Interesting case conference

11/19/12

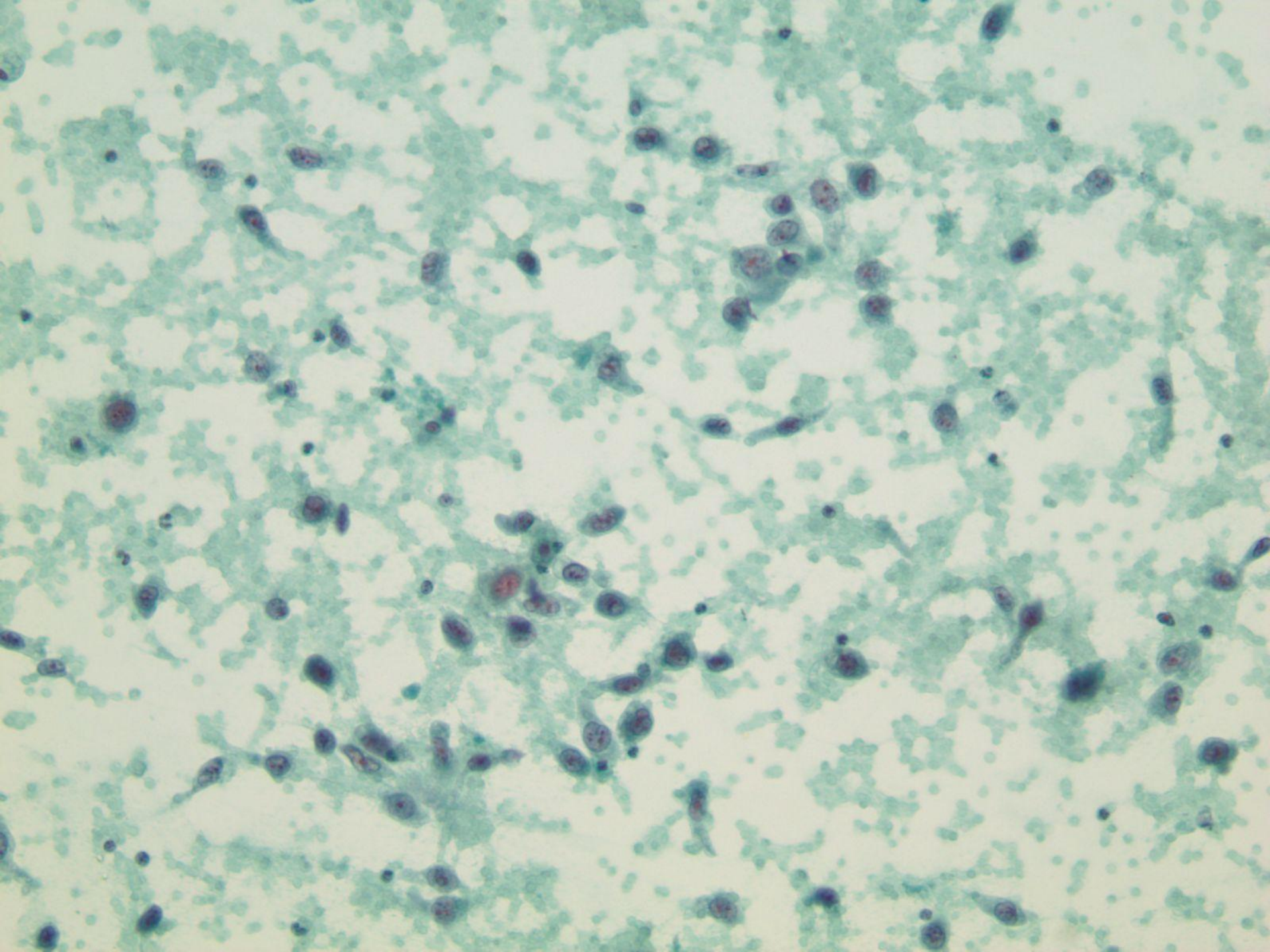
51 year old female

