Interesting case conference

11/19/12
51 year old female
-no significant medical history
-presented to ER with abdominal pain/distention and ascites
-imaging showed large pelvic mass as well as findings suggestive of carcinomatosis

=> referred for CT guided fine needle aspiration
ADDITIONAL IMMUNOSTAINING RESULTS

Negative for CK7, CK20, S-100, HMB 45, MITF, PR, CD34, desmin, SMA, PAX8, WT-1, calretinin

Positive for ER (3+), smooth-muscle actin

p53=> wild type staining
Final Diagnosis:
Positive for MART-1 positive malignancy, favor perivascular epithelioid cell tumor (PEComa).
Perivascular epithelioid cell tumor (PEComa):
- comprise a family of tumors (e.g., angiomyolipomas, clear cell sugar tumors) of perivascular epithelioid cell origin
- may be associated with tuberous sclerosis complex
- unpredictable natural history. Uterus is the most prevalent reported site of involvement of PEComas in the female genital tract.
Perivascular epithelioid cell tumor (PEComa):
- epithelioid to spindle cells with eosinophilic to clear cytoplasm
- fine vascular network mimicking clear cell RCC
- positive immunostaining for markers of both melanocytic and myoid differentiation (SMA, HMB45, Mart1/Melan-A)
- vast majority of PEComas have been described in females and therefore hormones may play a role in their pathogenesis. Estrogen receptor (ER) and progesterone receptor (PR) positivity can be observed in PEComas.