### **Nail Pathology Difficult Cases**

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CTA Pathology

and

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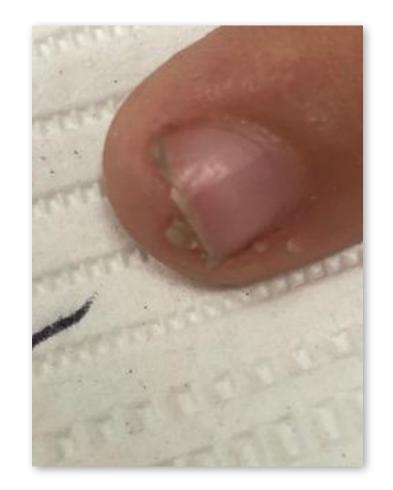
Oregon Health and Sciences University

Portland, Oregon, USA





Right index finger of 8 y/o male



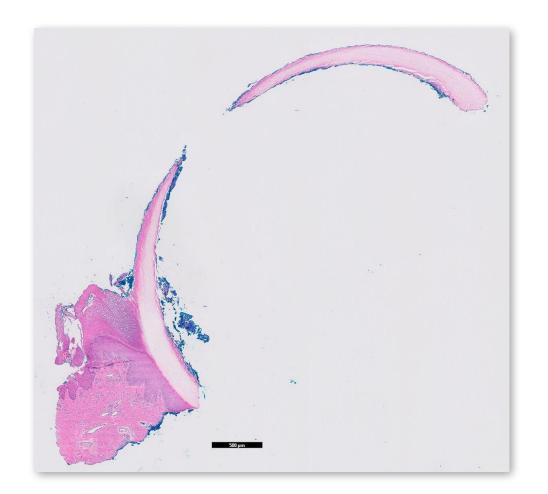






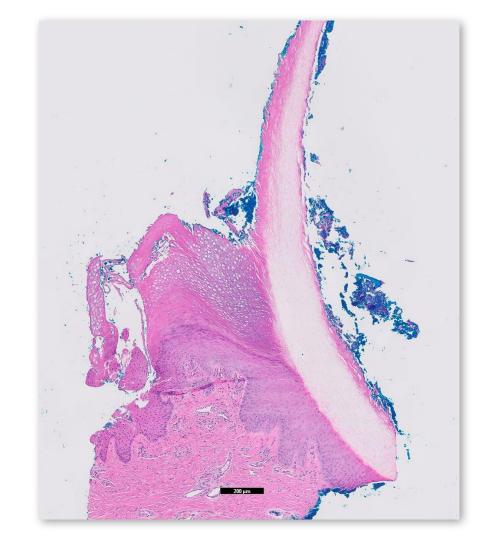






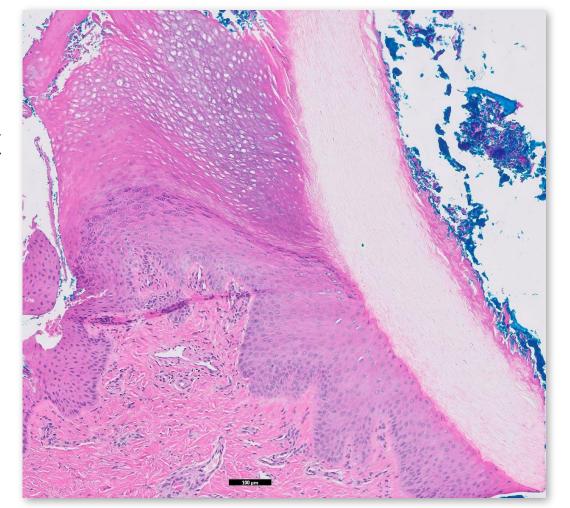






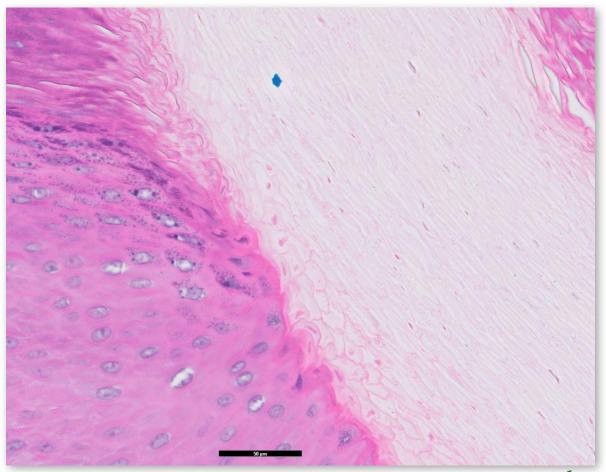
















#### **Onychoheterotopia (Ectopic Nail)**

- •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.





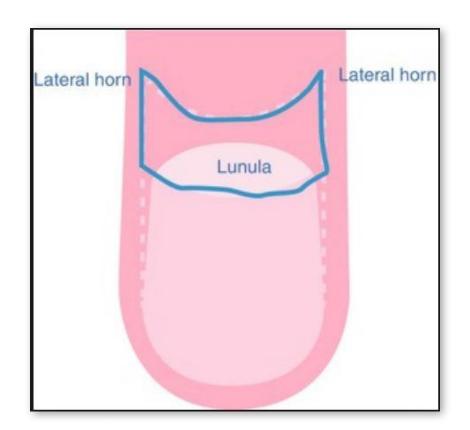
#### Traumatic ectopic nail







#### **Matrix horns**







#### **Onychoheterotopia (Ectopic Nail)**

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





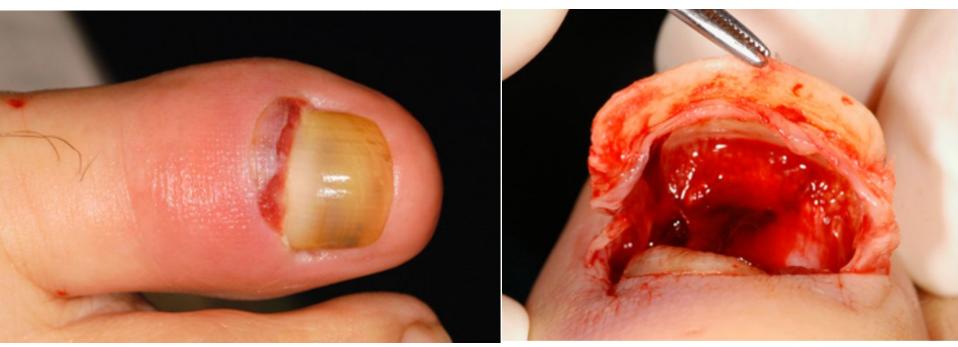
#### **Onychoheterotopia (Ectopic Nail)**

- Differential:
  - Retronychia
  - Residual nail unit after incomplete excision





## Retronychia

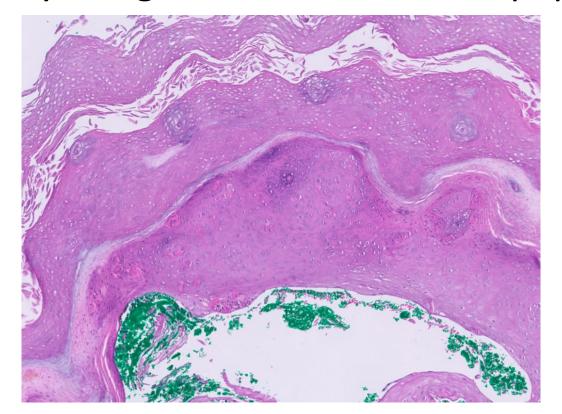


Baumgartner M and Haneke E. Retronychia: Diagnosis and Treatment. Dermatologic Surgery 36(10):1610-4, 2010





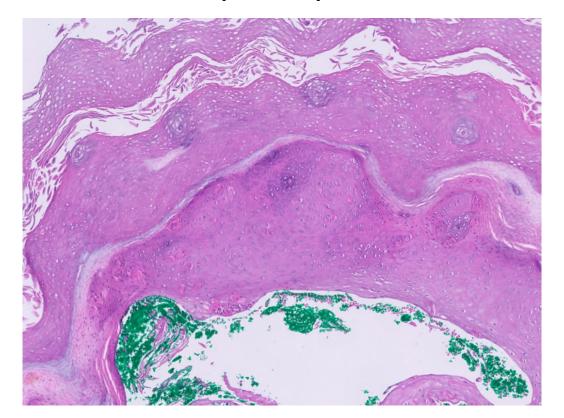
#### Case—73y/o F great toe—"keratotic papule"







