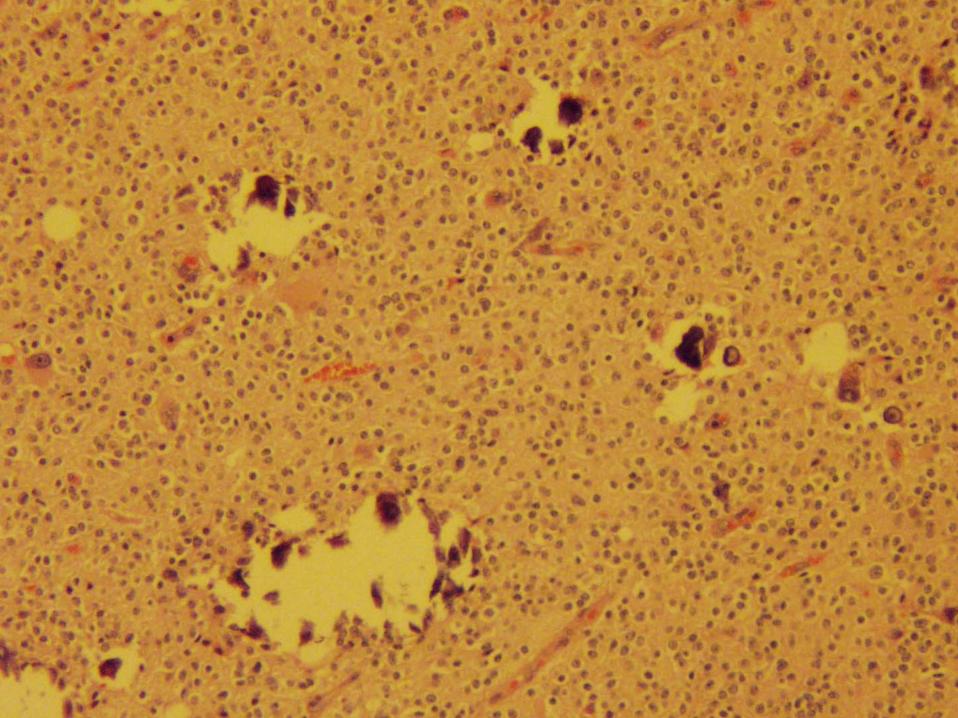
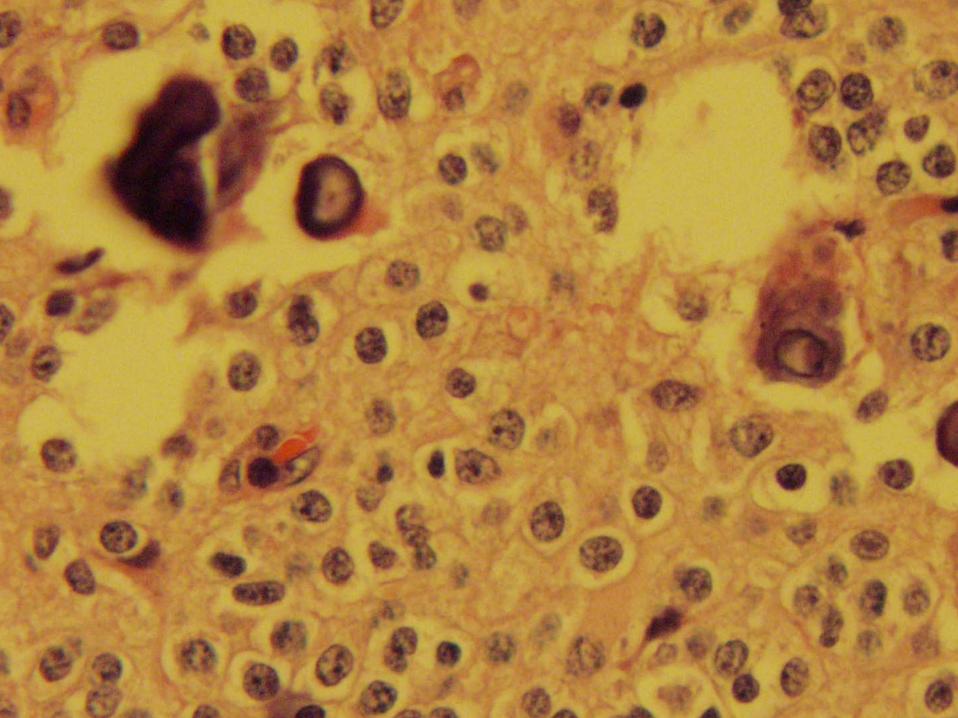
2003 PIP-B Cases

Paul K. Shitabata, M.D.

APMG

- 29M, 2mo hx HA and new onset Sz
- MRI with 7.2 cm hypointense nonenhancing mass in left frontal lobe
- Frontal lobectomy





Oligodendroglioma, WHO Grade II

WHO GRADE	Histopathology
	Deleted
IJ	Low cellularity, round uniform nuclei, absent or scant mitotic figures
	Anaplastic High cellularity with nuclear variation and hyperchromasia, brisk mitotic rate, endothelial proliferation or necrosis
IV	Highly anaplastic=GBM

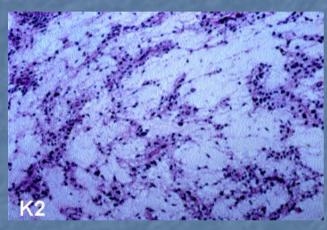
Oligodendroglioma

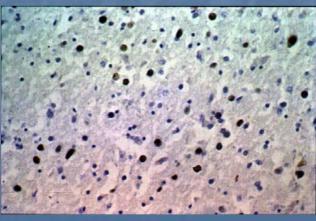
- Perinuclear halos are artifact of delayed fixation
- Occasional minigemistocytes with plumper eosinophilic cytoplasm
- Chicken wire vasculature
- Nodular growth pattern on background of a diffusely infiltrating tumor

Oligodendroglioma

- >5mf/10hpf associated with decreased survival
- Ki-67 labeling index variable, suggestion of 5% for low vs. high risk
- Highly anaplastic oligos do not correspond to GBM
- Allelic loss of 1p and 19q associated with chemosensitivity (40-70% of cases)
 - Correlates best with with classical morphology

