Rad Path 10/16/17: Mediastinum, Lung, and Chest wall

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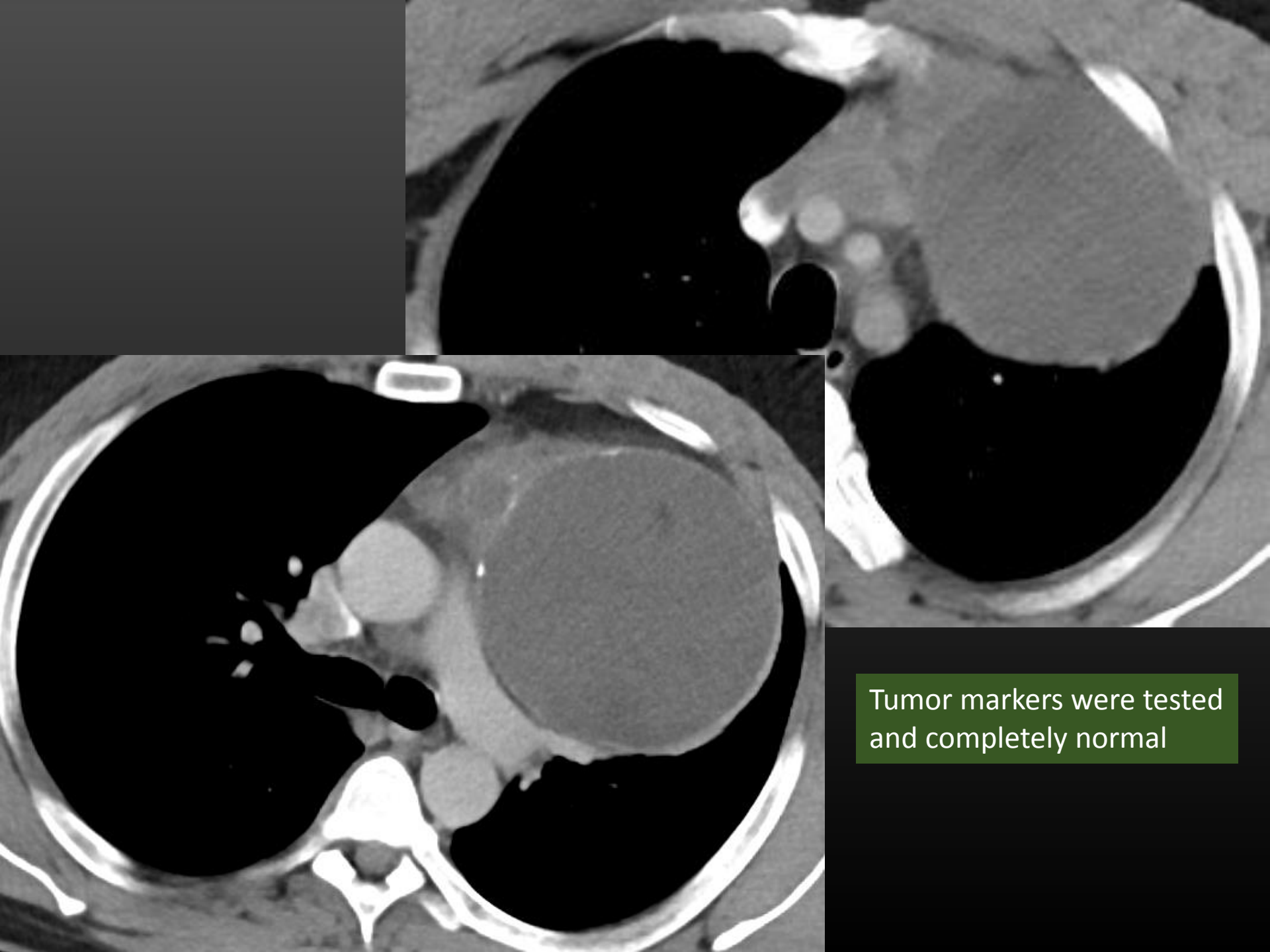
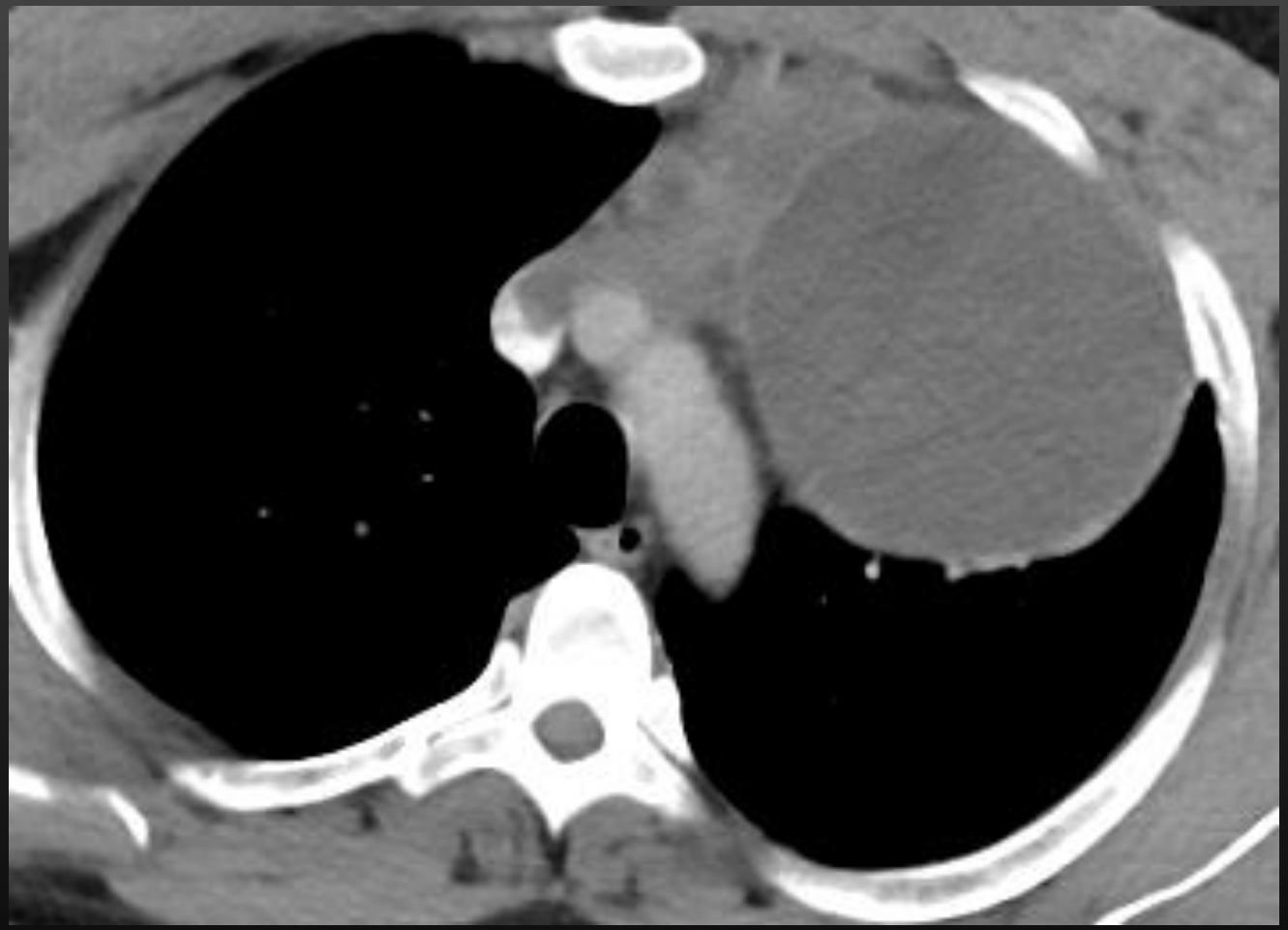
Brigham and Women's Hospital

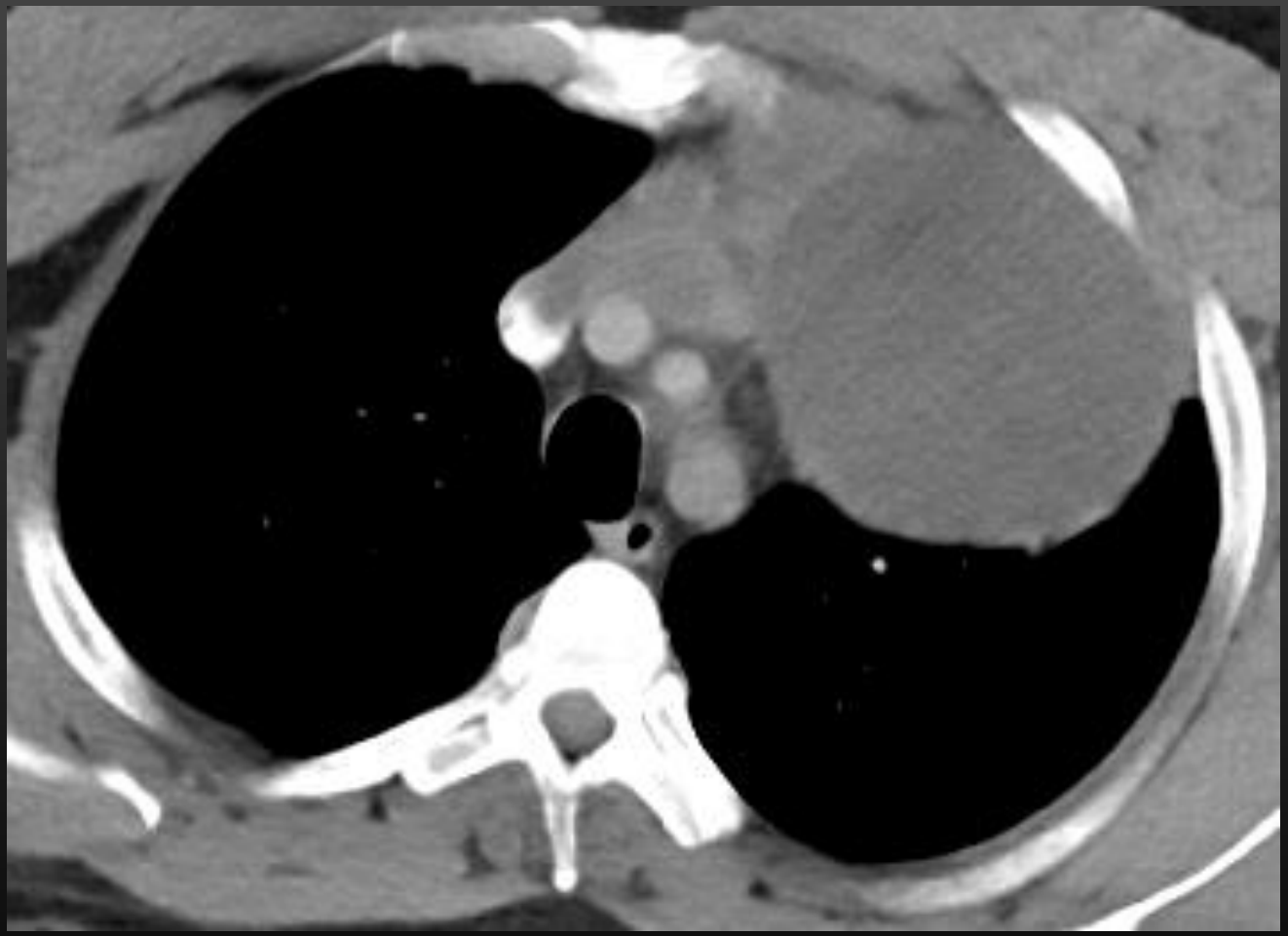
Case 1

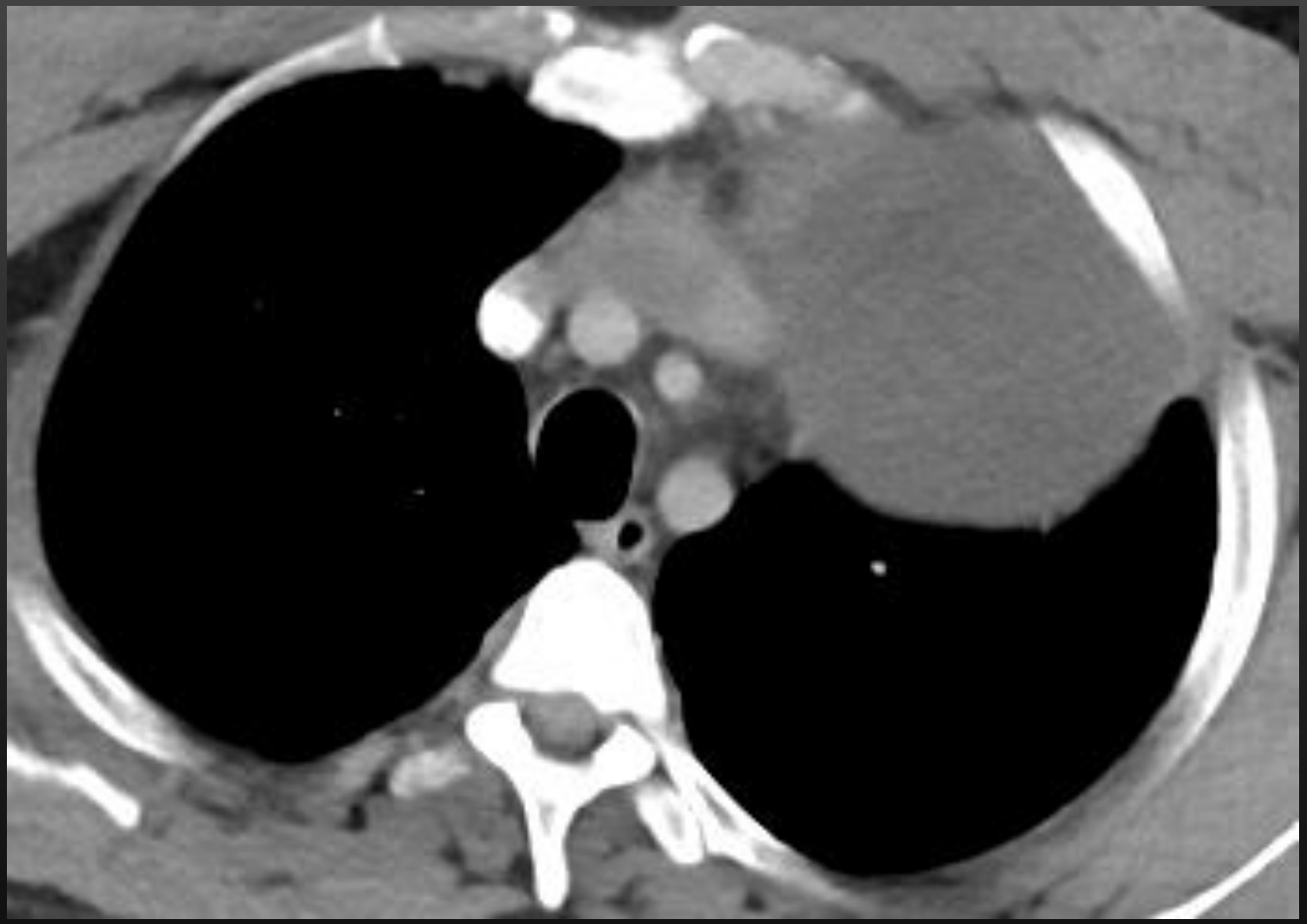
32M with acute onset neck stiffness and a “thickened neck blood vessel”