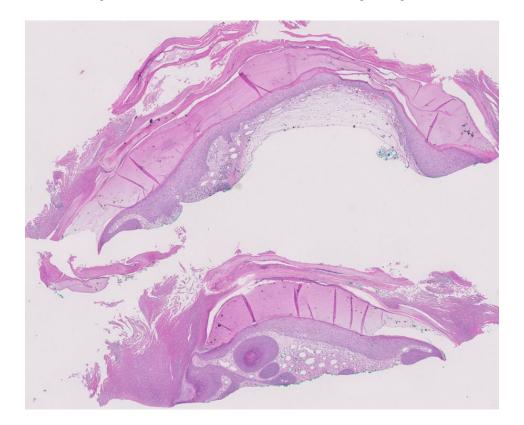
### Case—Dx: Verruca (Wart)







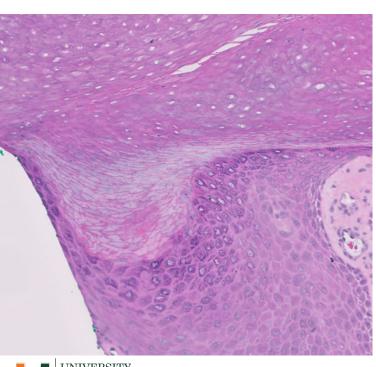
#### Case—Lesion persists—rebiopsy







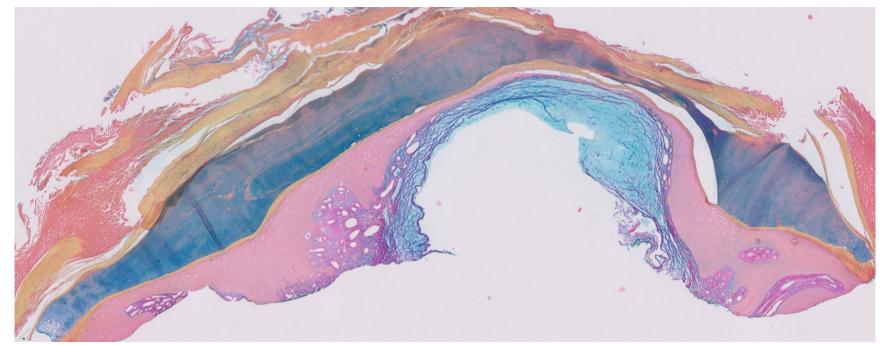
### Case—Lesion persists—rebiopsy







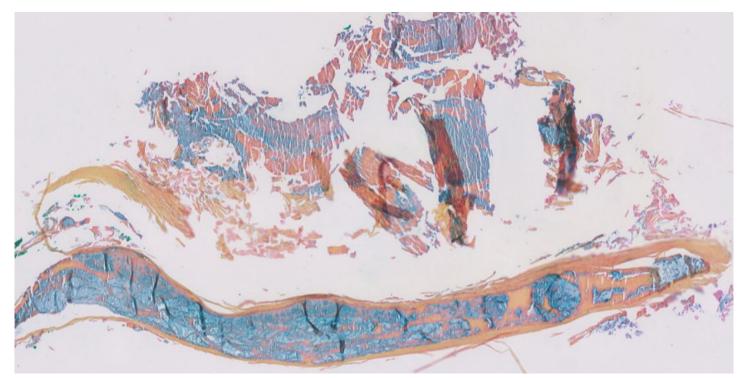
#### Case—Dx: Digital mucous/myxoid cyst







#### Case—Dx: Digital mucous/myxoid cyst

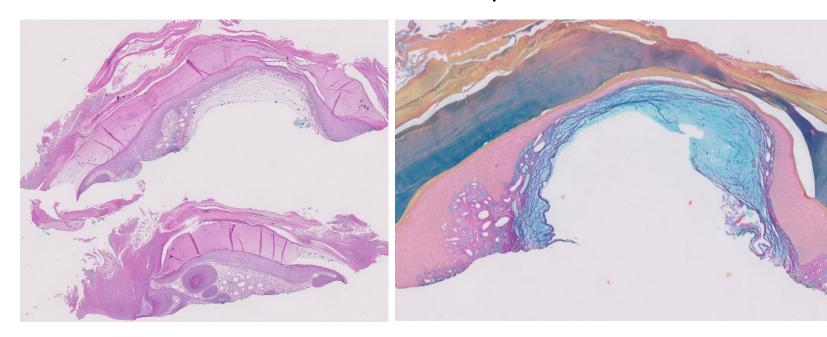






### Digital mucous/myxyoid cyst

•Older lesions look like scar—required mucin stain





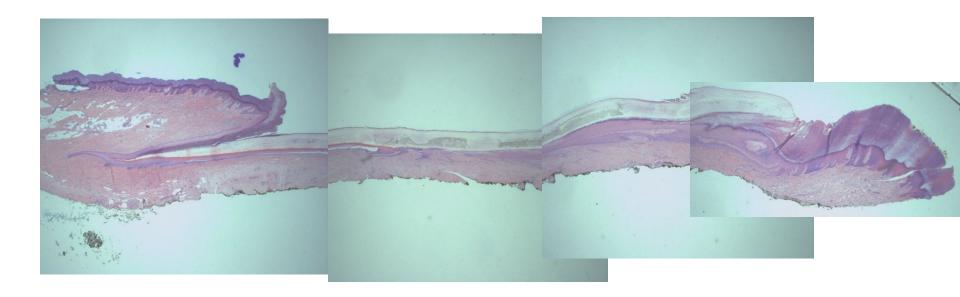


## Hutchinson's Sign





### **Hutchinson's Sign**







#### **Hutchinson's Sign**

- <u>J Am Acad Dermatol.</u> 2001 Feb;44(2):305-7.
- Two kinds of Hutchinson's sign, benign and malignant.
- <u>Kawabata Y</u>, <u>Ohara K</u>, <u>Hino H</u>, 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 precise diagnosis of subungual melanoma.