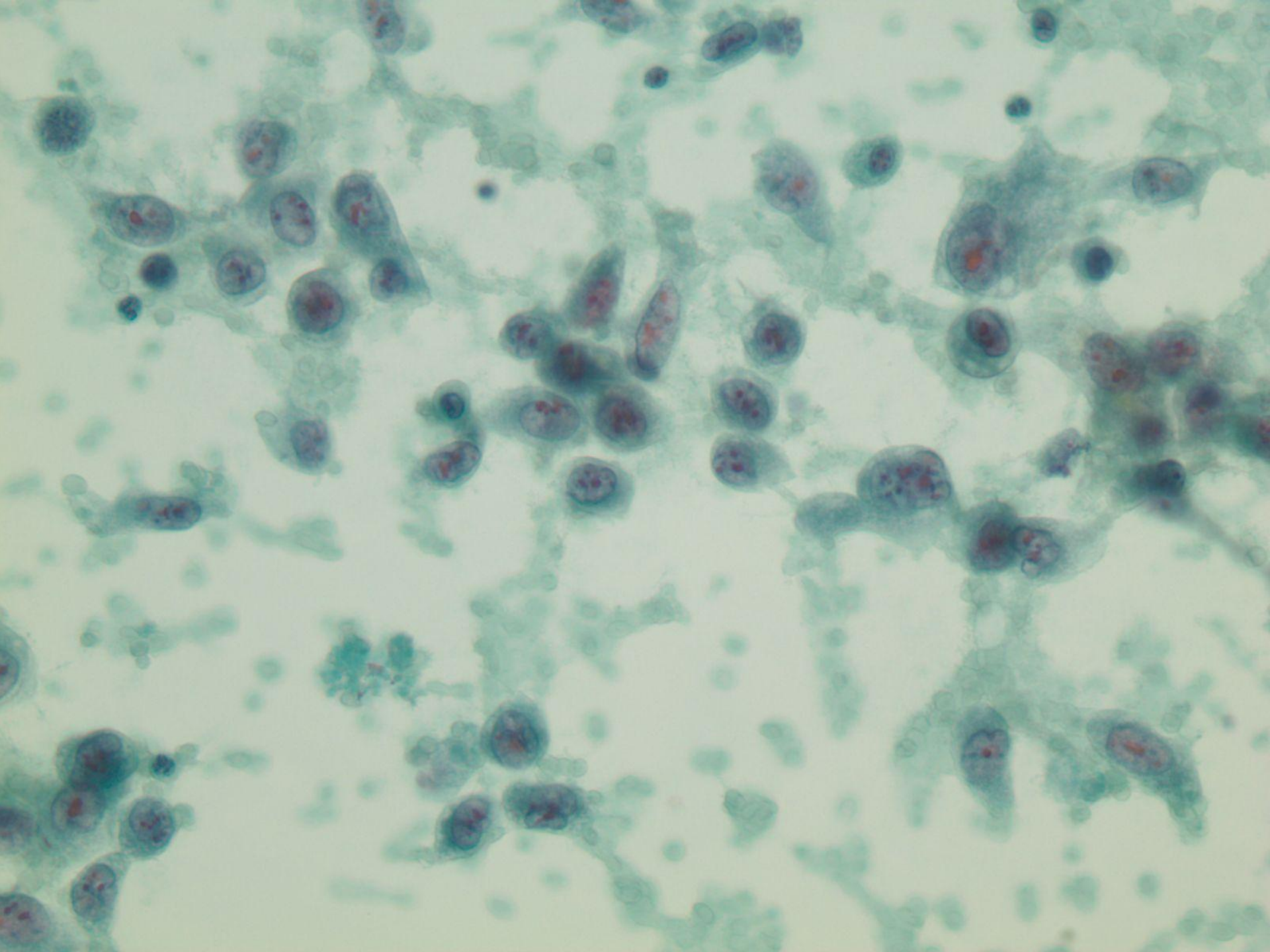
-no significant medical history

