Clinicopathologic Self-Assessment
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I have no relationship with industry relevant to this lecture

- Lippincott Williams Wilkins (Lever royalty)
- Myriad Genetics: Consultant
Case 1

• 70 year old man presents with a new 4mm skin colored papule of the lower eyelid/upper cheek. No bleeding. The biopsy is read by a dermatopathologist as:
  ❖ Nodular hidradenoma. The tumor involves the deep margin.
What is your next step?

A. Reassure the patient that the lesion is benign and no more treatment is needed
B. Inform the patient that the lesion is malignant and more surgery is needed
C. Call the dermatopathologist about the diagnosis
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C. Call the dermatopathologist about the diagnosis
• Four more dermatopathologists review the slide:
  - 2 report a benign diagnosis
  - 2 report a malignant diagnosis

• You decide to have the lesion excised.
Diagnosis

• Endocrine mucin producing sweat gland carcinoma

• Cytokeratin positive, and
  ❖ “Diagnosis aided by special stains”
Endocrine mucin-producing sweat gland carcinoma
Cytokeratin and special stains positive
“Diagnosis aided by special stains”
What type of stains were positive?

A. Melanocytic (S100, HMB, MelanA)
B. Amyloid (congo red, crystal violet, thioflavin T)
C. Vascular (CD31, CD34, ERG)
D. Neuroendocrine (NSE, chromogranin, synaptophysin)
E. Lymphoid (CD3, CD20, CD45)
What type of stains were positive?

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Endocrine Mucin-Producing Sweat Gland Carcinoma

• Low grade sweat gland carcinoma
• Eyelid region of elderly women

**Under-recognized by pathologists**

- Especially when partially biopsied
Endocrine Mucin-Producing Sweat Gland Carcinoma

- Described in 1997
- Mean age of presentation: 70 yrs
- Female: Male, 3:1
- Eyelid, upper cheek
- Slow growing skin colored papule
Endocrine Mucin-Producing Sweat Gland Carcinoma

• Solid and cystic component
• Formation of ducts
• Mucin production is subtle
• Cytological atypia is mild

• Pathology is similar to solid papillary carcinoma, breast
Endocrine Mucin-Producing Sweat Gland Carcinoma

• Positivity with neuroendocrine markers:
  - Neuron specific enolase
  - Chromogranin
  - Synaptophysin
  - CD56
  - CD57
Endocrine Mucin-Producing Sweat Gland Carcinoma

- Reported to occur with mucinous eccrine carcinoma
- May be a precursor lesion to mucinous carcinoma
Endocrine Mucin-Producing Sweat Gland Carcinoma

• Clinical Course:
  - Recurrence if incompletely removed
  - May progress to mucinous carcinoma (metastatic potential)

• Treatment
  - Surgical Excision
  - Mohs Micrographic Surgery
  - Exclude primary breast tumor
Case 2

• 35 year old man with rash for 4 years
• Burning areas of thighs and buttocks
• Otherwise healthy
• FH: sister with systemic lupus erythematosus
• Superficial and deep lymphocytic infiltrate

• Perivascular and periadnexal
Focal interface changes
Increased mucin
Perieccrine lymphocytes
28 year old healthy woman

Burning lesions on thighs and buttocks

Present 2 months
All lymphocytes
The pathologist says it looks like lupus erythematosus. What is your next step?

A. Ask about work and hobbies
B. Ask about exposure to ticks
C. Check renal function
D. Check CPK/aldolase
E. Ask about rash in others at home
Work up

- Normal labs
  - CBC, electrolytes
  - CPK/aldolase
  - C3, C4
  - ANA, Anti-Smith, SSA, SSB
  - RF, CRP, Cryoglobulins, quantiferon gold
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Equestrian Perniosis

- Both patients avid horseback riders
- Rash only occurred in winter months/cold weather
Equestrian Perniosis

• AKA
  ❖ equestrian cold panniculitis, cold-associated perniosis
• Papules, plaques, nodules
• Upper outer thighs/buttocks
Equestrian Perniosis

• Related to extended cold exposure
  ❖ At least 2 hours
• Compounded with wind & tight fitting pants
  ❖ May add to decreased blood flow
• No known systemic associations (SLE)
  ❖ Hypothesized that it may be a forme fruste of lupus
Besides equestrians, what other patient population might have a similar problem? Patients with

A. Chronic migraines
B. Chronic low back pain
C. Diabetes
D. Inflammatory bowel disease
E. Metastatic melanoma on nivolumab
Consider the diagnosis in non-equestrians

- Truck drivers, hikers, outdoor winter athlete, ice pack use
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Equestrian Perniosis: Treatment

- Avoidance of cold/warmer clothing
- Loose clothing in affected area
- Oral prednisone
- Antimalarials
Take home messages

• Nodular hidradenoma near eye:
  ❖ *Think* endocrine mucin-producing sweat gland carcinoma
  ❖ Complete excision

• Thigh/buttock lesions + lupus pathology
  ❖ *Think* equestrian perniosis
  ❖ Ask about cold exposure
Thank you

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