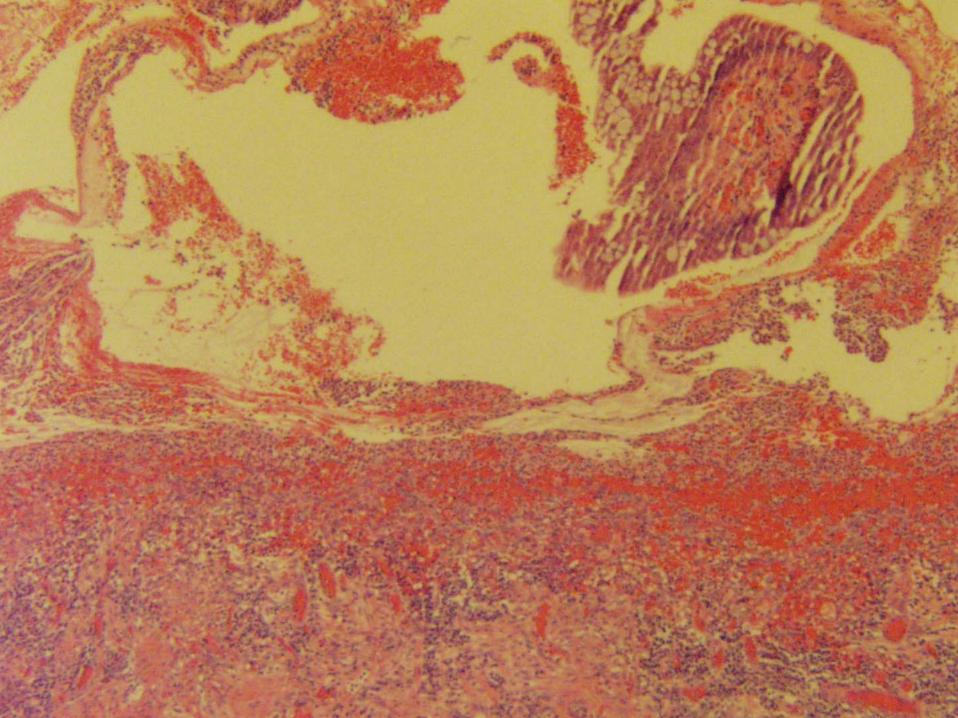
Oligodendroglioma, WHO Grade II DDX

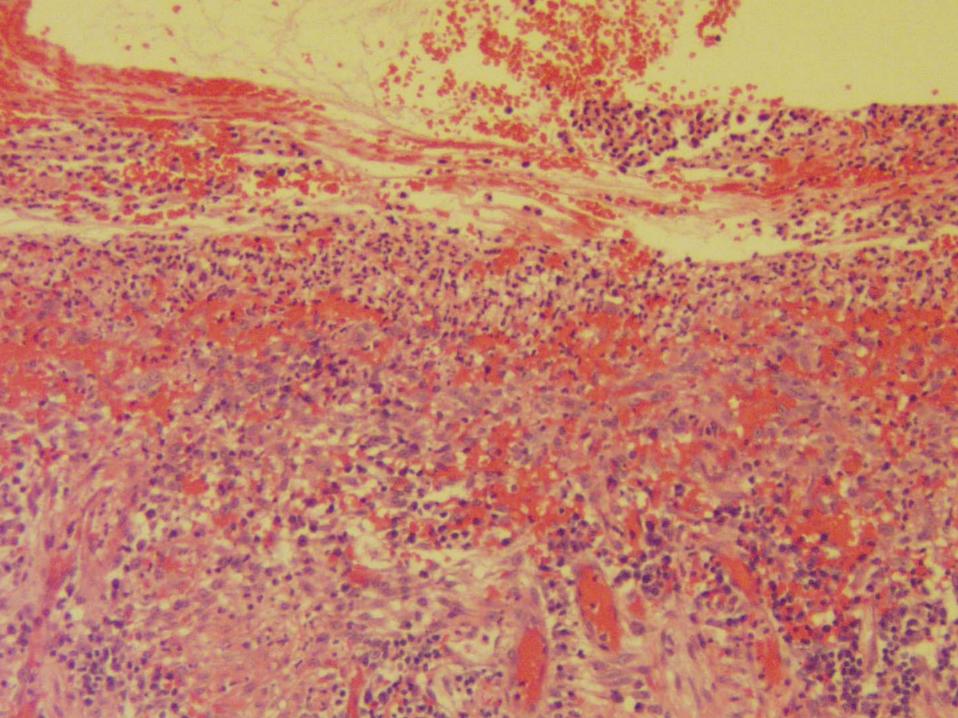


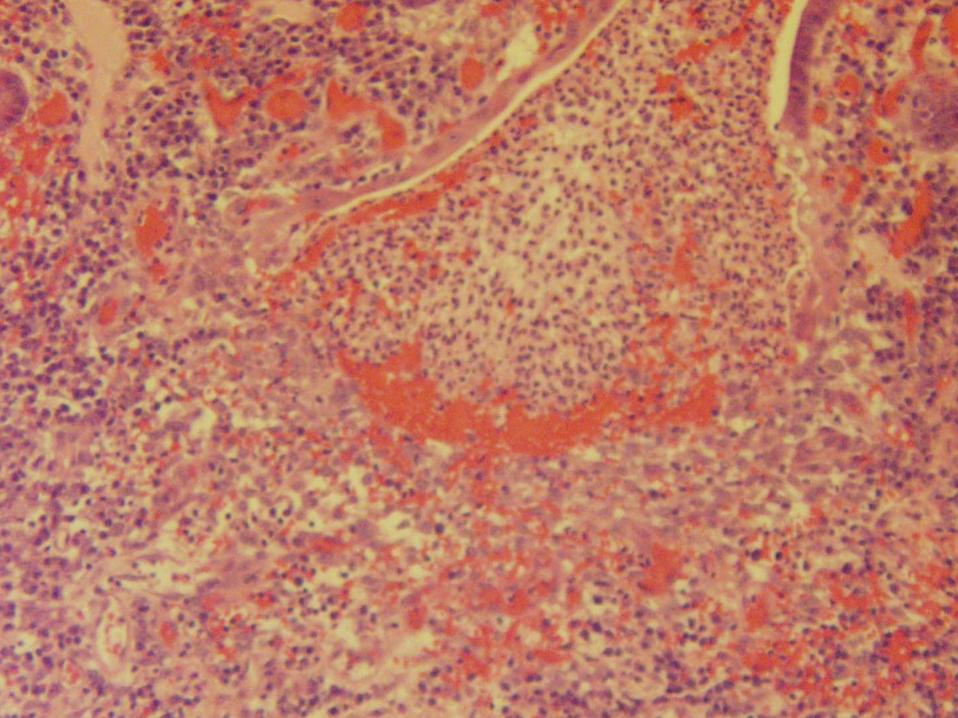


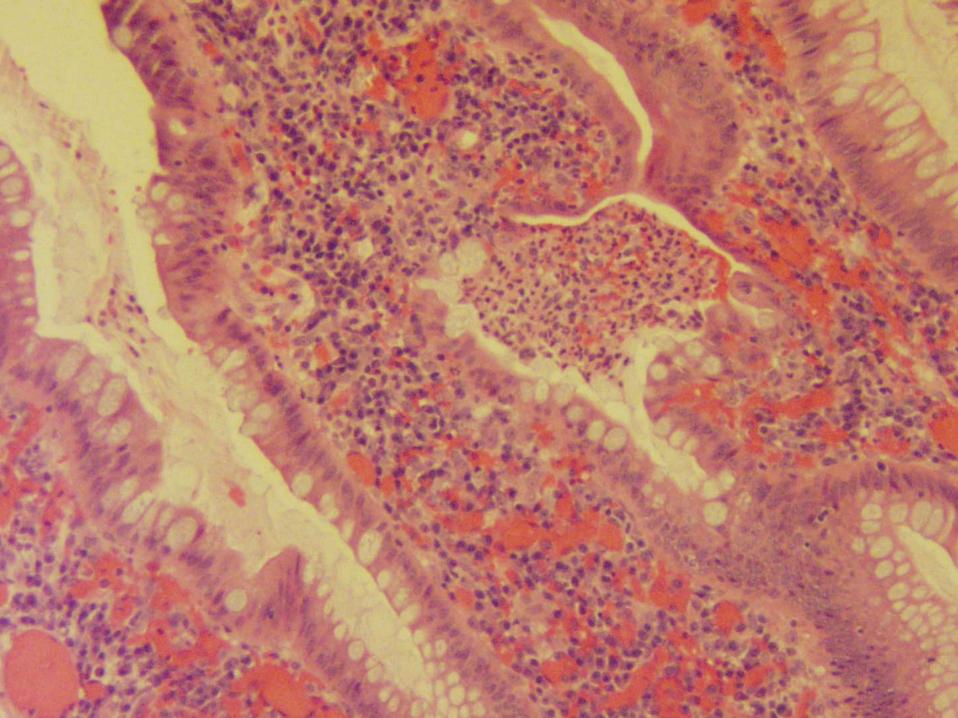
- Dysembryoplastic neuroepithelial tumor
 - Small round cells with delicate fibrillary processes and fine capillaries with pools of mucin and floating neurons
 - Midline region of the septum pellucidum
- Progressive multifocal leukoencephalopathy-JC virus infection

- 54M, 24 hr hx fever, abdominal cramping, severe bloody diarrhea
- Recent travel history
- Steroid and Abx tx with progressive deterioration over 3 days
- Subtotal colectomy with patchy hyperemia and granularity in cecum and 15 cm beyond ileocecal valve to distal resection margin
- Multiple fissures and granularity









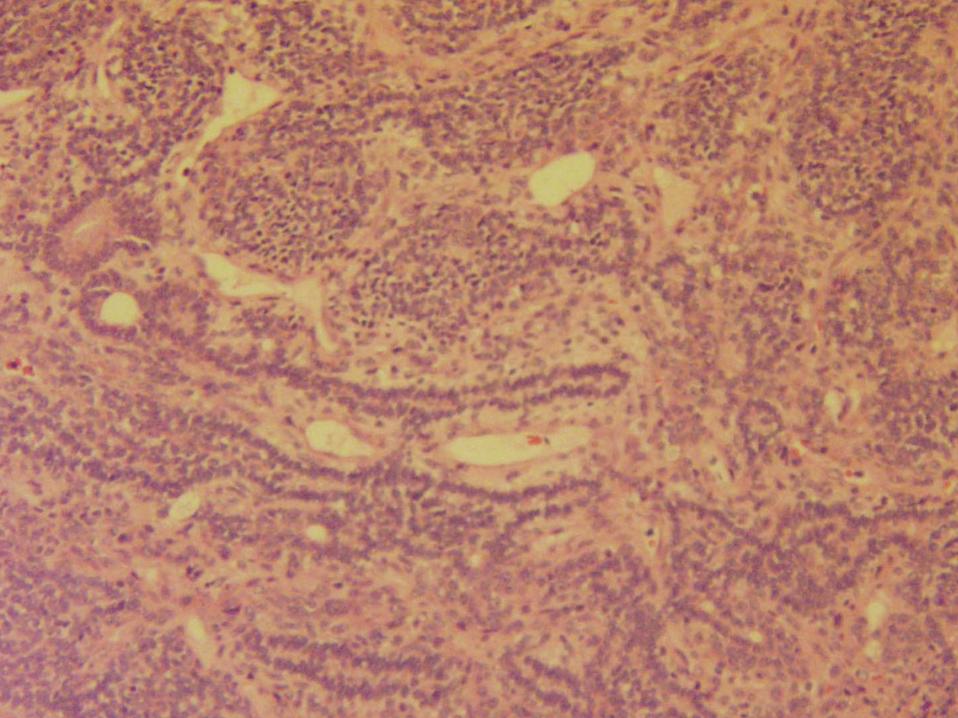
Fulminant Phase of Chronic Ulcerative Colitis

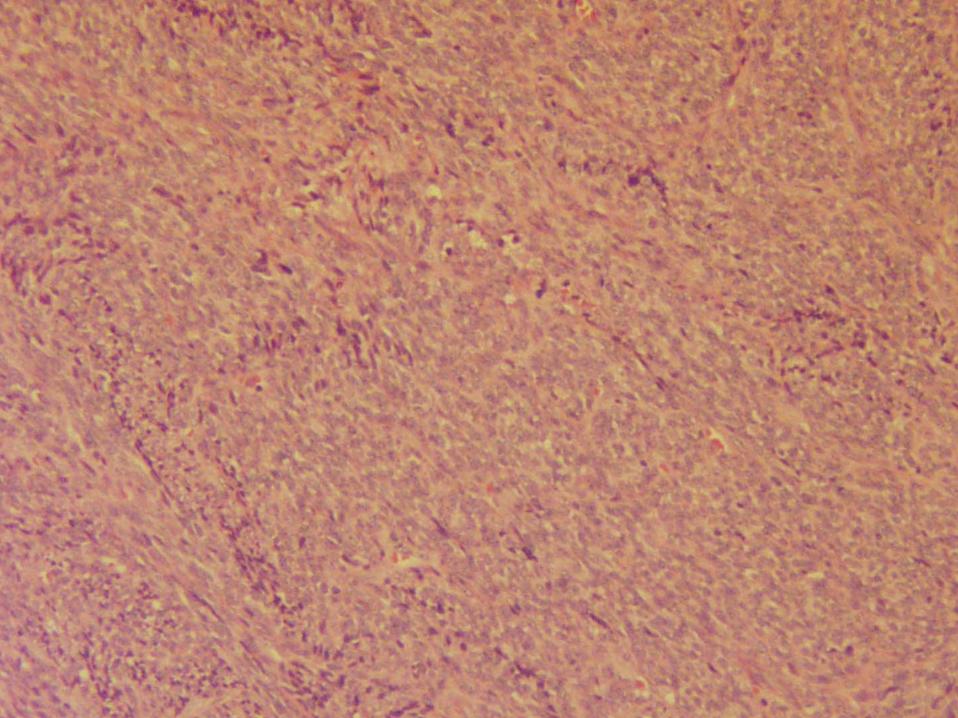
- May be associated with fissuring ulceration of colon
- Not usually associated with:
 - Granulomas, segmental jejunal involvement, skip lesions, or dense lymphoid aggregates in submucosa

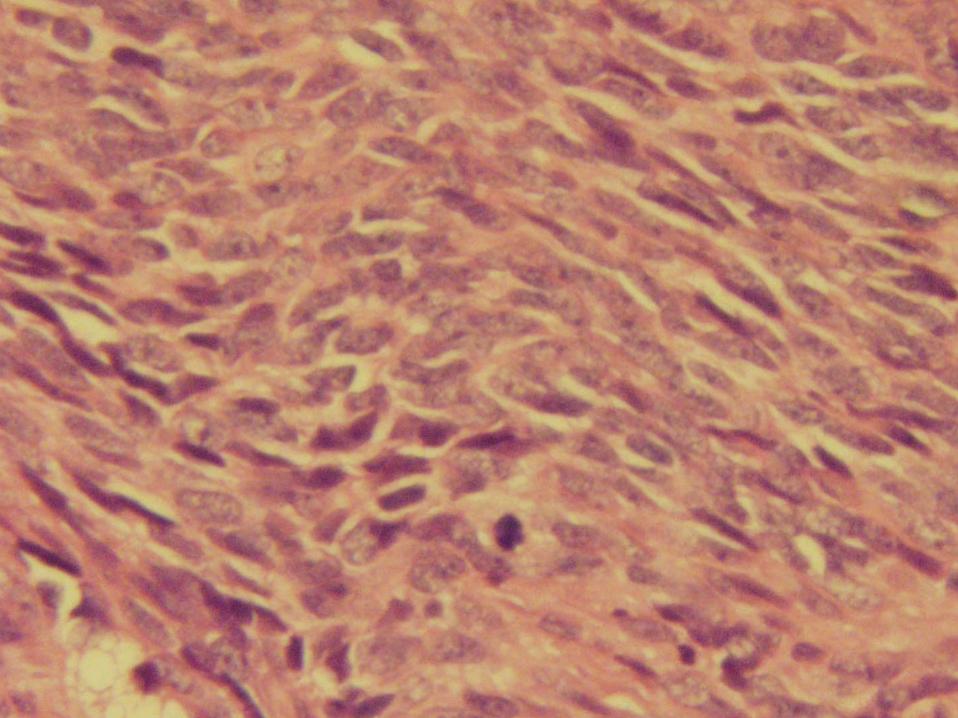
Fulminant Phase of Chronic Ulcerative Colitis