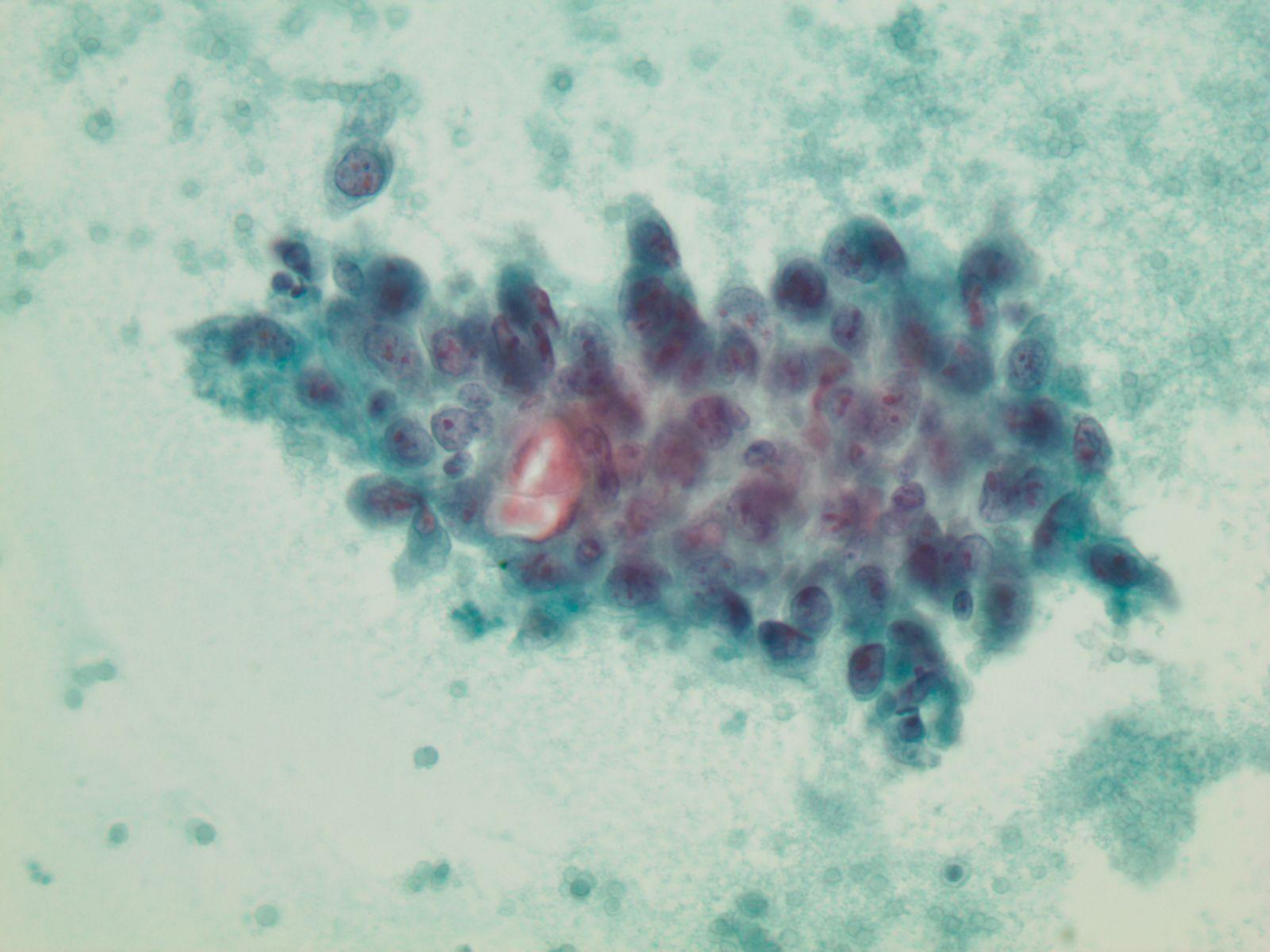
-presented to ER with abdominal pain/distention and ascites