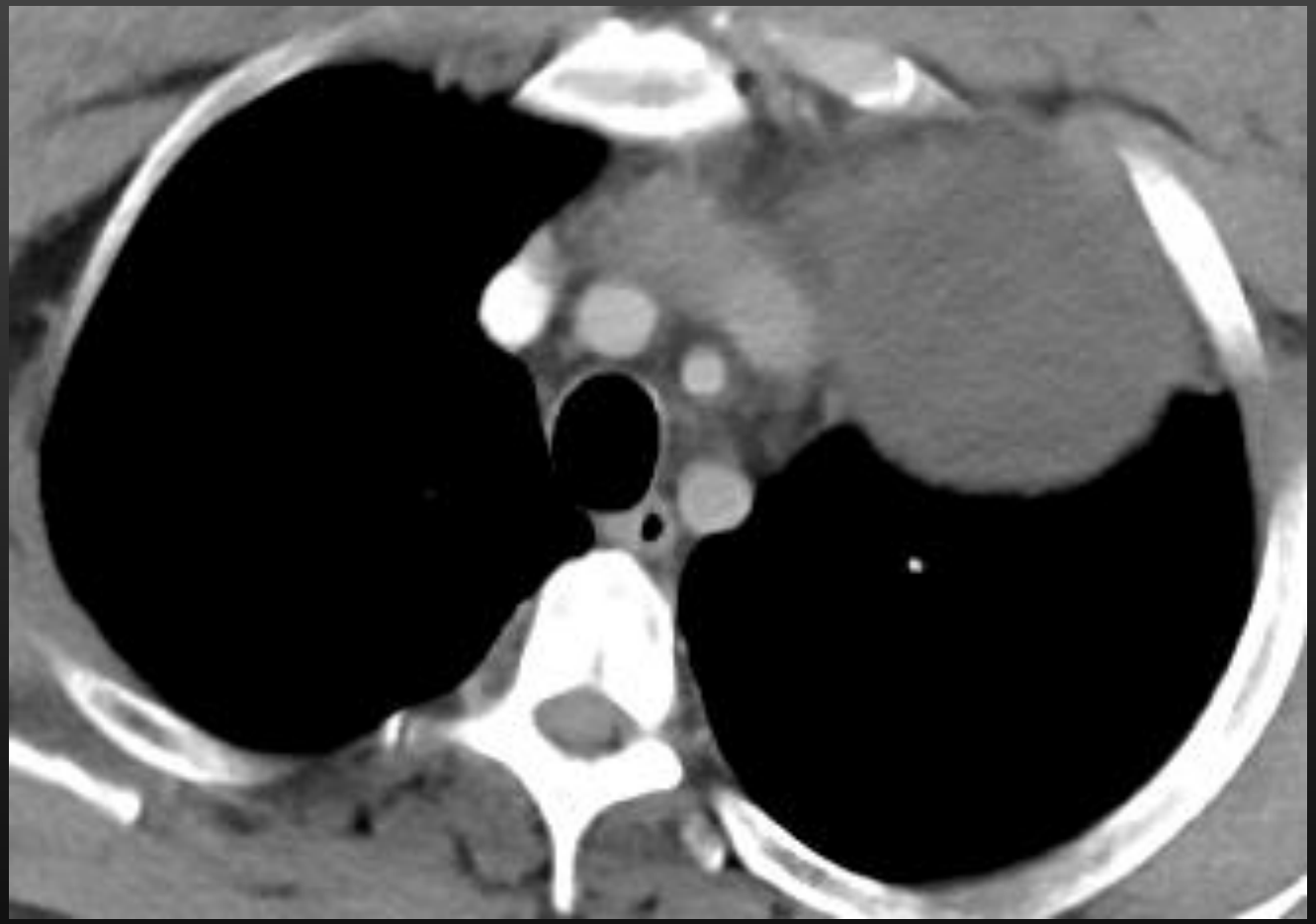


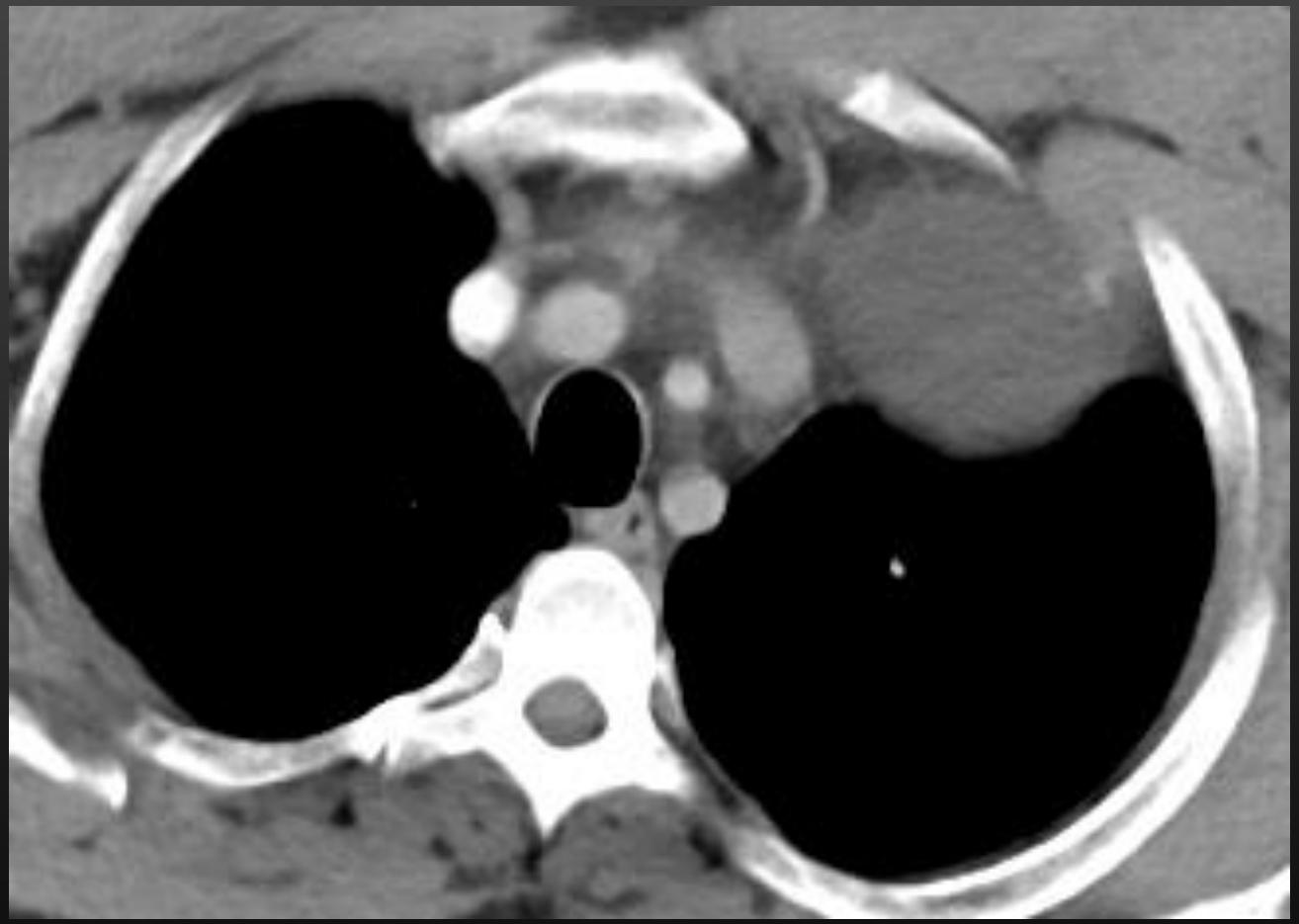
Tumor markers were tested and completely normal

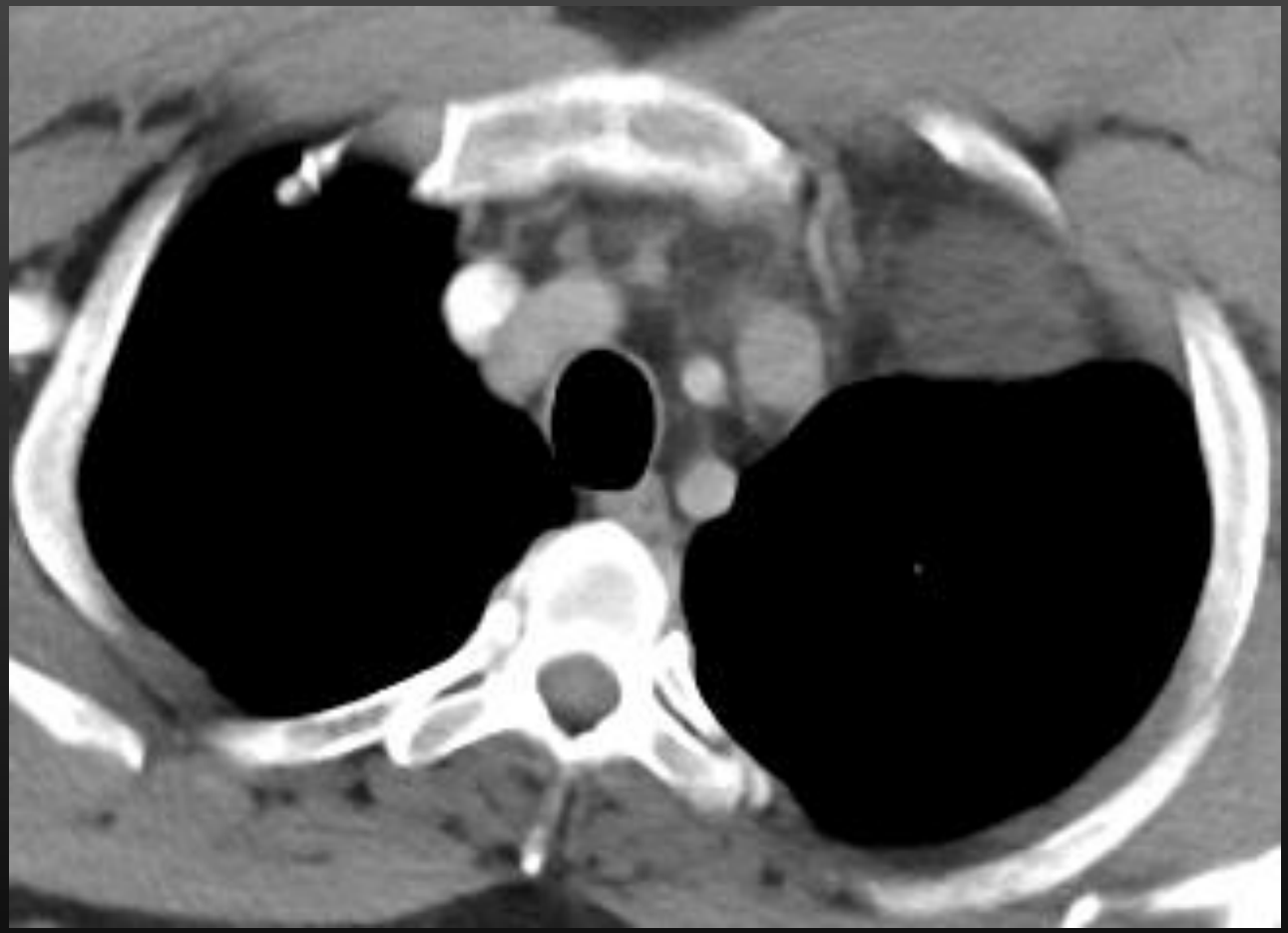












pathology
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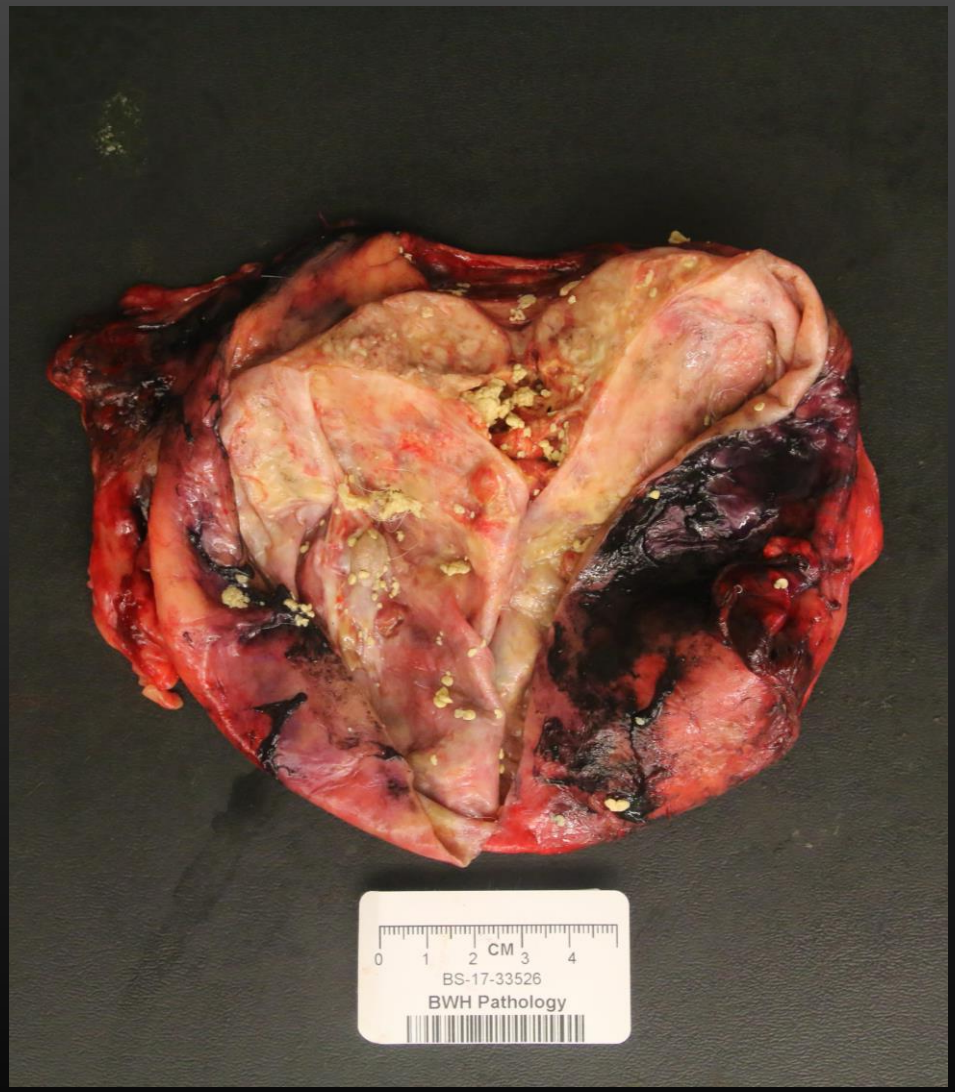
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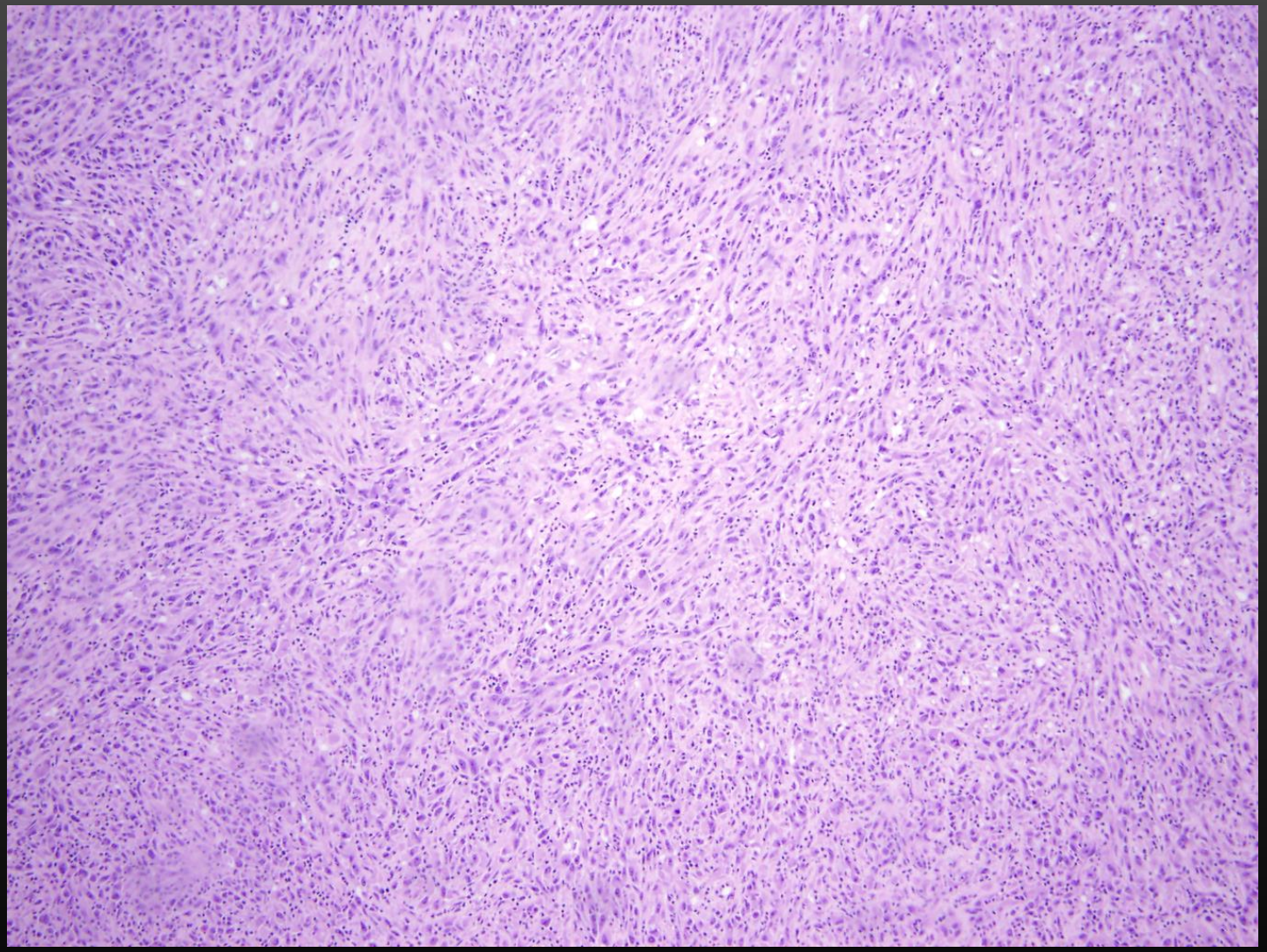
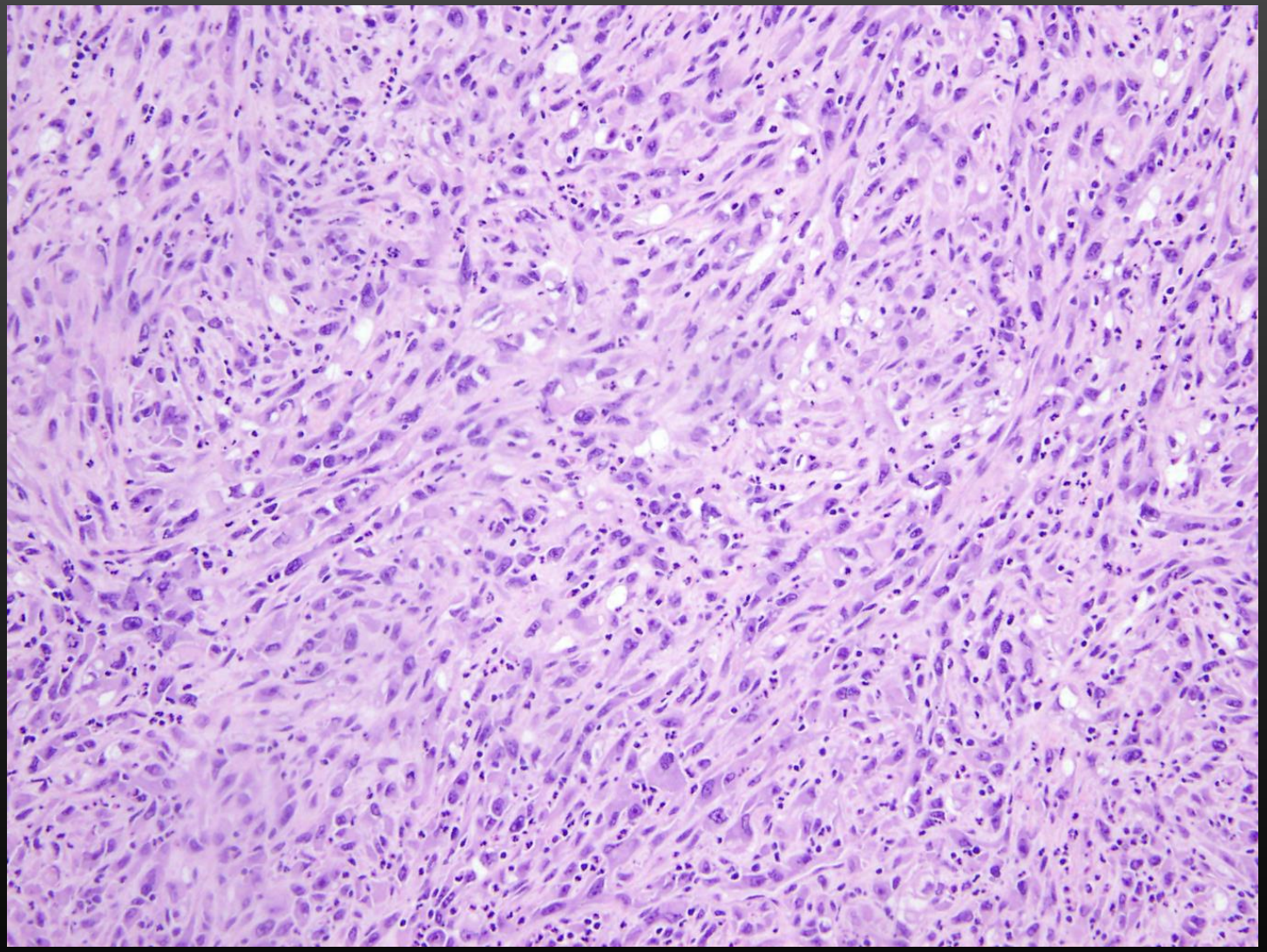




Benign teratoma

- Younger patients, age < 50, both F/M
- Usually asymptomatic
- Encapsulated and well defined on CT
- Heterogeneous: intralesional fat, fluid, calcification
- Fat-fluid level – highly specific but uncommon
- Can be mostly cystic
- May hemorrhage

- Vs. malignant teratomas → large, multinodular, poorly defined
- Both can grow in size

Mediastinal masses

- Slightly over ½ are in the anterior mediastinum
- ¼ middle
- ¼ posterior

Differential: anterior mediastinal masses

- Most common: Thymoma
  - Highest incidence in middle aged patients
- Other neoplasms
  - Thymic carcinoma, thymic carcinoid
  - Benign teratomas
  - Malignant germ cell tumors
    - Seminomas, nonseminomatous germ cell tumors (NSGCTs –include malignant teratomas)
  - Substernal extension of thyroid neoplasms
  - Ectopic parathyroid adenomas (small)
  - Lymphoma
- Non-neoplastic
  - Thymic cysts
  - Pericardial cysts, bronchogenic/foregut duplication cysts
  - Neurogenic tumor
  - Vascular abnormalities
  - Substernal extension of thyroid goiters
  - Infectious lesions
  - Morgagni hernia

Anterior mediastinal masses

- Image modalities
  - CECT is traditionally the first choice
    - Equal or superior to MRI in diagnosis of anterior mediastinal masses *except for thymic cysts* [2]
  - If cystic mass → MRI
    - Great for differentiating cystic from solid, thymic hyperplasia from thymic tumors
  - FDG-PET
    - Okay for staging cancer, but misleading since normal and hyperplastic thymus + inflammatory lesions are often FDG-avid

Approaching the Patient with an Anterior Mediastinal Mass: A Guide for Radiologists

Brett W. Carter, MD, Meinoshin Okumura, MD,† Frank C. Detterbeck, MD,‡ and Edith M. Marom, MD**