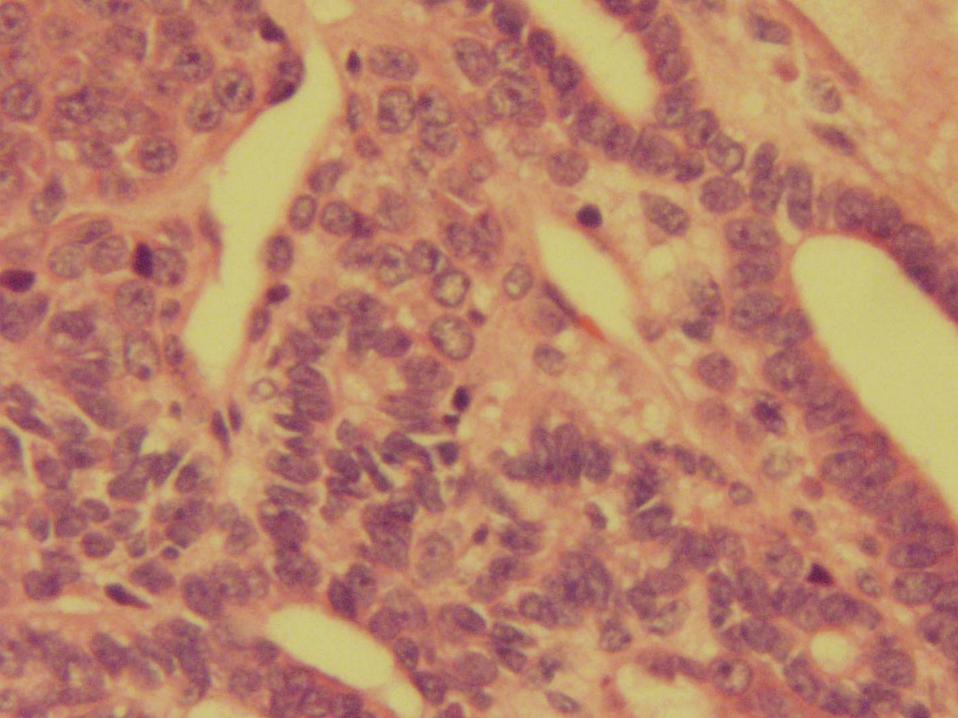
- Distinction from Crohn's colitis important for selection of appropriate surgical resection
 - Reconstructive pouch procedure contraindicated in Crohn's due to high recurrence and complication rate
- DDX:
 - Ischemic colitis
 - Acute and segmental with superficial mucosal necrosis
 - Diverticular disease-associated colitis
 - May be indistinguishable
 - NSAIDs
 - May exacerbate IBD and by itself produce similar findings
 - Disease exacerbation common in poor responders to salicylate treatment

- 53F, asymptomatic with firm pelvic mass extending midway to umbilicus
- TAH-BSO
- Right ovary replaced by firm 14.5 cm 770 gram mass
- Solid yellow-tan cut surface with scattered cysts and focal necrosis









Sertoli-Leydig Cell Tumor

- 0.5% of all ovarian tumors
- 2nd-3rd decades (mean 25 yrs)
- Virilization in 1/3 of cases
 - May have excess estrogen production
 - 50% without endocrine changes
- Majority unilateral
- Range 1-20 cm

Sertoli-Leydig Cell Tumor

- Well differentiated tumors
 - Well formed hollow tubules with fibrous or edematous stroma
- Poorly differentiated tumor
 - Lobulated, ranging from ribbons and solid tubules to diffuse spindle cell sheets
 - Sarcomatoid with numerous MF
- Heterologous elements in 20%
- Inhibin positive

Sertoli-Leydig Cell Tumor

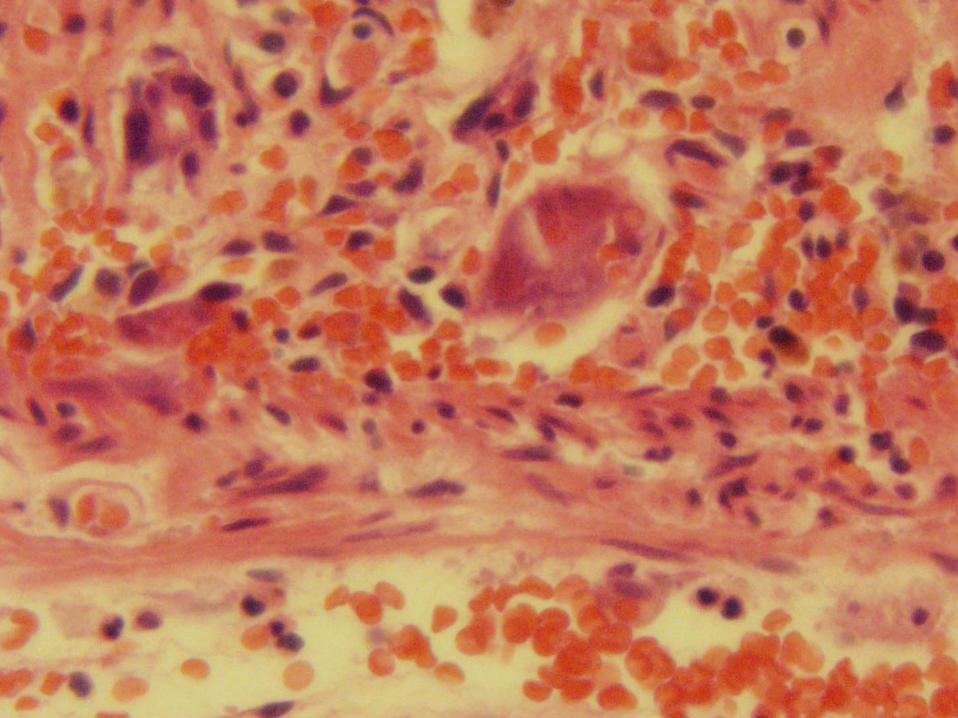
DDX:

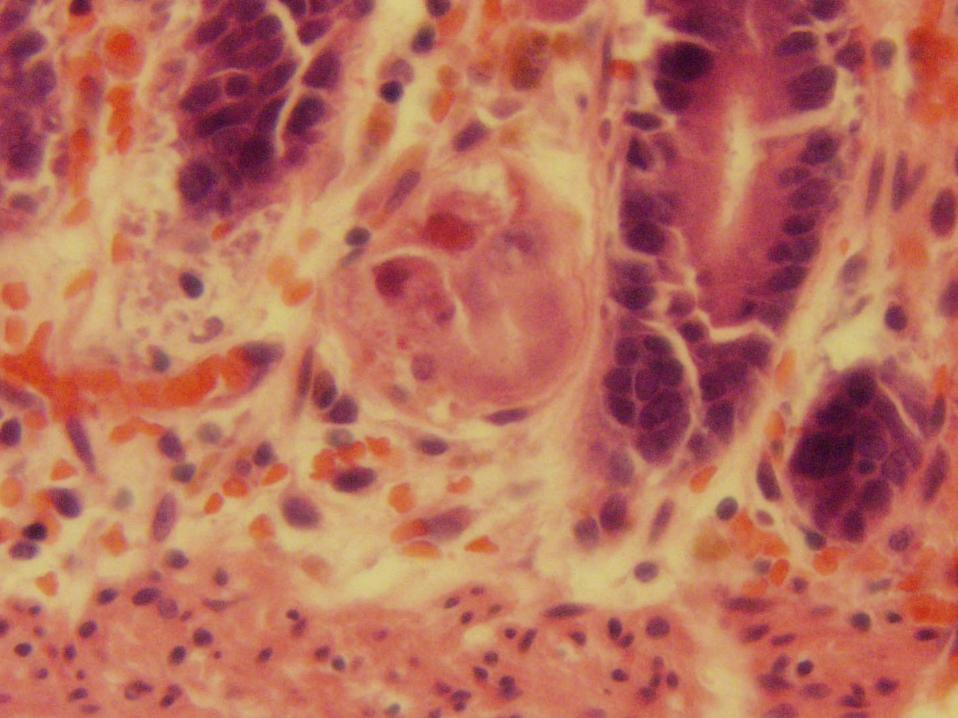
- Adult granulosa cell tumor-inhibin+
- Endometrioid carcinoma-squamous nests
- Metastatic carcinoma-usually bilateral

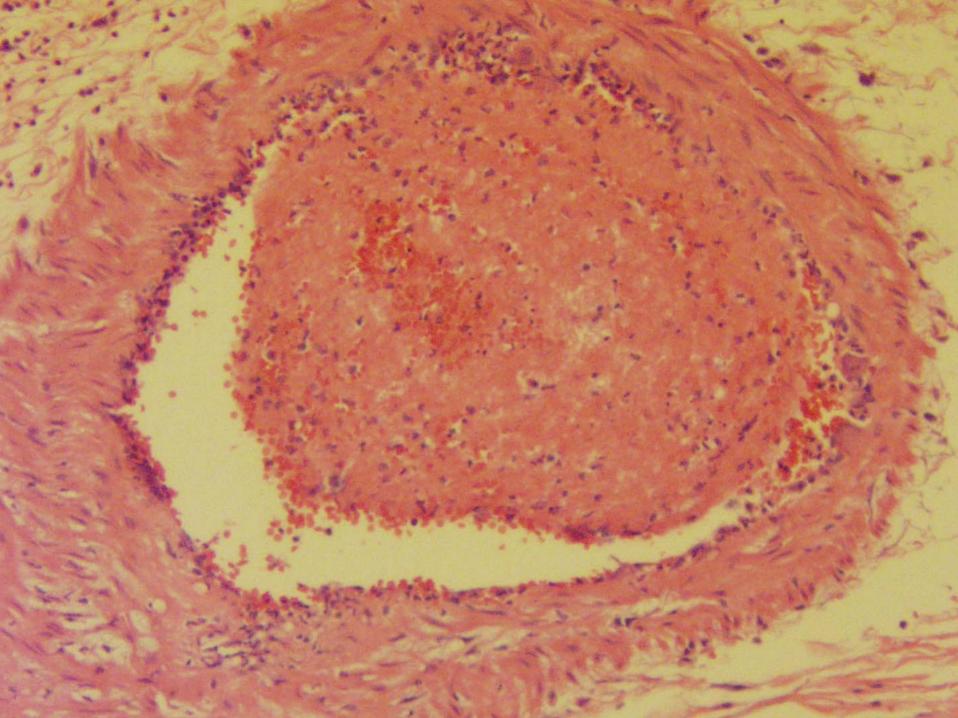
PGX and TX

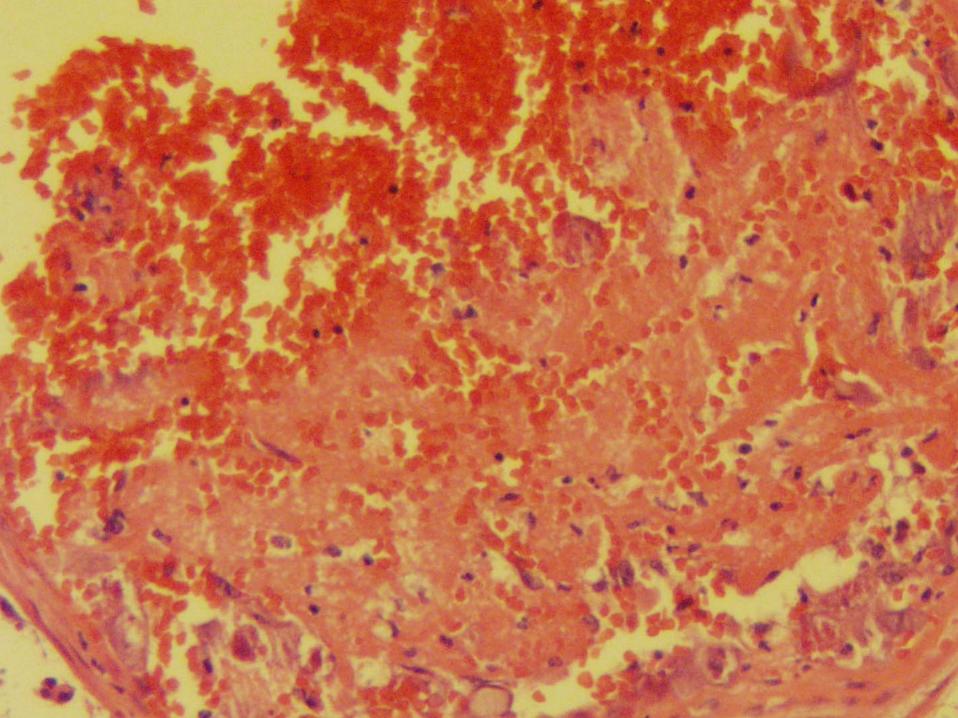
- Majority stage IA, excellent prognosis with 10YRS 92%
- Unilateral SPO
- TAH/BSO for older patients, high tumor state, or malignant heterologous elements