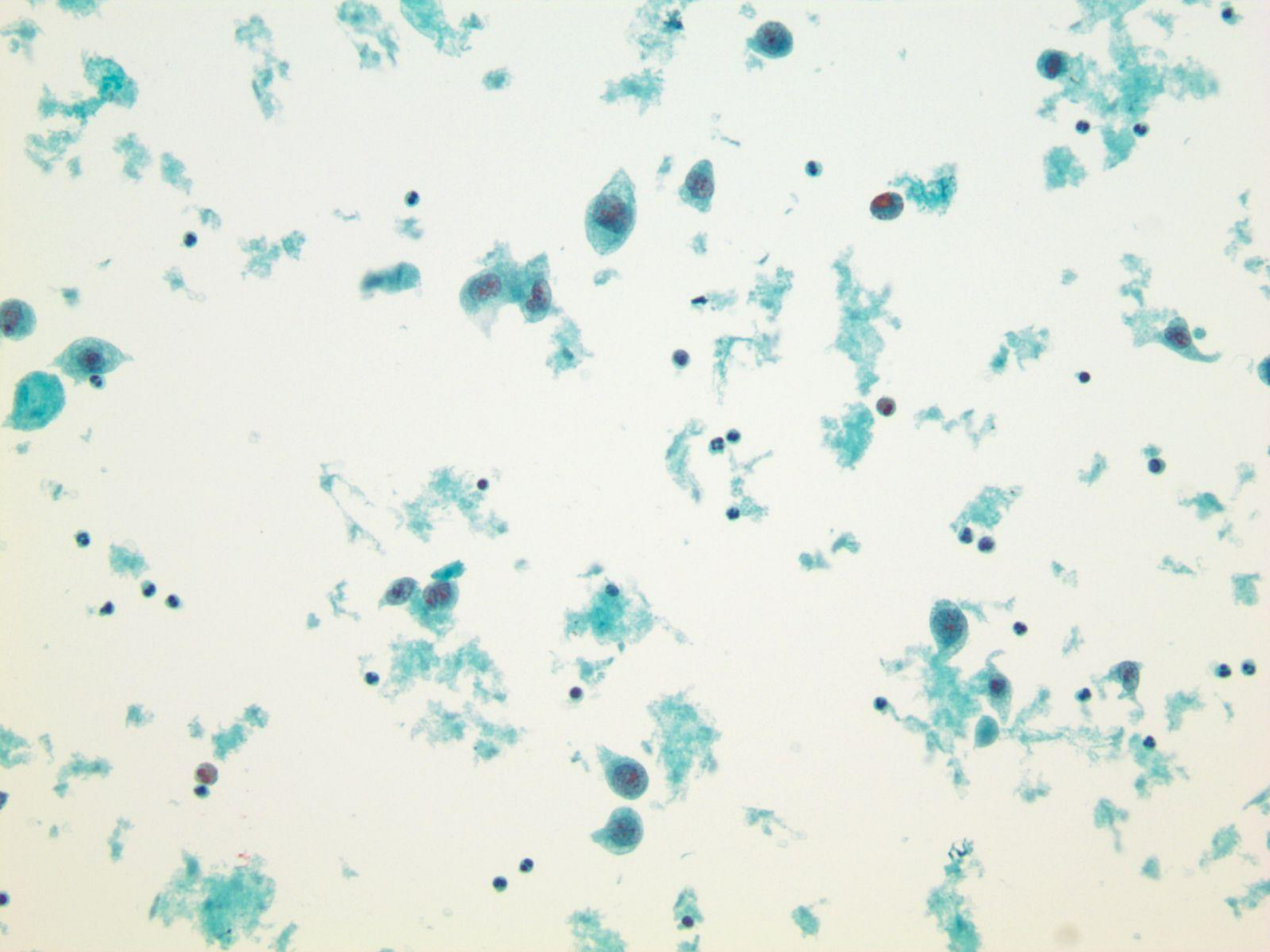
-imaging showed large pelvic mass as well as findings