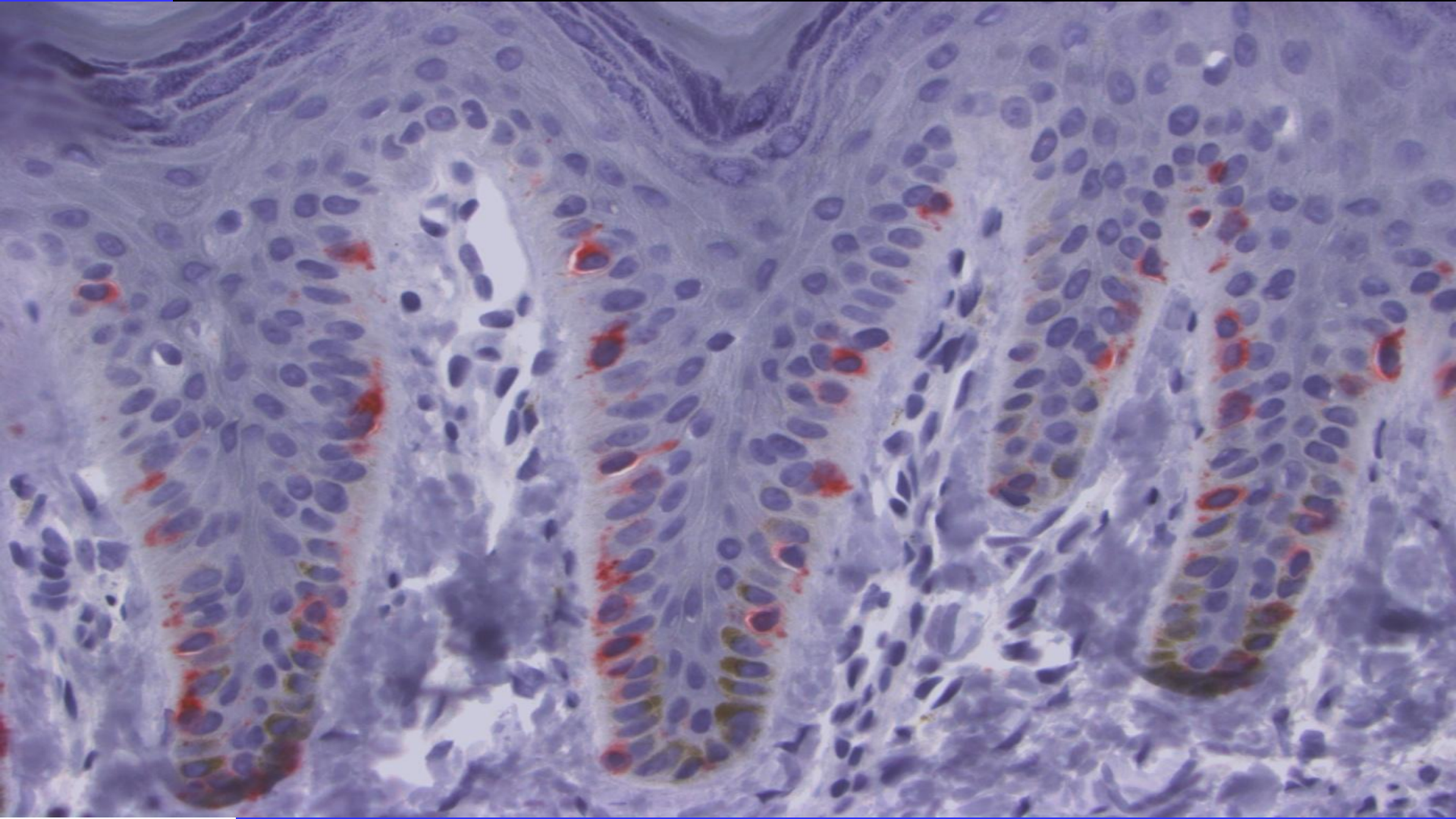
Proximal Nail Fold

Proximal Nail Fold

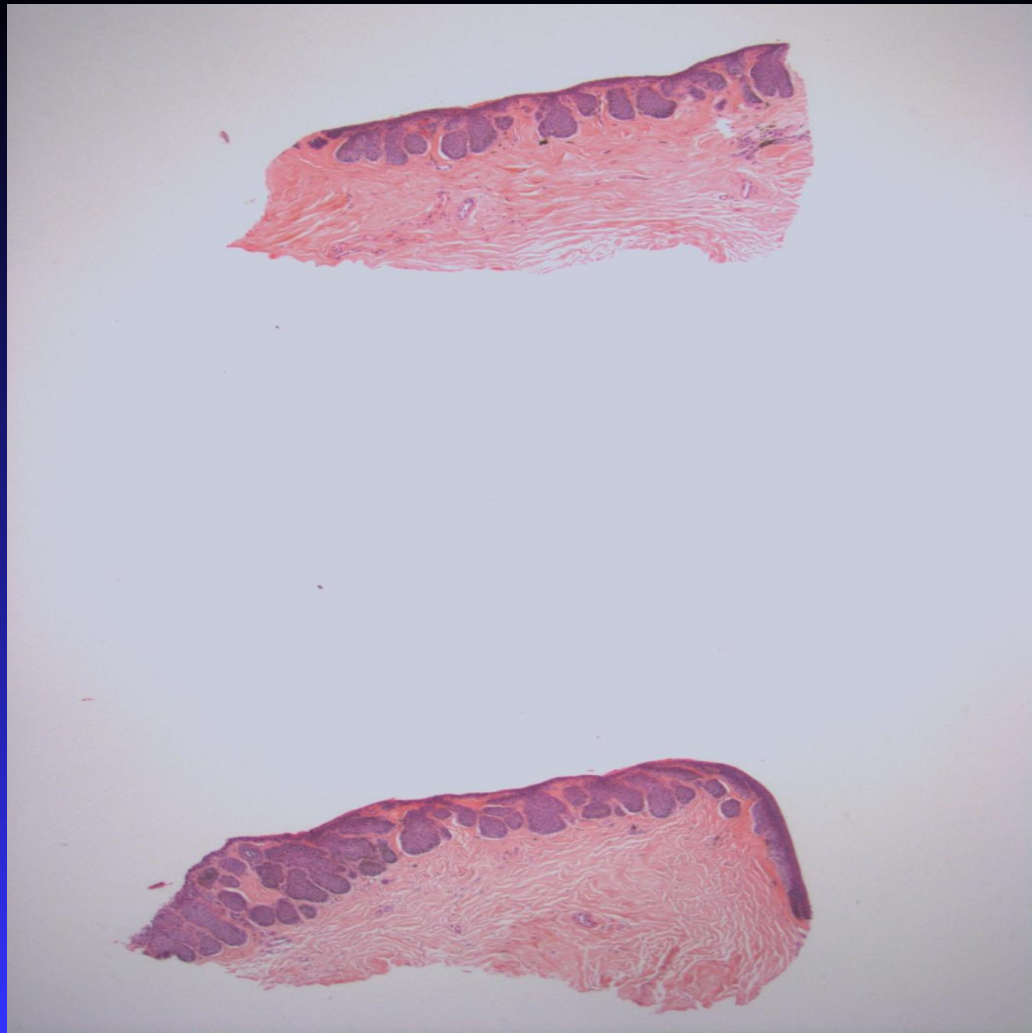


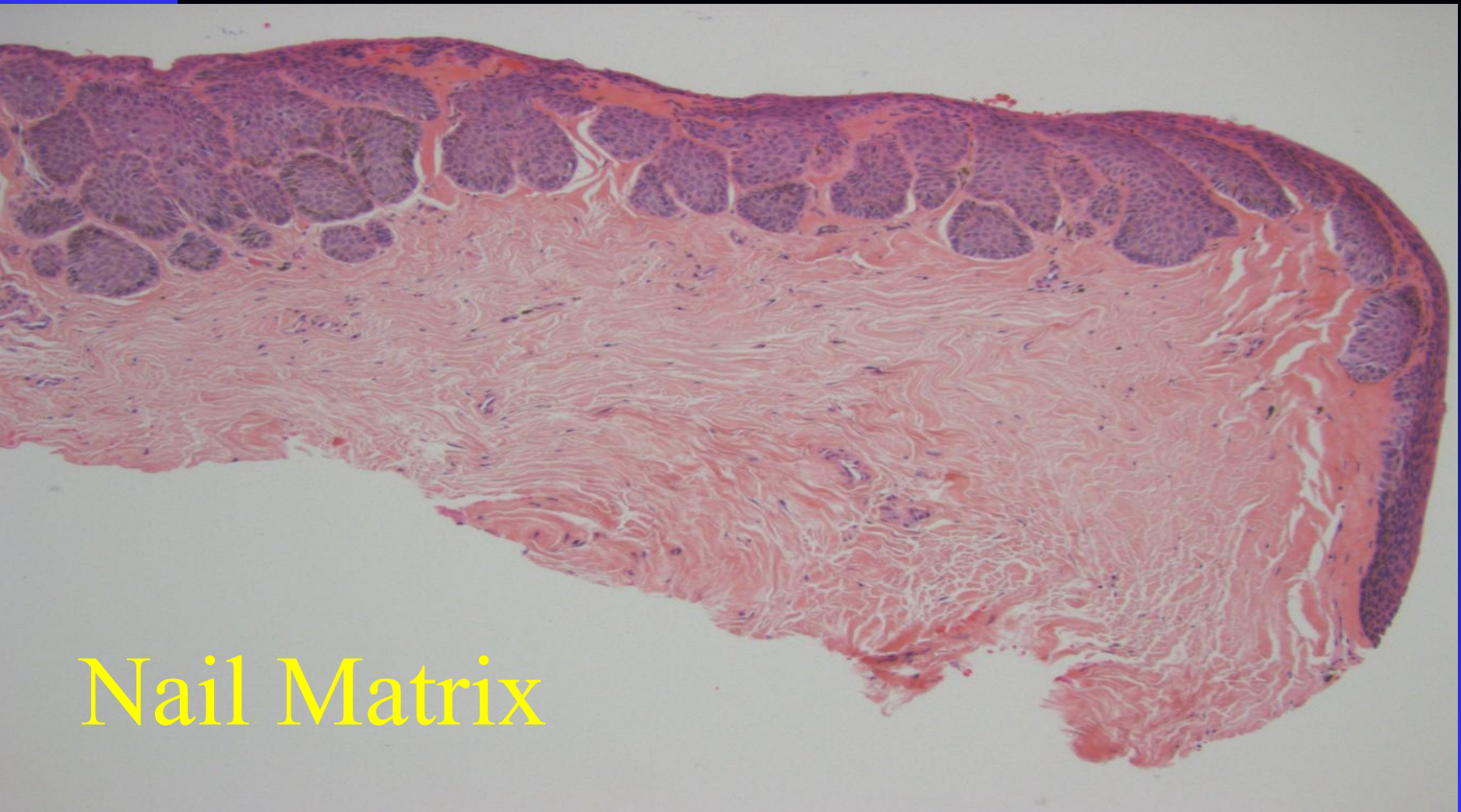

Fontana-Masson Stain


MelanA IHC Study

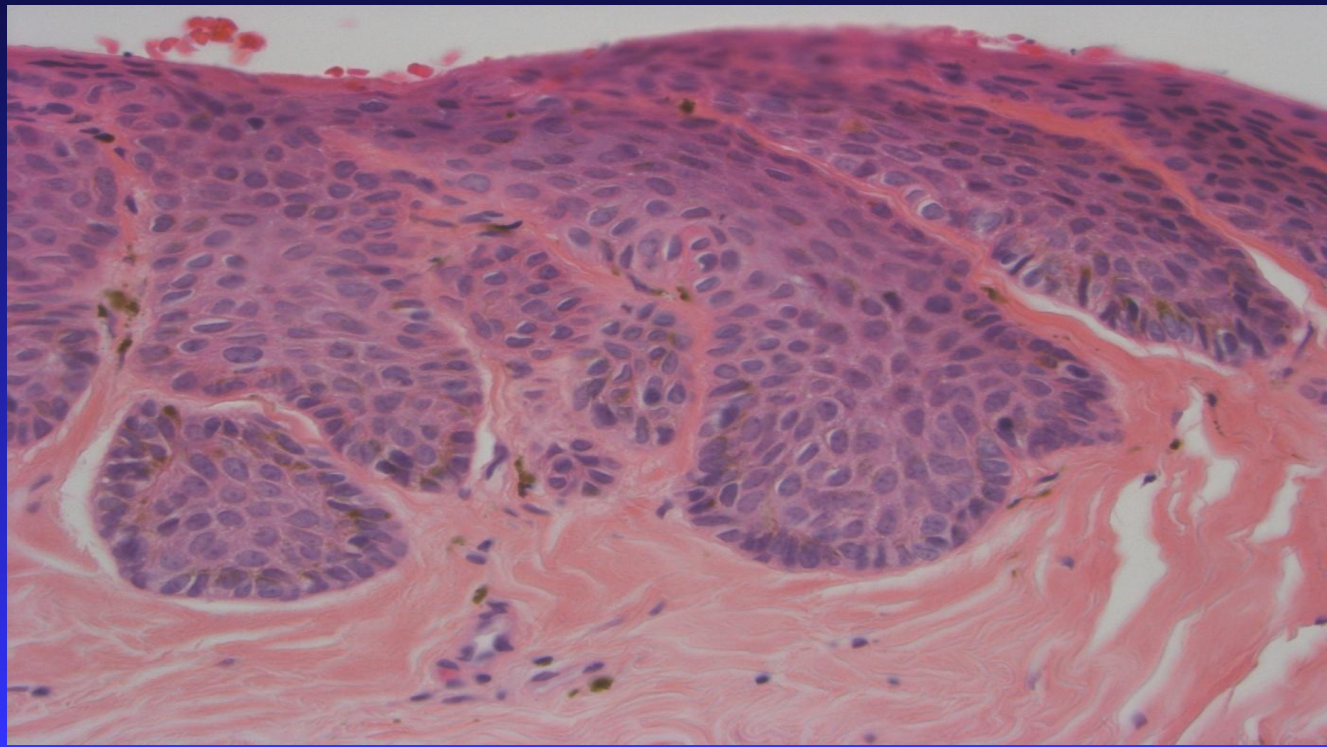


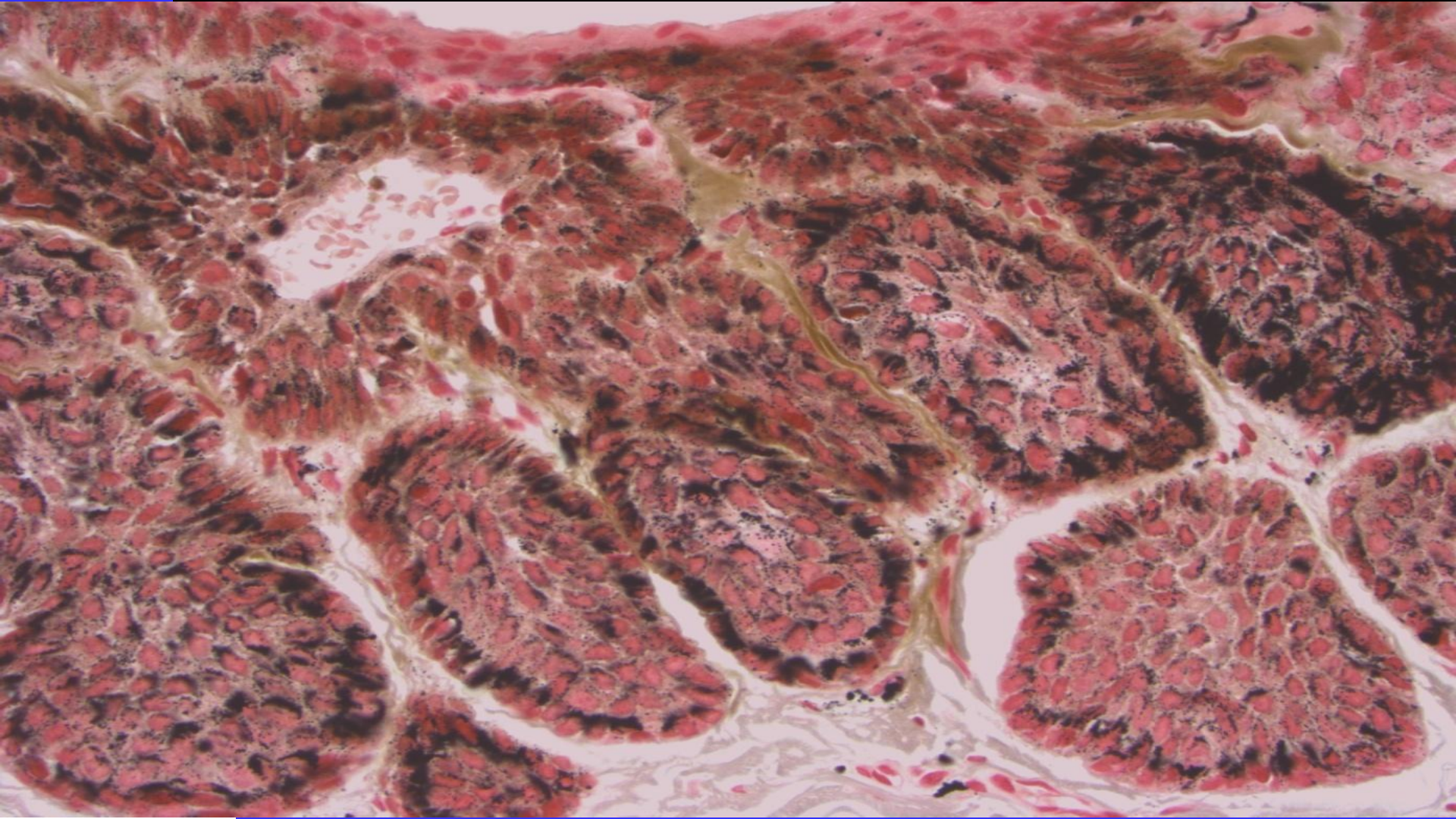
Nail Matrix

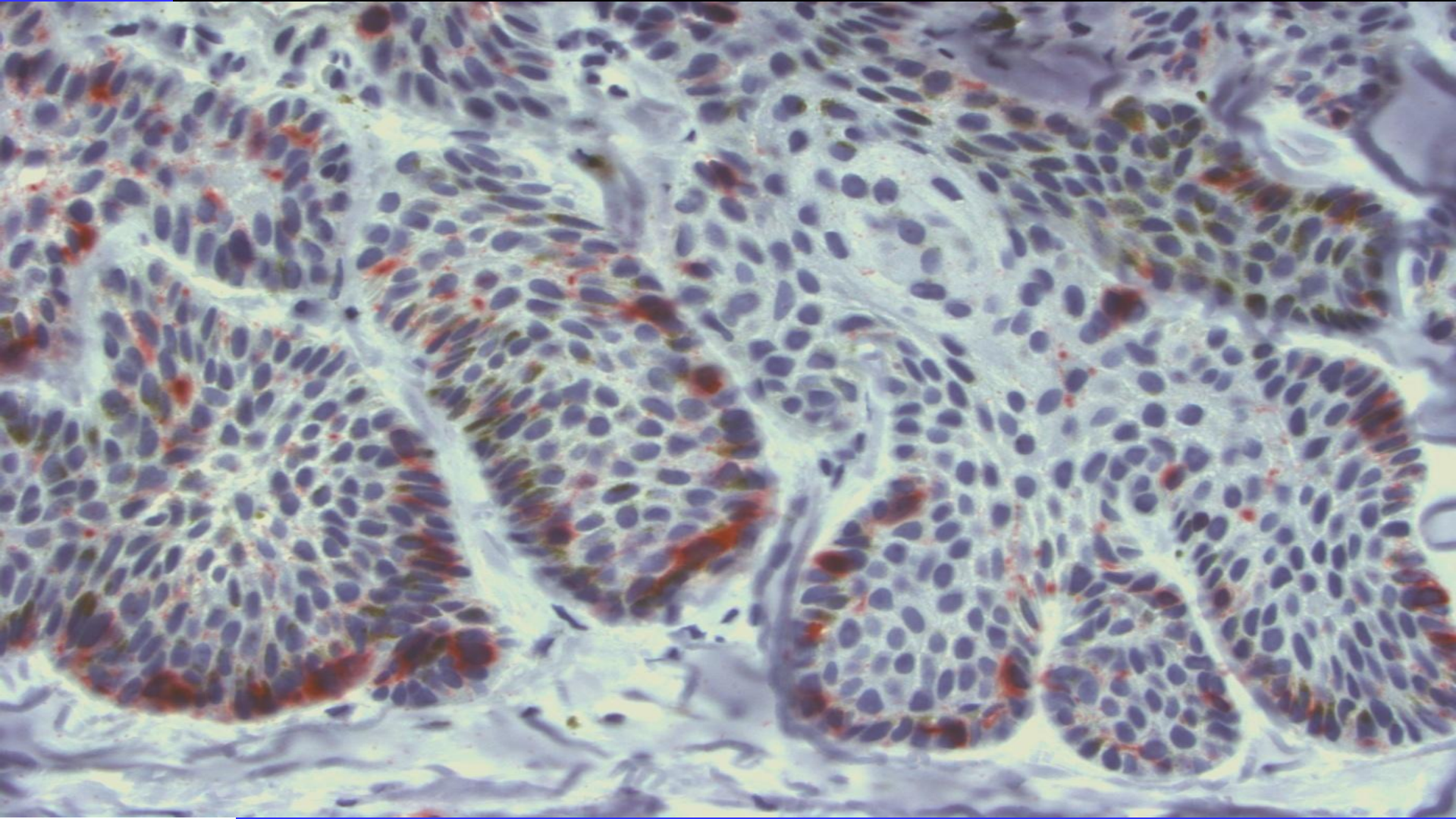


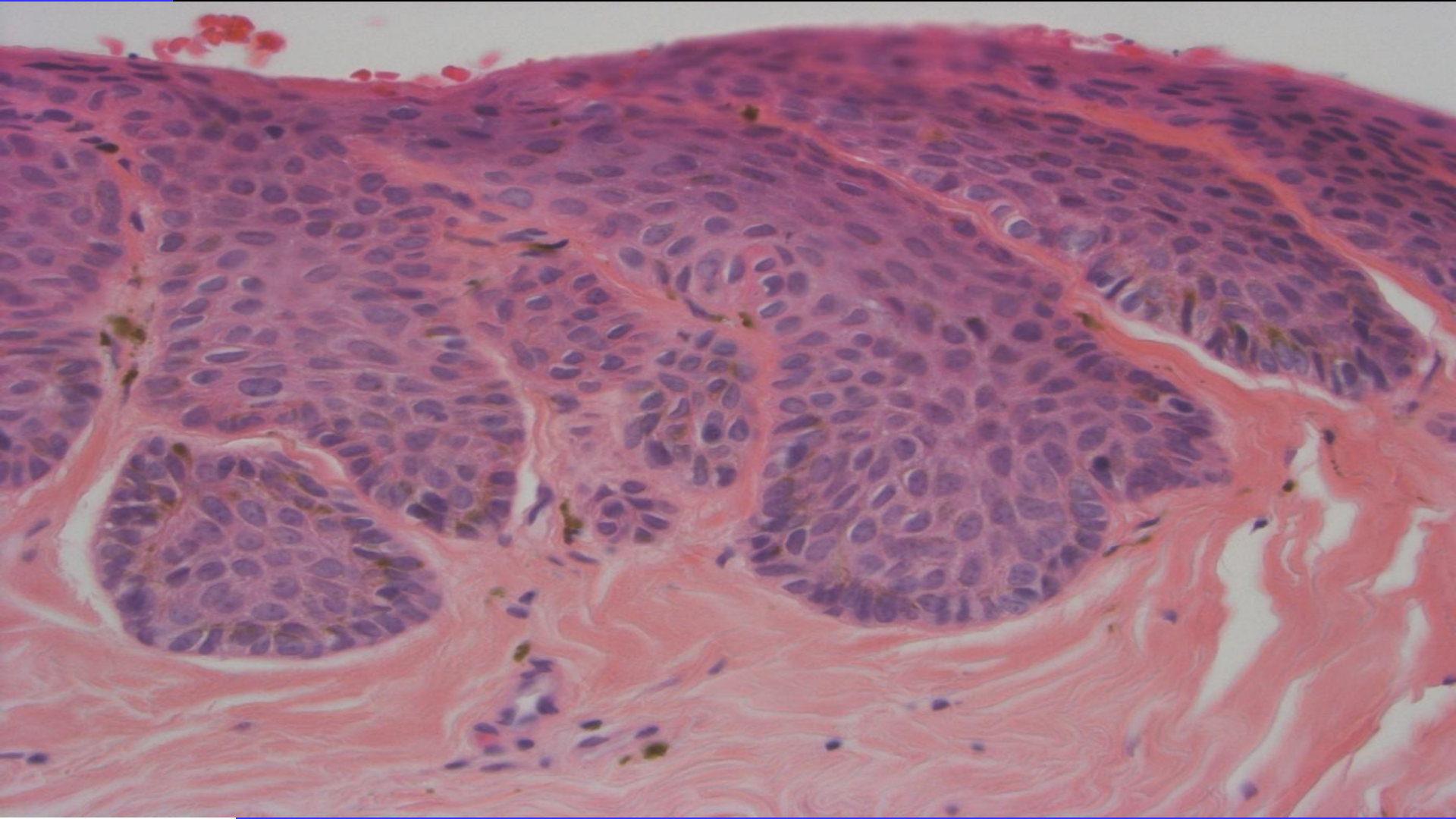

Nail Matrix

Onychocytic matricoma with a Hutchinson's sign









Onychocytic Matricoma

Am J Dermatopathol. 2012 Feb;34(1):54-9. doi: 10.1097/DAD.0b013e31822c3d8b.

Onychocytic matricoma presenting as pachymelanonychia longitudinal. A new entity (report of five cases).

Perrin C[1], Cannata GE, Bossard C, Grill JM, Ambrossetti D, Michiels JF.

Author information

Abstract

Among the tumors of the epidermal appendages, only rare tumors have been proved as differentiating in the direction of the nail. Beside onychomatricoma, we report a new matrical tumor of the nail: onychocytic matricoma (acanthoma of the nail matrix producing onychocytes). The main differential diagnosis of onychocytic matricoma is seborrheic keratosis. However, if attention is paid to the nature of the different layers of the tumor and the peculiar microanatomy of the