## Dr. Rich's Differential Diagnosis

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





# **Biopsy**

- Matrix sampled
- Proximal nail fold sampled

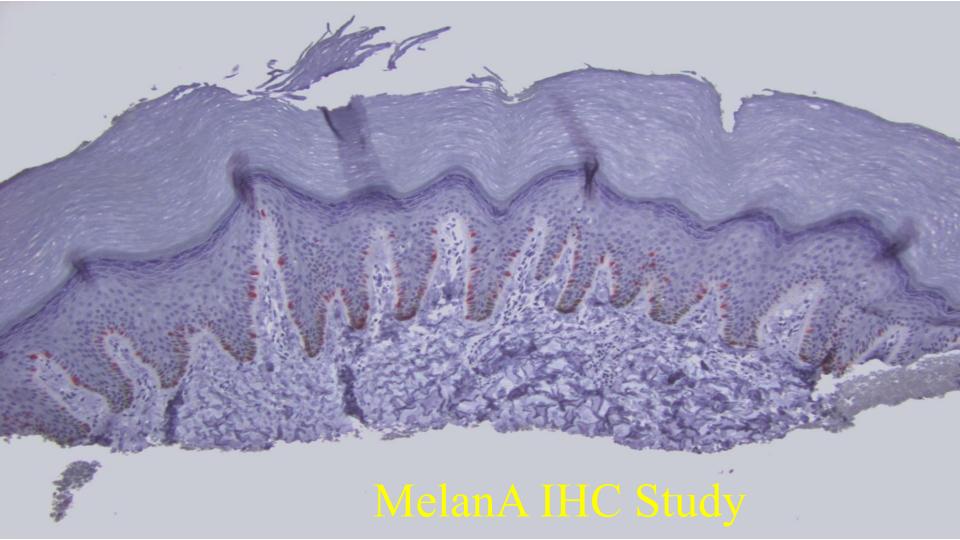


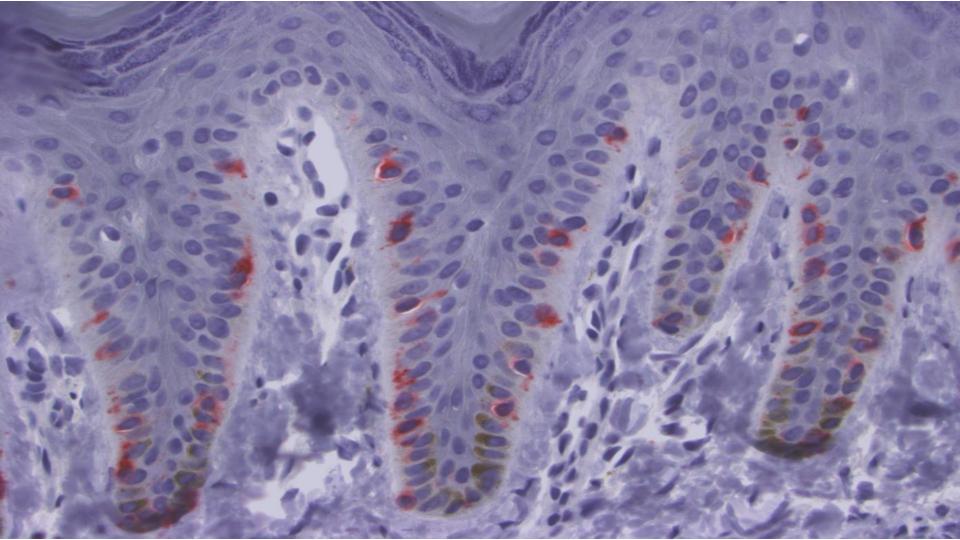




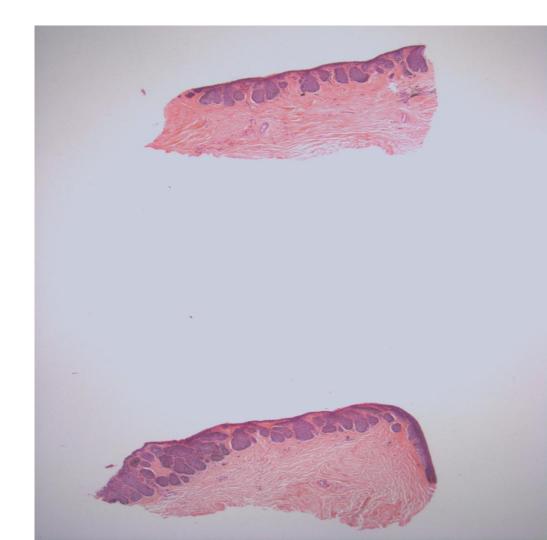








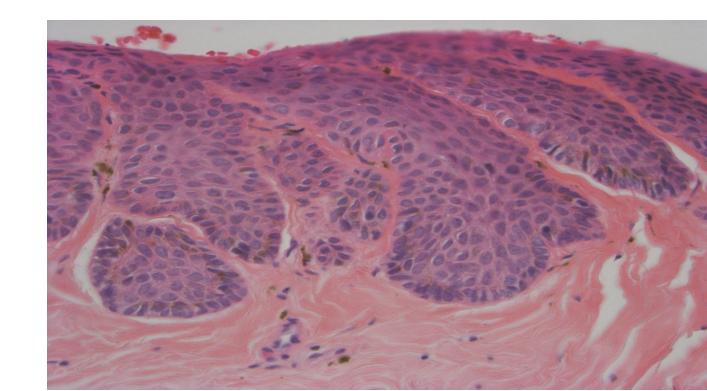
#### Nail Matrix



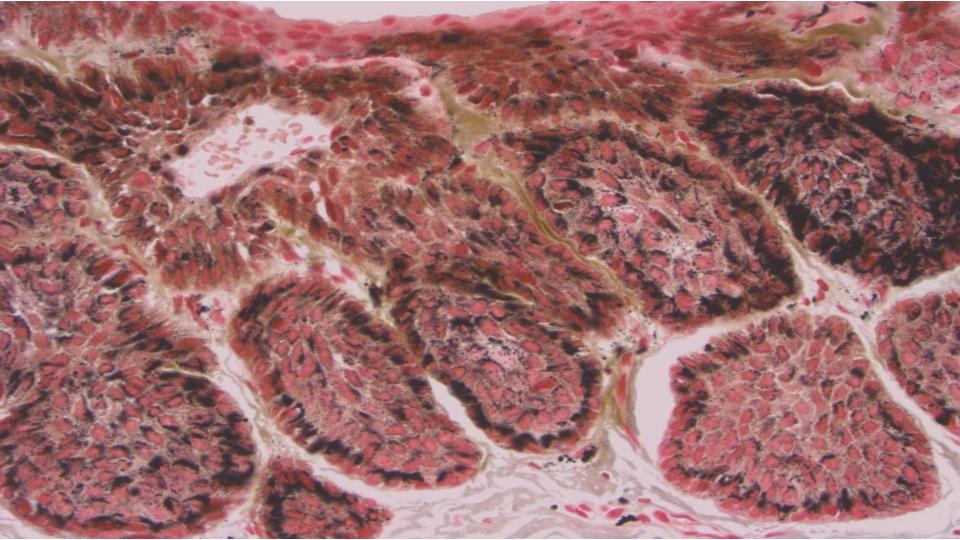


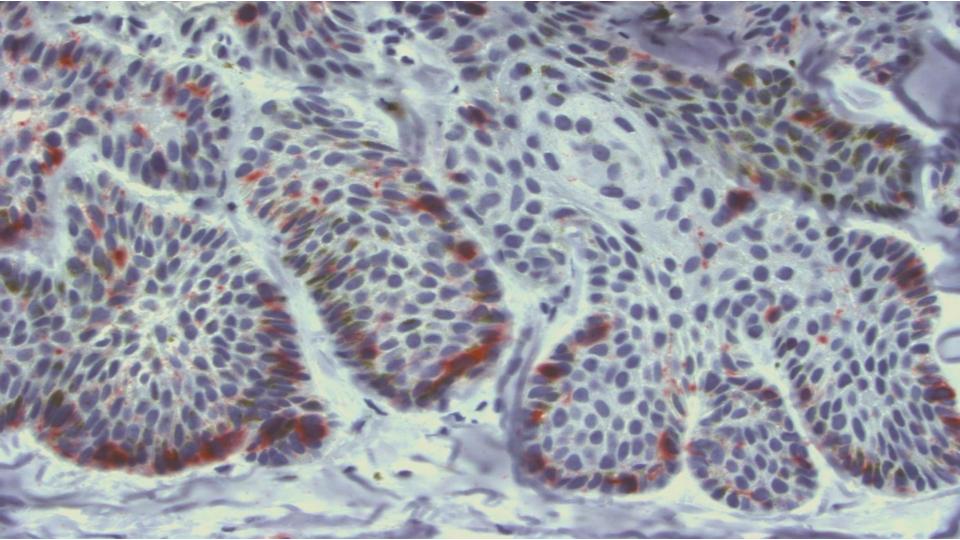


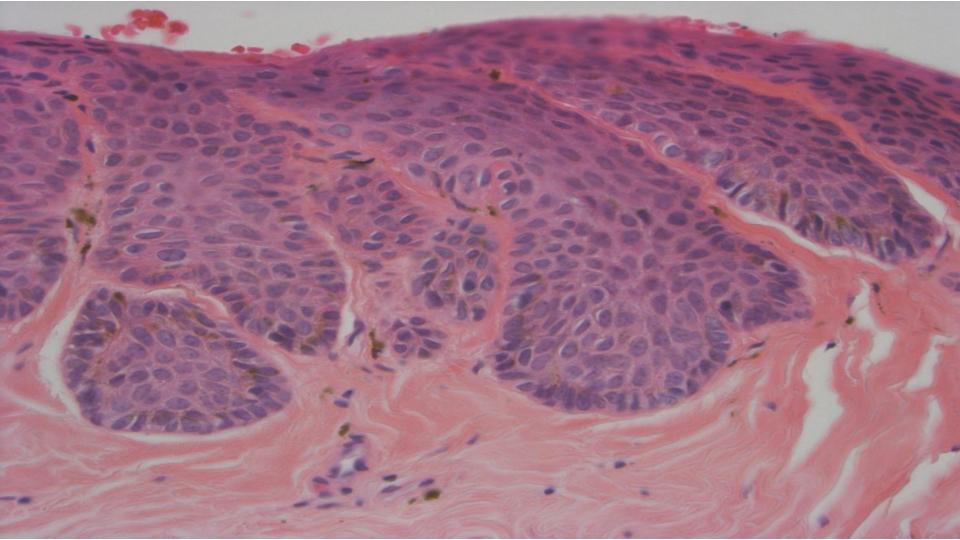
#### Onychocytic matricoma with a Hutchinson's sign











#### 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<sup>1</sup>, 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 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





# Melanonychia When did it appear?

All recently developed melanonychias in adult fair-skinned individuals over 30 years are suspicious.