Journal of Thoracic Oncology® • Volume 9, Number 9, Supplement 2, September 2014

	% of Anterior Mediastinal Masses	Level of Confidence	Diagnosed With[a]	Confirm With/Next Steps
Highly Characteristic Lesions				
Hyperdense and enhancing lesion with connection to thyroid → Goiter	20-40% age >40	Certain	Imaging	-
Heterogeneous with fat, fluid, soft tissue, & calcification → Benign teratoma	25% age 10-19 10-15% age 20-49	Very high	Imaging	-
Well-circumscribed, round/oval/saccular, and homogeneous mass located near thymic bed on CT → Consider thymic cyst and evaluate with MRI				
If purely cystic → Thymic cyst and follow-up with MRI	<5%	Very high	Imaging	MRI
If cystic but with soft tissue components → Multilocular cyst or cystic thymoma	<5%	Uncertain	Imaging, Clinical	Resection
If purely cystic and located in cardiophrenic angle → Pericardial cyst	Low	Very high	Imaging	-

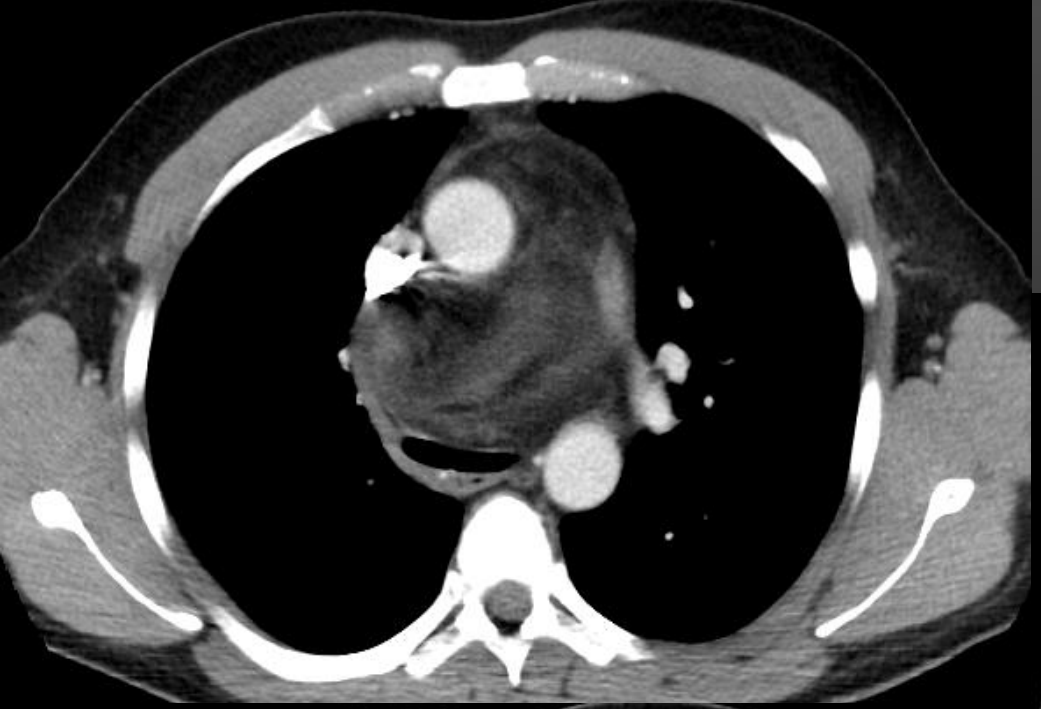
Calcifications are NOT helpful in distinguishing benign and malignant anterior mediastinal lesions!

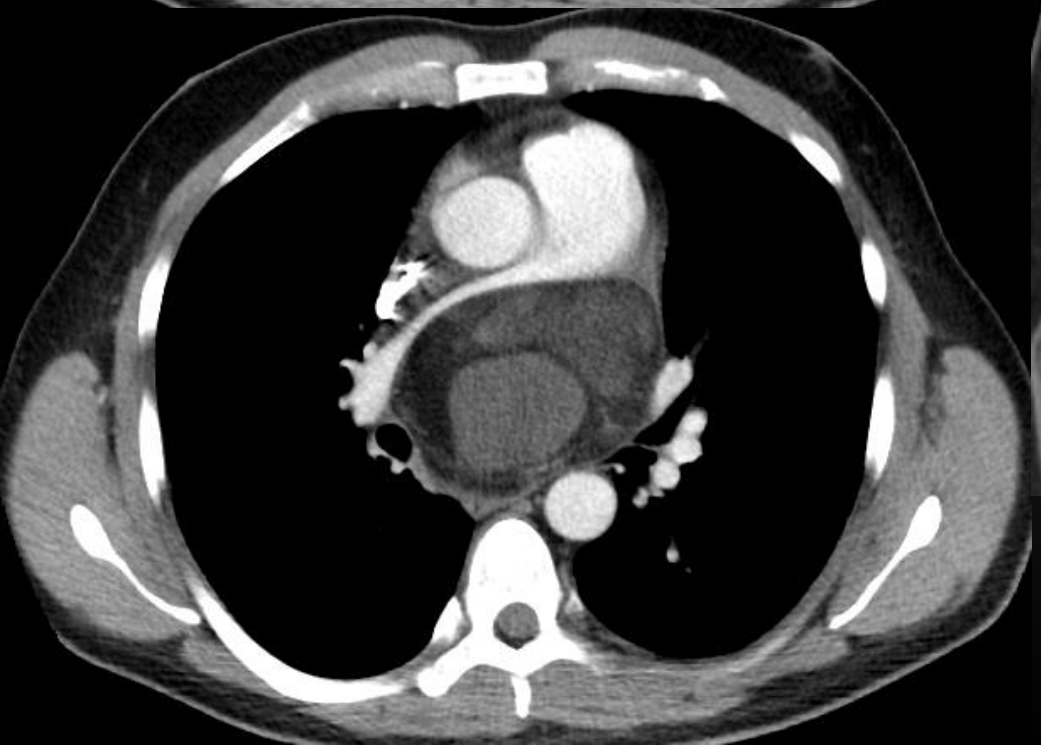
Suggestive on Imaging; Requires Clinical Context				
Lobular, homogeneous or slightly heterogeneous mass→ Thymoma *Context: Pt with Myasthenia gravis or other paraneoplastic syndrome*	15-25% age 20-39 ~50% age >40 5-10% age 20-39 ~20% age >40	Moderate Certain	Imaging +Clinical	± Biopsy ± Biopsy
Multiple markedly enlarged or matted lymph nodes / masses in anterior mediastinum ± neck, ± encasing but respecting vessels → HD, MLC-NHL *Context: "B" symptoms and ↑ LDH*	20-50% ♀ age <40 20-25% ♂ age <40 <10% ♂♀ age >40 Same	High Very high	Imaging + Clinical	Core biopsy Core biopsy
Large mass with pleural effusion, rapid onset, "B" symptoms → LB-NHL	15% age 10-19 5-10% age 20-39	High	Imaging, Clinical	Cytology
Large heterogeneous mass, especially with lung metastases → NSGCT *Context: Young ♂ with rapid onset of symptoms*	10-25% ♂ age <40 < 5% ♂ age >40 2-5% ♀ age <40	Moderate	Imaging, Clinical	Labs, ± biopsy
Homogeneous or slightly hetrogeneous mass ± lung metastases → Seminoma *Context:: young ♂, intermediate onset*	5-10% ♂ age <40 0-2% ♀ age <40	Moderate	Imaging, Clinical	Biopsy
Large heterogeneous mass, local invasion, lymphadenopathy, ± distant metastases → Thymic carcinoma or carcinoid	<2% age 20-39 ~5% age >40	Uncertain	Imaging	Biopsy
Low attenuation, symmetric enlargement of thymus *Context:: Pt treated with chemo, RT, or steroids → Thymic hyperplasia*	Uncommon	Very high	Clinical, Imaging	± MRI

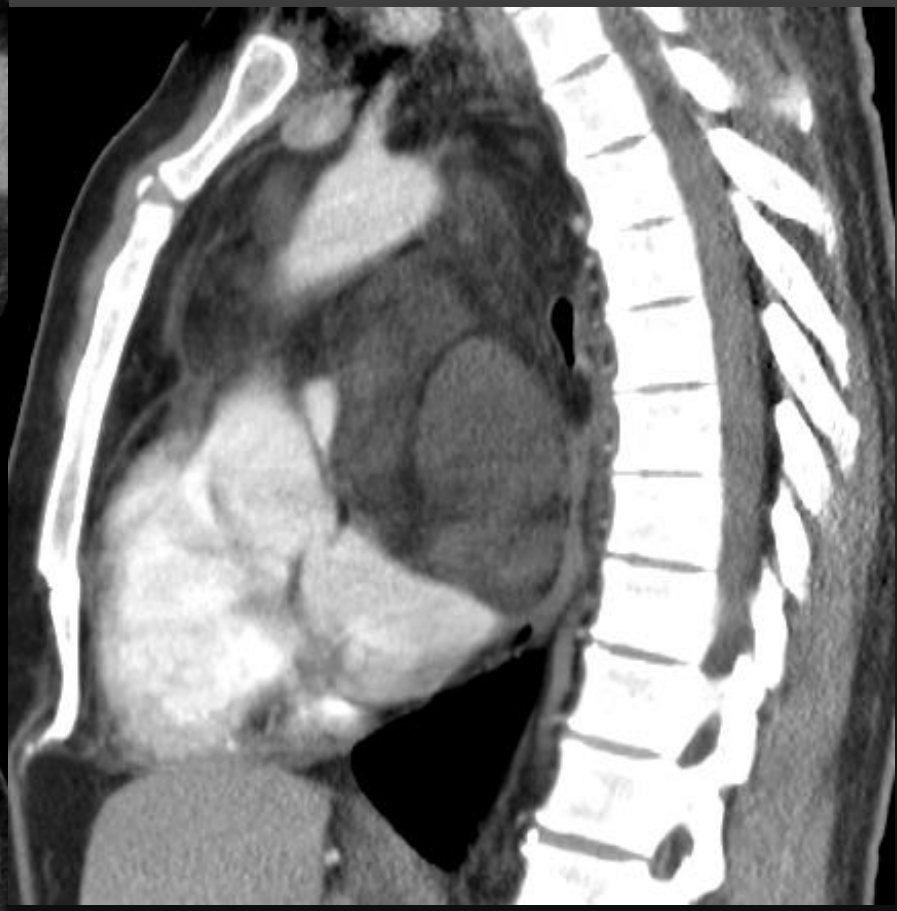
Rare Characteristic Features				
Lobular, homogeneous or slightly heterogeneous mass and with subpleural implants → Thymoma (invasive)	<5%	Very high	Imaging	Biopsy
Large fatty mass with small amount of soft tissue & vessels, connection with thymus → Thymolipoma	<5%	Very high	Imaging	-
Lobulated, encapsulated lesion consisting almost entirely of fat → Lipoma	2%	Very high	Imaging	-
Fatty lesion with aggressive features such as soft tissue components, invasion, lymphadenopathy, or metastatic disease → Liposarcoma	Very rare	Moderate	Imaging	Biopsy

Case 2

34M with numbness and tingling in his fingers







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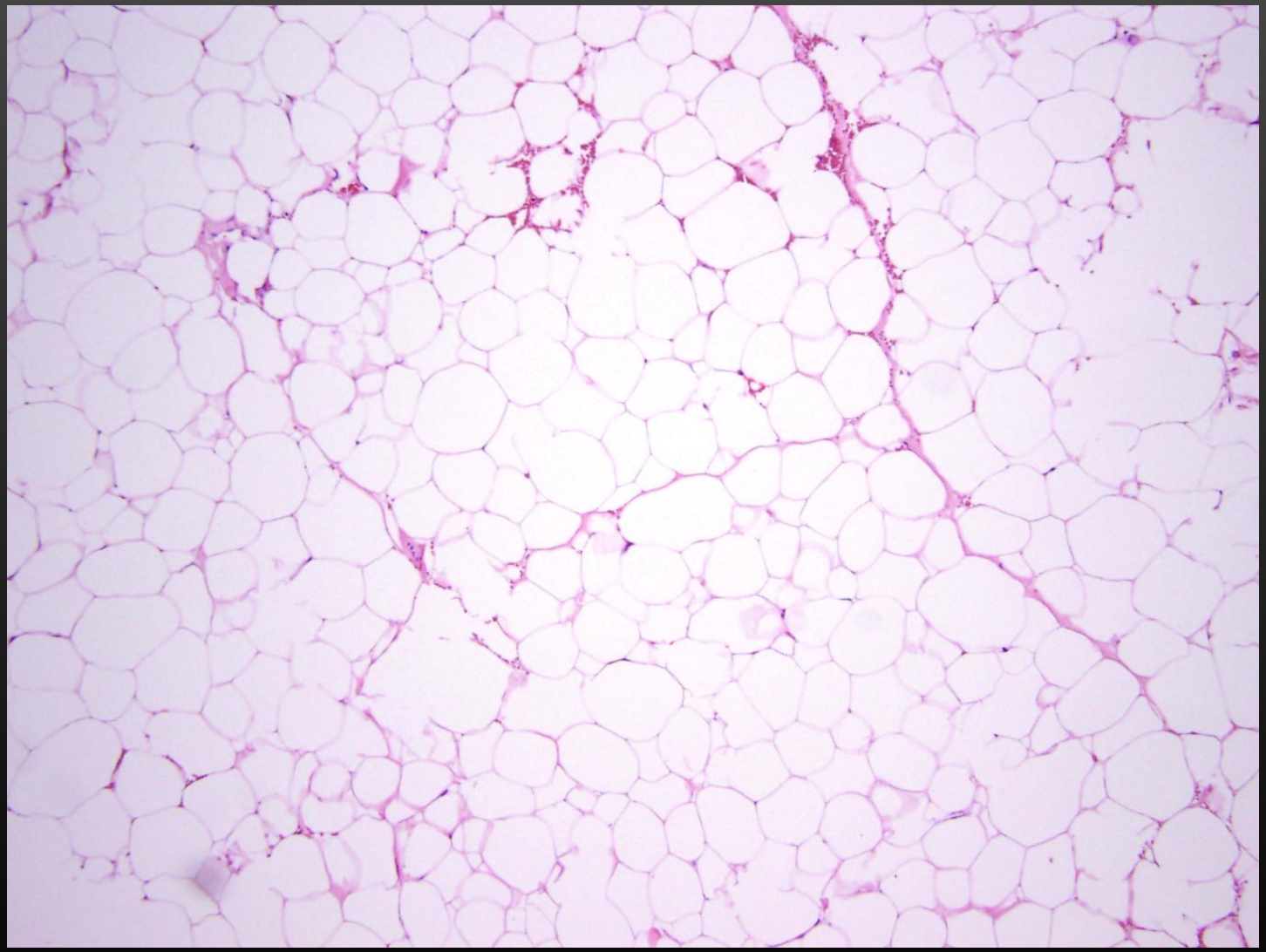
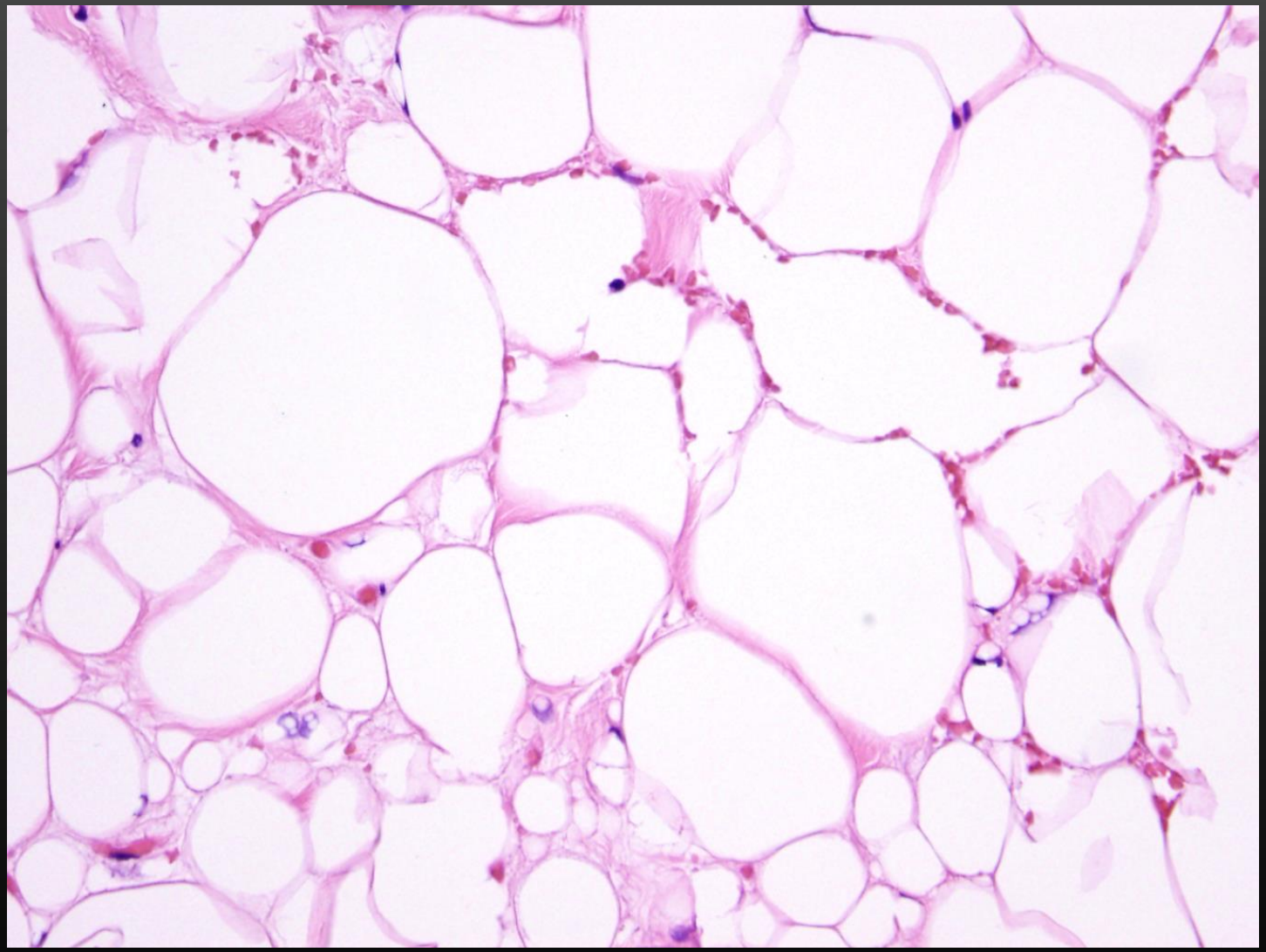