nail matrix, the differentiation is not difficult. Onychocytic matricoma is a localized (monodactylous) longitudinal melanonychia which is slightly raised. The term pachymelanonychia is used to define the 2 clinical features of the tumor. Pachyonychia indicate a localized thickening of the nail plate, and melanonychia indicate its longitudinal pigmented band. Onychocytic matricoma is composed of a basal compartment with a varying admixture of prekeratogenous cells and keratogenous cells. Endokeratinization originating in the deep portion of the tumor and nests of prekeratogenous and keratogenous cells in concentric arrangement are a characteristic feature. Three major patterns can be identified as follows: acanthotic, papillomatous, keratogenous type with retarded maturation. Given the peculiar thickening of the nail plate observed both in pigmented onychomatricoma and onychocytic matricoma, the term pachymelanonychia longitudinal could be proposed to specify clinically these 2 lesions, which the clinician sometimes mistakes for melanoma.

Onychocytic Matricoma

http://archderm.jamanetwork.com/article.aspx?articleid=1819583

Observation | March 2014

Onychocytic Matricoma: A New, Important Nail-Unit Tumor Mistaken for a Foreign Body FREE

Karolyn A. Wanat, MD[1]; Erika Reid, MD[1]; Adam I. Rubin, MD[1]

[1]Department of Dermatology at the Hospital of the University of Pennsylvania, Philadelphia

JAMA Dermatol. 2014;150(3):335-337. doi:10.1001/jamadermatol.2013.6358.

Onychocytic matricoma (OCM) is a benign acanthoma of the nail unit that presents with localized thickening of the nail plate and melanonychia.[1] This newly described entity has suggestive clinical features and distinctive histopathologic changes.

REPORT OF A CASE

A man in his 40s presented with a history of traumatic injury to the nail unit, after which he noted a dark line under the nail, which he assumed to be a splinter. It persisted for 3 years without any notable change. The patient reported removing portions of it when he would clip the nail back.

Physical examination demonstrated a 2-mm-wide black longitudinal streak extending to the distal lunula with localized nail plate thickening on the right second digit (Figure 1A and B). Dermatoscopic findings were consistent with a foreign body under the nail (Figure 1C and D). Nail clippings of the nail plate were performed to sample the distal portion of the lesion and demonstrated parakeratosis associated with pigmentation.