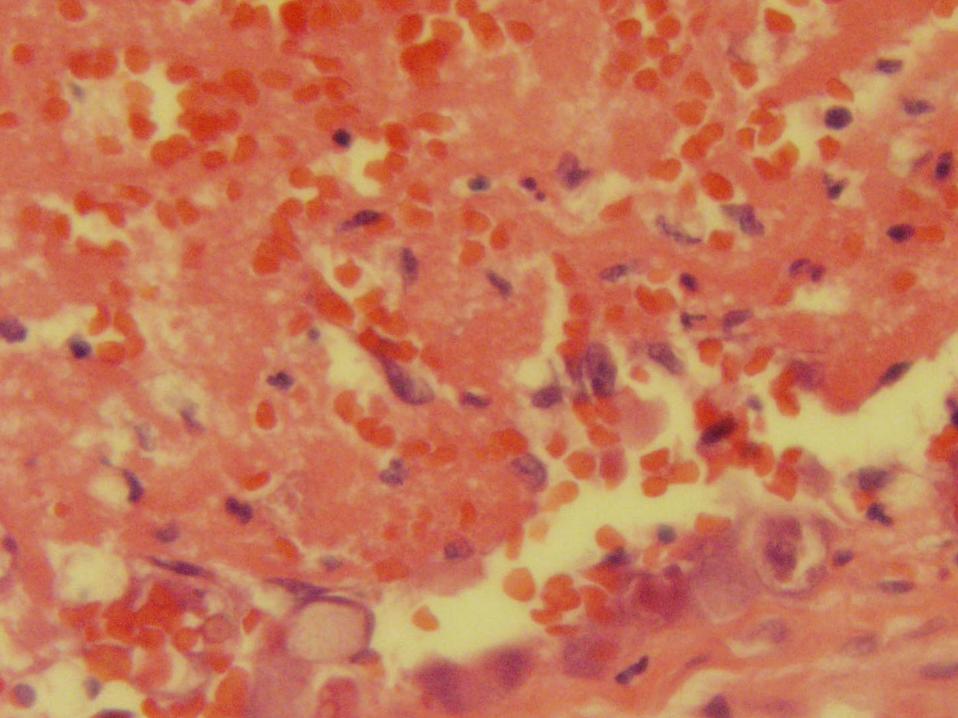
- 64M, s/p cadaveric renal transplant 6 months prior
- 2 wk hx. Malaise and 3 d hx fever and diarrhea
- After admission, sudden onset of diffuse abdominal pain, nausea, bloody diarrhea
- Extensive mucosal ulceration and edema from sigmoid colon to cecum
- Total colectomy











Cytomegalovirus Colitis

- Ulceration of upper GI tract and colitis
- Patchy and right sided
- May have pseudomembranes
- Exuberant granulation tissue may rarely mimic a neoplastic mass

Cytomegalovirus Colitis

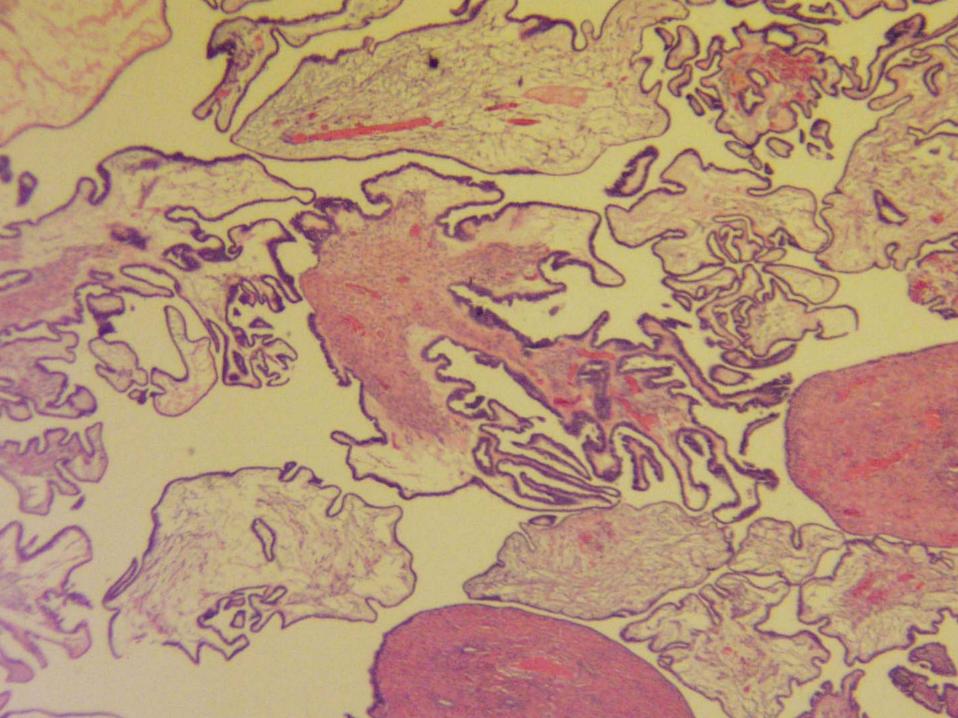
Confirmatory tests:

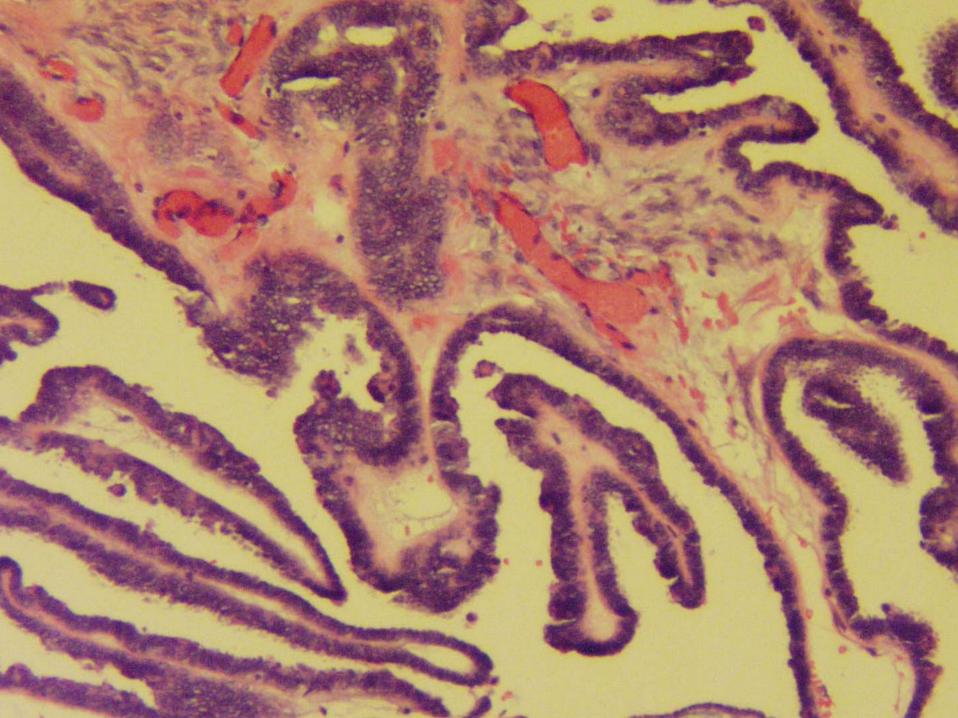
- Viral culture of fresh tissue
- CMV on biopsy sections
- IPOX
- In-situ hybridization
- Stool viral culture not helpful

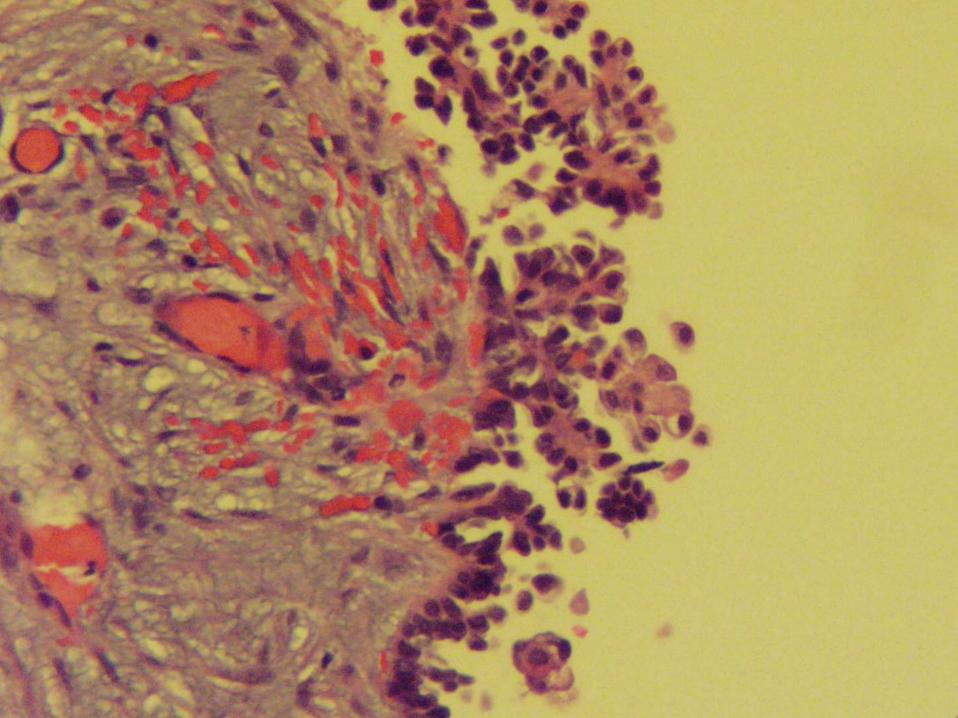
DDX:

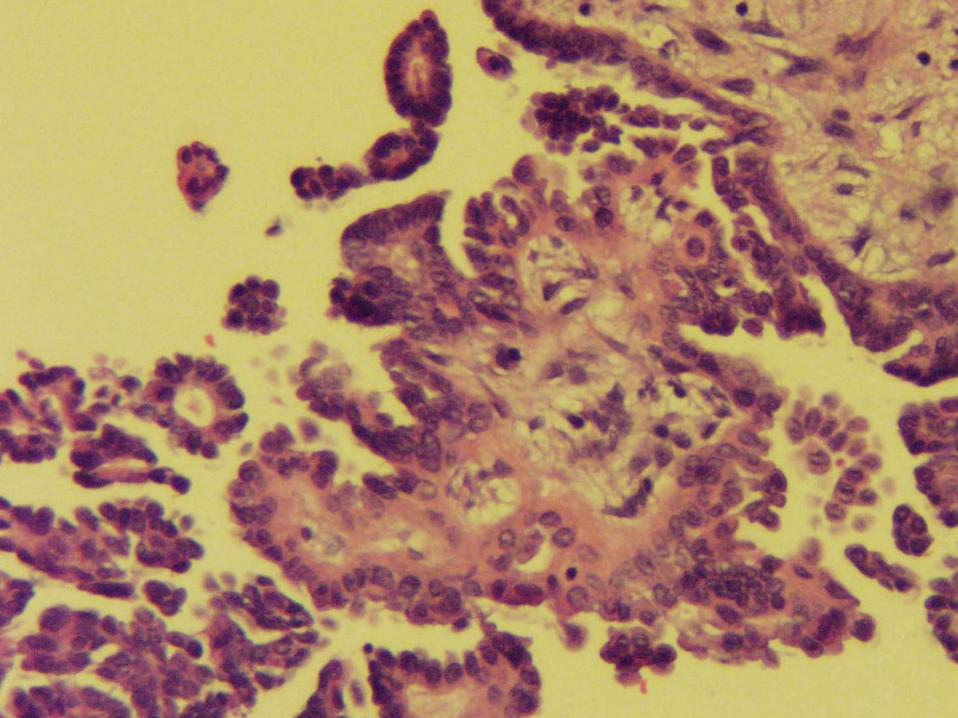
- Hemorrhagic colitis E. coli 0157:H7-hemolytic uremic syndrome
- Adenovirus-amphophilic crescent or targetoid nuclear inclusions
- Ischemic colitis-long distance running, contraceptives, pseudoephedrine, danazol
- C. difficile