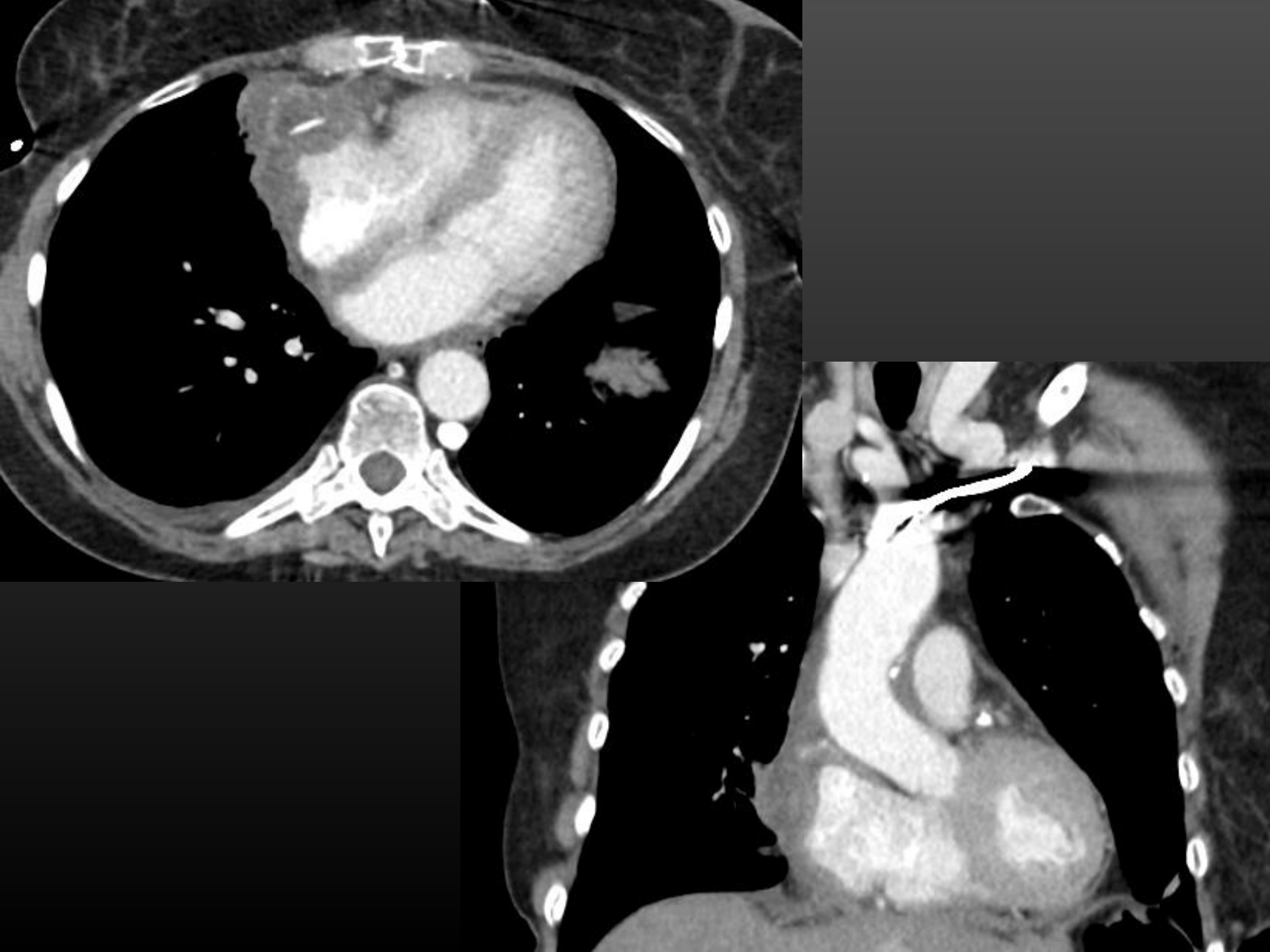
Mediastinal lipoma vs. liposarcoma

- Lipoma
  - ~2% of primary mediastinal neoplasms
  - Mostly fat + small amount of soft tissue and vessels

- Liposarcoma
  - Very rare
    - 0.5-1.9% of all liposarcomas are found in mediastinum
  - Aggressive features → increased soft tissue components, local invasion, adenopathy, mets
  - Treat by surgical resection
  - No established chemotherapy (poor response)
  - 40% recur after surgery

Case 3

64F heart transplant patient with right-sided chest pain, which is reproducible on exam



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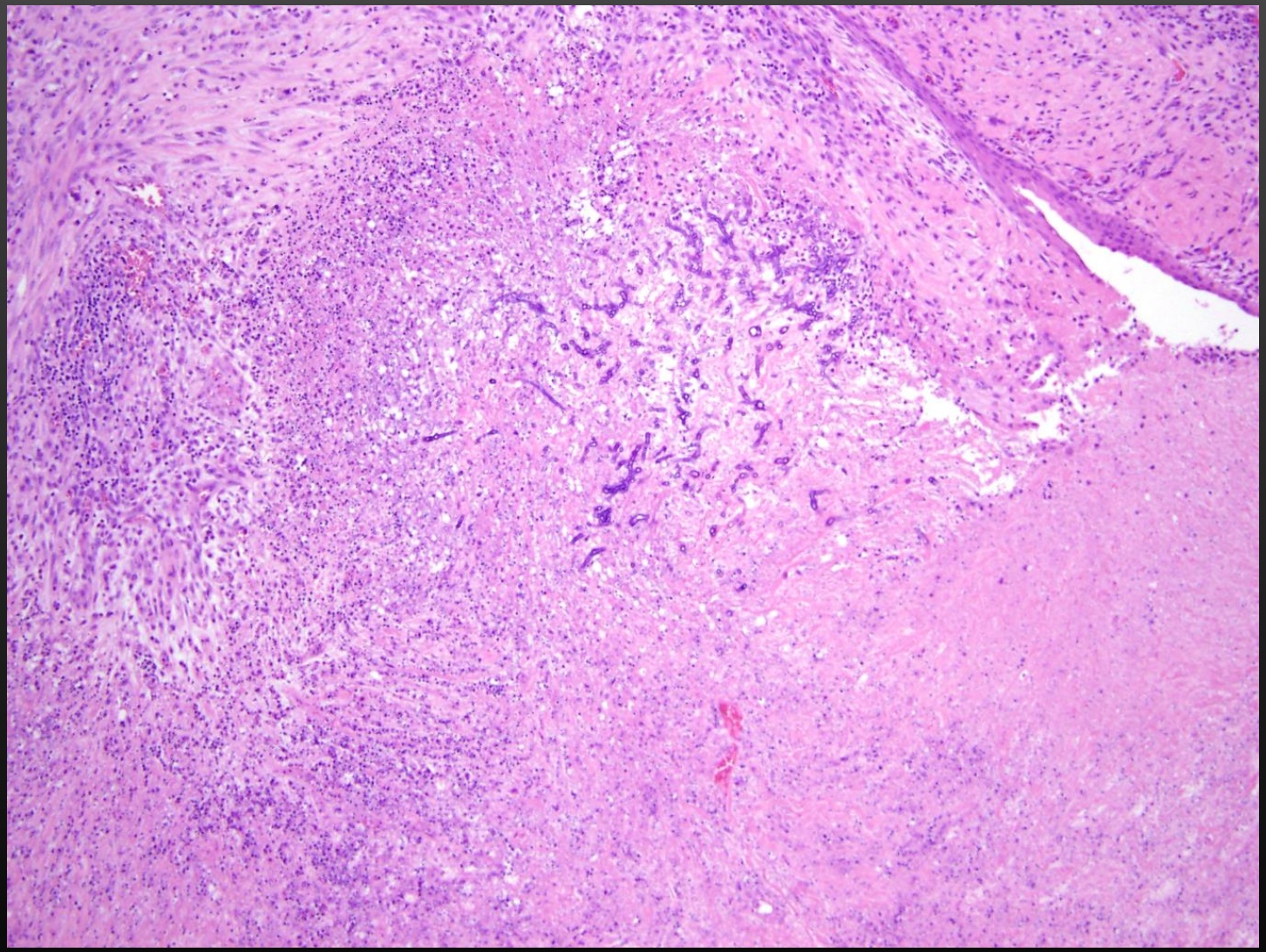
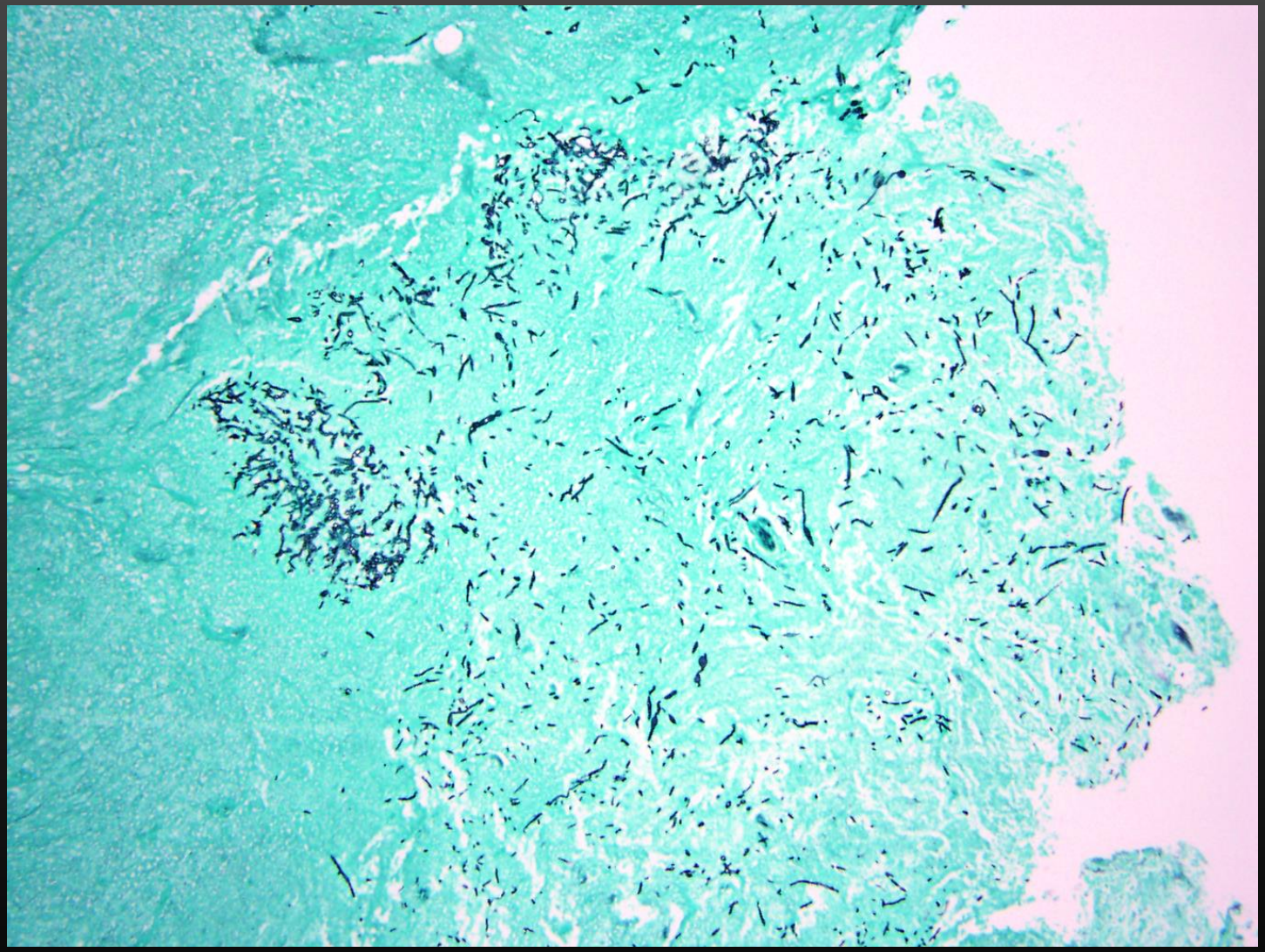




Surgical debridement with culture/histology consistent with Aspergillus fumigatus

Unfortunately, despite ~ 4 months of antifungal treatment, patient developed respiratory failure, enlarging pleural effusions and hypotension. Code status changed to comfort measure only.

Autopsy results:

PERICARDIAL FUNGAL ABSCESS with infiltration of ~90% of the thickness of right atrial wall, and extension into mediastinum and right-sided pleura

Post mortem histology with numerous fungal hyphal forms consistent with Aspergillus fumigatus

Mediastinal invasive aspergillosis

- Predisposing factors for invasive aspergillosis
  - Prolonged neutropenia, chronic granulomatous disease, high dose steroids, post-transplantation, leukemia, cytotoxic therapy recipients, AIDS
- CT: necrotic mass infiltrating into adjacent tissues
  - No clear differentiation from mediastinal tumors
- Diagnosis requires high clinical suspicion

Quiz

Mediastinal invasive aspergillosis only occurs in immunocompromised patients.

A. True
B. False

Quiz

Mediastinal invasive aspergillosis only occurs in immunocompromised patients.

A. True
B. False

Occurs even in immunocompetent patients!

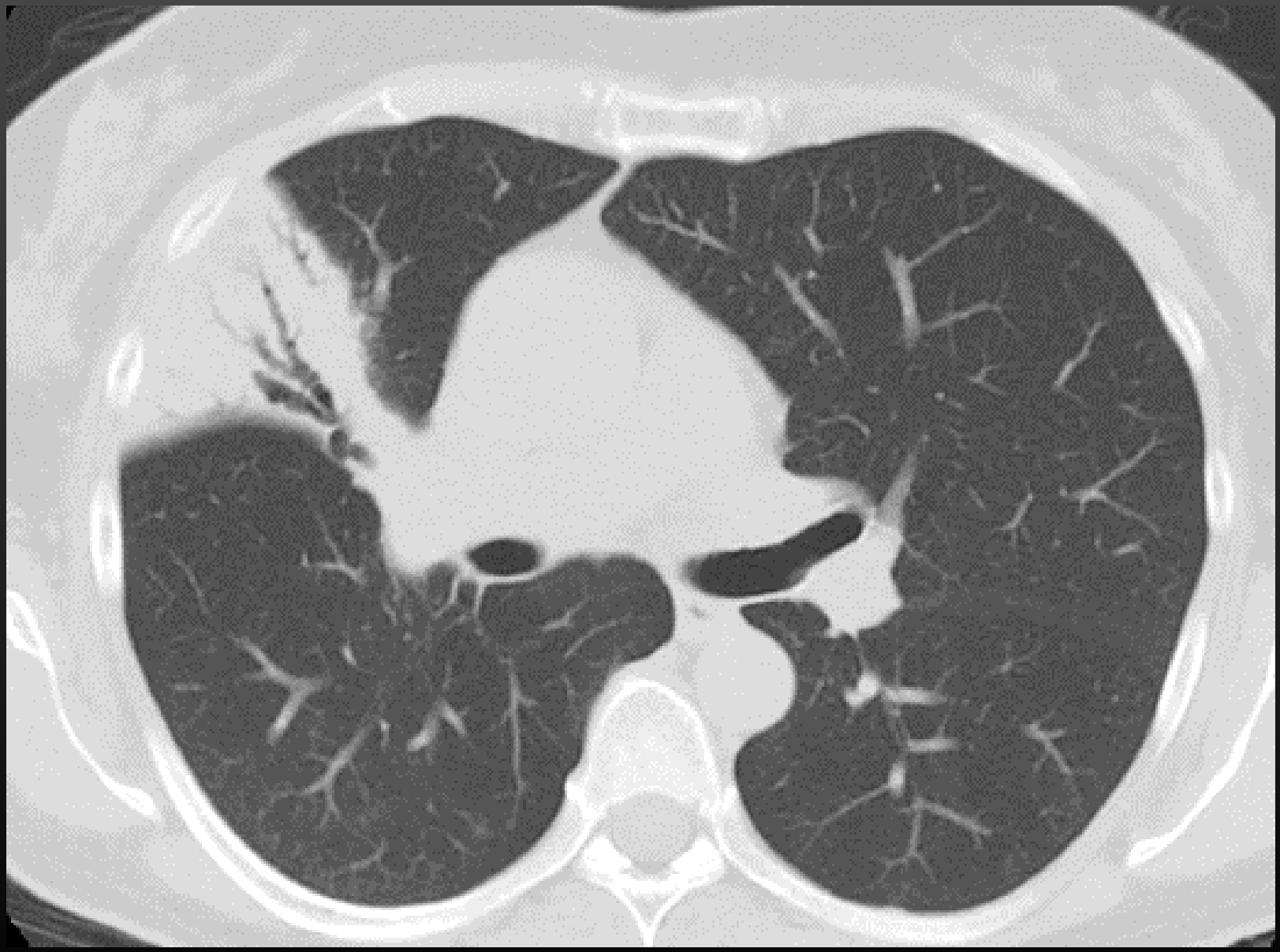
Mediastinal invasive aspergillosis

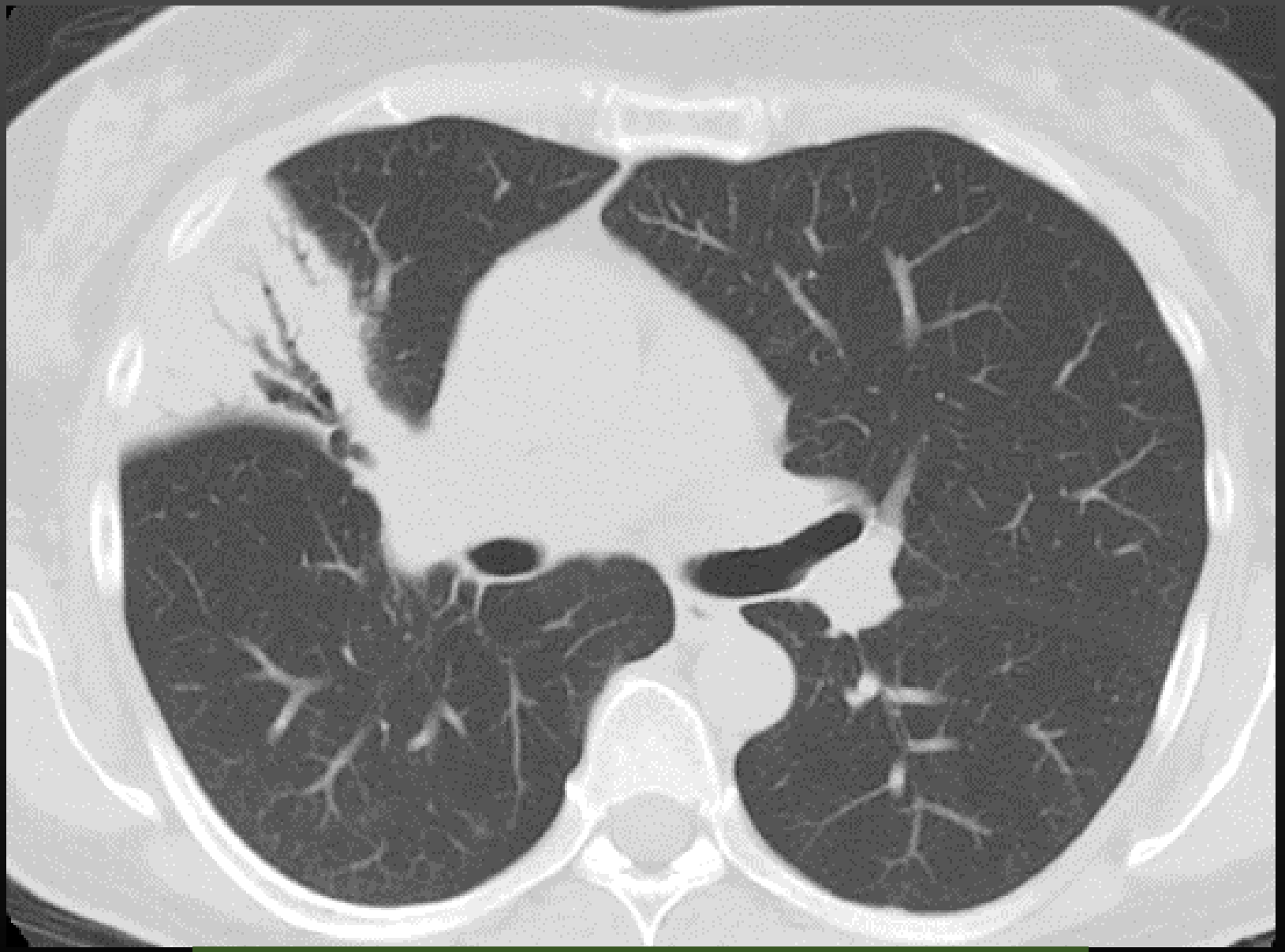
- Sensitivity of galactomannan assay
  - High in immunocompromised patients or with fast disease progression
  - Very low in immunocompetent patients or in non-invasive Aspergillus infection
- High mortality, 30-50% despite adequate antifungals
  - Very poor prognosis with cardiac involvement, usually due to delayed diagnosis
- Surgical resection is the only curative treatment