Onychocytic matricoma vs Nail unit seborrheic keratosis

- Semantic difference
- Seborrheic keratosis is very common
- More important is to make sure this is not subtle, pigmented squamous cell carcinoma
- Onychocytic matricoma is a difficult name

COVID-19

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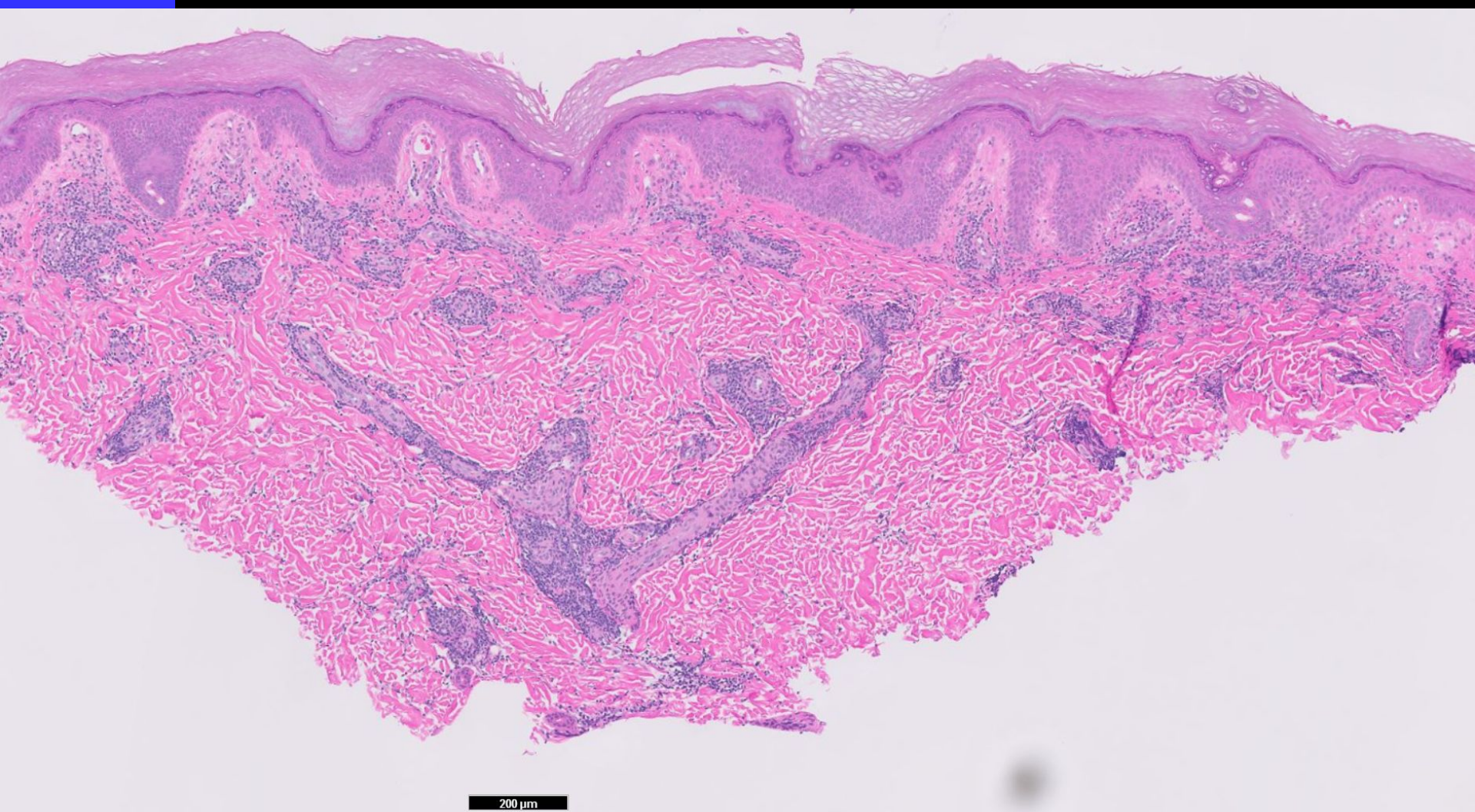
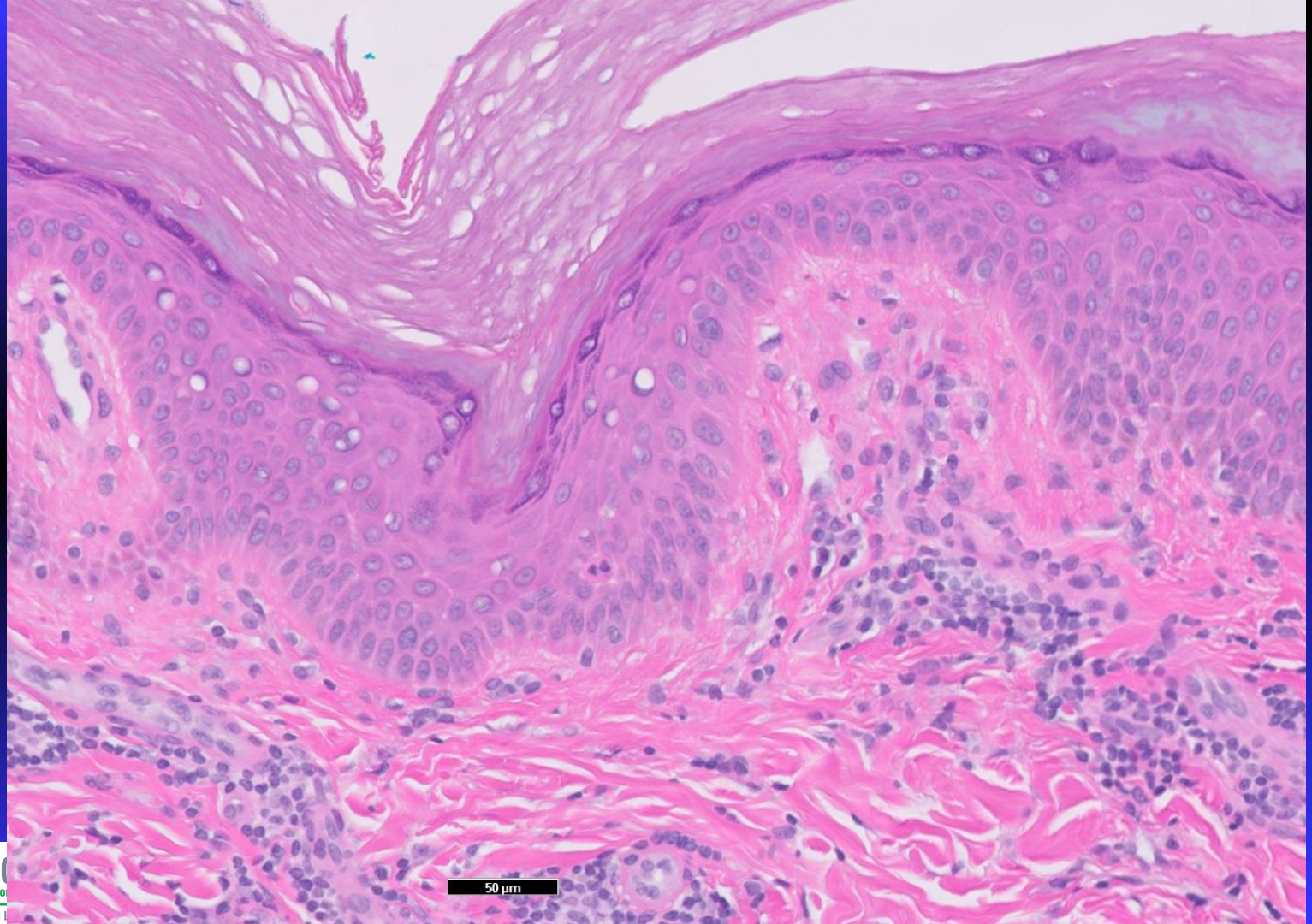
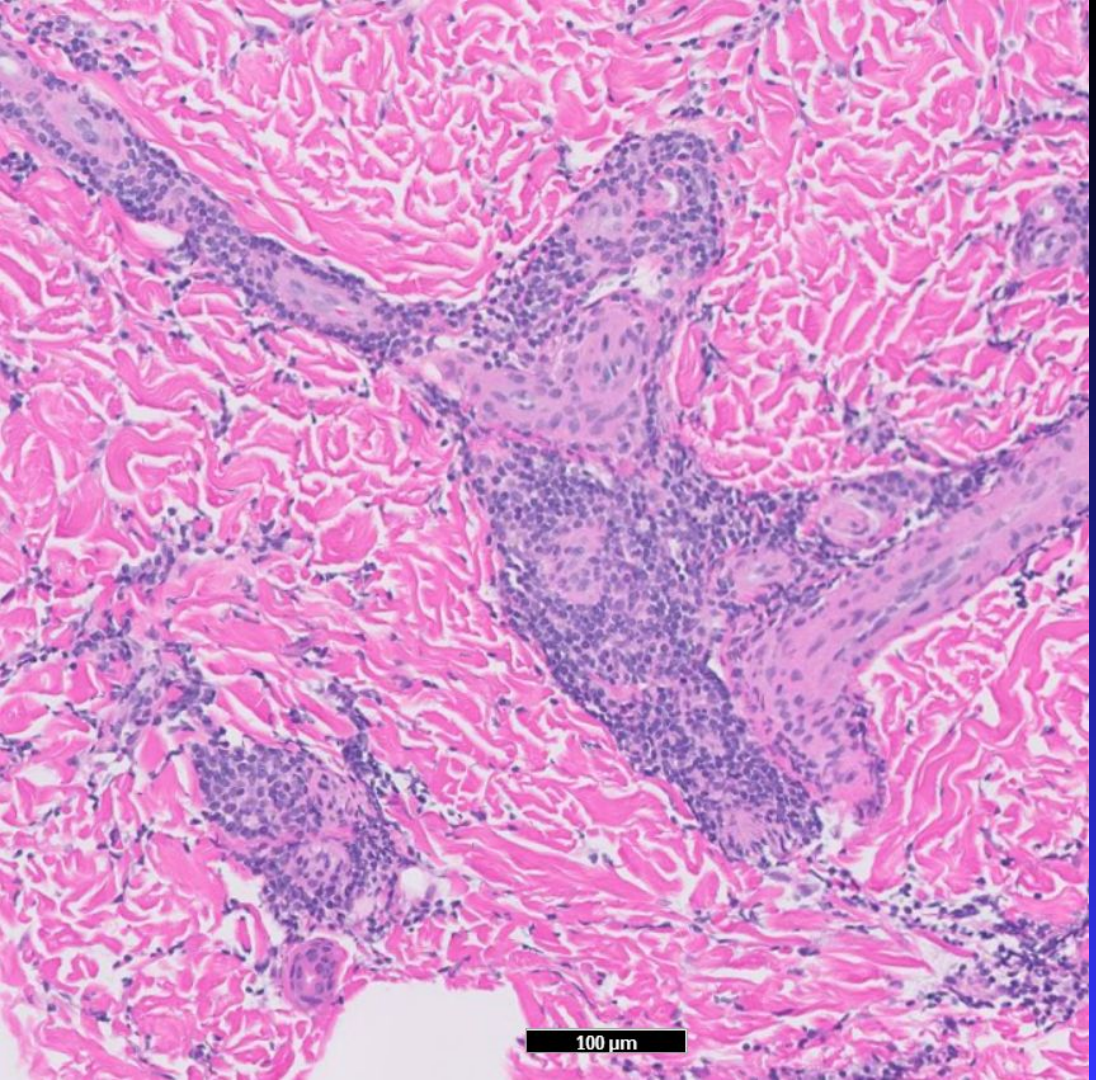
COVID-19