- 51F, vague abdominal pain
- Pelvic mass with laparotomy
 - 205 gram, 10.5 cm unilocular cystic mass replacing right ovary
 - Left ovary, abdominal and pelvic surfaces WNL









Serous Borderline Cystadenofibroma

- Extensive epithelial stratification
 - Some suggest at least 10% of tumor should show stratification and budding to be correctly diagnosed
- Usually < 5mm of confluent micropapillary or cribriform growth
- Detachment of epithelial cell clusters
- Epithelial cell atypia
- MF usually <4/10 hpf</p>
- Microinvasion
 - Defined as focus of stromal invasion occupying <10mm2 with no single focus exceeding 3 mm in greatest dimension</p>
 - Occurs in 2-10% of tumors
 - Commonly overlooked
- Epithelial peritoneal implants
 - Noninvasive implants seen in 1/3 of cases
 - Current thought that invasive implants may represent inadequate sampling of a micropapillary serous carcinoma or serous carcinoma

Serous Borderline Cystadenofibroma

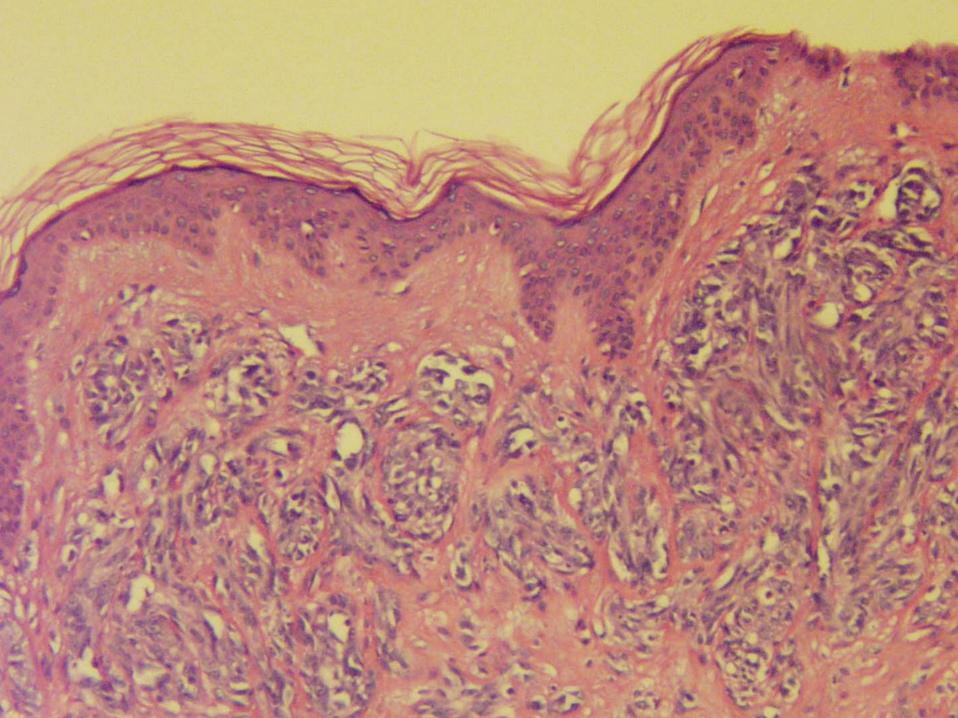
IPOX

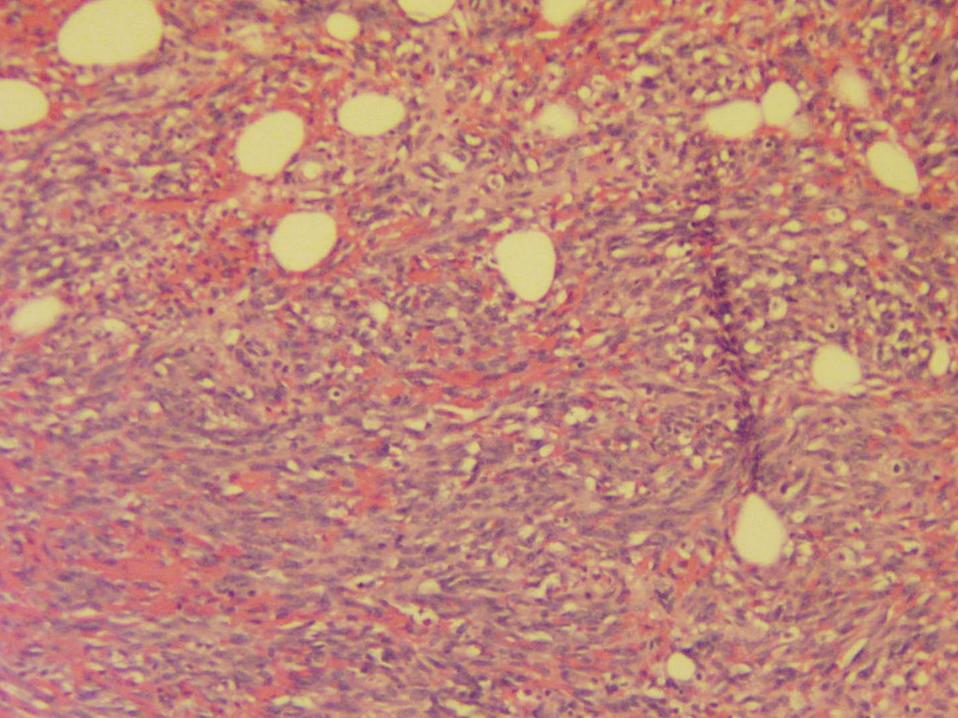
Most surface epithelial tumors are CK7+, EMA+ and ER/PR +

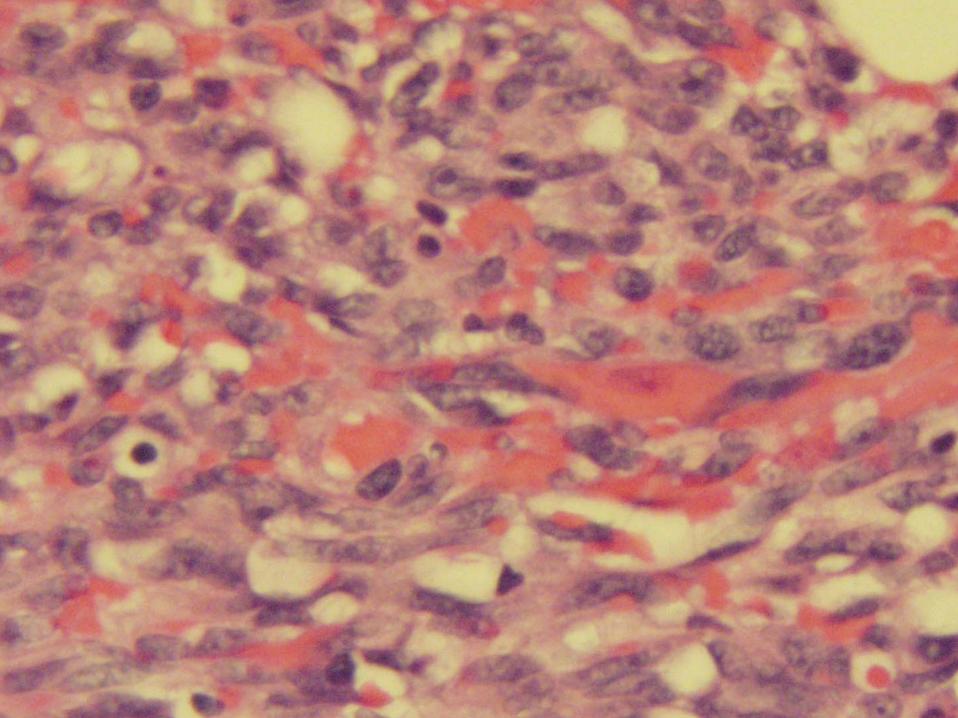
DDX:

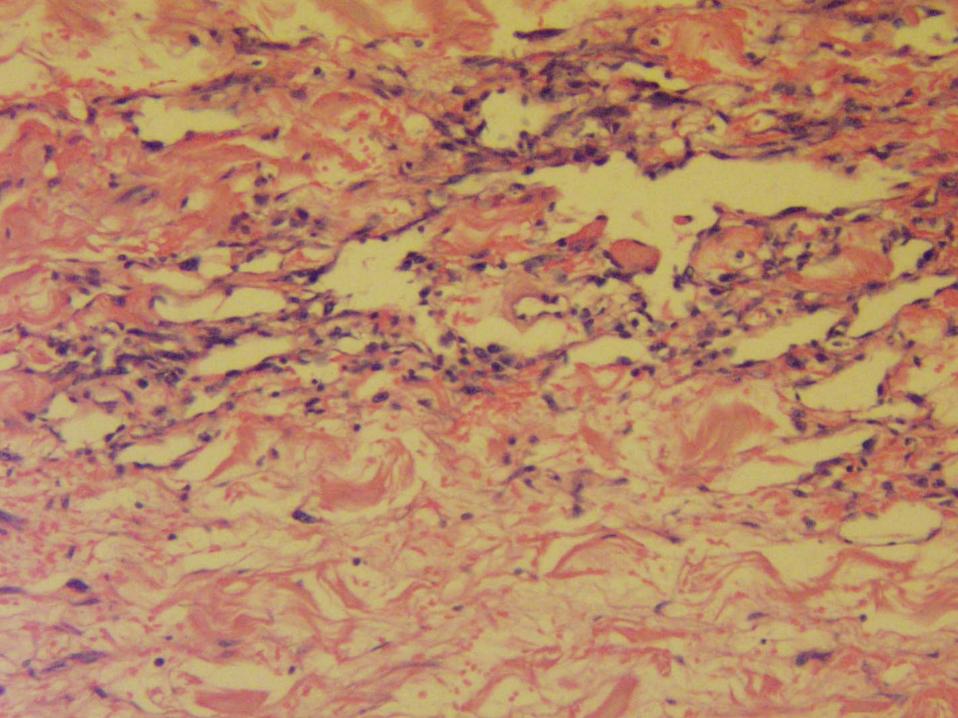
- Serous cystadenoma-most common surface epithelial tumor
- Micropapillary serous carcinoma
 - More cytologic atypia and higher degree of epithelial stratification
 - Small delicate papillae with a filigree pattern instead of hierarchihcal branching
 - Micropapillary and cribriform areas >5 mm.

