Greatest Clinicopathologic Cases

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DISCLOSURE OF RELATIONSHIPS WITH INDUSTRY

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S0013 Greatest Clinicopathologic Cases

• None relevant to this talk
• Other:
  ❖ Royalties Lippincott Williams Wilkins
    ▪ Lever’s Histopathology of the Skin
Case 1

- A 75 year old man present with 9 month history of a rash that is progressively getting worse
Punch Biopsy
No definite diagnosis from pathologist

- Patient returns 6 weeks later
- No history of contactants
- No radiation history
- No sun exposure
Two more biopsies
Second biopsy
Third biopsy
What is your diagnosis?

A. Angiolymphoid hyperplasia
B. Angiomatosis
C. Angiosarcoma
D. Dermatomyositis
E. Well’s syndrome
What is your diagnosis?

A. Angiolymphoid hyperplasia
B. Angiomatosis
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Reactive Angiomatosis: Ischemia

- Severe atherosclerotic disease
- AV fistula for hemodialysis
- Calciphylaxis
- Cryoproteinemia
- Large pendulous breasts
- Anti-cardiolipin antibodies
- GVHD
Clinical Follow-up

- Pathology felt not to represent angiosarcoma
- Clinical differential diagnosis included AESOP Syndrome
The skin changes in AESOP are associated with which underlying process

A. Plasmacytoma  
B. Pneumonia  
C. Prostate Carcinoma  
D. Pulmonary Embolism  
E. Pulmonary Fibrosis
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AESOP Syndrome

Adenopathy and
Extensive
Skin patch
Overlying a
Plasmacytoma
AESOP Syndrome

- Adenopathy and Extensive Skin Patch Overlying Plasmacytoma

- Rare variant of POEMS:
  - Polyneuropathy
  - Organomegaly
  - Endocrinopathy
  - Monoclonal Protein
  - Skin Changes
AESOP Syndrome

• 2003: 4 patients reported as a syndrome
  ❖ One patient described in 1938
• Slowly enlarging erythematous to violaceous patch of trunk
• Underlying plasmacytoma of the bone
• 3 patients had polyneuropathy and 2 had POEMS
  ❖ Polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, skin changes

Lipsker et al. Medicine 82:1, 2003
The AESOP (Adenopathy and Extensive Skin Patch Overlying a Plasmacytoma) Syndrome
Report of 4 Cases of a New Syndrome Revealing POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal Protein, and Skin Changes) Syndrome at a Curable Stage

Dan Lipsker, Murielle Rondeau, Gilbert Massard, and Edouard Grosshans
AESOP Syndrome

- Plasmacytomas most commonly in the bones of the thorax
- Clinical lesions blanche because of the vascular proliferation seen on biopsy
- Vascular proliferation likely due to cytokine-induced cutaneous angiogenesis by the plasmacytoma
AESOP Syndrome

- The cutaneous lesions may precede the plasmacytoma diagnosis by up to a year
- Recognition helps detect the plasmacytoma potentially in a treatable/curable stage
Our patient: Clinical follow up

• No plasmacytoma was identified; normal SPEP
• Biopsy shoulder joint showed synovial sarcoma
Synovial Sarcoma

• Malignant mesenchymal tumor with partial epithelial differentiation
• Predominates in older children/young adults
• Second most common sarcoma in children
• Occurs any site
  ❖ Joints are rare
  ❖ Misnomer, does not arise from synovium
  ❖ Rarely reported in skin
Synovial Sarcoma

- Cell of origin is unknown
- Translocation involving Chromosomes X and 18
  - T(X;18)(p11.2, q11.2)
- Large histological spectrum
  - Typically biphasic: spindle and epithelial cells
- Survival
  - 5 year: 60%
  - 10 year: 50%
Case 2

- A 52 yo woman presents for second opinion of a scalp mole
- She is unsure of the duration
- She brings the pathology report
Pathology Report

- Junctional dysplastic nevus with atypia
- If this is part of a larger lesion, the biopsy may not be representative
52 year old woman, pigmented lesion 1.5cm
What is your next step?

A. Assure the patient it is a benign mole
B. Ask the pathologist to do more melanoma stains
C. Refer the patient to surgeon for wide excision
D. Send the block for genetic testing (CGH)
E. Take another biopsy
What is your next step?

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MART-1 (red)
What other stain might be helpful?

A. Fontana
B. HMB-45
C. PAS
D. S100
E. SOX-10
What other stain might be helpful?

A. Fontana
B. HMB-45
C. PAS
D. S100
E. SOX-10
PAS stain
Additional information

- There is associated hair loss
- Patient has a history of lupus erythematosus
Final Diagnosis

• Discoid lupus erythematosus

• What happened to the dysplastic nevus?
Immunohistochemical staining in lichenoid tissue reaction

- “pseudomelanocytic” nests misdiagnosed as melanoma in situ
- “nests” positive with melanocytic marker MART-1 / Melan A
Melan-A–Positive “Pseudomelanocytic Nests”: A Pitfall in the Histopathologic and Immunohistochemical Diagnosis of Pigmented Lesions on Sun-Damaged Skin

Helmut Beltraminelli, MD,*† Laila El Shabrawi-Caelen, MD,* Helmut Kerl, MD,* and Lorenzo Cerroni, MD*
Nests with numerous SOX10 and MiTF-positive cells in lichenoid inflammation: pseudomelanocytic nests or authentic melanocytic proliferation?

Claudine Yap Silva, Lynne J. Goldberg, Meera Mahalingam, Jag Bhawan and Deon Wolpowitz

Department of Dermatology, Dermatopathology Section, Boston University School of Medicine, Boston, MA, USA
An Immunohistochemical Analysis of Pseudomelanocytic Nests Mimicking Melanoma In Situ: Report of 2 Cases

Kimberly M. Nicholson, MD and Pedram Gerami, MD

Take home message

- Lichenoid tissue reaction can mimic melanocytic lesions
- Clinical-Path Correlation!
Thank you

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