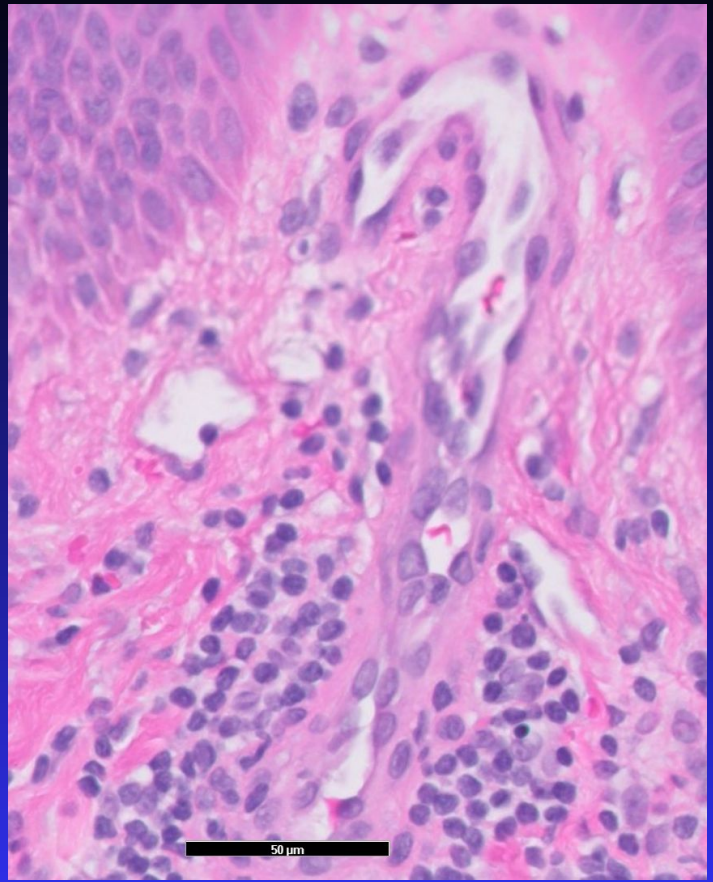
- Mee's lines

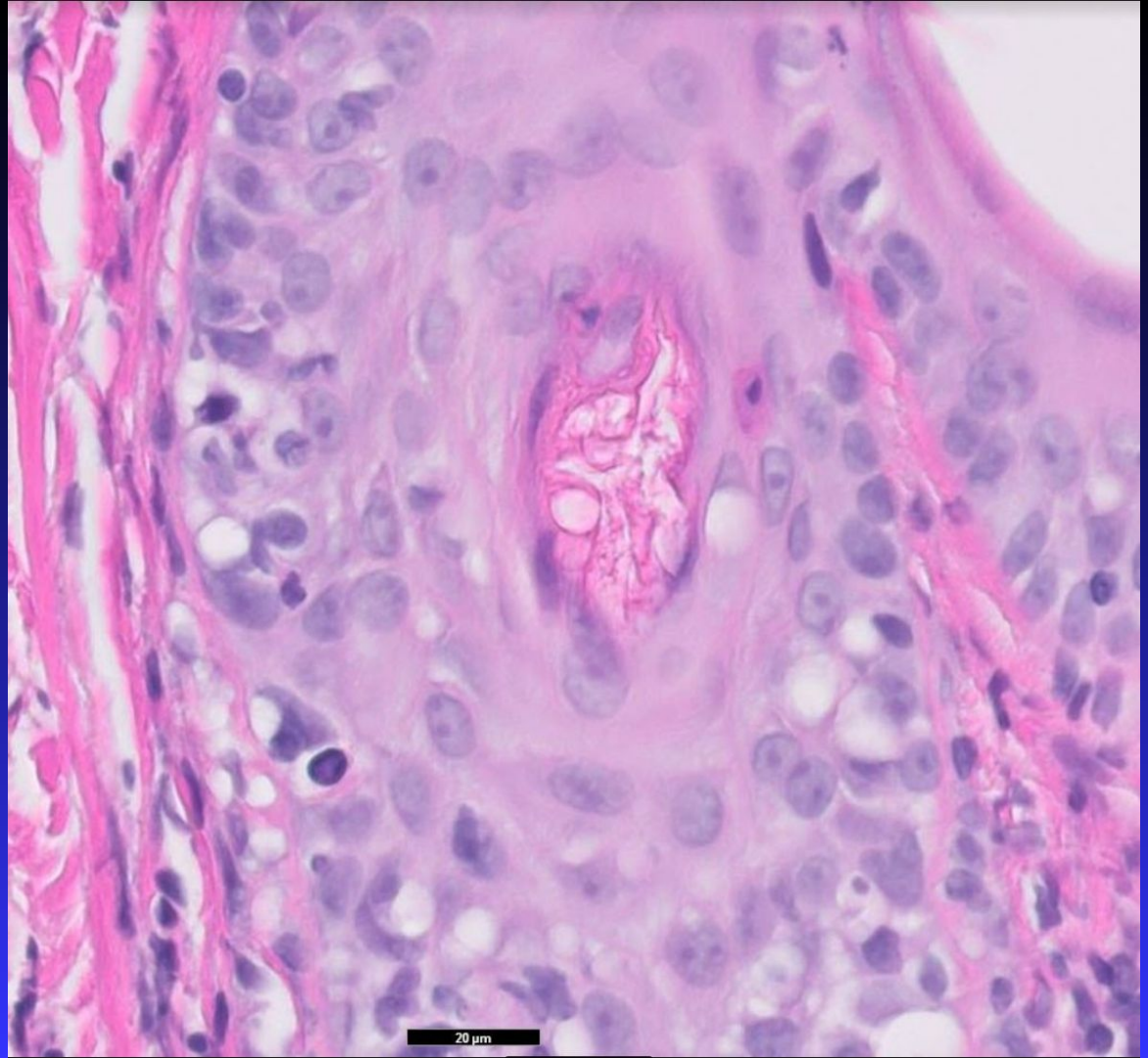
31 y/o male with acute onset of toe papules


200 µm


50 µm


100 µm


50 µm



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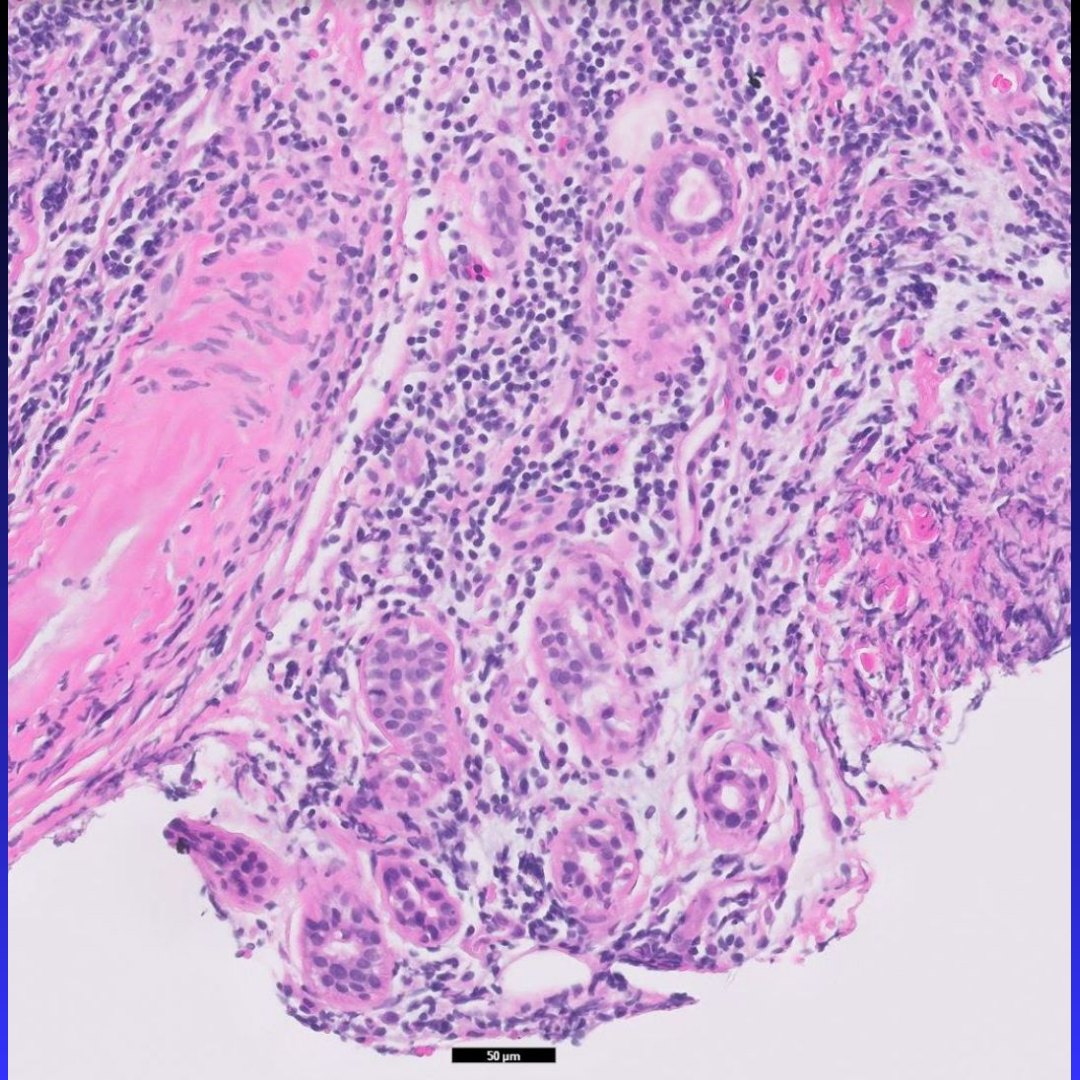