Exception: functional melanonychia!





# Melanonychia of recent onset: Which age at onset?

Babies and children: benign

Adolescents: usually benign

Adults < 30 years: probably benign

Adults > 30 years: suspicious

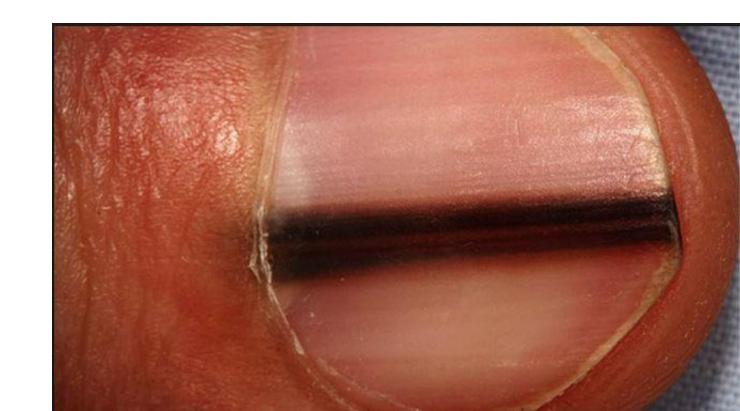
Adults > 40: probably malignant

Adults > 50: usually malignant





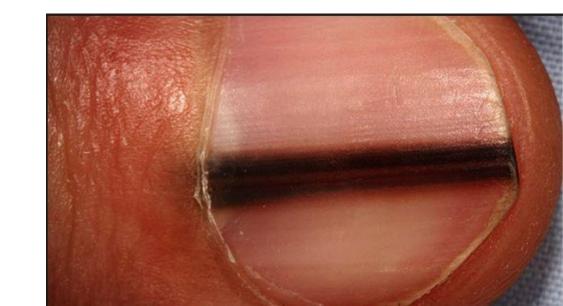
# 60 y/o left thumbnail





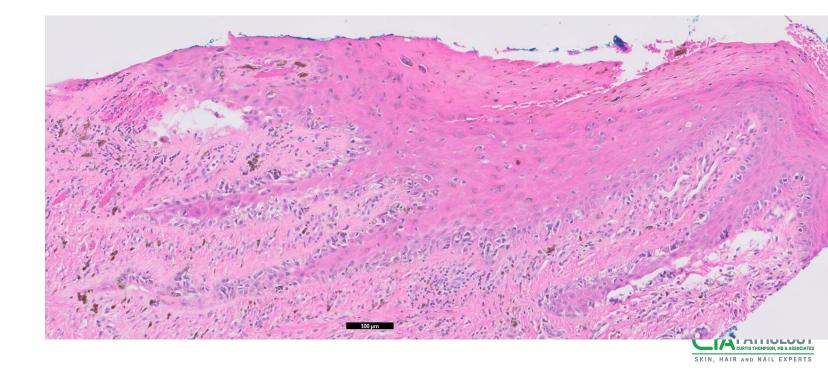
#### 60 yo left thumbnail

- New pigment
- Older age
- Skin type



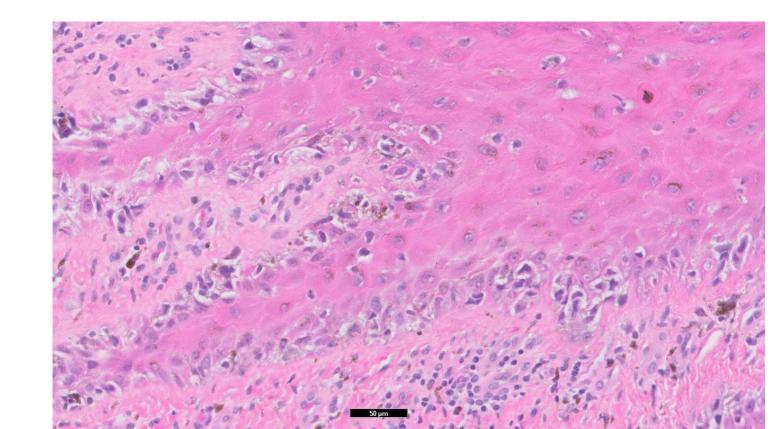


## 60 y/o left thumbnail



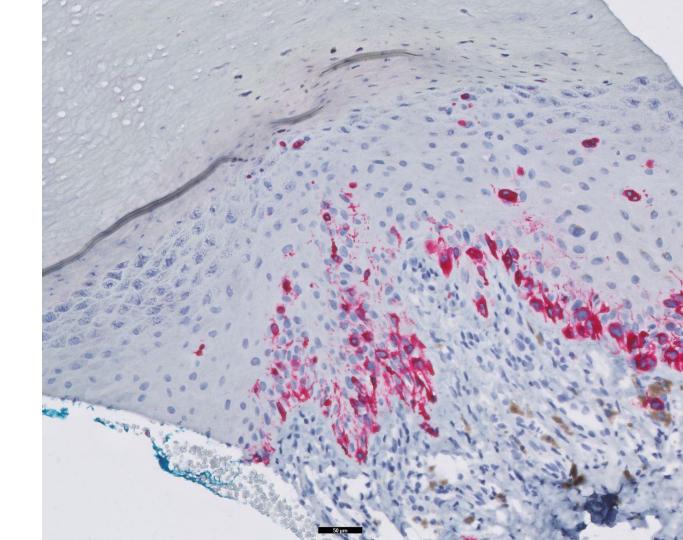


# 60 y/o left thumbnail



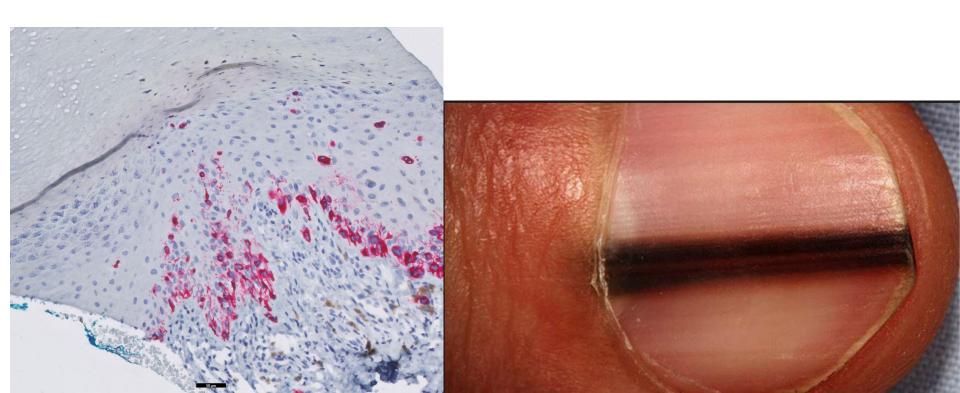


# 60 y/o

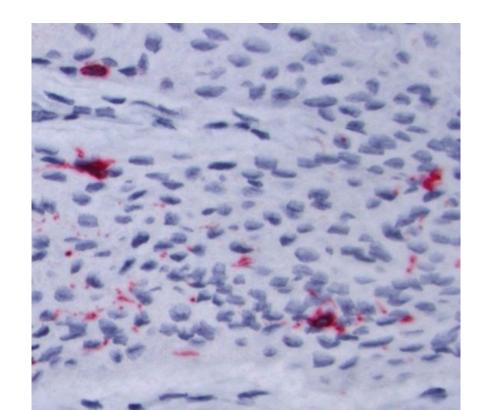




#### Melanoma in-situ



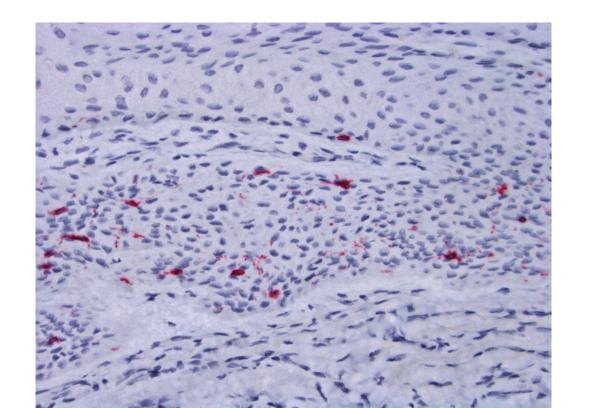
#### Dendritic melanocytes in B9 and malignant