Case 4

67F with incidentally found lung abnormality

- No improvement after azithromycin





Bronchoscopy with FNA showed numerous lymphocytes

pathology
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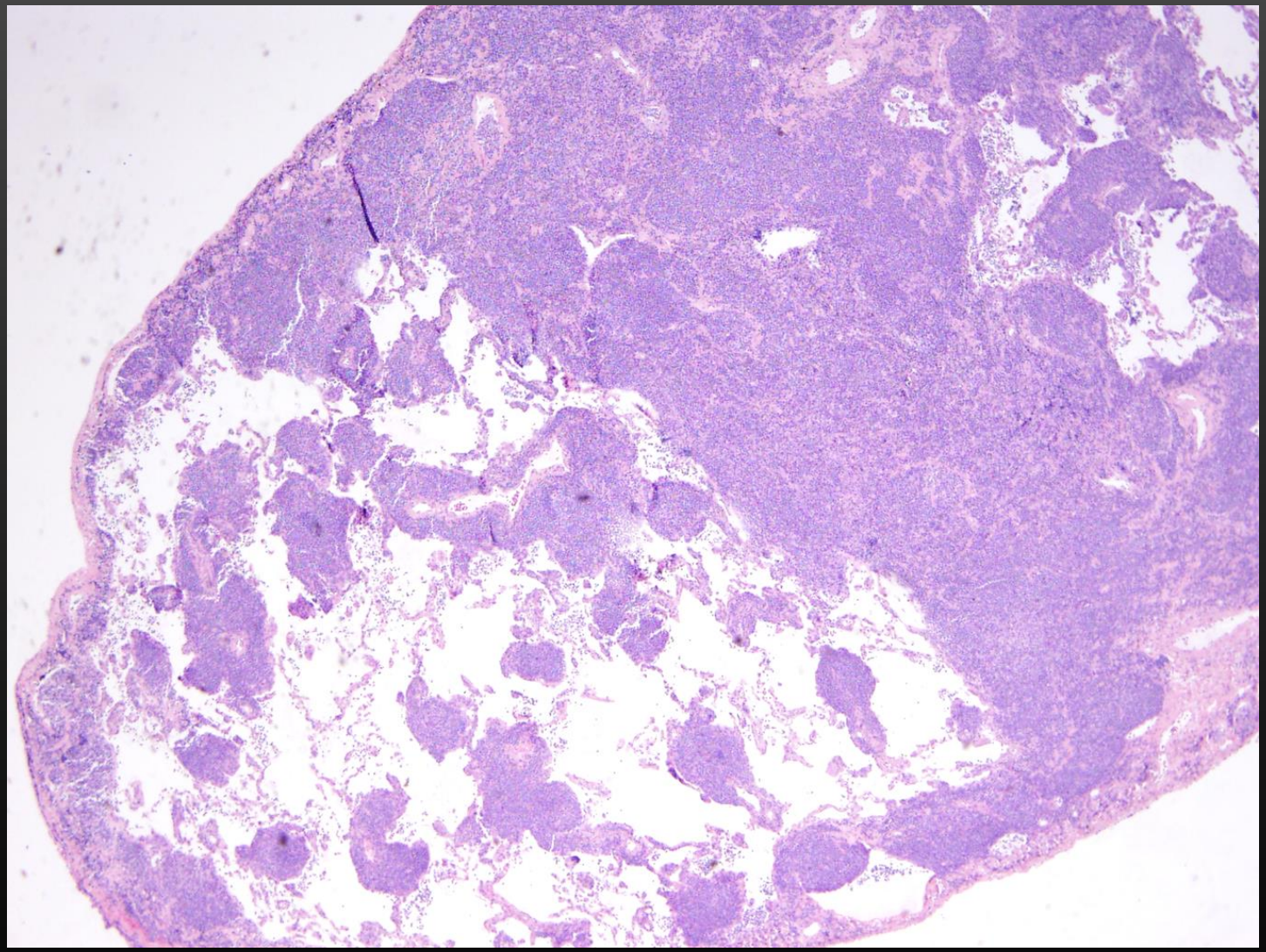
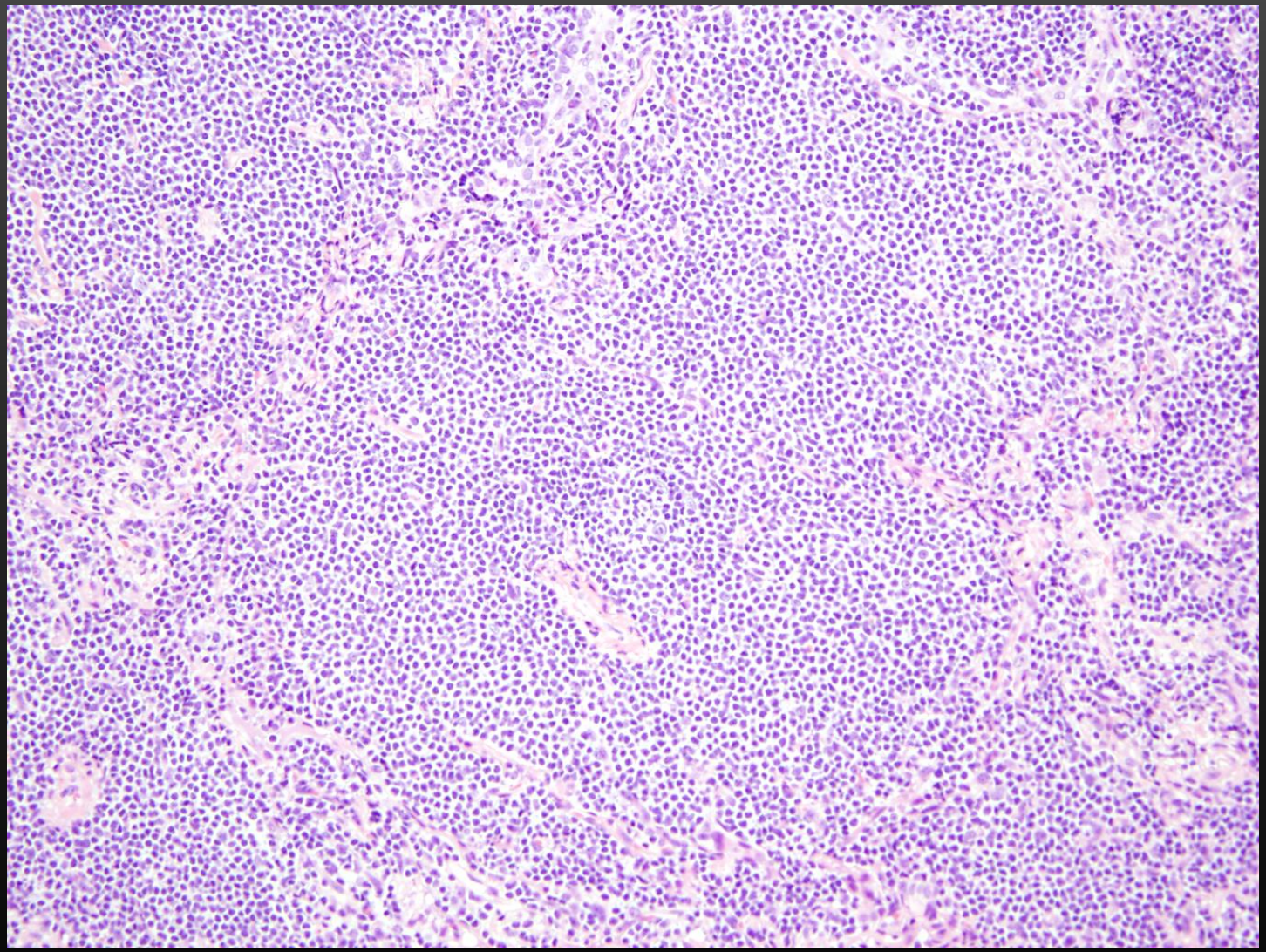




Primary pulmonary lymphoma

- Types
  - MALT lymphoma (low-grade marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue)
  - Other NHLs (e.g. DLBCL)
  - Hodgkin lymphoma

- Most common (90%) = MALT lymphoma
  - Though only 3-4% of extranodal lymphomas
  - <1% of all lung malignancies

MALT lymphoma

- Often asymptomatic, indolent
- Patients usually in 6th decade
- Can coexist with pulmonary amyloidosis, LIP
- Association with
  - Autoimmune diseases (Sjogrens, RA, CVID)
  - Chronic inflammation from smoking
  - Infections such as HIV, hepC

Histopathologic findings	Sheets of monoclonal lymphocytes
	Plasma cells and germinal centers present; Dutcher bodies present
	Lymphangitic spread at the periphery of lesions
	Can invade adjacent airways, vessels, pleura
	May be associated with trisomy 3, bcl-10 mutations, and t(11;18)(q21;q21)

Radiographics. 2016 Jan-Feb;36(1):53-70.

MALT lymphoma

- Imaging
  - Nodules, masses, nodular/masslike area of GGO and/or consolidation
  - Multiple/bilateral > 70%
  - Bronchovascular distribution
  - Air bronchograms
  - Hilar and mediastinal adenopathy seen in 30%
  - Rare
    - Present as patchy large areas of GGO throughout
    - Or infiltrate along airways → mosaic attenuation
  - ½ are FDG-avid
- Pleural effusions, BM involvement are uncommon
- Monoclonal gammopathy seen in 40% patients

MALT lymphoma

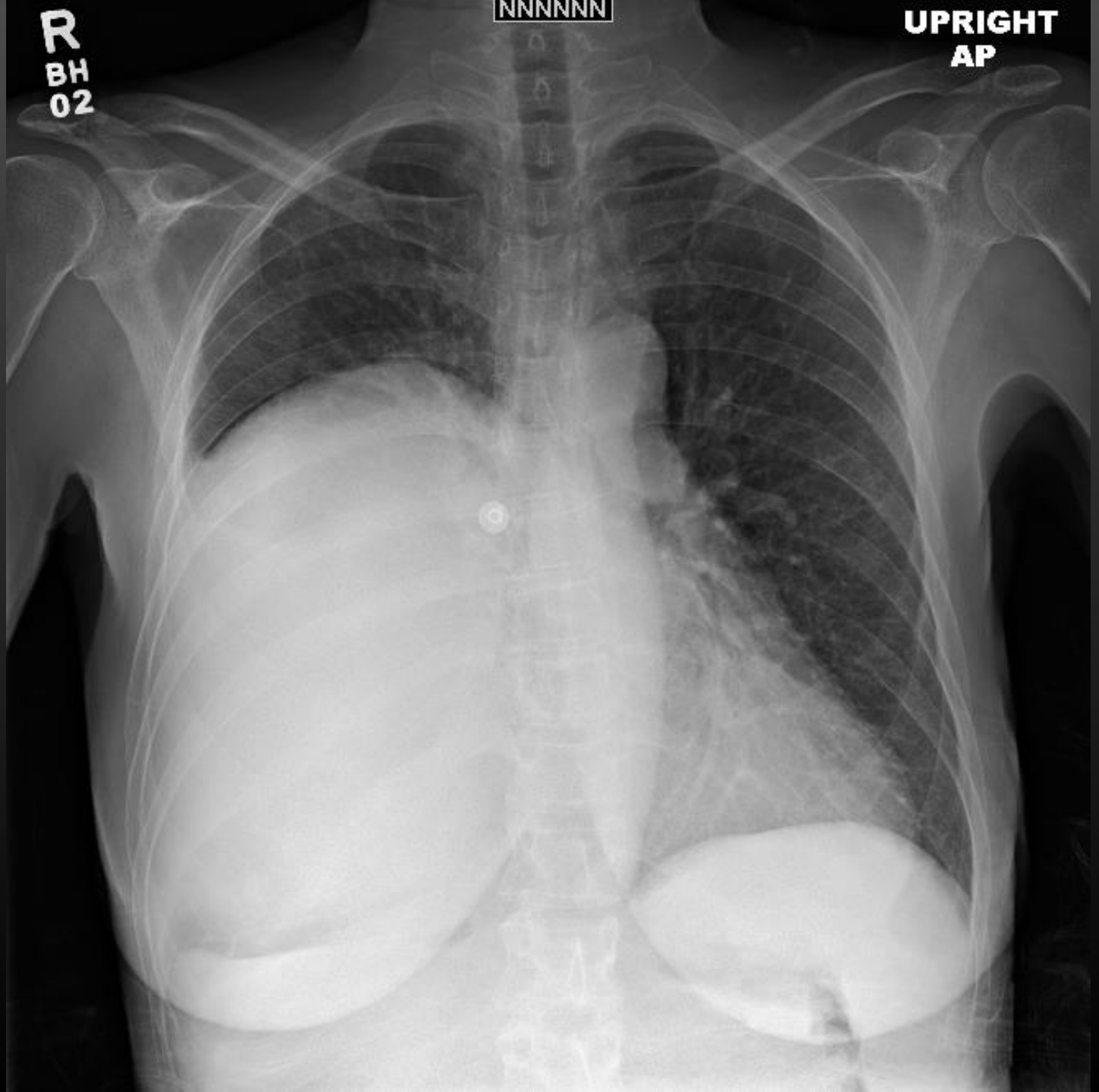
- DDx for MALT lymphoma
  = other entities that manifest in a peribronchovascular distribution
  - Wegener granulomatosis
  - Sarcoidosis
  - Perilymphatic spread of metastatic disease
  - Infection

- Clinical history is helpful
- Diagnosis needs tissue sample with immunohistochemistry
- Treatment
  - Chemo or immunotherapy (rituximab)
  - Surgery and radiation if localized disease
- Good prognosis
- But recur in up to 50% in lungs or other areas containing MALT (e.g. GI tract)

Case 5

62F with recurrent severe hypoglycemia

- Past smoker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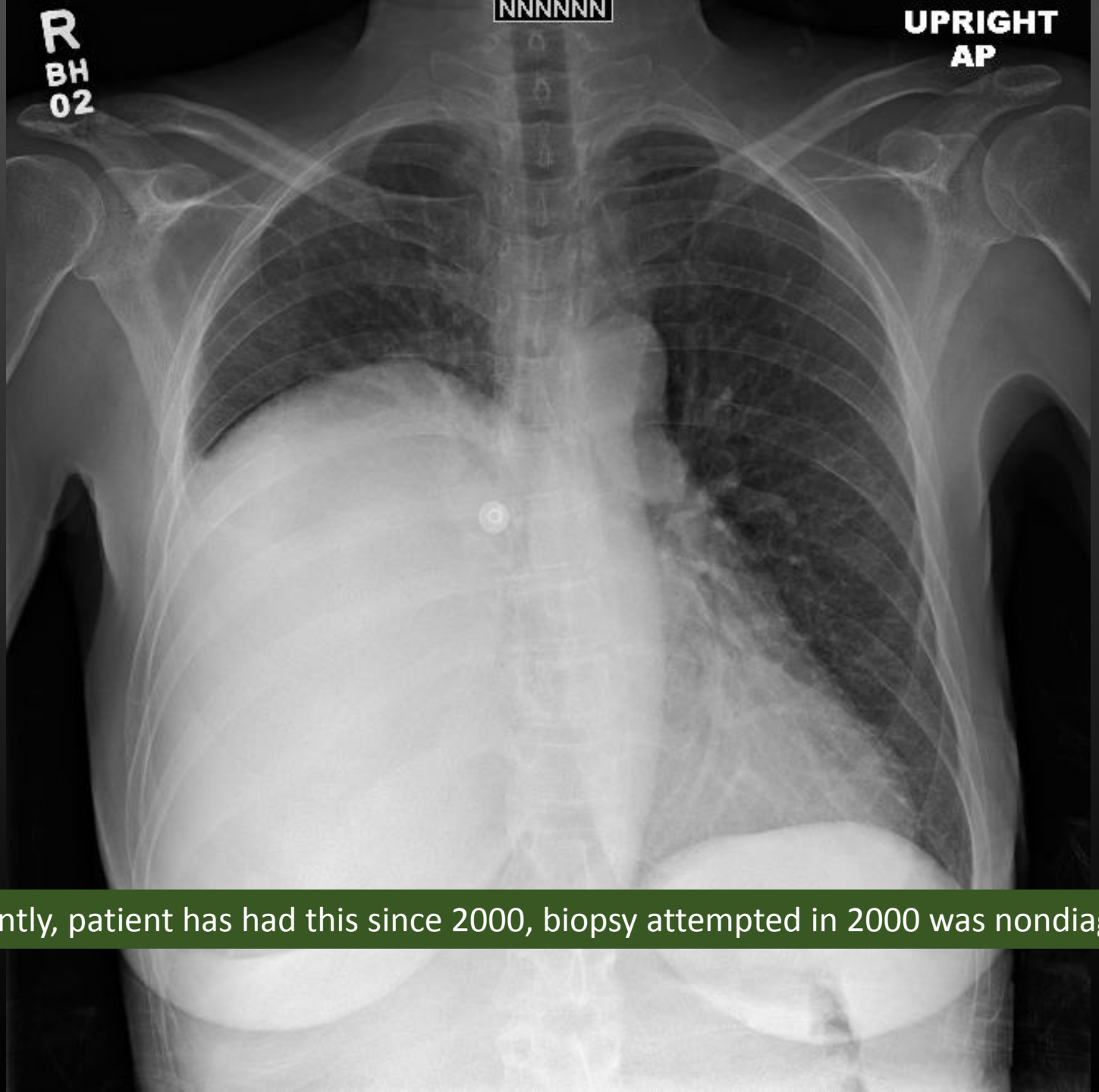
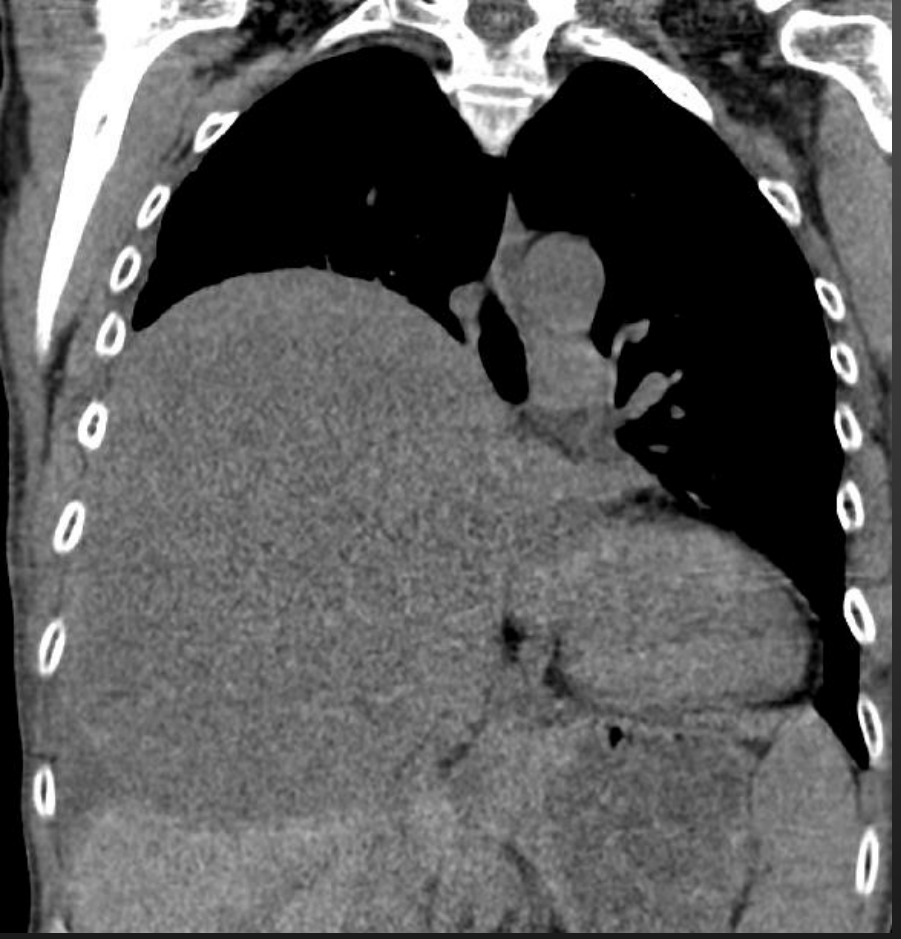
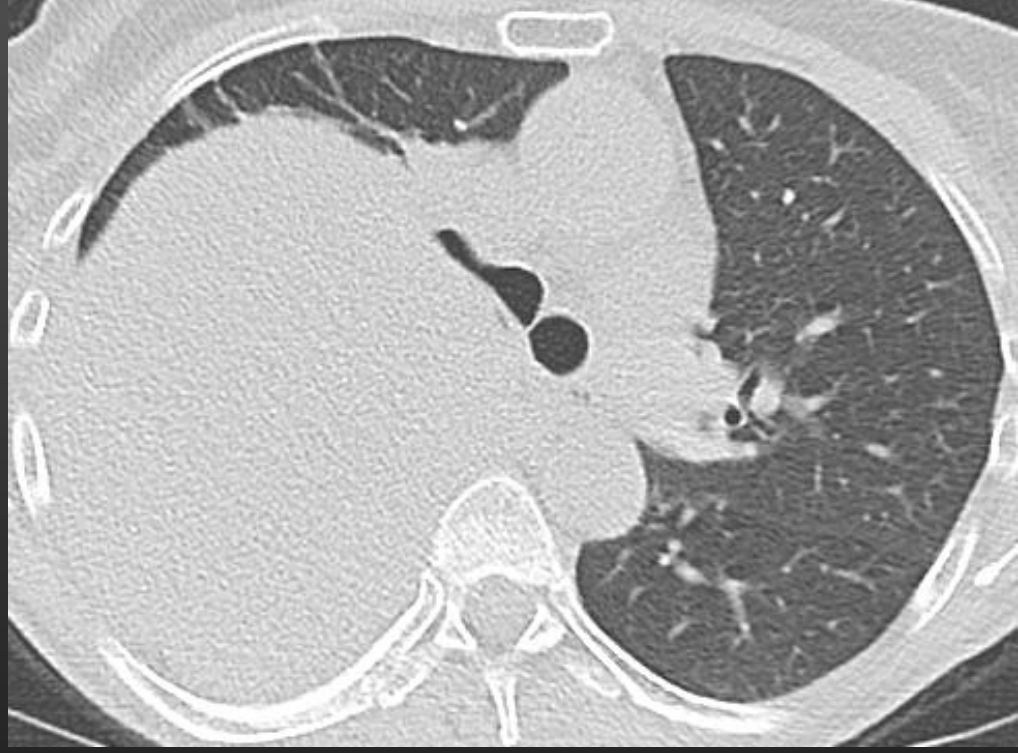
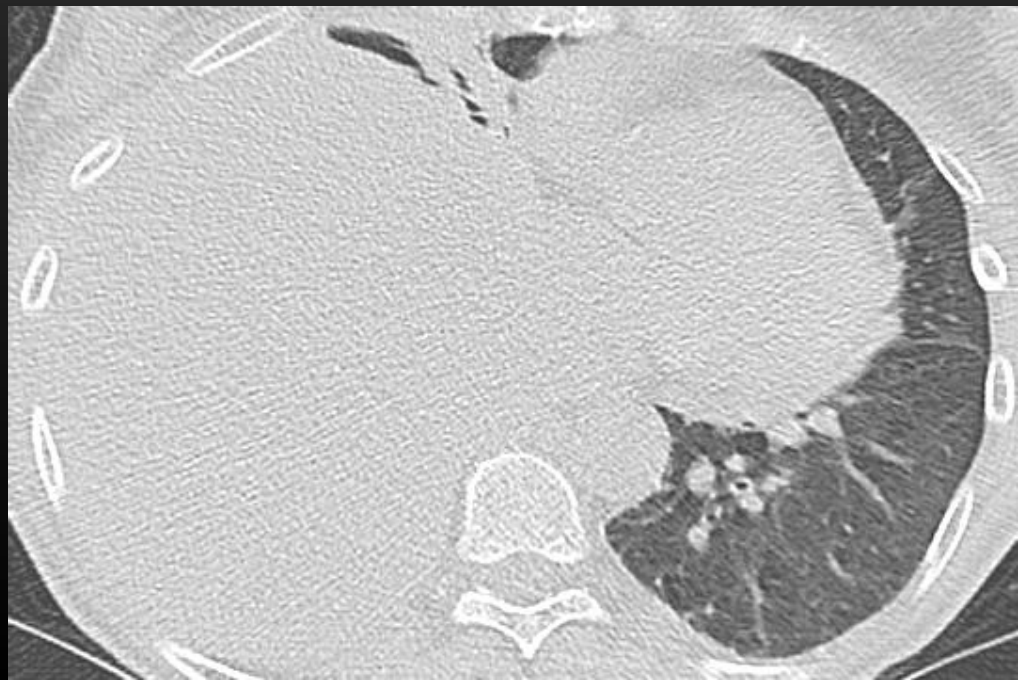