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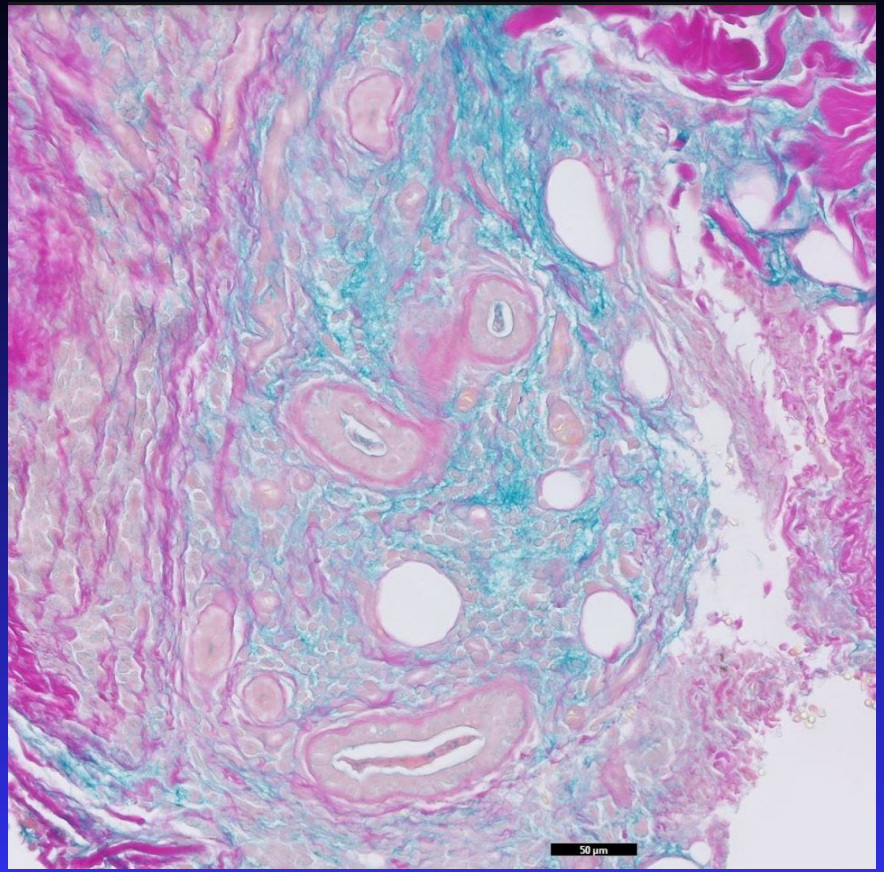

Mucin stain

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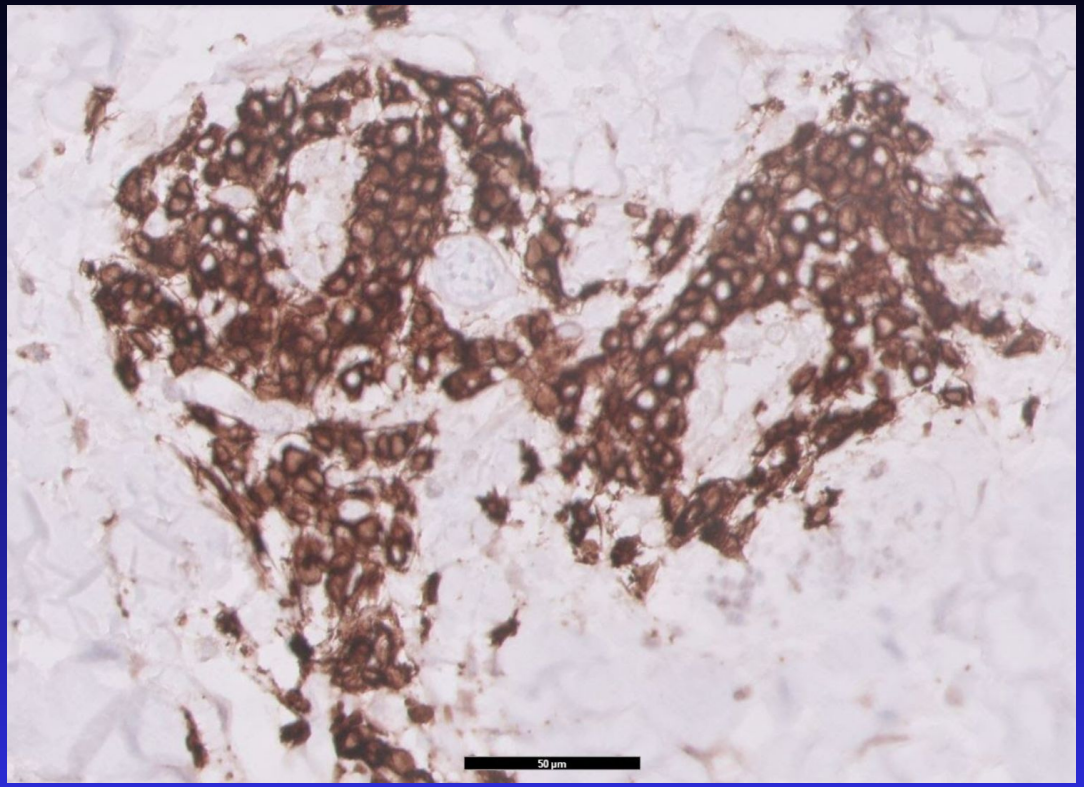

CD123 (Plasmacytoid Dendritic Cell Marker)

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Parvovirus B19 simulating SLE

- Accumulation of nucleic acids after apoptosis

- Transient ANA titer and other serologies
  - RF, anti-DS-DNA, anti-phospholipids,
  - Ribonucleoprotein, Sjögren syndrome A/B
  - Topoisomerase scl-70

- When the ANA is positive, the patient is no longer considered to be infectious.

Parvovirus B19 simulating SLE

Clinical feature	Parvovirus B19 infection	Lupus
Course	Self-limiting	Persistent
Severity	Mild	Mild to severe
Persistent fevers	Rare	May be present
Anemia	Secondary to bone marrow suppression	Secondary to autoimmune hemolysis
Reticulocyte count	Low in presence of bone marrow suppression	Normal to high in presence of evidence of hemolysis
Splenomegaly	Rare	May be present
Discoid lesions, alopecia	Absent	May be present
Oral ulcers	Rare	May be present
Raynaud phenomenon	Absent	May be present
Neurologic (seizures, psychosis, chorea) and ocular symptoms	Rare	May be present
Gastrointestinal involvement (peritonitis, pancreatitis, obstruction/pseudo-obstruction)	Rare	May be present
Cardiac involvement	Rare	May be present
Renal involvement	Rare	May be present

ORIGINAL STUDY

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Comparative Analysis of Chilblain Lupus Erythematosus and Idiopathic Perniosis

Histopathologic Features and Immunohistochemistry for CD123 and CD30

Wang, Michael L. MD, PhD[*]; Chan, May P. MD[*,†] Author Information

The American Journal of Dermatopathology: April 2018 - Volume 40 - Issue 4 - p 265-271
doi: 10.1097/DAD.0000000000000945

BUY

Metrics

Abstract

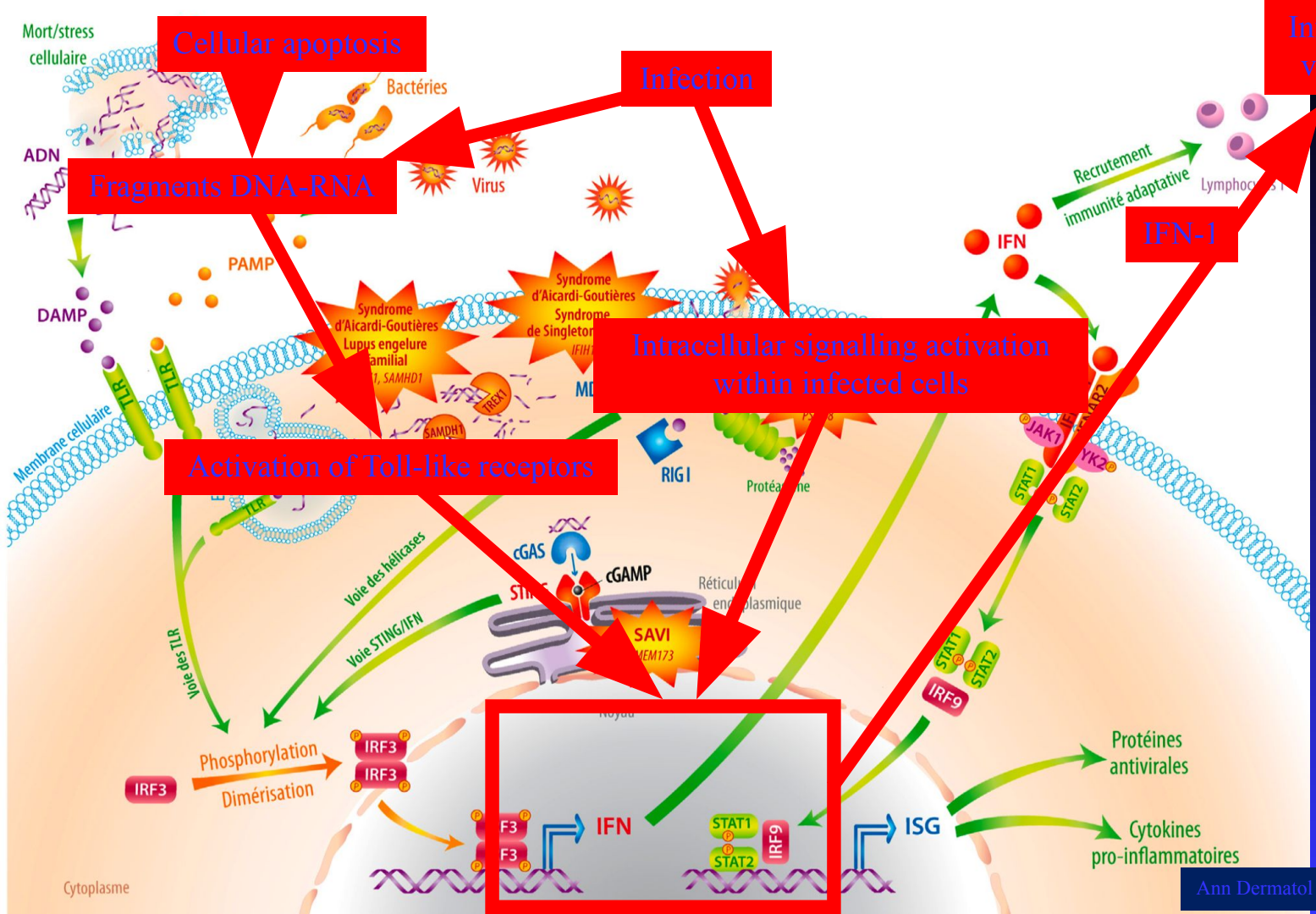
Abstract:

Distinction of chilblain lupus erythematosus (CLE) from idiopathic perniosis (IP) could predict an underlying connective tissue disease; however, histopathologic discrimination of the two is difficult. Increased $CD123^+$ plasmacytoid dendritic cells and $CD30^+$ lymphocytes have been demonstrated in various forms of cutaneous lupus erythematosus and IP, respectively. To our knowledge, CD123 and CD30 have not been examined in CLE. Our objective was to identify helpful histopathologic and

Chilblains vs Chilblain lupus erythematosus

- Chilblain LE is a manifestation of chronic cutaneous LE (CCLE)
  - Discoid lesions on hands/fingers and feet/toes
  - Subungual (nail bed) hyperkeratosis
  - Atropic digital ulcers similar to those seen in systemic sclerosis
  - Proximal nail fold capillary alterations (dermoscopic)
  - Histopathology—Interface dermatitis

- Chilblains—digits are normal
  - Swelling = Papillary dermal edema
  - No interface dermatitis

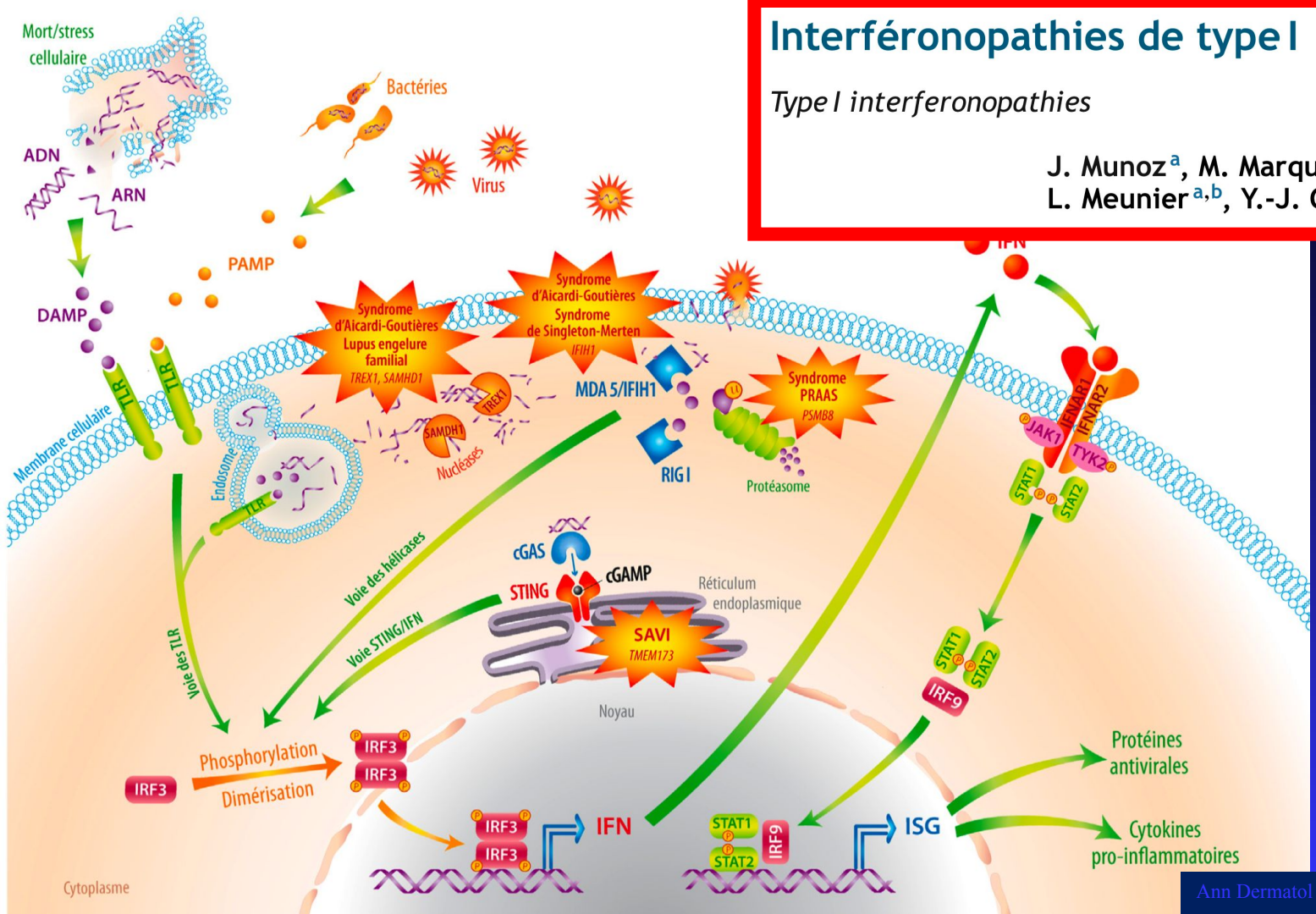


Ann Dermatol Venereol 2015; 142: 653—663

Interféronopathies de type I

Type I interferonopathies

J. Munoz[a], M. Marque[a], M. Dandurand[a], L. Meunier[a,b], Y.-J. Crow[c,d], D. Bessis[e,*,f,g]



Ann Dermatol Venereol 2015; 142: 653—663

Type I Interferonopathy	Major cutaneous findings	Major extra-cutaneous findings	SLE-like
Aicardi-Goutières syndrome (AGS)	Chillblains, digital amputations, ear tissue loss, panniculitis	Severe neurological disease with developmental delay and intracranial calcification	+++
Familial chillblain lupus (FCL)	Chillblains, digital amputations, ear tissue loss	-	+
Spondylenchodrodysplasia (SPENCD)	Chillblains, digital amputations	Skeletal dysplasia, neurological developmental delay with intracranial calcification	+++
Stimulator of interferon genes (STING) - associated vasculopathy with onset in the infancy (SAVI)	Chillblains, digital amputations, ear tissue loss	Interstitial lung disease	+++

Ann Dermatol Venereol 2015; 142: 653—663

Cutaneous histopathological findings of Aicardi–Goutières syndrome, overlap with chilblain lupus

Athanassios Kolivras[1], Alec Aeby[2], Yanick J. Crow[3], Gillian I. Rice[3], Ursula Sass[1] and Josette André[1]

J Cutan Pathol 2008; 35: 774–778

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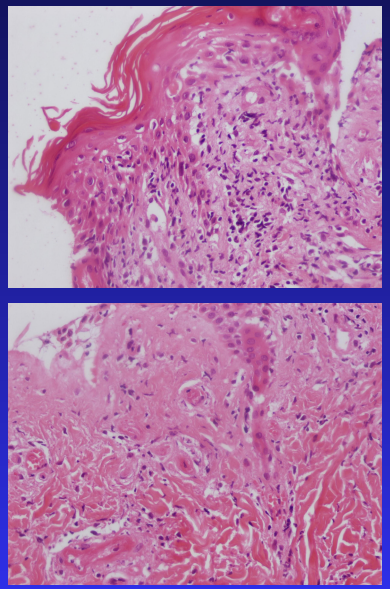

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Aicardi-Goutières syndrome

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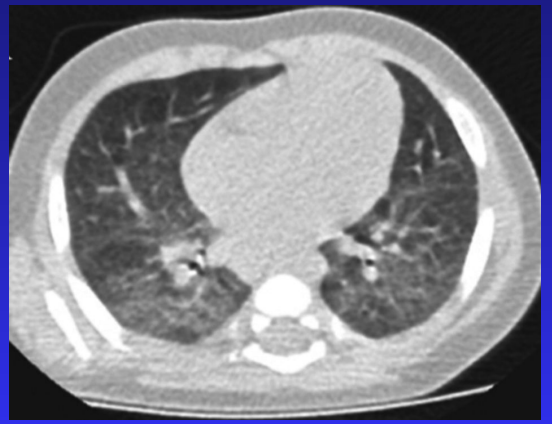
Familial chilblain lupus and Aicardi-Goutières syndrome are allelic phenotypes of the same disease

Aicardi-Goutières syndrome

Familial chilblain lupus

Ann Dermatol Venereol 2015; 142: 653—663

Stimulator of interferon genes (STING) - associated vasculopathy with onset in the infancy (SAVI)