#### Margin assessment may be challenging

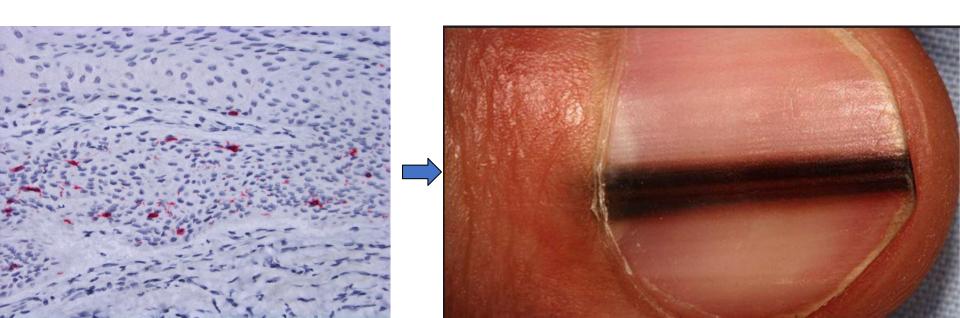






#### **Clinical presentation important**

• New lesion in older patient



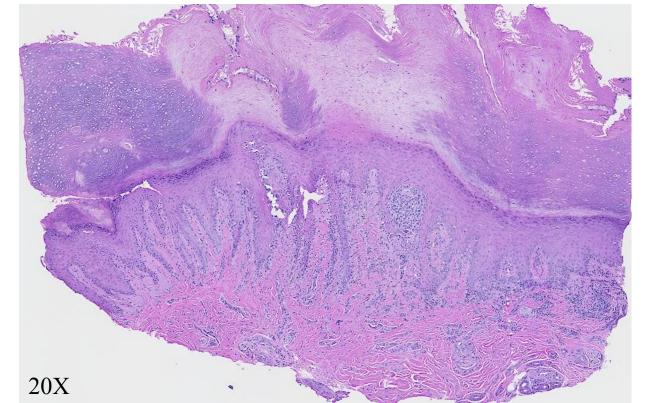
#### **Challenging Case**

- ■61 y/o female
- Right thumbnail
- ■Patient of Dr. Zaiac

#### Clinical Information

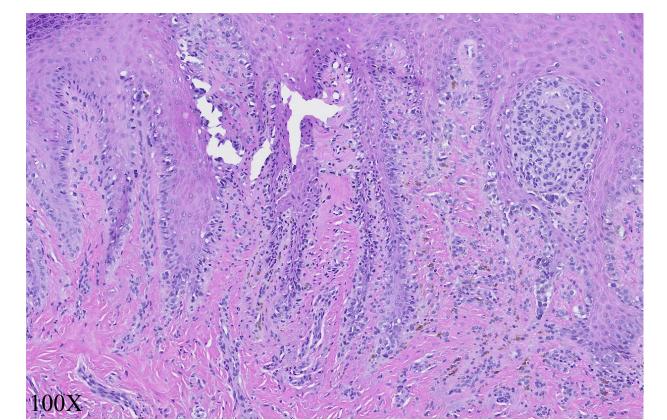
Morphology: irregular brown papule DDX: Atypical Junctional Melanocytic Neoplasm





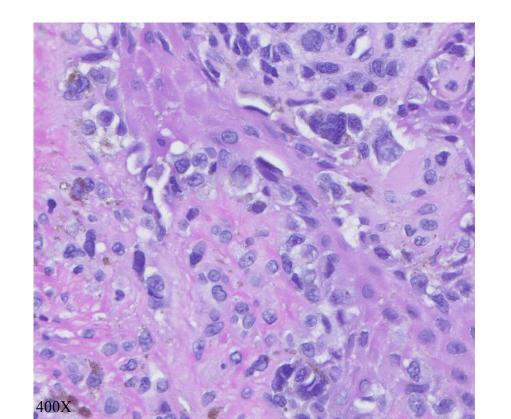








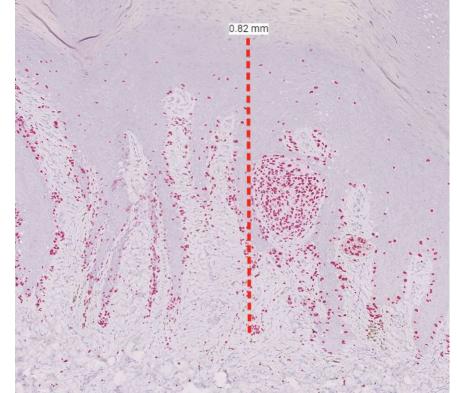








- Breslow 0.82mm
- Stage pT1a

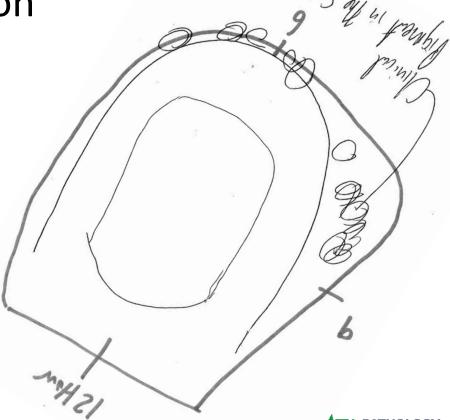






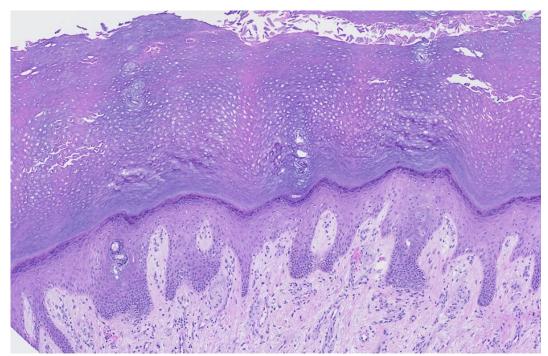
61 y/o female Re-Excision

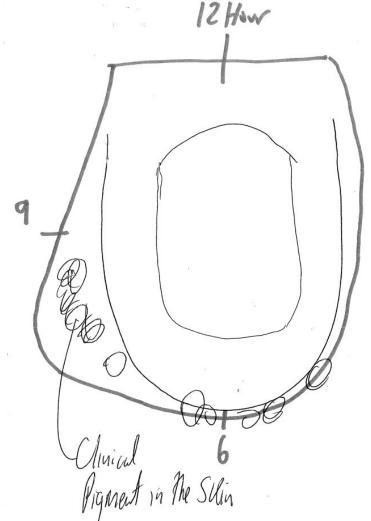






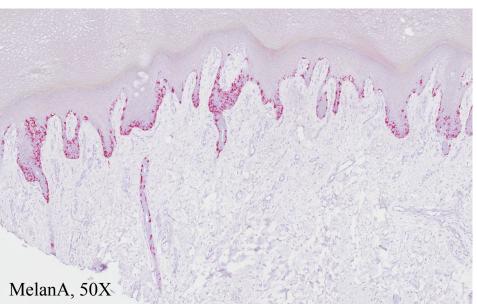
#### 61 y/o female Re-Excision

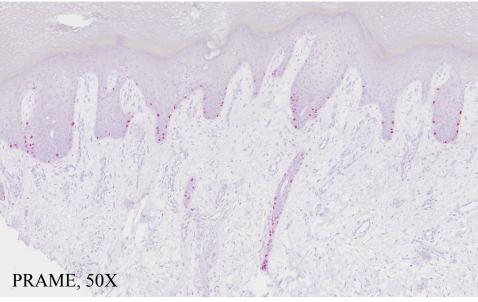






#### 61 y/o female Re-Excision







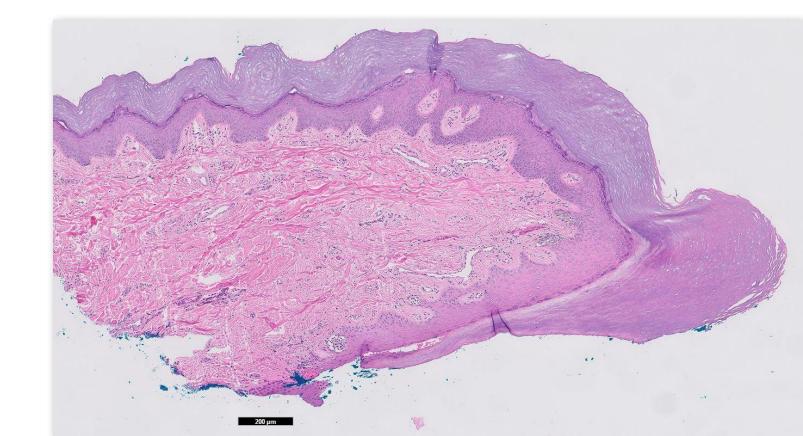


■10 y/o male with pigmented nail of great toe. It appeared at age 3.