Apparently, patient has had this since 2000, biopsy attempted in 2000 was nondiagnostic







pathology
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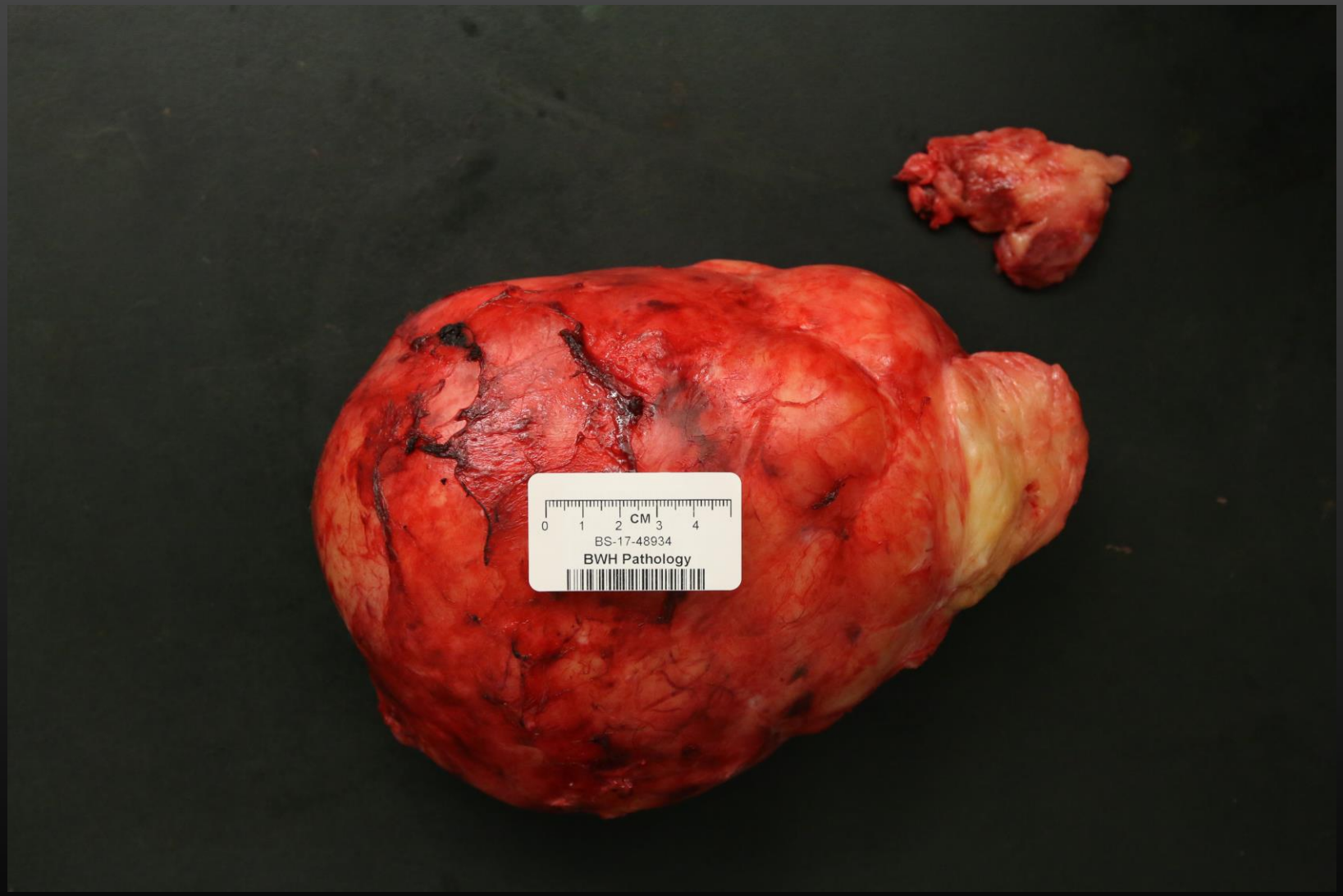
Gave to Lisa R. for AIRP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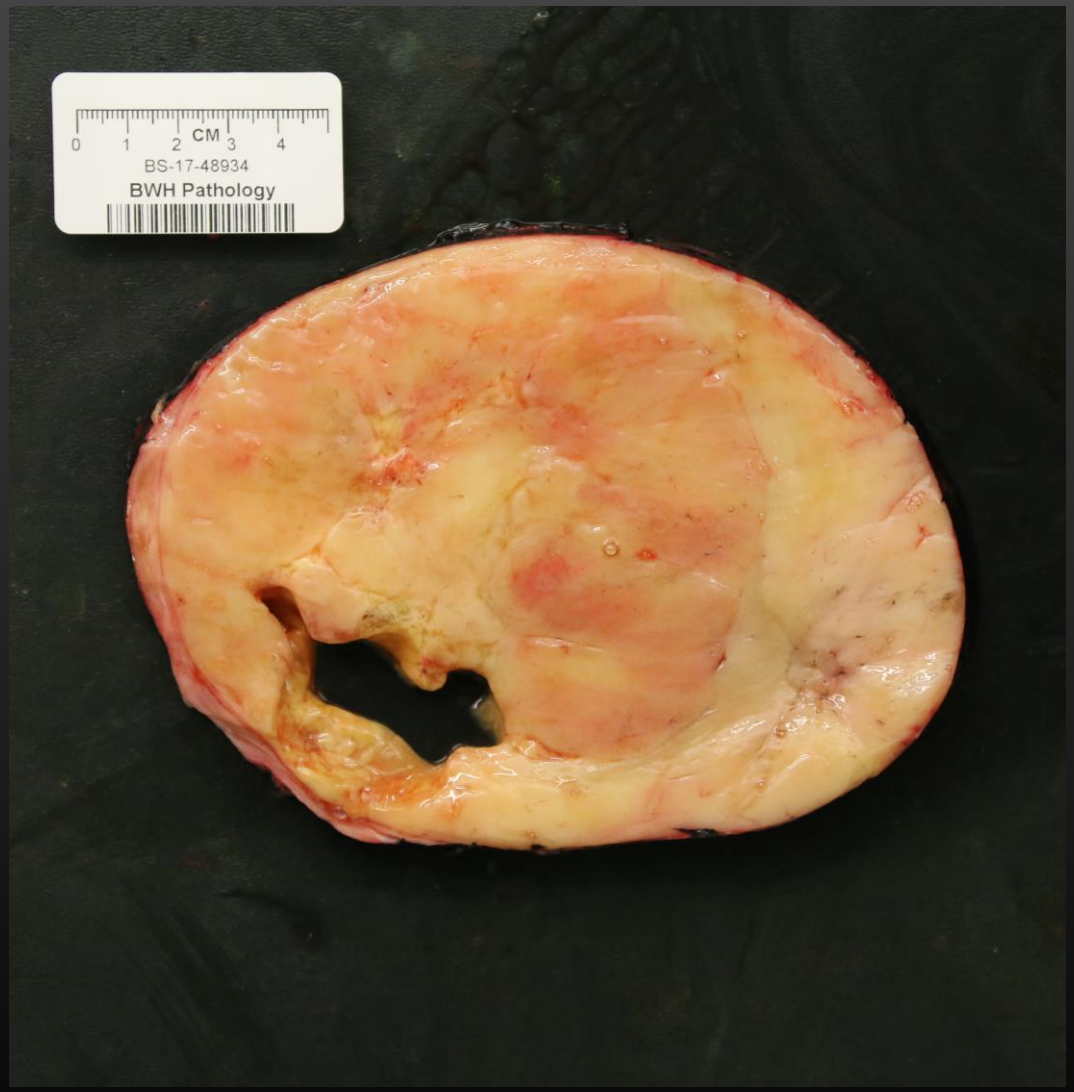
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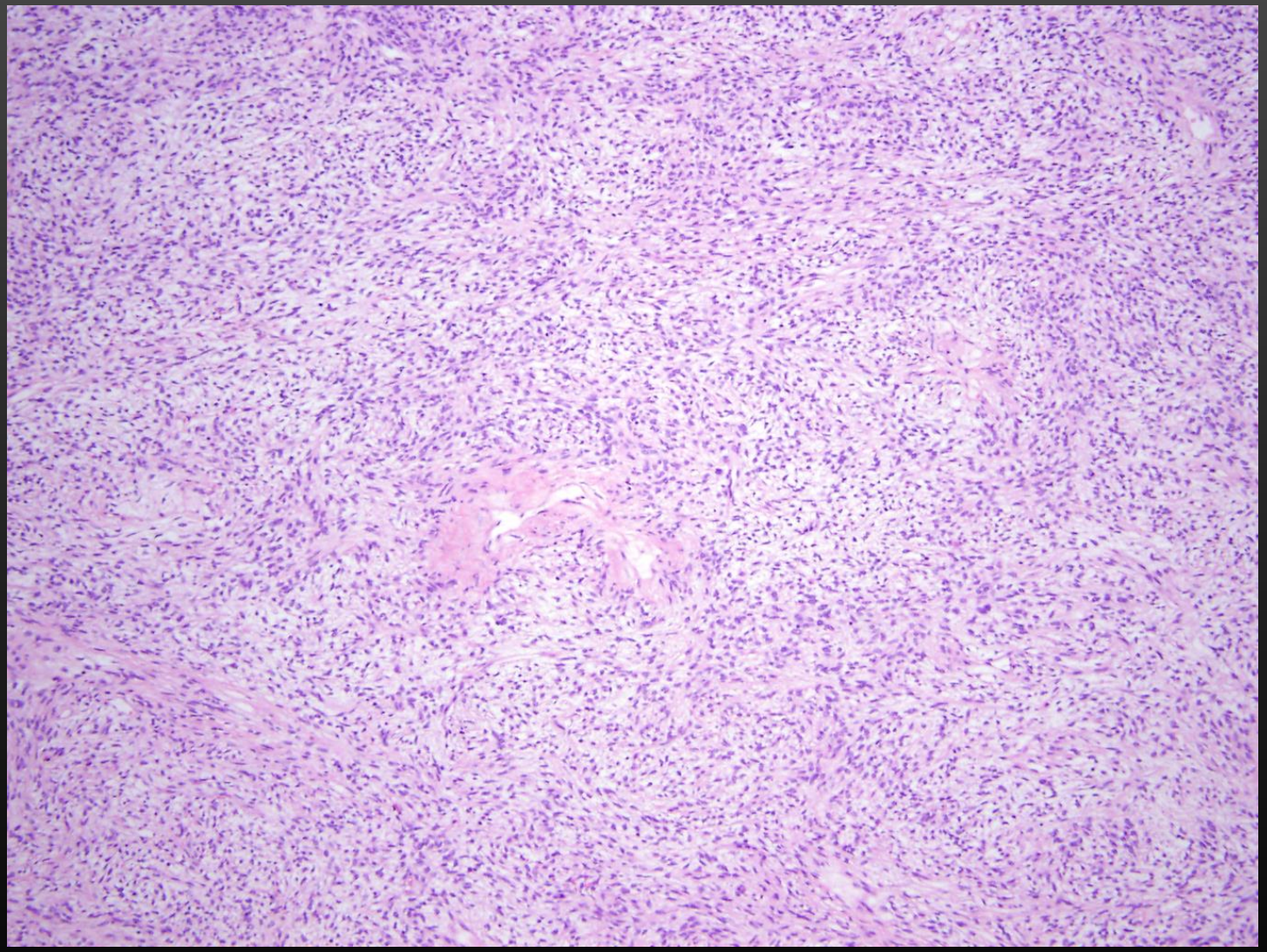

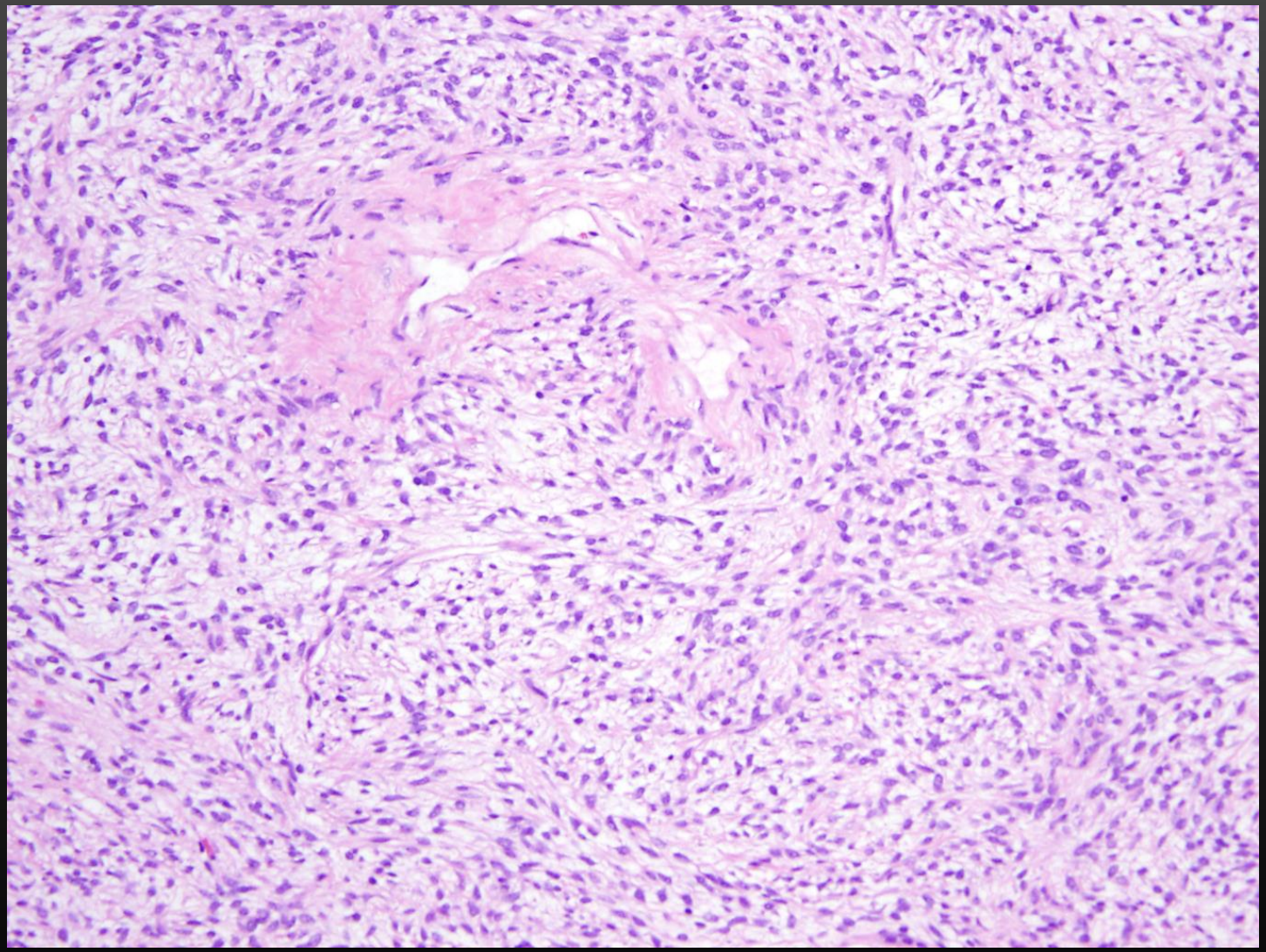

Solitary fibrous tumor of pleura

- 4% of all pleural neoplasms
- ~~Mesothelial origin,~~ mesenchymal origin
  - Not associated with asbestos exposure and usually good prognosis
- 80% arise from visceral pleura
  - May also derive from parietal pleura, other serosal membranes (peritoneum, pericardium), and nonserosal sites (lung parenchyma, mediastinum)
- Malignancy is defined by pathologic findings (cellularity, mitosis, atypia…), associated pleural effusion, atypical location, and invasion
  - 12 – 33% are malignant
- M=F, peak incidence in 6^{th}-7^{th} decades of life
- Usually asymptomatic, unless large