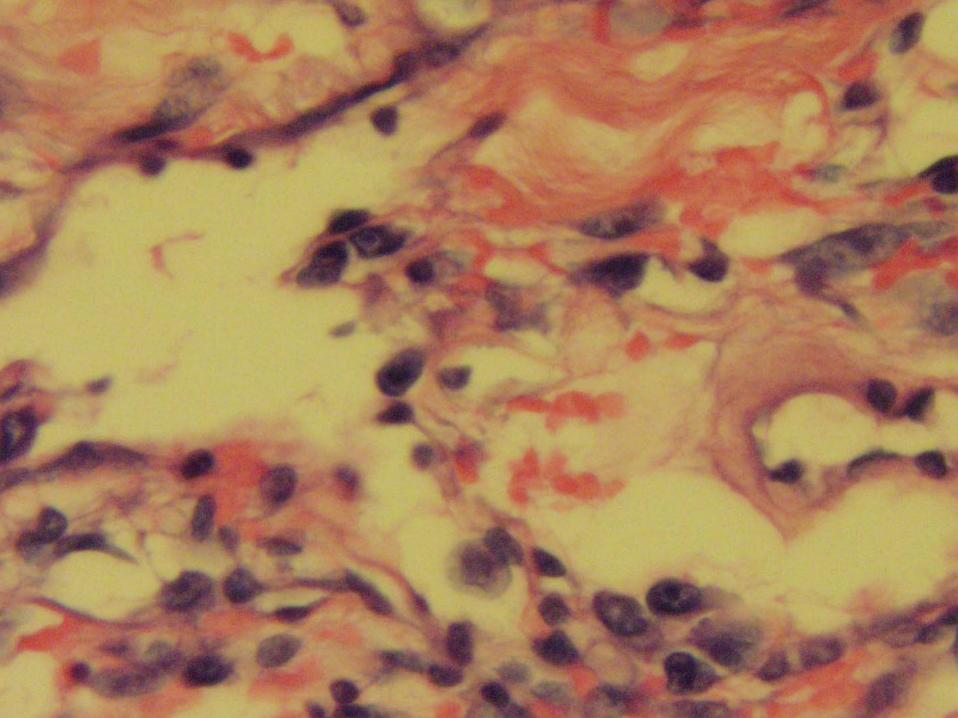
- 77F, multiple gray-purple areas over left breast
- Total mastectomy with multiple hemorrhagic lesions with most of skin surface







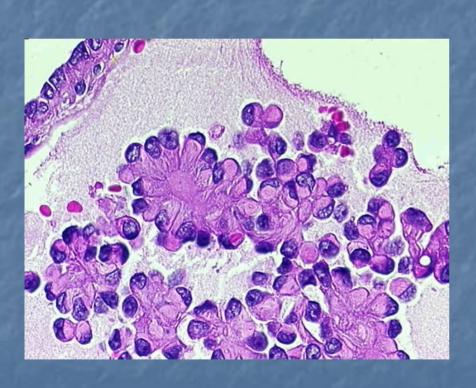




Angiosarcoma

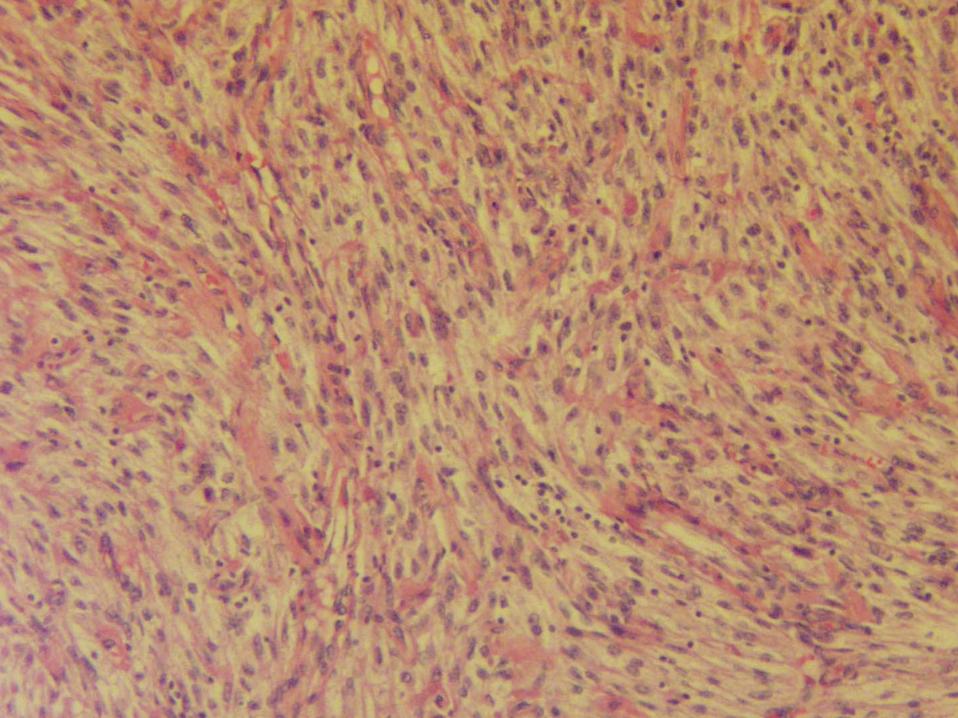
- Stewart-Treves syndrome
 - Associated with chronic lymphedema in post mastectomy patients
- Sites of defunctionalized AV fistulas in renal transplant patients
- Foreign materials
- Post-radiation

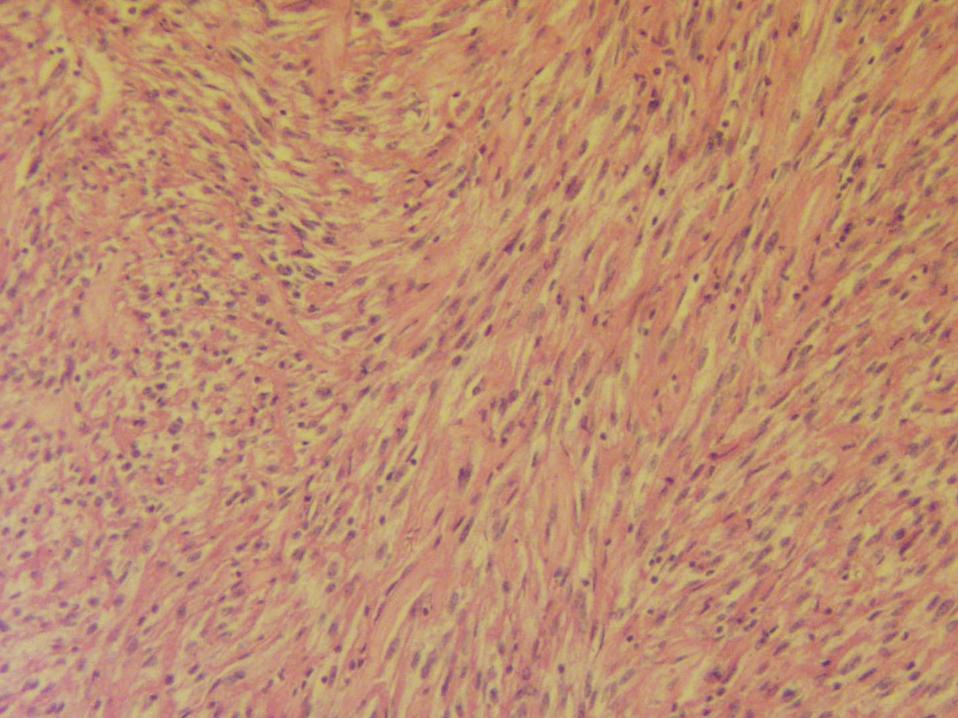
Angiosarcoma DDX

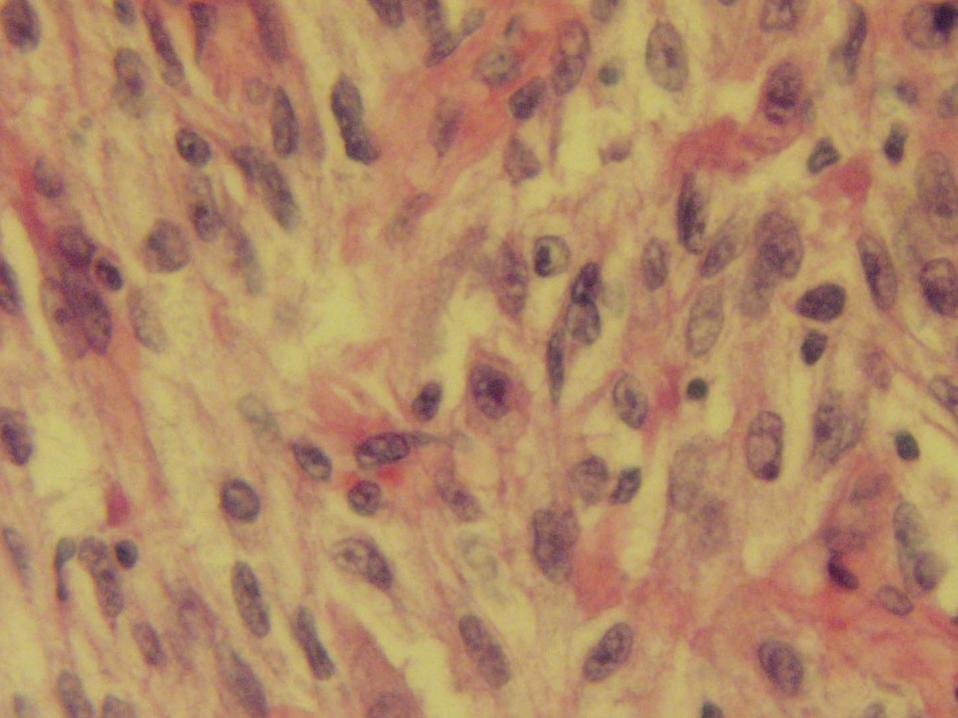


- Kaposi's sarcoma-HHV8+
- Dabska's tumor
 - Intermediate grade malignant cutaneous neoplasm presenting as diffuse swelling or intradermal tumor in children
- Spindle cell hemangioma
 - Distal extremities of children/young adults
 - Dilated thing walled cavernous spaces with solid cellular areas

- 18M, 6 cm mass from hepatic ligamentum teres
- 190 gm, 6 cm firm mass
- No hemorrhage or necrosis