suggestive of carcinomatosis

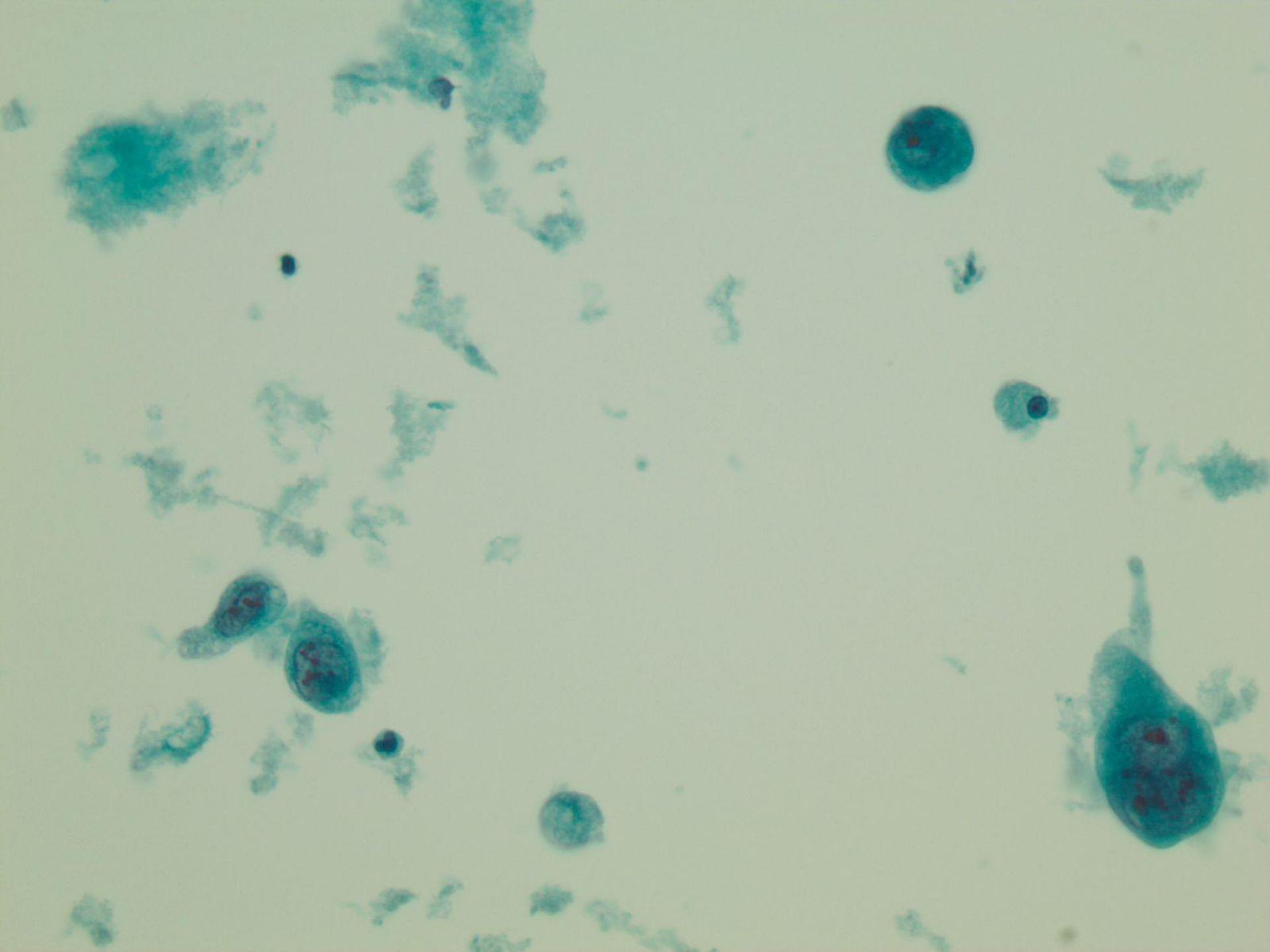
=> referred for CT