Quiz

The association of paraneoplastic hypoglycemia with pleural solitary fibrous tumor is termed the

A. Pierre-Marie-Bamberg syndrome
B. Mankowsky syndrome
C. Doege-Potter syndrome
D. Klempere-Rabin syndrome

Quiz

The association of paraneoplastic hypoglycemia with pleural solitary fibrous tumor is termed the

A. Pierre-Marie-Bamberg syndrome
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D. Klempere-Rabin syndrome

Solitary fibrous tumor of pleura

- Paraneoplastic syndromes
  - Hypertrophic osteoarthropathy (20%) = Pierre-Marie-Bamberg syndrome
    - Due to production of hyaluoronic acid, or hepatocyte growth factor
  - Hypoglycemia (2-4%) = Doege-Potter syndrome
    - Due to production of insulin-like growth factor II
  - Serous pleural effusion (10%)

Quiz

The following tumors in the chest can also present with hypoglycemia, EXCEPT for

A. Mesothelima
B. Leiomyosarcoma
C. Squamous cell carcinoma
D. Liposarcoma
E. Metastatic hepatocellular carcinoma

Quiz

The following tumors in the chest can also present with hypoglycemia, EXCEPT for

A. Mesothelima
B. Leiomyosarcoma
C. Squamous cell carcinoma
D. Liposarcoma
E. Metastatic hepatocellular carcinoma

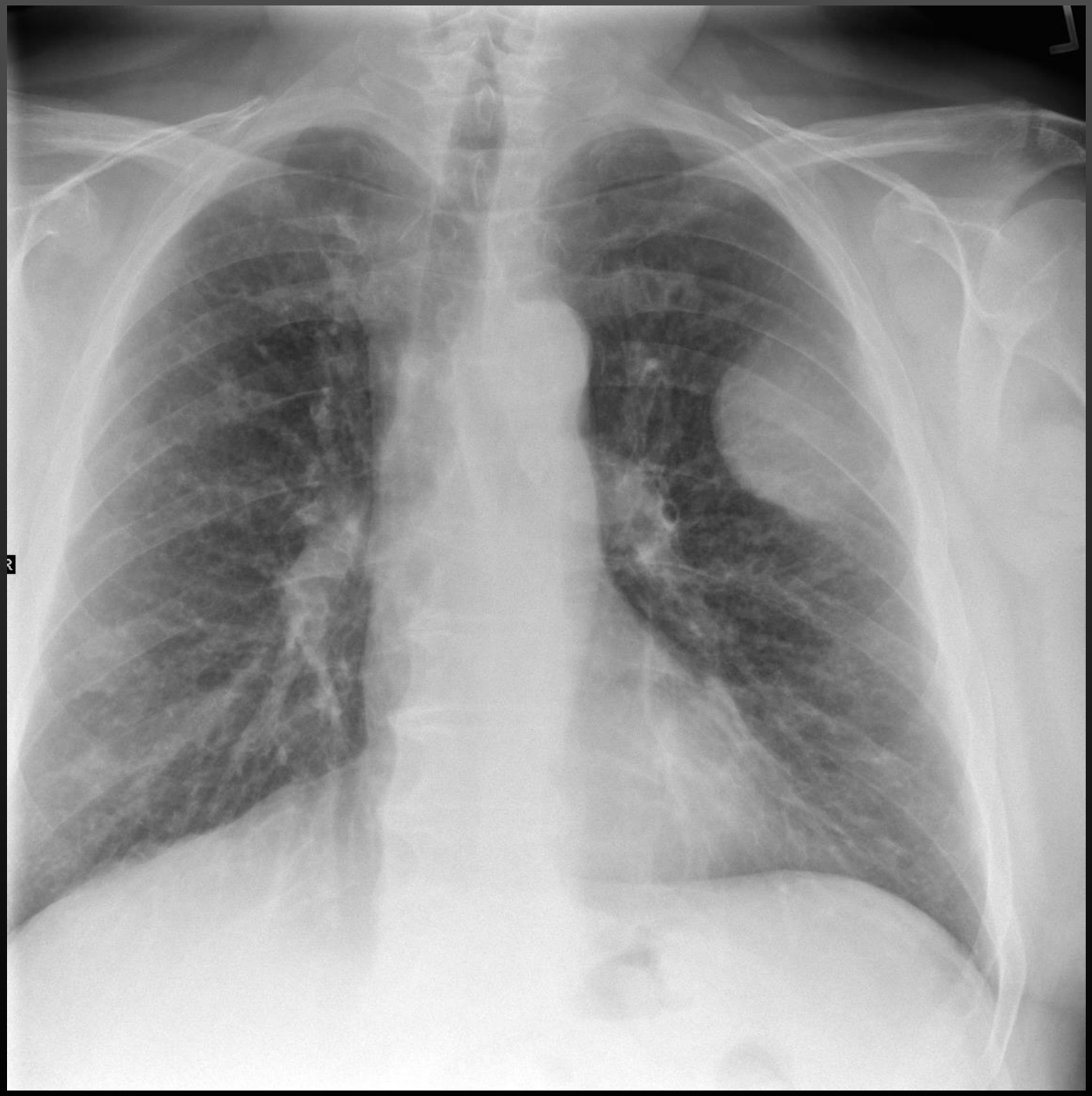
Solitary fibrous tumor of pleura

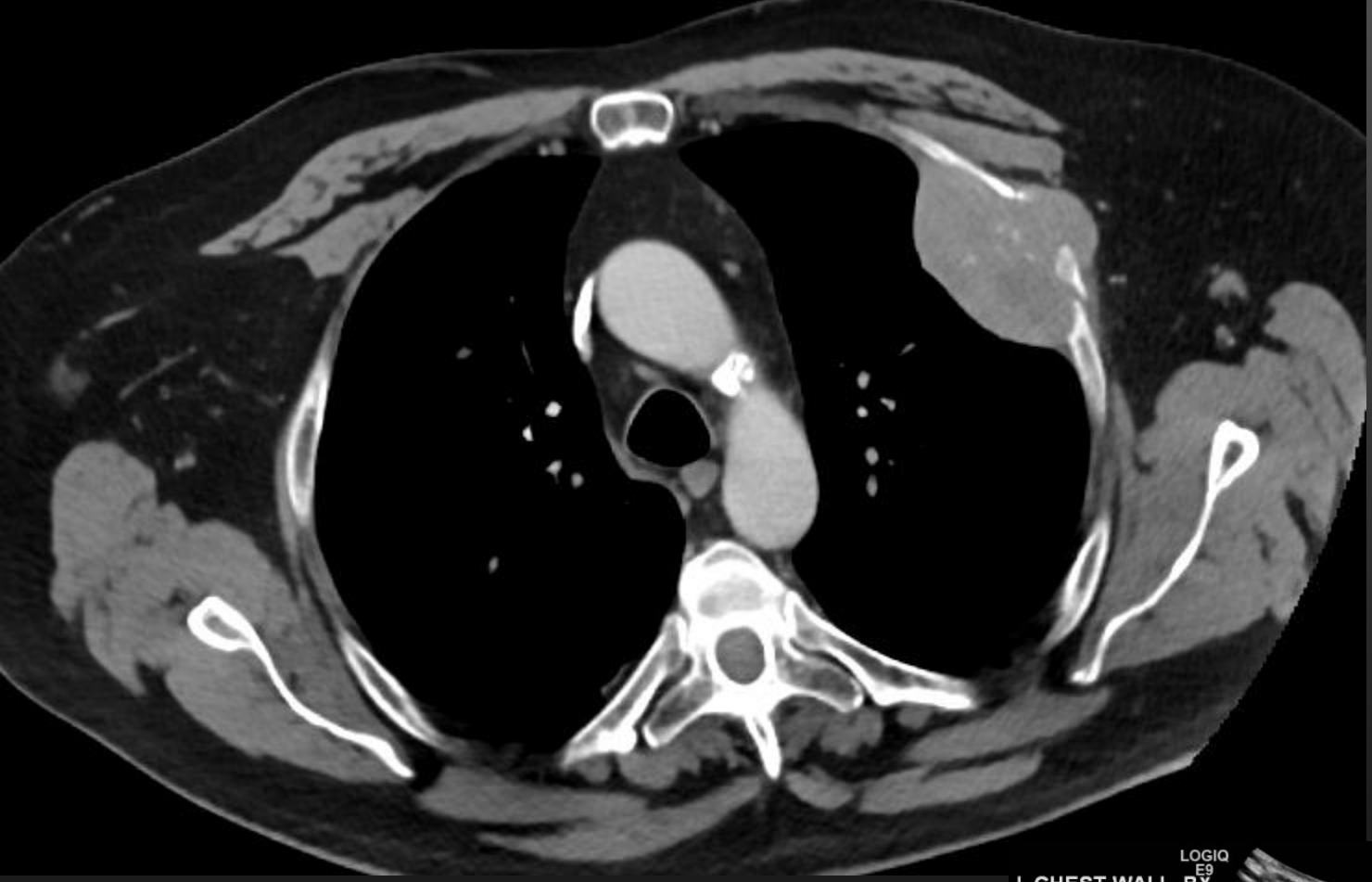
- CT
  - Well circumscribed, homogeneous density
  - CT cannot tell between benign and malignant --- FDG PET can (high uptake)

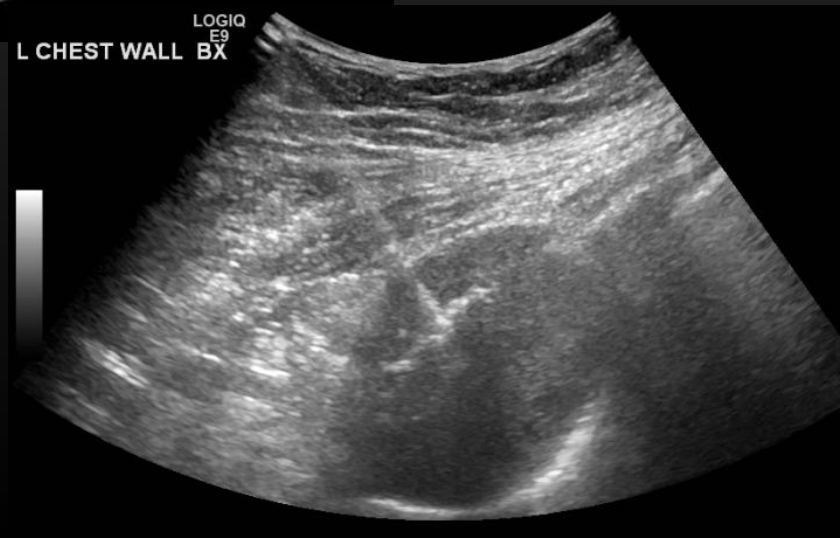
- CT guided FNA is often inconclusive (45% accuracy)
- Diagnose by histologic and immunohistochemical analysis
- Treat by complete surgical excision
- Local recurrence uncommon in benign lesions (8%), but high rate for malignant lesions (up to 63%)
- Most recurrences occur within 24 months of initial resection
- But late recurrence possible → need long term follow up of 15-20 yrs!

Case 6

- 63M presenting to PCP office with cough, dyspnea, left anterior chest wall pain



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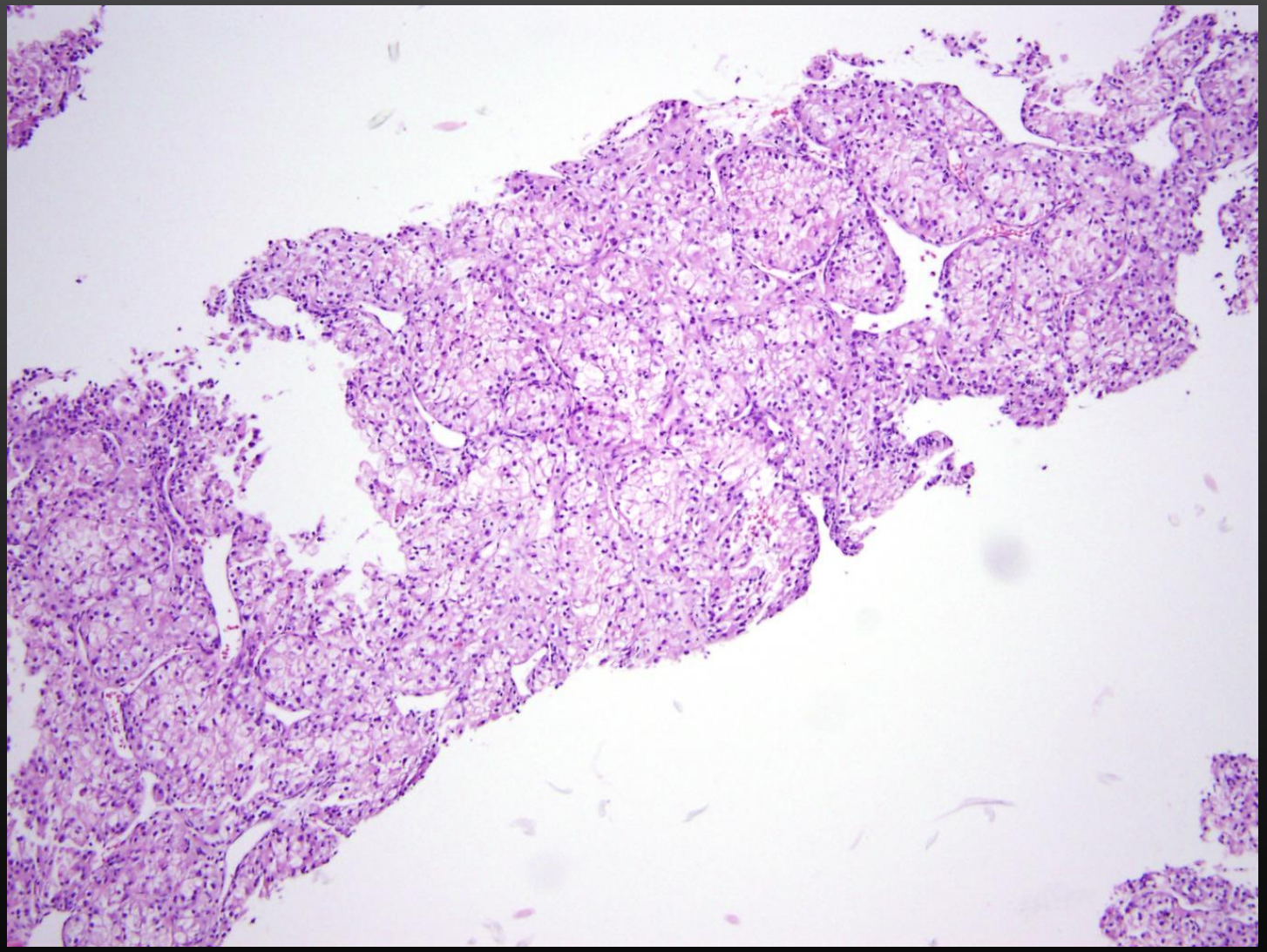
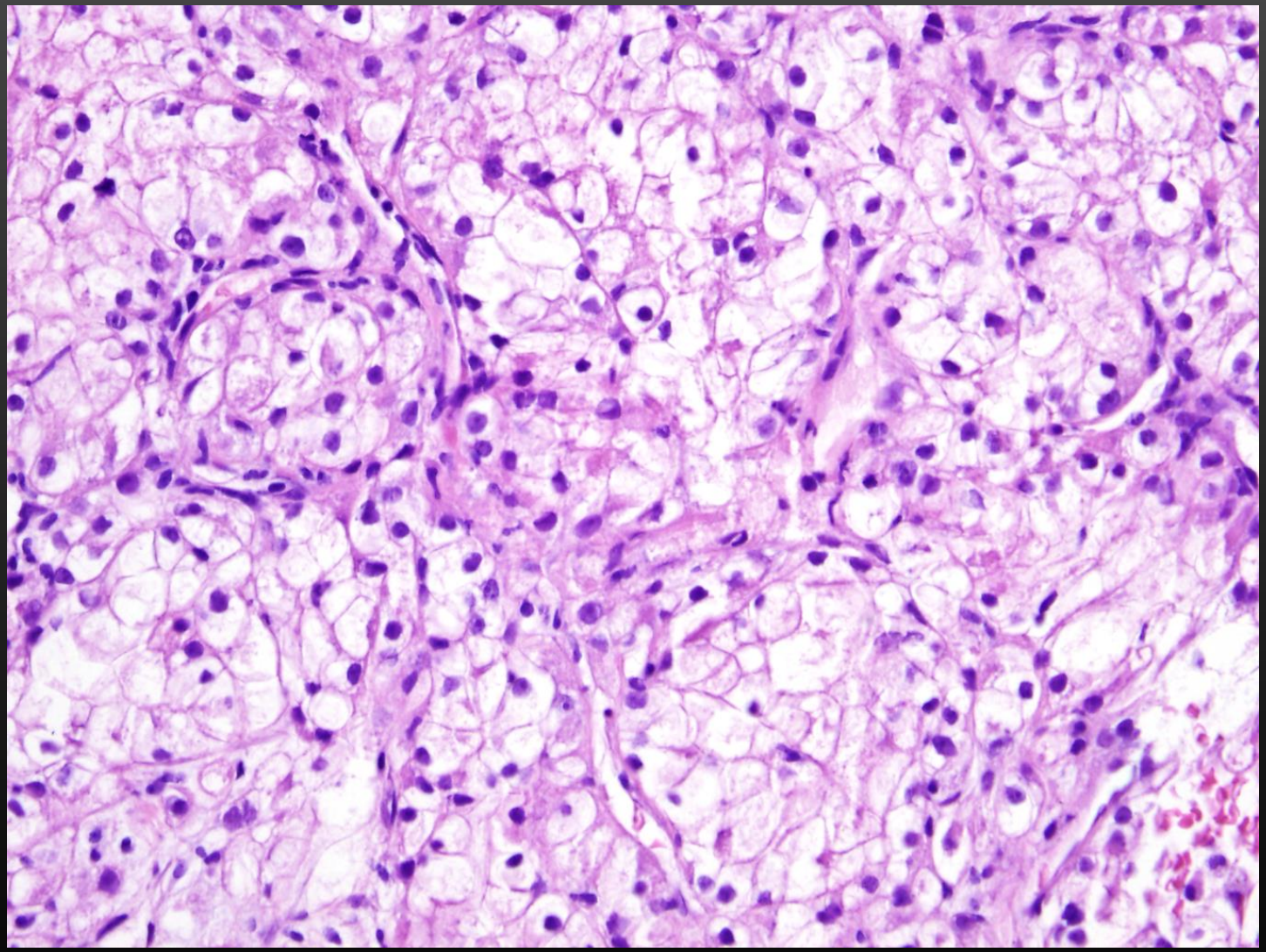
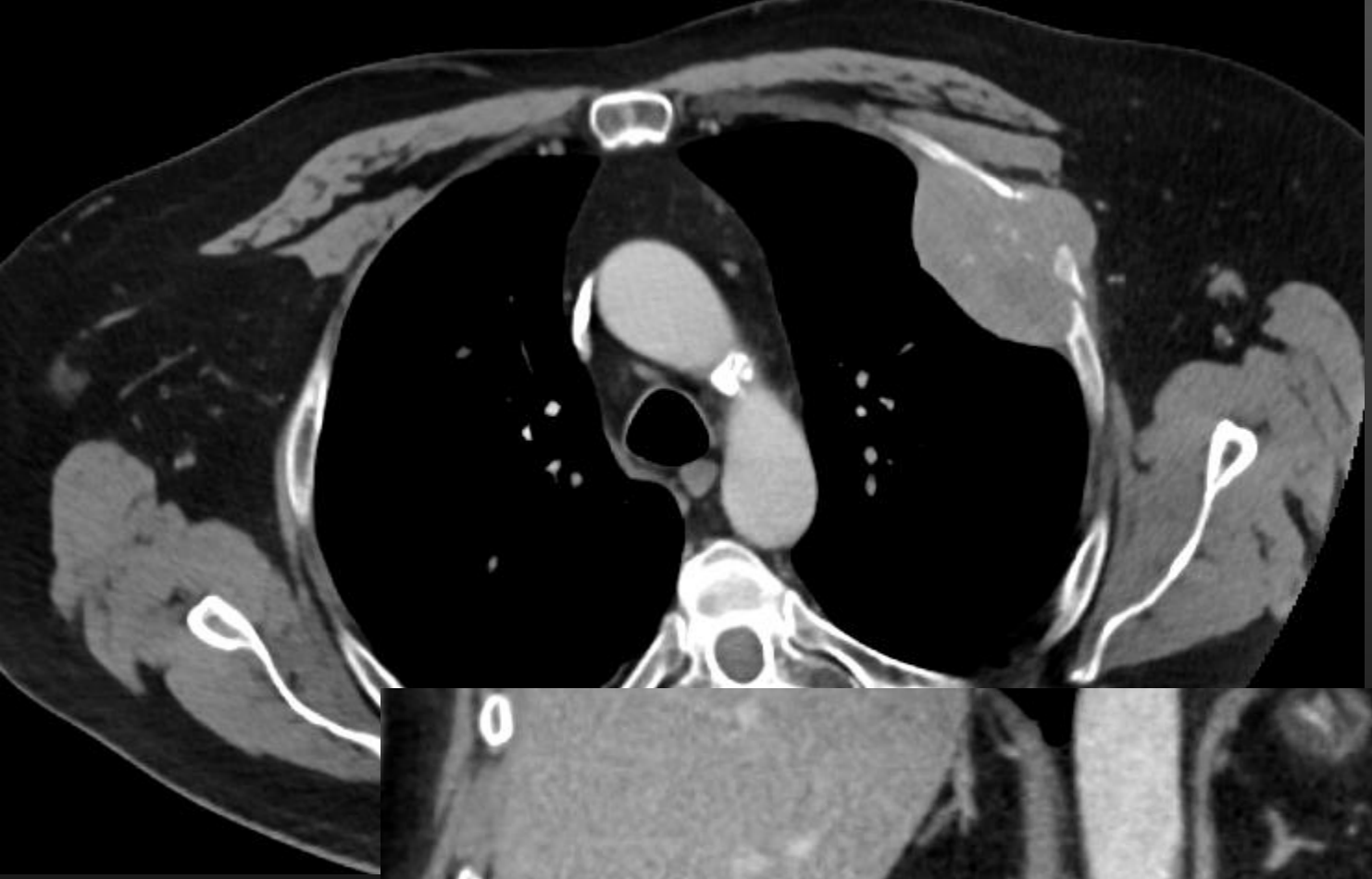
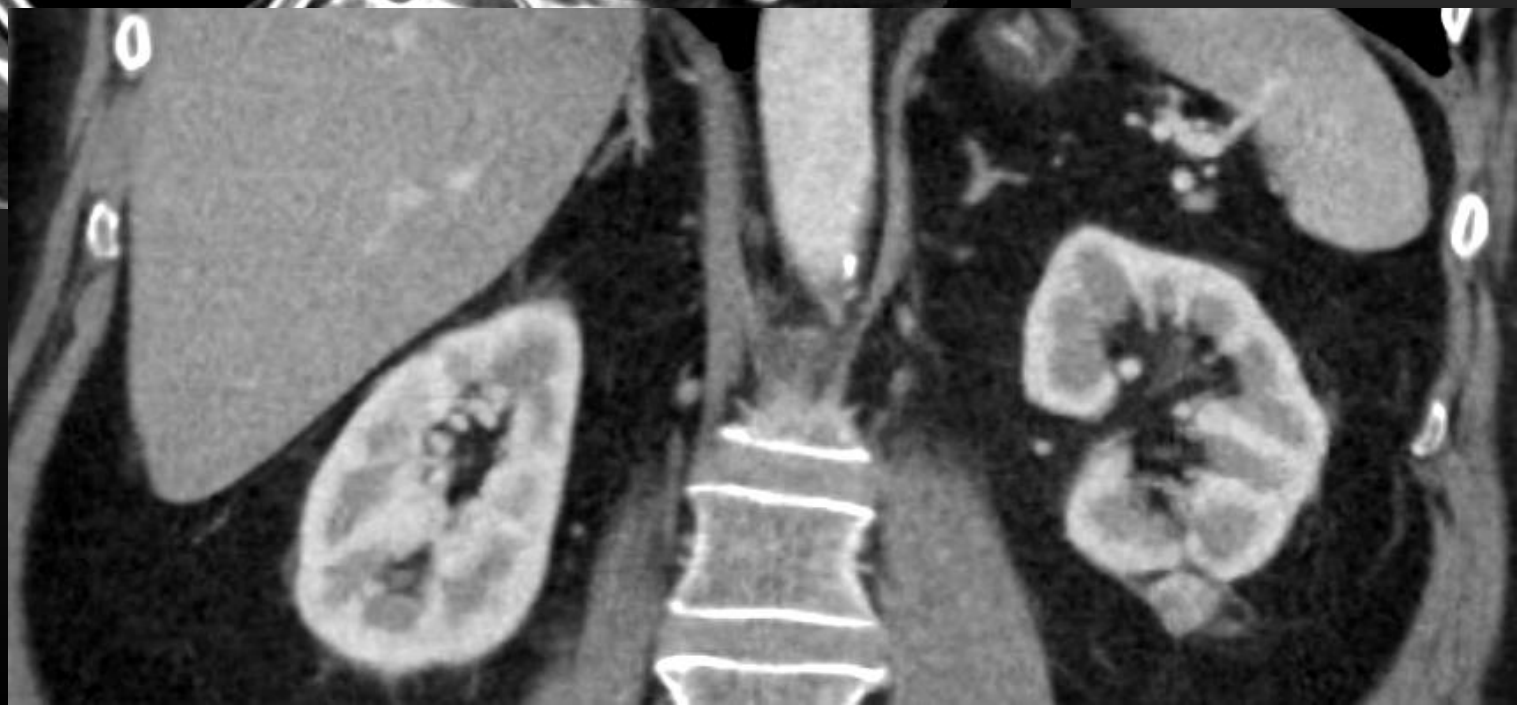
BS-16-56742