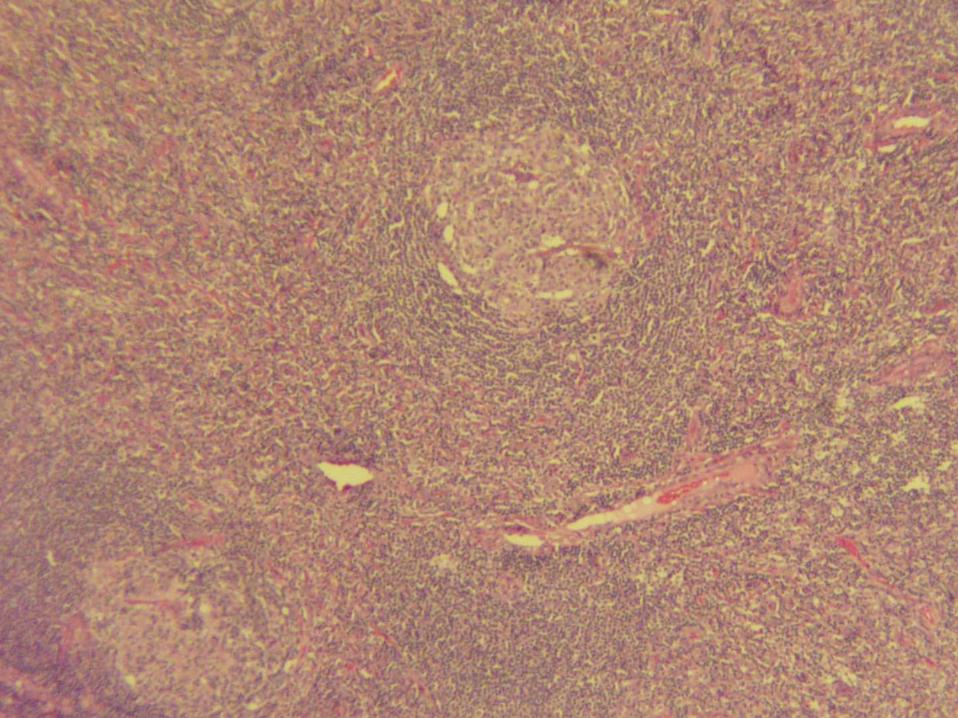


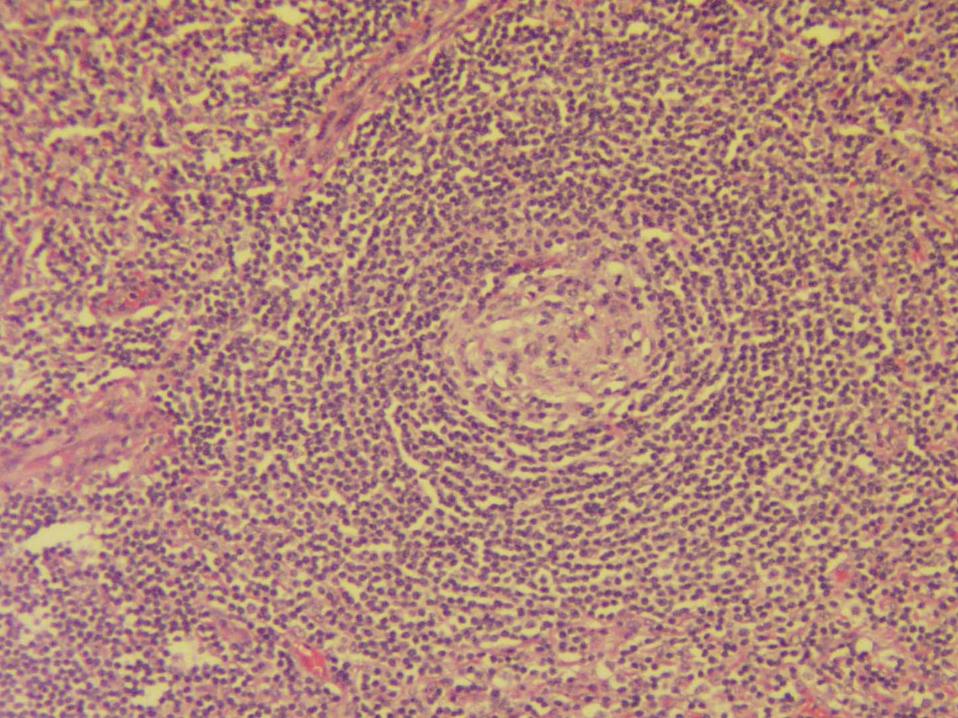


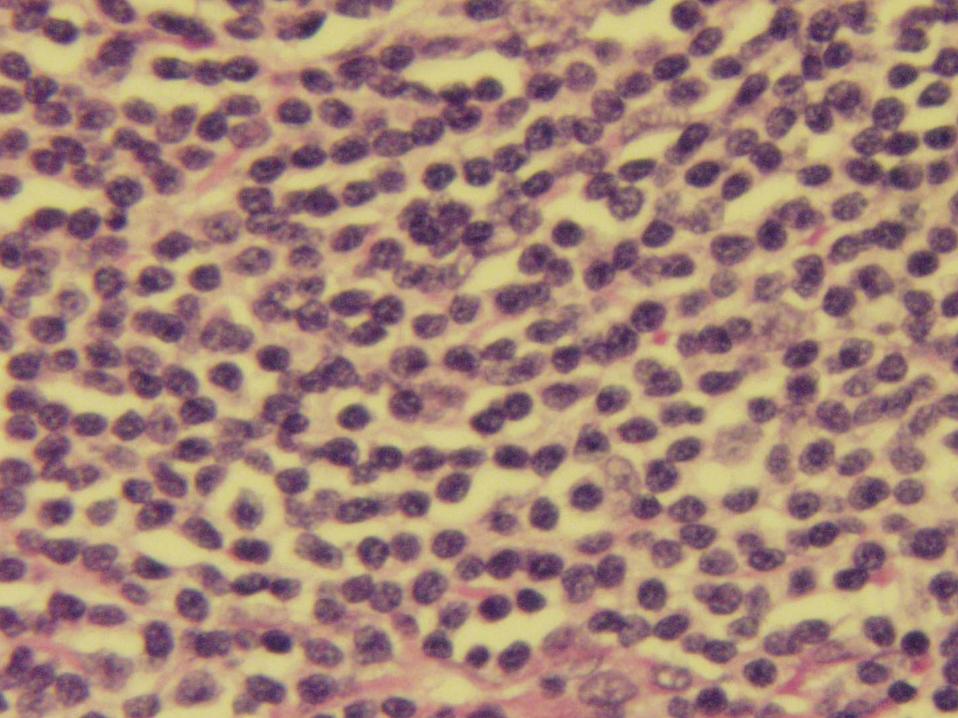
Clear Cell Myomelanocytic Tumor of the Hepatic Falciform Ligament

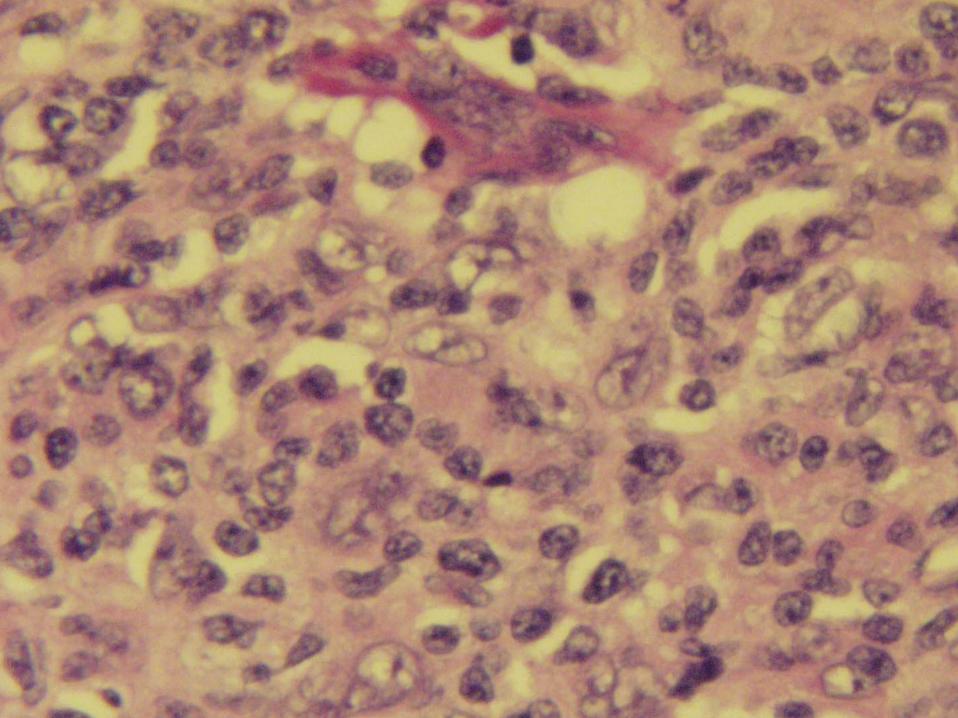
- PEComa (Perivascular epithelioid cell tumors)
 - Clear cell tumors with HMB-45 reactivity and premelanosomes
 - Co-expressed SMA and melanocytic markers
 - Most express tuberin but CCMMT lacks
- DDX:
 - Desmoplastic SRBCT-polyphenotypic but HMB45 negative
 - Clear cell sarcoma
 - Has epithelioid areas and \$100 positive
 - GIST

- 20F, asymptomatic with solitary anterior chest wall mass
- 8.5 cm circumscribed mass with red-tan cut surface









Castleman Disease

- Prominent interfollicular proliferation of blood vessels and small lymphocytes
- Obliteration of lymph node sinuses, abnormal follicles with regressive germinal centers and onion-scaling lymphocytes

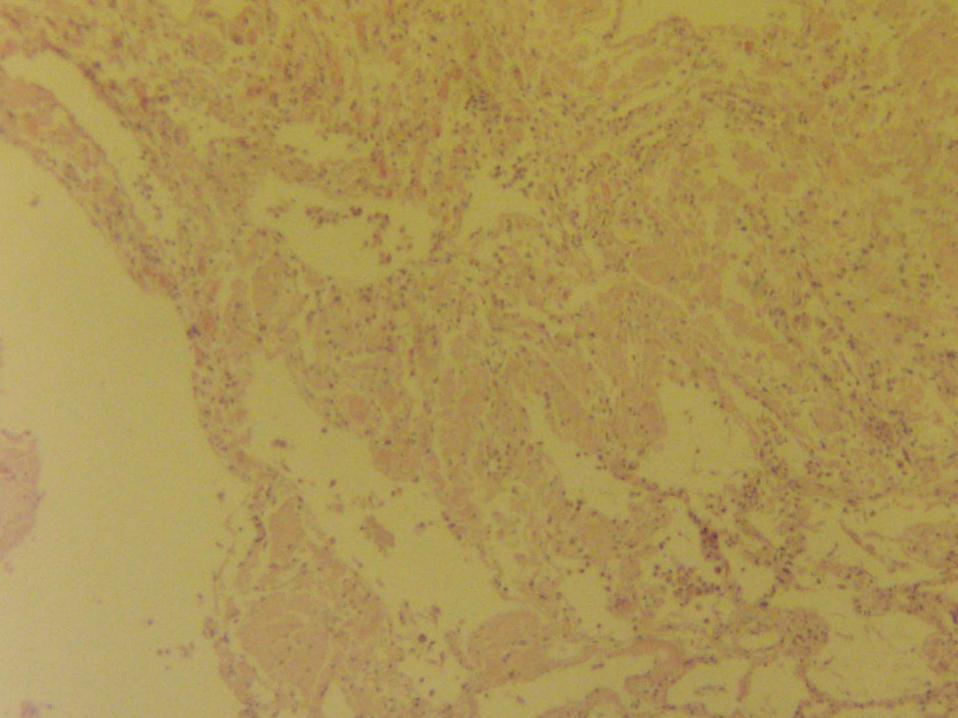
Castleman Disease

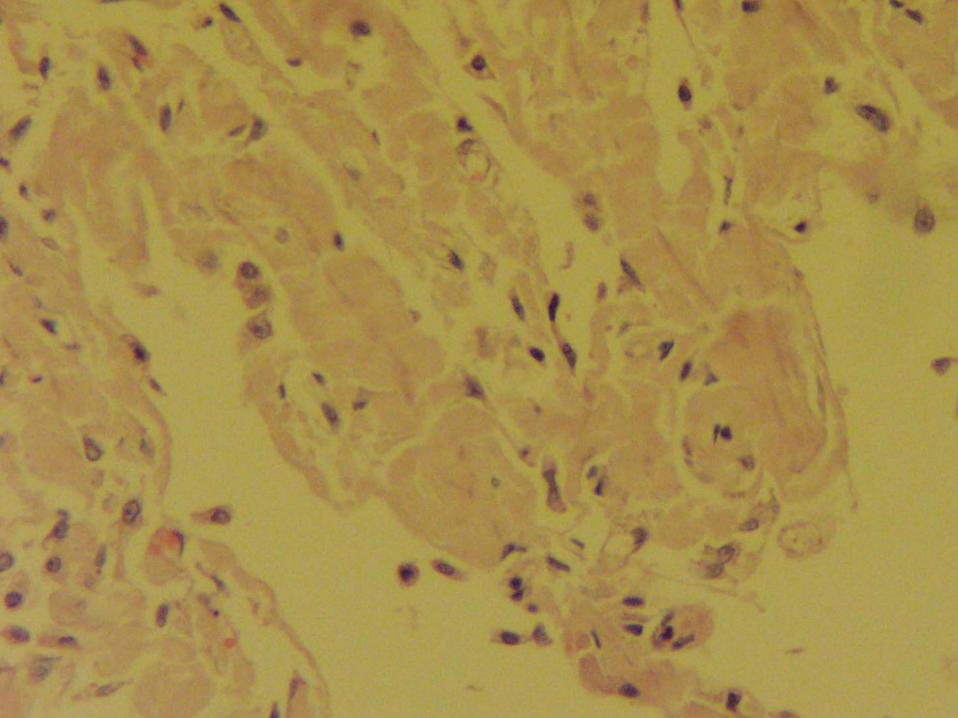
- Localized
 - Hyaline vascular
 - **80-90%** of cases
 - Young adults, F>M
 - Mediastinum, cervical region, abdoment
 - Extranodal presentation common
 - Plasma cell variant
 - Systemic symptoms common
 - ESR elevated, anemia, polyclonal hyper IgG
 - Multiple and intrabdominal lymph nodes
- Systemic
 - Plasma cell variant
 - Thought to be due to IL-6 production
 - Grouped with atypical lymphoproliferative disorders