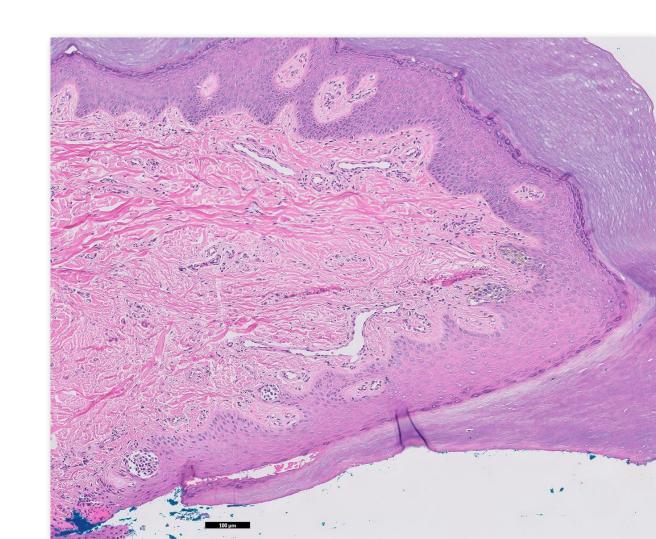




# 10 y/o boy

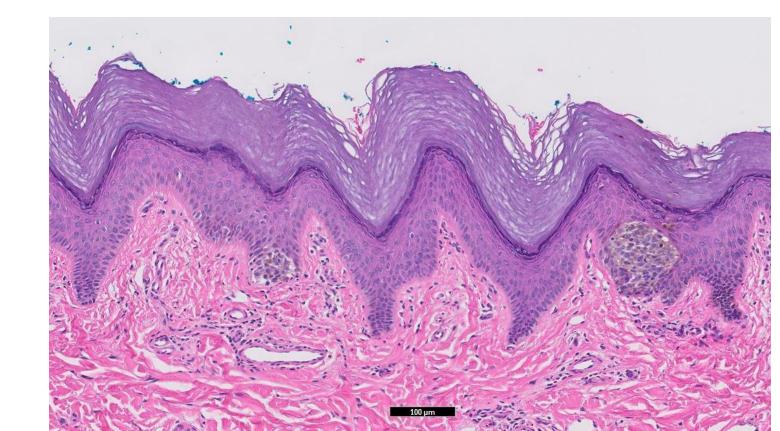






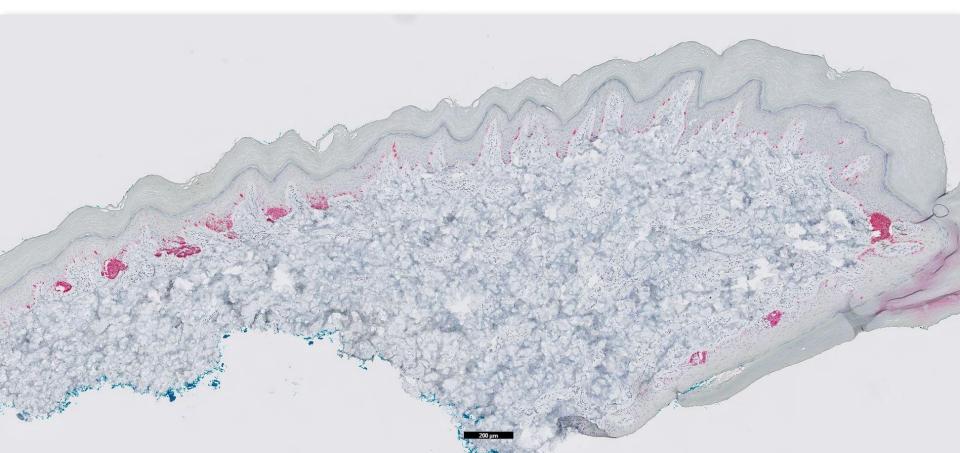


#### **Proximal nail fold**

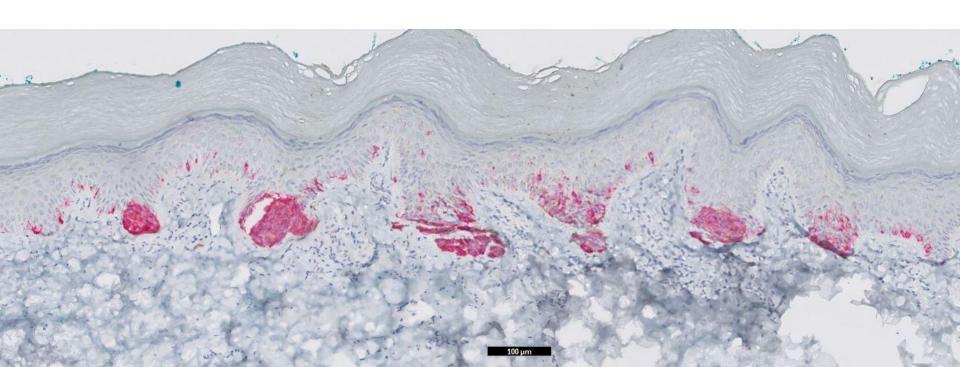




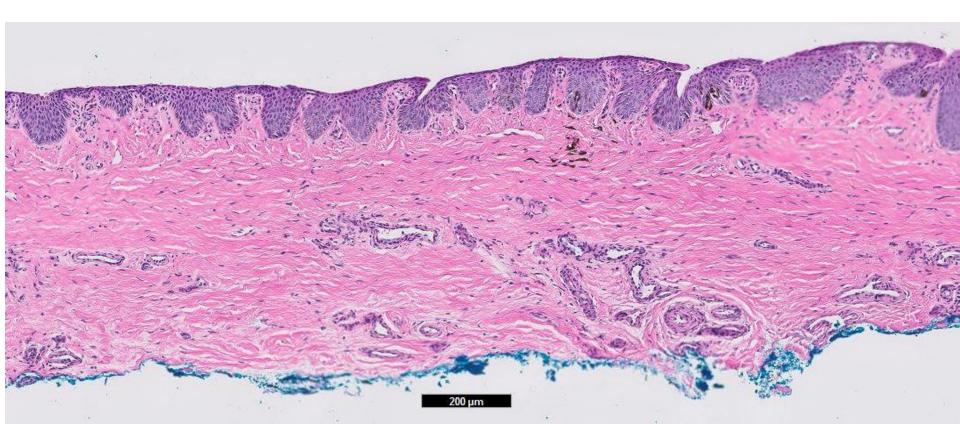
# 10 y/o



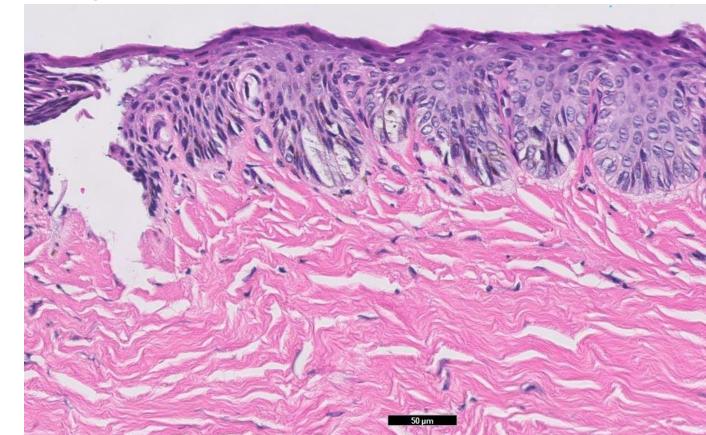
#### **Proximal nail fold**



# Nail bed/matrix

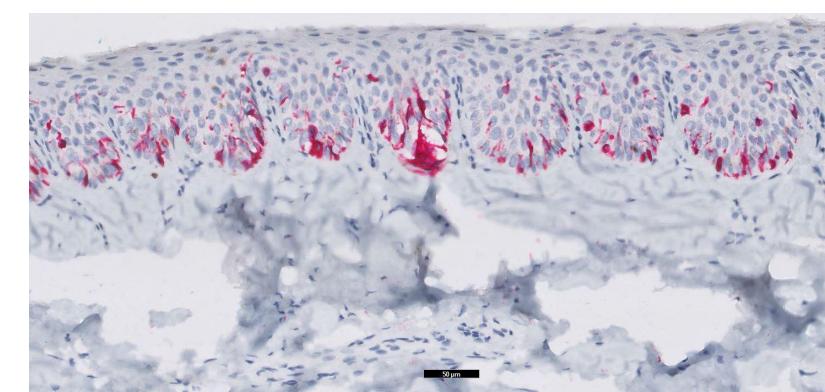


## 10 y/o nail bed/matrix



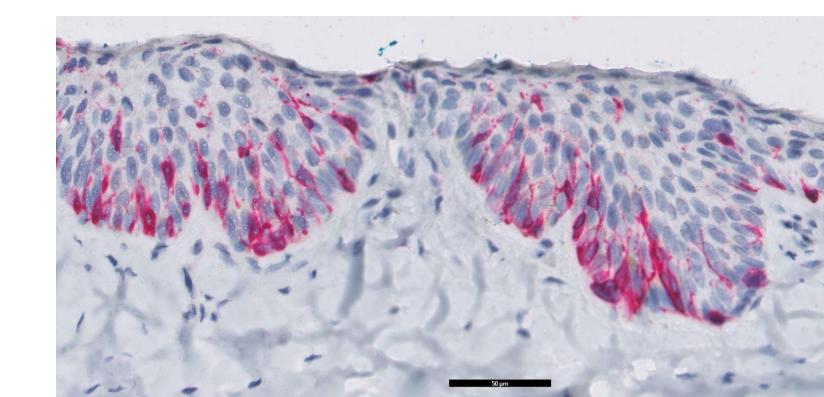


## Nail bed/matrix



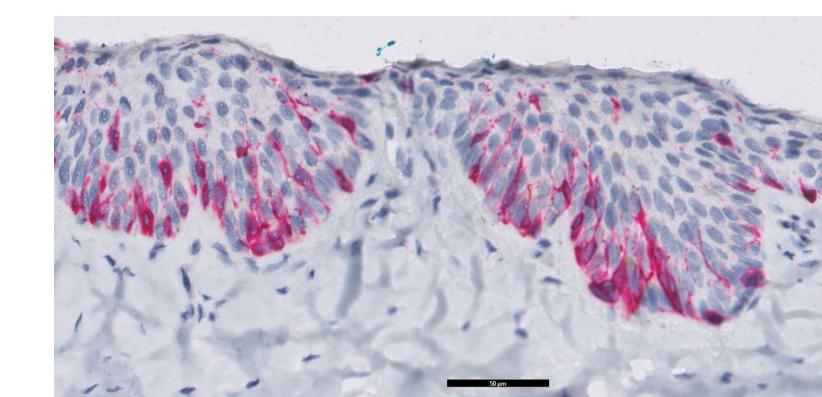


## Nail bed/matrix



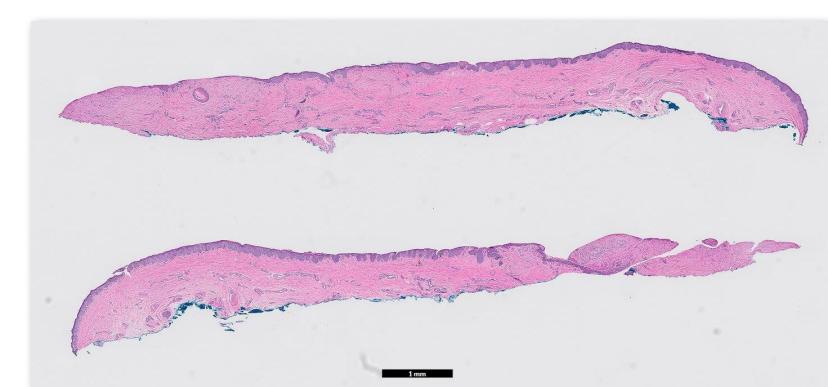


#### Benign nevus in 10 y/o



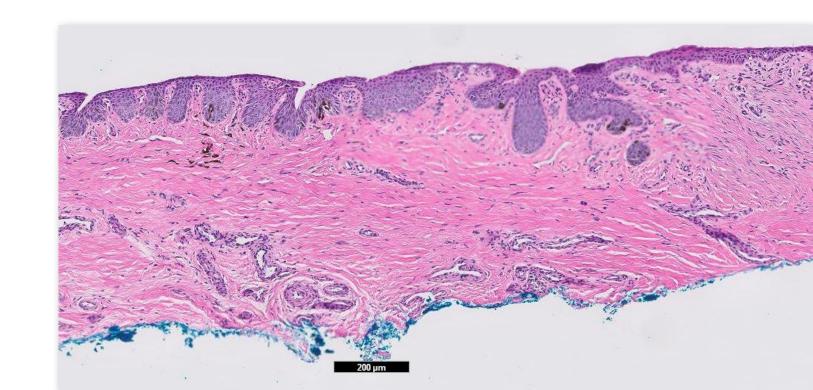


# 13 y/o male



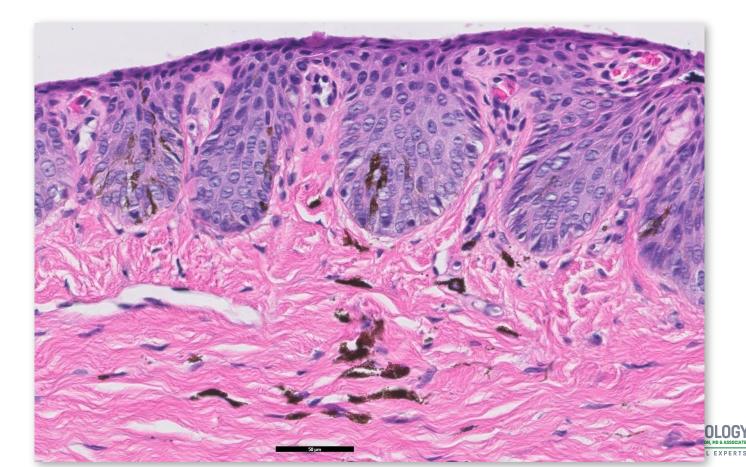


#### 13 y/o male



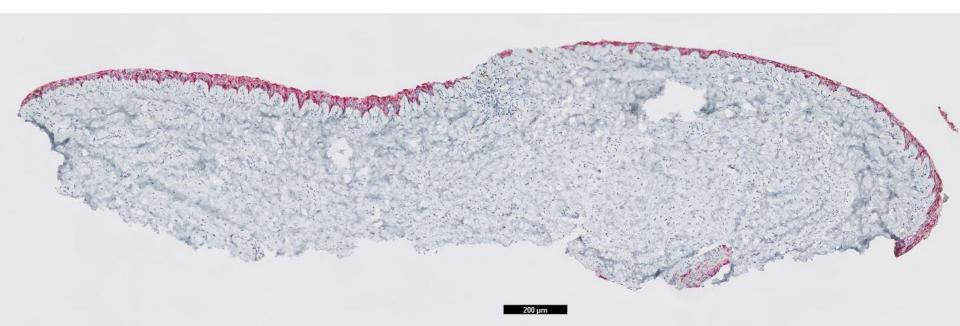


#### 13 y/o male

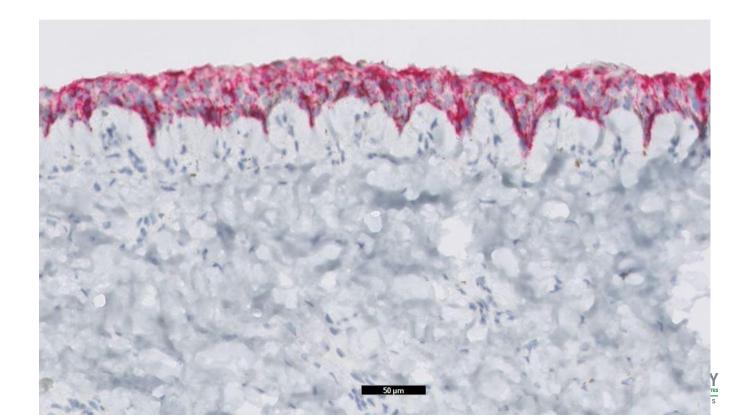




