**Case 1**
- 13 yo febrile male with syncope and progressive weakness
- Painful purpuric lesions of palms and soles
- Facial rash for 1 month prior

**Work-up**
- Anemia
- Transaminitis
- Proteinuria
- Increased ESR
- Hypocomplementemia
- Normal platelets
- Normal anti-myeloperoxidase and anti-proteinase antibodies

**Disclosures**
- I do not have any relevant relationships with industry.
What is the most likely diagnosis?

A. Disseminated gonococcemia  
B. Henoch-Schönlein purpura  
C. Levamisole induced vasculitis  
D. Libman-Sacks endocarditis  
E. Rocky Mountain Spotted Fever

Work-up

- Positive Antibodies
  - DsDNA
  - SSA
  - Sm
  - RNP
  - Cardiolipin

Libman-Sacks Endocarditis

- Manifestation of SLE
- Verrucous, non-bacterial thrombotic endocarditis
- Usually silent but can result in embolic phenomena
Libman-Sacks endocarditis is most associated with:
A. ANCA positivity
B. Antiphospholipid antibody
C. Cold agglutinins
D. Cryoglobulinemia
E. Myelodysplastic disorder

Take Home Points
- Acral purpuric lesions due to pauci-inflammatory fibrin thrombi
- Coagulopathy
- Embolic (infectious endocarditis, cholesterol, atrial myxoma, aseptic endocarditis ...)

Case 2
- 21 yo scalp cyst

What is the most likely diagnosis?
A. BAPoma
B. Epithelioid blue nevus
C. Melanoma
D. Proliferative nodule
E. Spitz nevus

Addendum Comment
From UCSF Dermatopathology/Dr. Timothy H. McCalmont

DIAGNOSIS: POLYPOID COMBINED MELANOCYTIC NEVUS, INCLUDING A COMPONENT OF "BAPOMA":
We completed BAP-1 immunostaining as part of our evaluation, and a loss of nuclear BAP-1 is apparent, thereby solidifying the above diagnosis.
BAPomas/Weisner’s nevus (MBAITs)
- Flesh colored dome shaped papules
- Epithelioid cells
- Dermal
- May have common nevus component and frequent TILs
- IHC loss of nuclear expression

Second Lesion

BAP1

Weisner Nevus/BAPoma
Which of the following has an established association with the BAP1 tumor syndrome?

A. Acute myeloid leukemia  
B. Cervical cancer  
C. Mesothelioma  
D. Myelodysplastic syndrome  
E. Prostate cancer

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**BAP1 Tumor Syndrome**

- BCC
- Breast
- Ovarian
- Pancreatic
- Colon
- Meningioma, paraganglioma, neuroendocrine
- Hepatic cholangiocarcinoma

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**Incidence?**

- BCC: 31%
- Cutaneous melanoma: 13%
- Ovarian cancer: 22%
- Pancreatic cancer: 67%
- Kidney cancer: 31%
- Lung cancer: 4%
- Mesothelioma: 22%
- Meningioma, paraganglioma, neuroendocrine: 31%
- Hepatic cholangiocarcinoma: 67%

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**Results:**

- **Test name:** BAP1 Hereditary Cancer Predisposition Syndrome Test
- **Genes Analyzed:** BAP1
- **Laboratory:** Invitae
- **Results:** POSITIVE, Likely Pathogenic Variant

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**BAP1 Tumor Syndrome**

- **Incidence?**

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**References**

Take Home Points

- Flesh colored dome shaped dermal tumors composed of epithelioid cells may be BAPoma/Weisner nevus
  - Look for loss of nuclear BAP1
  - Germline mutation can be associated with a cancer syndrome

Thank You