DISCLOSURE OF RELATIONSHIPS WITH INDUSTRY

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F107 Challenging Cases in Dermatopathology

DISCLOSURES

• None relevant to this talk
• Others:
  • Royalties Lippincott Williams Wilkins
    • Lever’s Histopathology of the Skin
  • Consultant Myriad Genetics
Case 1

• 72 year old man
• Idiopathic pulmonary fibrosis
• s/p double lung transplant 5 years ago
• Immunosuppression: prednisone, tacrolimus, mycophenolic acid

• 2 years severely itchy rash
• No gastrointestinal symptoms
Case 1: Clinical

- Thin velvety plaques with hyperpigmentation
- Neck, back, chest, axillae, buttocks, legs
What is the most likely etiology?

A. Infectious agent
B. Medication
C. Neoplasia (hematopoietic malignancy)
D. Contactant
E. Autoimmune (connective tissue disease)
Skin Biopsy
Site: L upper arm
Date: 2/2/17
What is the most likely etiology?

A. Infectious agent
B. Medication
C. Neoplasia (hematopoietic malignancy)
D. Contactant
E. Autoimmune (connective tissue disease)
Differential

- Graft vs host disease
- Chronic eczematous dermatitis
- Sezary Syndrome
- Dermatomyositis
- Other
Patient

“They told me I had a rare viral infection of the skin.”
Human Polyomavirus 7 associated rash

- Human Polyomavirus 7–Associated Pruritic Rash and Viremia in Transplant Recipients
- Jonhan Ho Jaroslaw J. Jedrych Huichen Feng et al
- The Journal of Infectious Diseases, Volume 211, Issue 10, 15 May 2015, Pages 1560–1565,
Diagnosis

• Immunoperoxidase stain to HPyV7 antigen
  • (+) epidermal keratinocytes; weaker in stratum corneum

• Electron microscopy
  • 39-46 nm icosahedral virions c/w polyomavirus

• Quantitative PCR
  • High HPyV-7 viral genome copy number
Human Polyomavirus 7–Associated Pruritic Rash and Viremia in Transplant Recipients
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Human papillomavirus-7 skin rash

• Resembles peacock plumage
Human Polyomavirus

• Family Polyomaviridae
  • 77 species of virus
  • 13 known to be infectious to humans

• Icosahedral virus

• Double stranded circular DNA
Polyomavirus History

• First polyomavirus described in 1952
  • Caused adenocarcinoma and leukemia when injected into mice
  • 1971 became known as BK virus (patient B.K.)

• Same year JC virus isolated from patient J.C. with Hodgkin’s lymphoma and progressive multifocal leukoencephalopathy (PML)

• 2008: Merkel cell polyomavirus
• 2010: trichodysplasia spinulosa virus
Polyomavirus

- JC virus: progressive multifocal leukoencephalopathy
- BK virus: nephropathy
  - BK and JC virus affect immunosuppressed patients; not known to cause skin disease
- Merkel cell polyomavirus: MC carcinoma
- Polyomavirus 8: trichodysplasia spinuloma
Viral Associated Trichodysplasia

Viral Associated Trichodysplasia

Trichodysplasia Spinulosa

Viral Associated Trichodysplasia

• Folliculocentric papules with keratotic spiculations
• Mid face and ears
• Less common on trunk and extremities
• Variable alopecia of eyelashes and eyebrows

• Path: dysmorphic follicles with enlarged trichohyalin granuleless
Dysmorphic hair follicles
Dysmorphic hair follicles
Prominent and enlarged trichohyalin granules
Prominent and enlarged trichohyalin granules
DDX: Wong-Type Dermatomyositis
Wong-type Dermatomyositis

HPyV 7
Take home

• Polyomaviruses are ubiquitous and asymptomatic in most humans
• Immunosuppression may lead to skin manifestations of viral infection
Case 2

• A 55 year old woman presents with a firm SQ nodule on the abdomen
• “rule out cyst”
Immunohistochemistry

• Cytokeratin stains negative
  • No epithelium

• S100 negative
What is your diagnosis?

A. Cutaneous endometriosis
B. Hidradenoma papilliferum
C. Cutaneous ciliated cyst
D. Adenomatosis in an accessory nipple
E. Cutaneous metaplastic synovial cyst
What is your diagnosis?

A. Cutaneous endometriosis
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Cutaneous Metaplastic Synovial Cyst

• Aka: Synovial Metaplasia

• **Definition of metaplasia**
  • transformation of one tissue into another

• Dermis/SQ tissue transforms into tissue that looks like synovium
Synovial Metaplasia

• Occurs in scars from trauma or surgery
• Not associated with joints
• Months to years after injury
• Firm nodules that may resemble a suture granuloma
Synovial Metaplasia: pathology

- Cystic space
- Surrounding connective tissue shows papillary fronds
- May be some cells lining the fronds
- Hyper and hypocellular areas of bland spindle cells
- No epithelium
Synoviocytes

- Cells lining spaces between developing bones (synovium)

- Type A: macrophages derived from bloodborne mononuclear cells
- Type B: likely locally derived fibroblasts
Synovial Metaplasia

- Immunohistochemistry
- Cytokeratin stains negative
- S100 negative
- Spindle cells + with vimentin, CD68
  - “fibrohistiocytic” markers
Synovial Metaplasia

- Adjacent to joint prosthesis
- Near silicone breast implants
- At pacemaker site
- Laparotomy scar
- Trauma
Synovial Metaplasia of the Skin

Juan G. Gonzalez, M.D., Robert W. Ghiselli, M.D., and Daniel J. Santa Cruz, M.D.

Cutaneous Metaplastic Synovial Cyst of the Cheek Generated by Repetitive Minor Trauma

Woo-Haing Shim, M.D., Seung-Wook Jwa, M.D., Margaret Song, M.D., Hoon-Soo Kim, M.D., Hyun-Chang Ko, M.D., Byung-Soo Kim, M.D., Moon-Bum Kim, M.D.

Metaplastic synovial cyst in male breast

Pratistadevi K. Ramdial, FCPath(Anat)SA a, *, Yetish Singh, MBChB a, Bhugwan Singh, FCS (SA) b
Synovial Metaplasia

- During wound repair, fibroblasts proliferate and foreign material resists penetration
- Fibroblasts create a synovium-like membrane
Synovial metaplasia

- Benign reactive change
- Confidently reassure the dermatologist
Thank you

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