guided fine needle aspiration

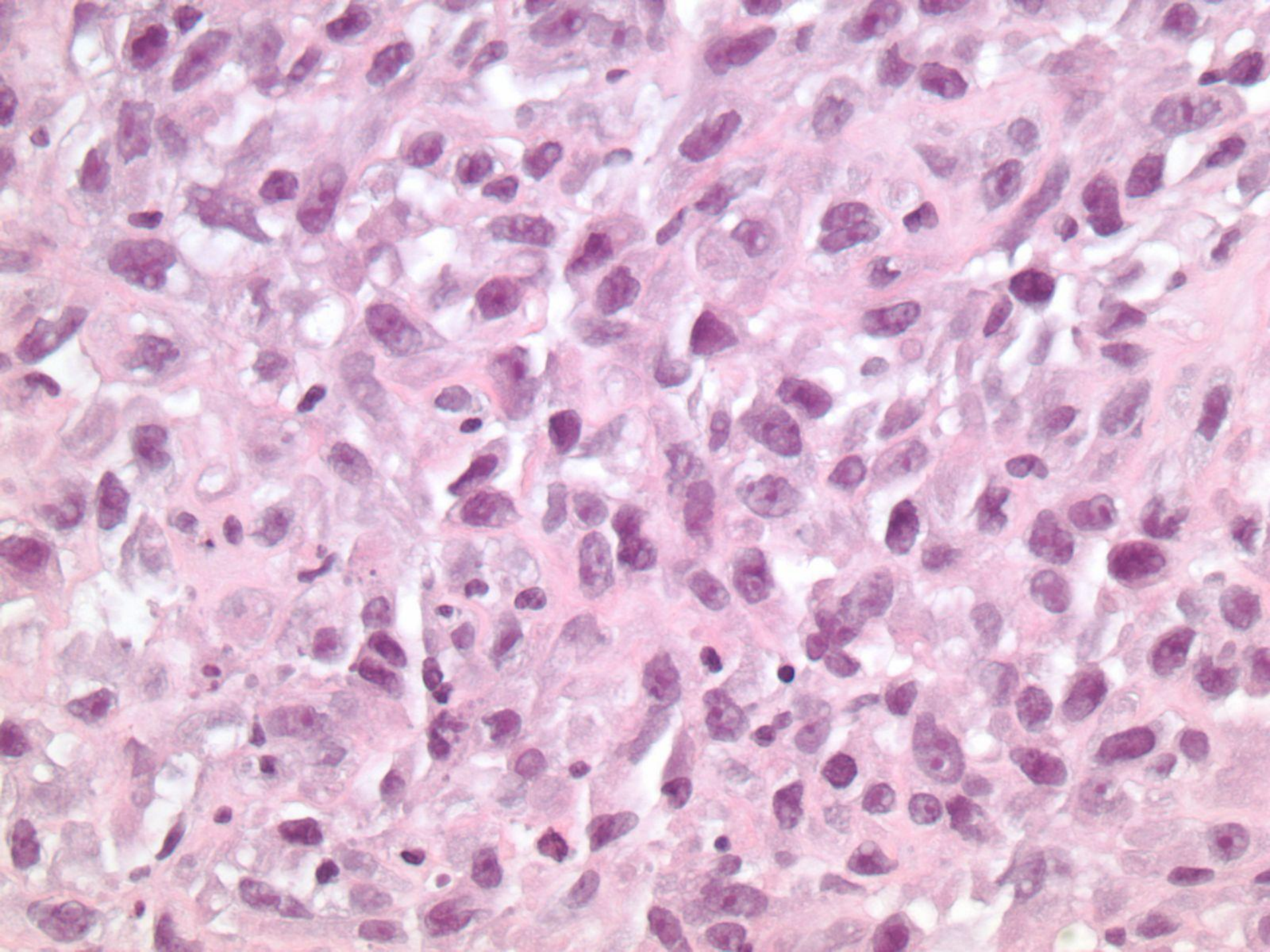


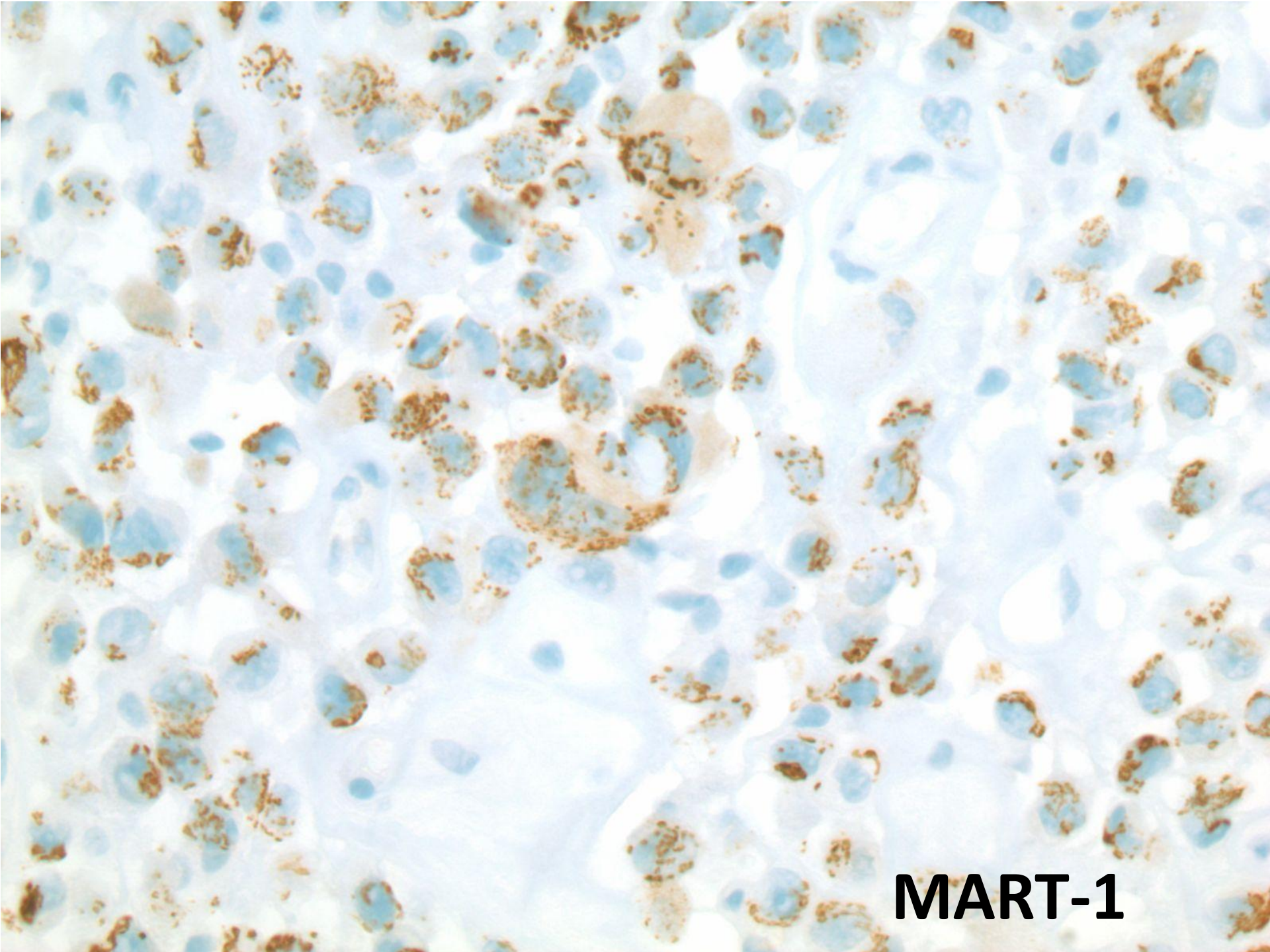











MART-1

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.