### 13 y/o nailbed

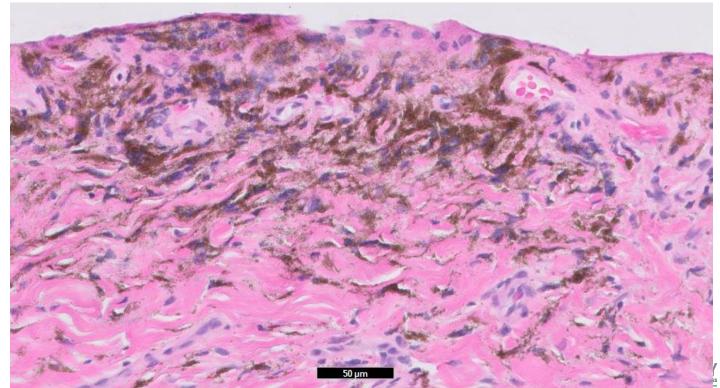


### 13 y/o nailbed





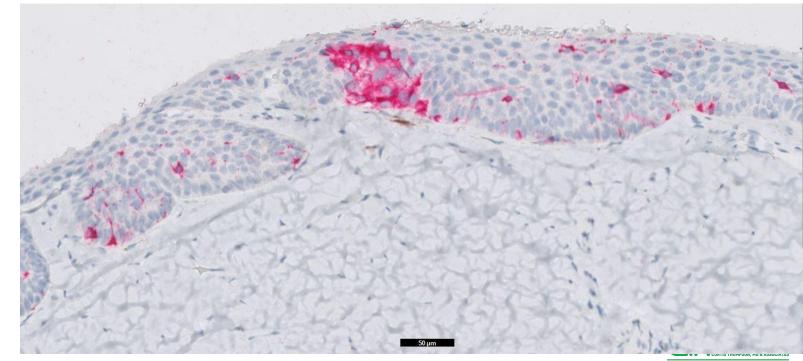
# 18 y/o





SKIN, HAIR AND NAIL EXPERT

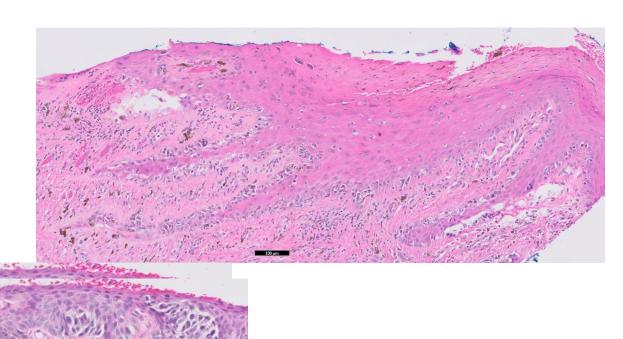
# 18 y/o nail





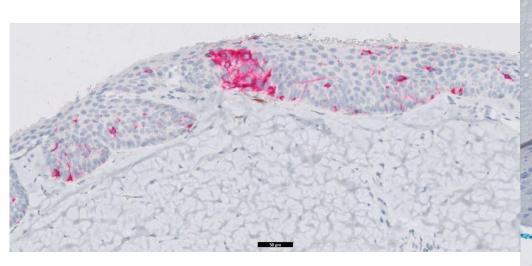
SKIN, HAIR AND NAIL EXPERTS

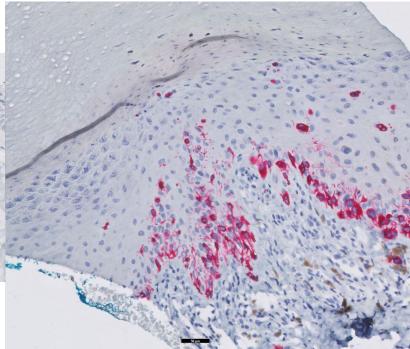
## Benign? Atypical? Malignant?





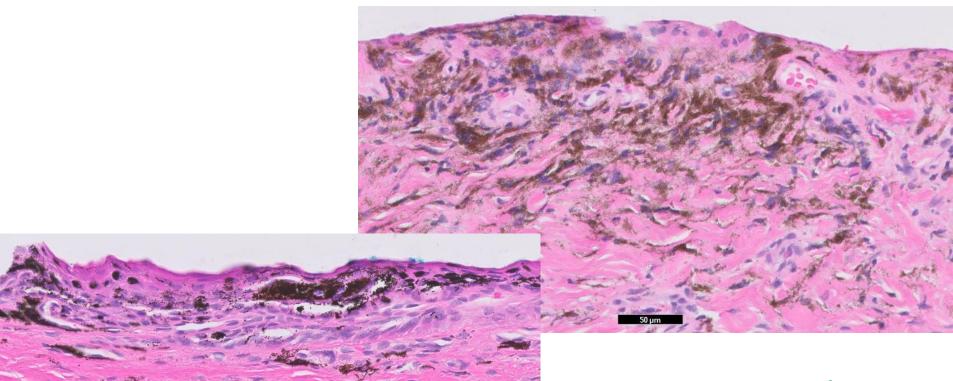
## Benign? Atypical? Malignant?







### **Benign? Atypical? Malignant?**





#### Pigmented lesion in a child

- Often type III skin type or above (worry about acral lentiginous MM)
- Concern to parents—pressure to biopsy
- Biopsy shows large, single, Spitzoid melanocytes (?Atypia?)





#### Pigmented lesion in a child

- Few pediatric melanocytic tumors sampled—nail experts do not sample