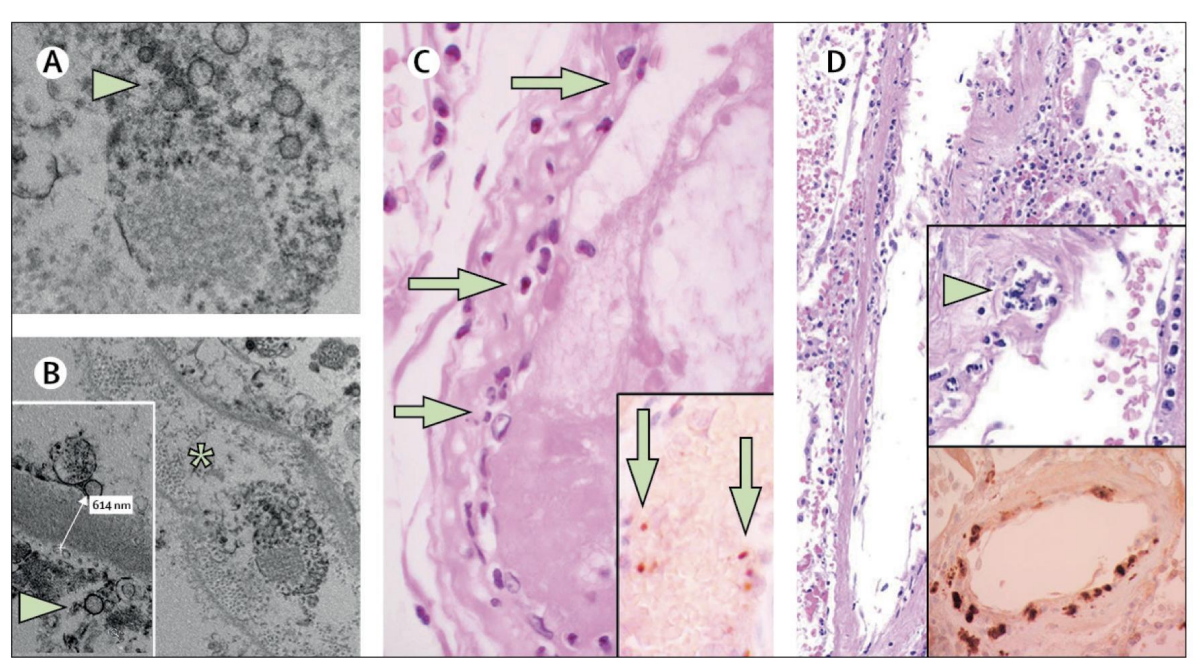
Ann Dermatol Venereol 2015; 142: 653—663
JAMA Dermatol 2015; 151: 872-877

SAVI

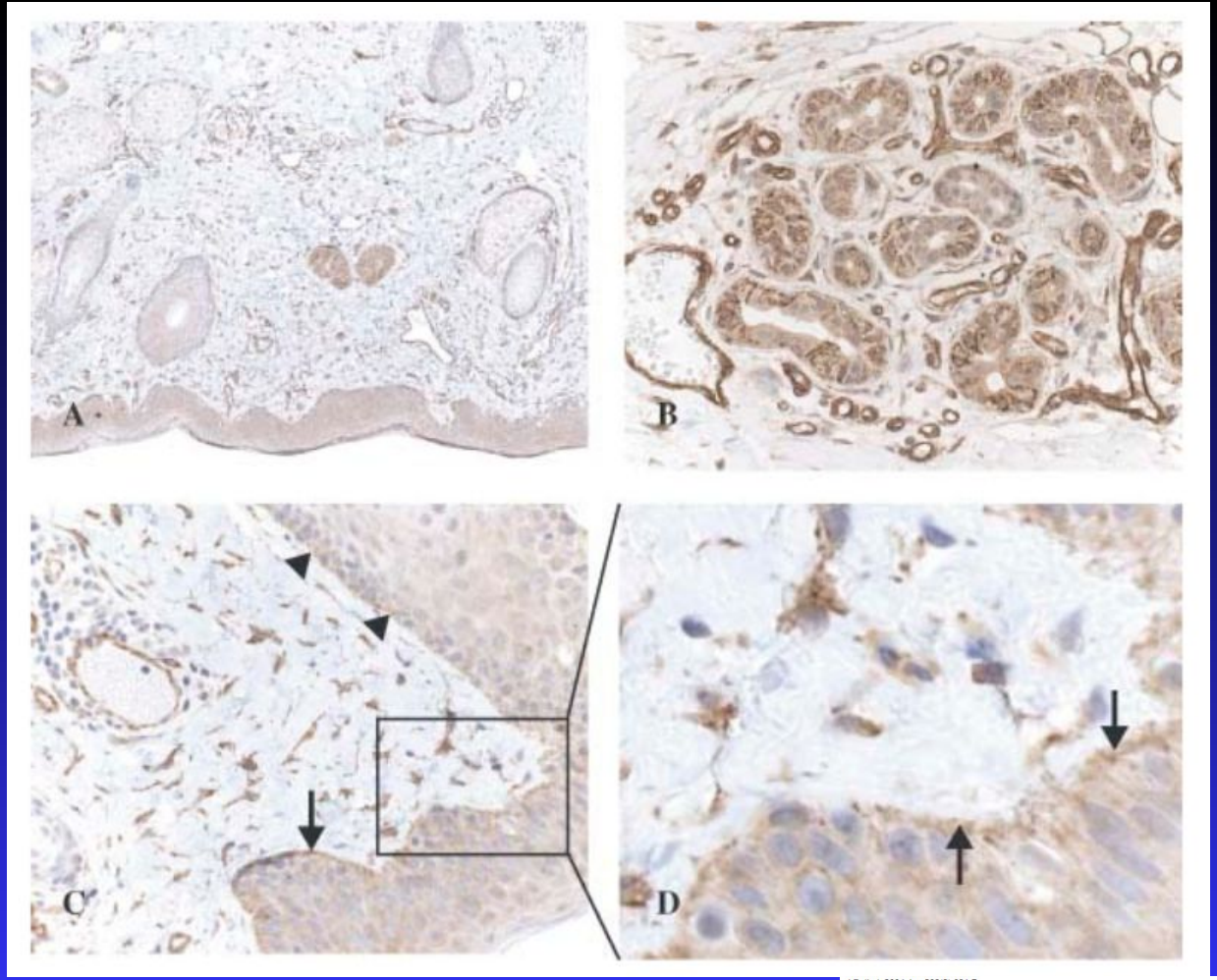
Overlapping features with AGS and chilblain lupus
Differential diagnosis = granulomatosis with polyangiitis

J Am Acad Dermatol 2016; 74: 186-9

Endothelial cell infection and endothelitis in COVID19



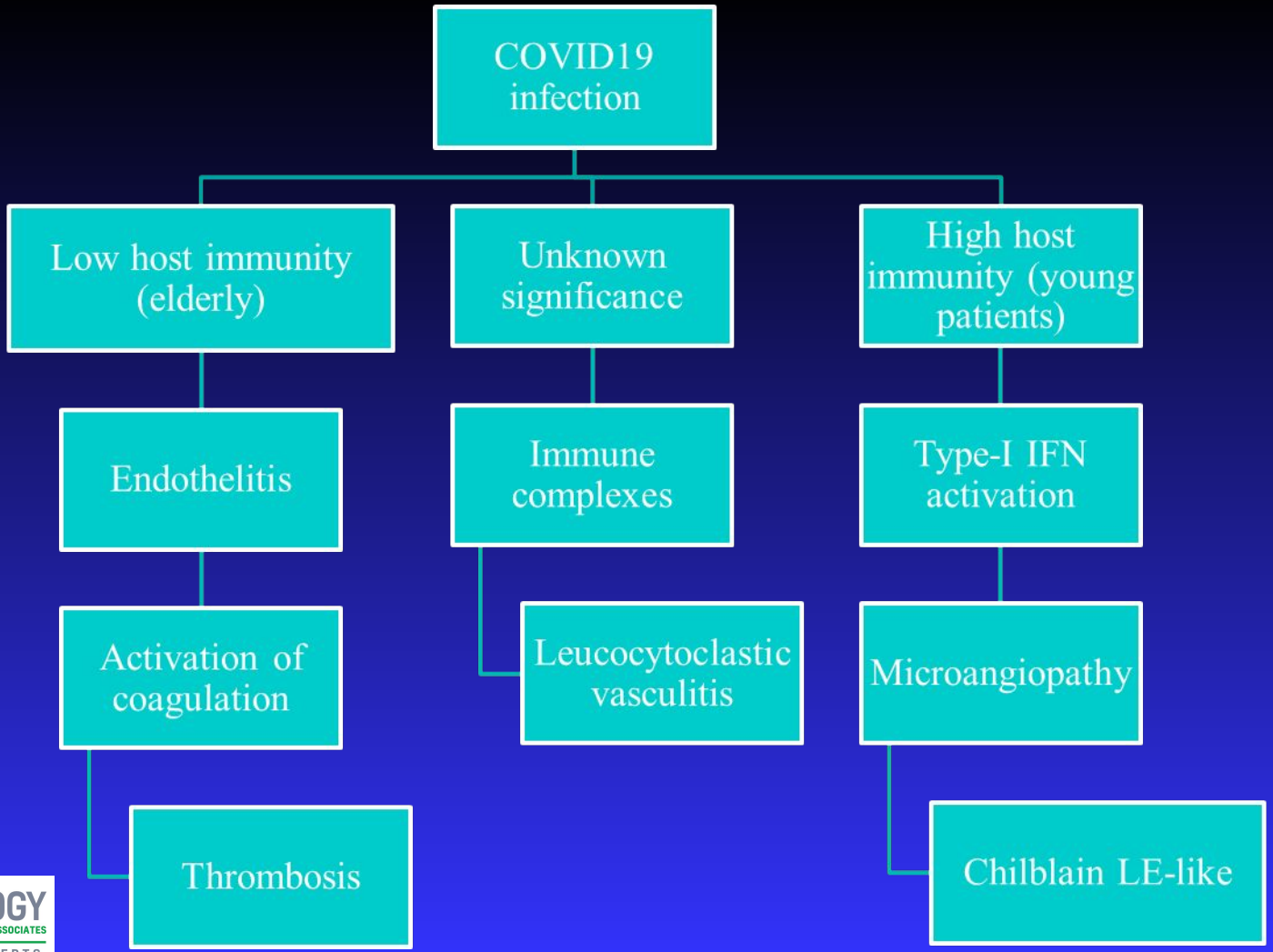
Varga Z, Flammer A, Steiger P, et al. Endothelial cell infection and endotheliitis in COVID-19. The Lancet 2020
https://dx.doi.org/10.1016/s0140-6736(20)30937-5



J Pathol. 2004 Jun;203(2):631-7.

Tissue distribution of ACE2 protein, the functional receptor for SARS coronavirus. A first step in understanding SARS pathogenesis.

Hamming I[1], Timens W, Bulthuis ML, Lely AT, Navis G, van Goor H.



Thanks!

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