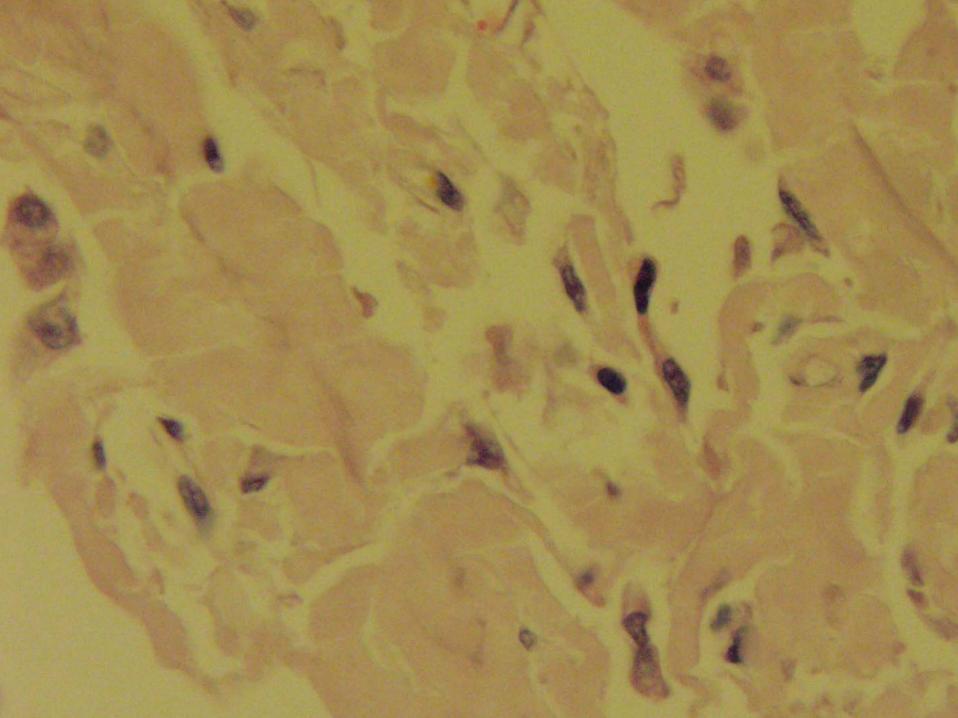
Castleman Disease DDX

- Angioimmunoblastic T-cell lymphoma
 - Vascular proliferation and interfollicular or diffuse proliferation of intermediate size atypical T-lymphocytes with clear cytoplasm, increased MF
- Hodgkin lymphoma
- HIV associated lymphadenopathy
 - Sinuses are patent and distended
 - Polymorphous interfollicular proliferation
 - Diffuse lymphadenopathy
- Mantle cell lymphoma
 - Effacement of lymph node architecture
 - Cytologically atypical mantle cells

- 50M, malabsorption, anasarca, progressive dyspnea following heart transplant, s/p 1 year
- Death from respiratory failure
- Lungs at autopsy with scattered pinpoint nodules and firm







Pulmonary Amyloidosis, Diffuse Parenchymal Type

- Usually associated with systemic amyloidosis or secondary to myeloma
- Usually does not cause severe respiratory impairment
- Most cases not familial and not associated with hemodialysis
- Histologic appearance indistinguishable from pulmonary involvement by systemic light chain disease

Pulmonary Amyloidosis, Diffuse Parenchymal Type

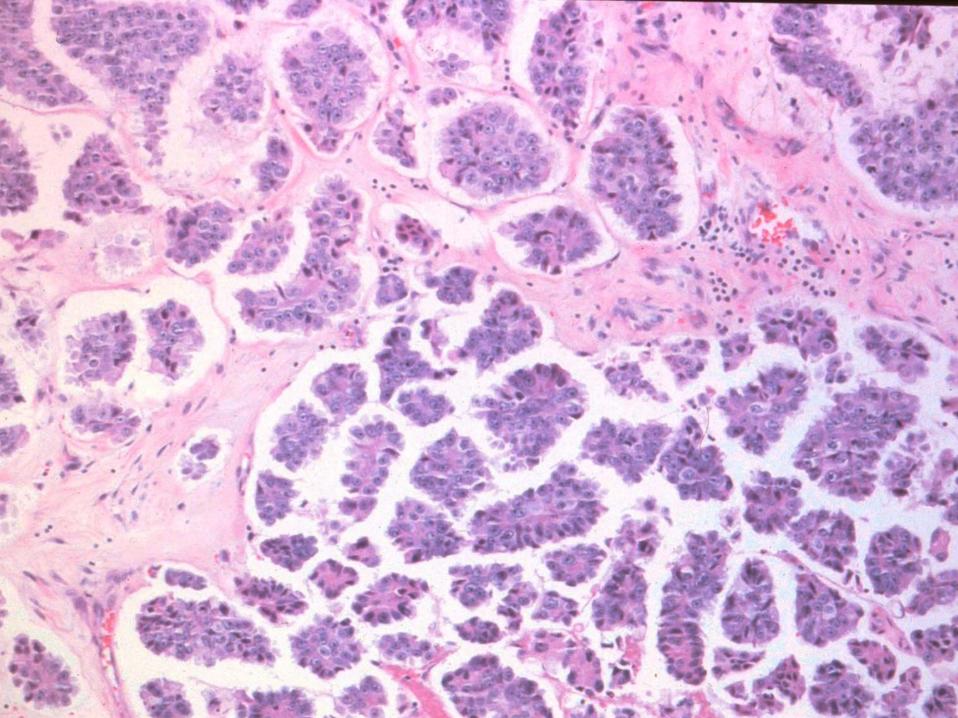
- Tracheobronchial
 - Rare, single or multiple
 - Mimic CA clinically
- Nodular (Amyloid tumor)
 - Single or multiple
 - May cavitate and present with hilar adenopathy
 - Calcification and ossification
 - Systemic amyloidosis usually not present

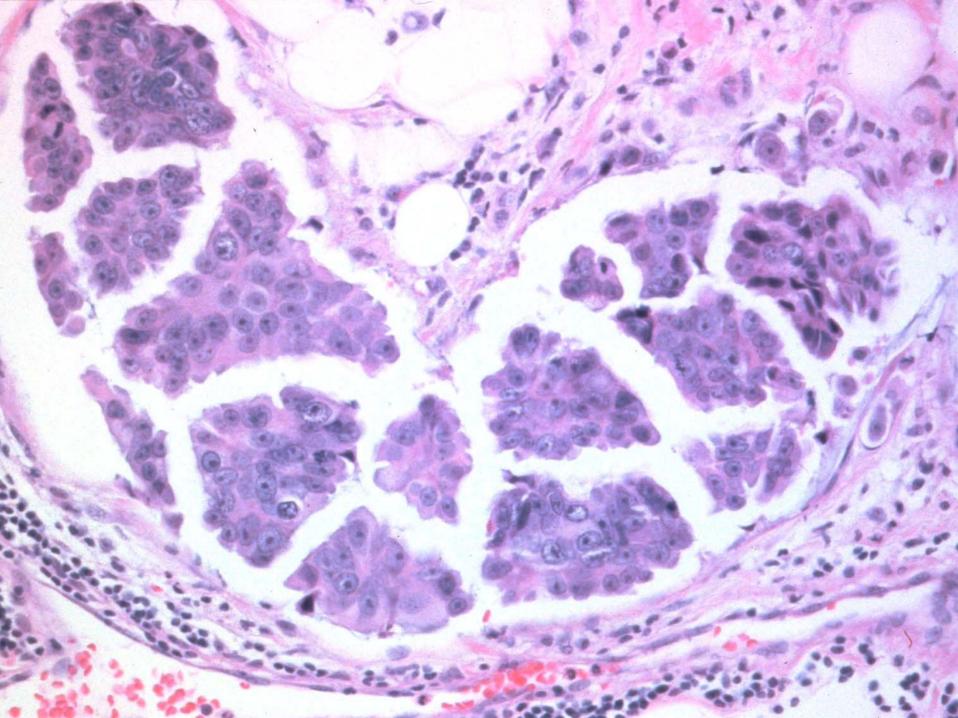
Pulmonary Amyloidosis, Diffuse Parenchymal Type

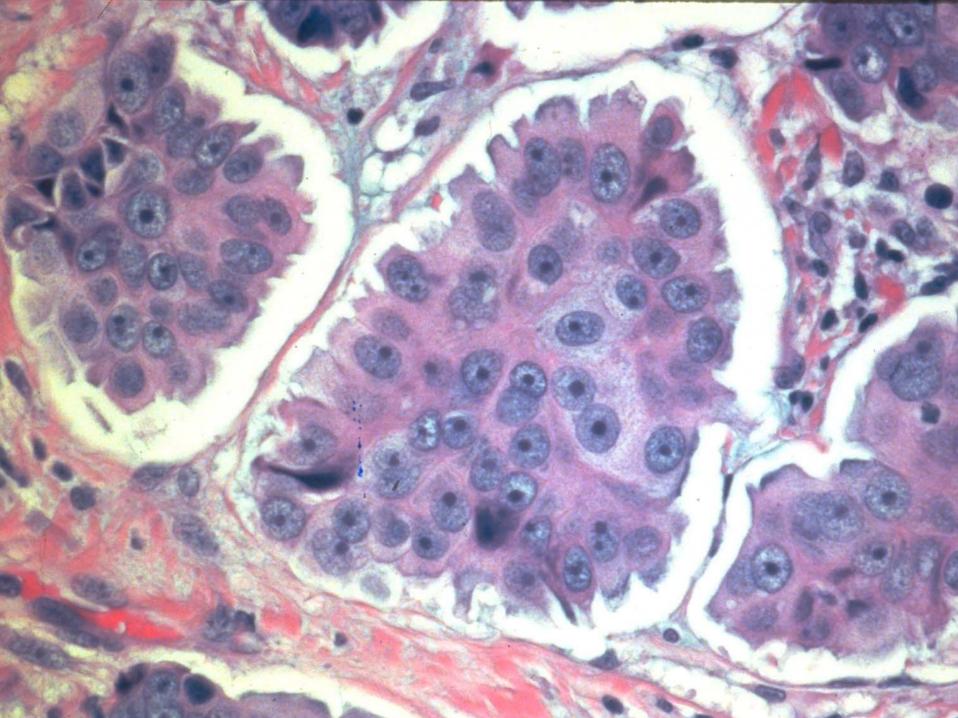
DDX:

- NSIP-diffuse pulmonary fibrosis without temporal heterogeneity or hyaline membrane formation
- UIP-patchy interstitial fibrosis with temporal heterogeneity with fibroblastic foci
- DAD-organizing hyaline membranes
- PAP-no significant interstitial changes

- 44F, palpable mass in UOQ, left breast
- Lumpectomy with 4.5 cm hard stellate mass







Invasive Micropapillary Carcinoma

- Frequently associated with lymph node mets (~100%)
 - May be in tumors <1.0 cm
- Distinctive histologic features:
 - Spaces surrounding micropapillary structures
 - Intermediate or high nuclear grade
 - Frequent tumor emboli
 - Numerous mitoses
 - No vascular papillary fronds
- IPOX
 - ER/PR+, Her2+

Invasive Micropapillary Carcinoma

- Uniform 8p-
 - Only 13-33% of invasive ductal CA, NOS
- Poor outcome
 - High combined histologic grade
 - ≥ 2 cm.
 - High mitotic rate
 - Lack of estrogen receptors