- Traditional criteria for benign vs atypical vs malignant do not apply





### Molecular markers—B9 vs malignant

Cyclin D1 and PRAME expression in distinguishing melanoma in situ from benign melanocytic proliferation of the nail unit

Young Jae Kim, Chang Jin Jung, Hyoungmin Na, Woo Jin Lee, Sung Eun Chang, Mi Woo Lee, Chan-Sik Park, Youngkyoung Lim 

⊗ Chong Hyun Won

Diagnostic Pathology 17, Article number: 41 (2022) Cite this article





#### Molecular markers—PRAME

- Benign--13 of 14 cases negative
- Melanoma In-situ—10 of 13 positive

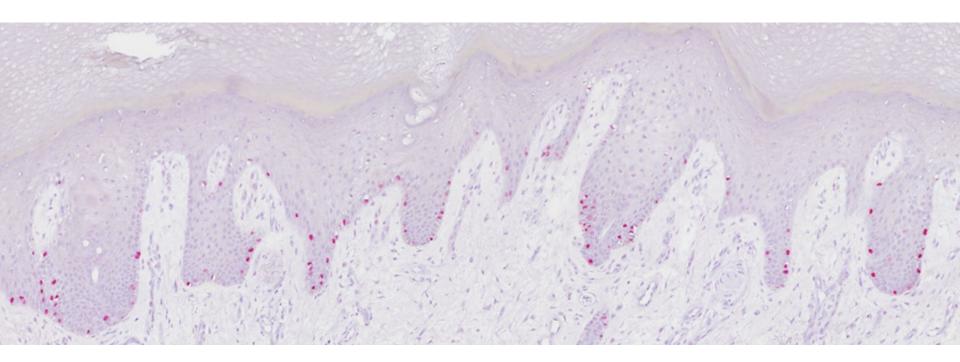
\*\*Very strong PRAME staining most c/w MIS

Cyclin D1 and PRAME expression in distinguishing melanoma in situ from benign melanocytic proliferation of the nail unit





#### Be careful with PRAME







#### Thanks!

curtis@ctapathology.com