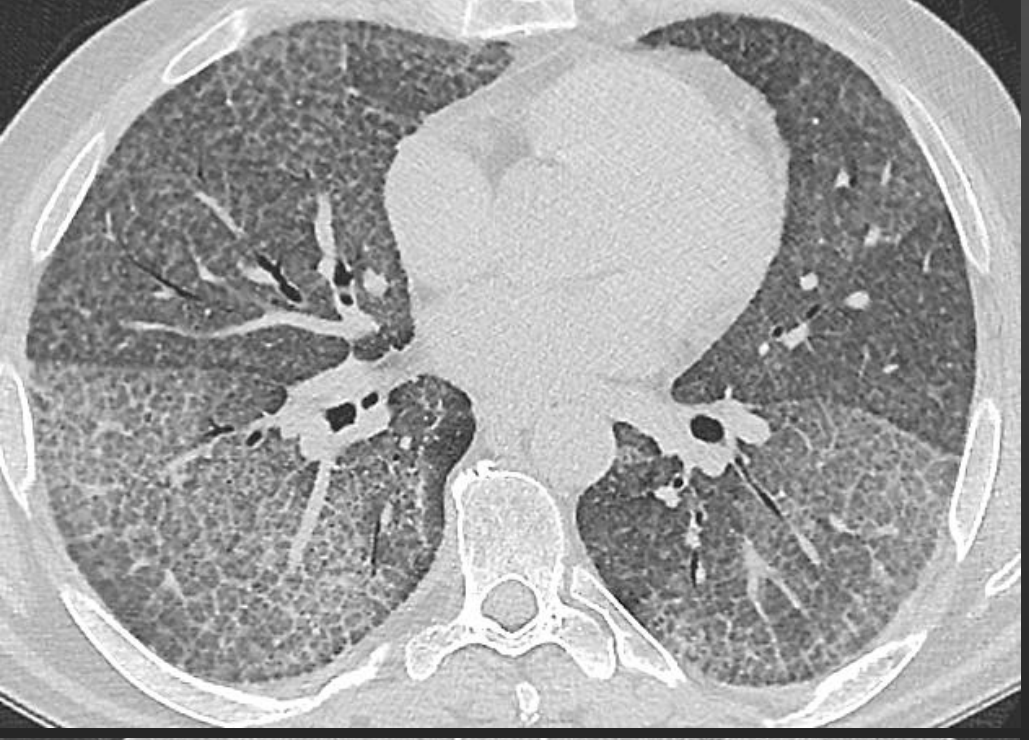


Differential:
solitary expansile lytic/lucent rib lesion

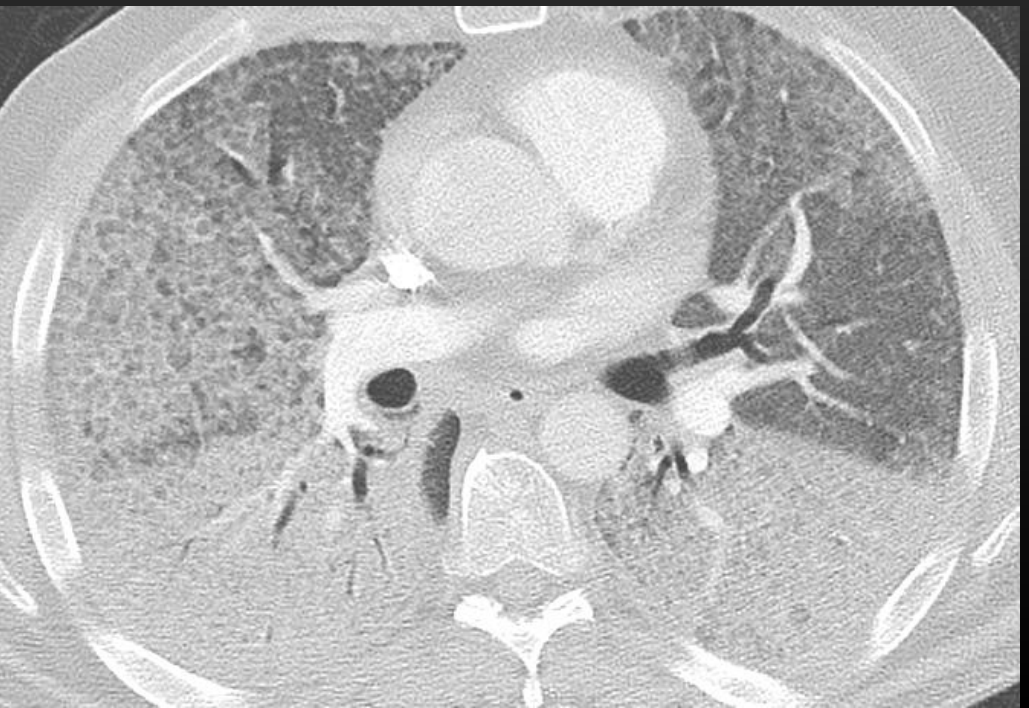
- Plasmacytoma
- Metastases (most common)
  - Multiple myeloma
  - RCC
- Chondrosarcoma (ring and arcs)
- Enchondroma (ring and arcs)
- Ewing sarcoma
- Fibrous dysplasia
- Aneursymal bone cyst
- Eosinophilic granuloma (Langerhans cell histiocytosis)

Bonus case!

42M with interstitial lung disease of unknown etiology, who presents with increased hypoxia

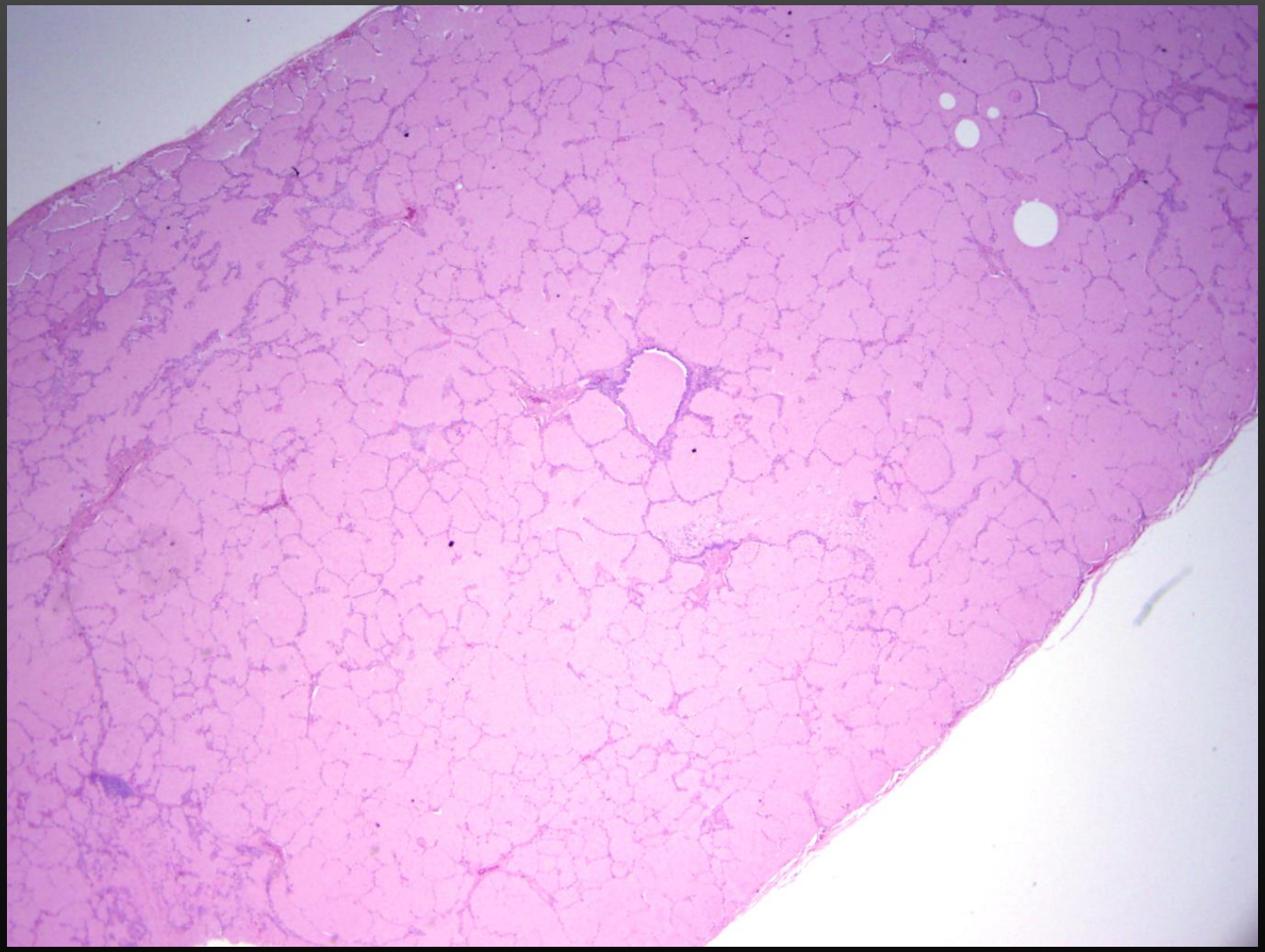


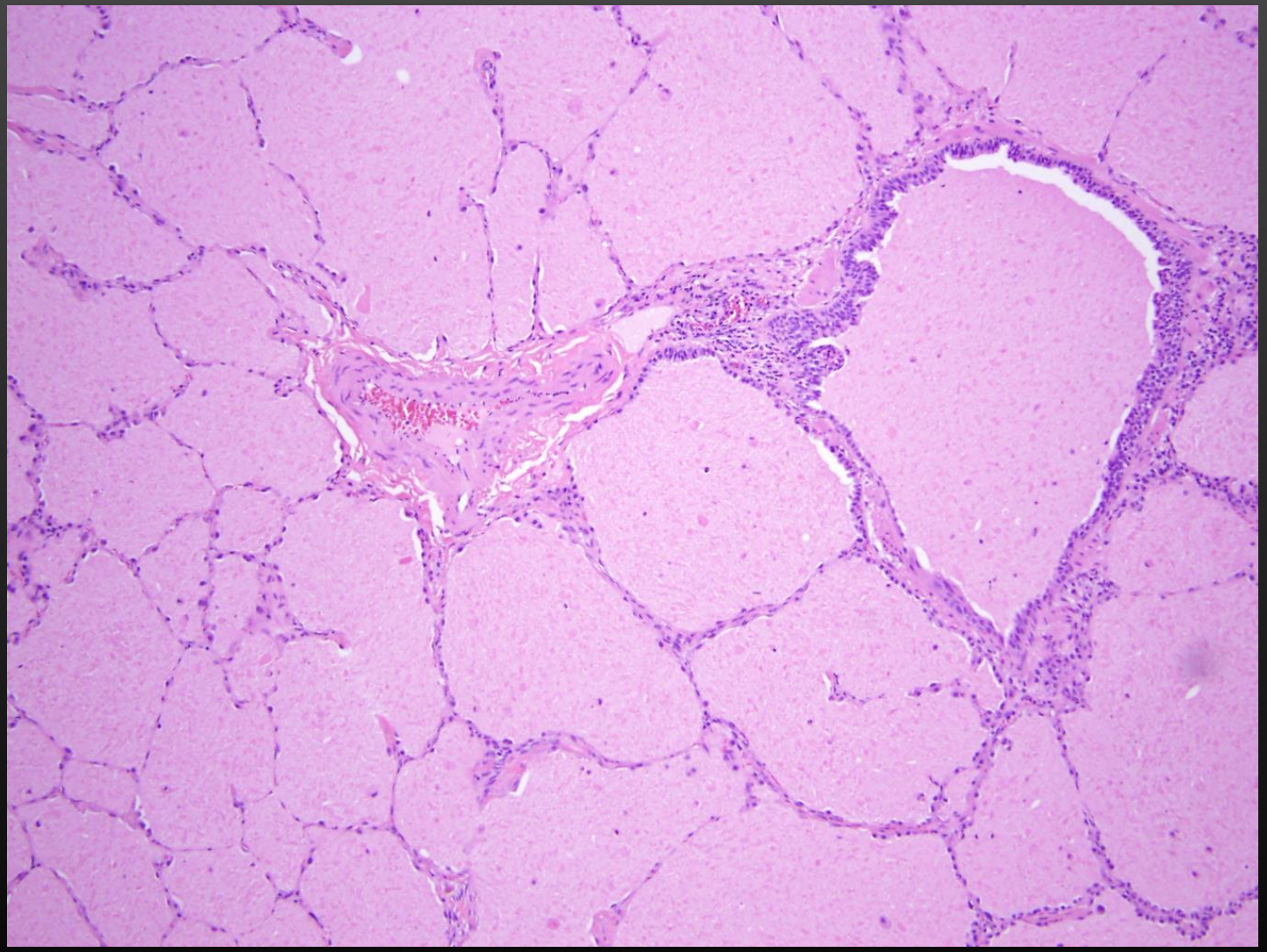
April



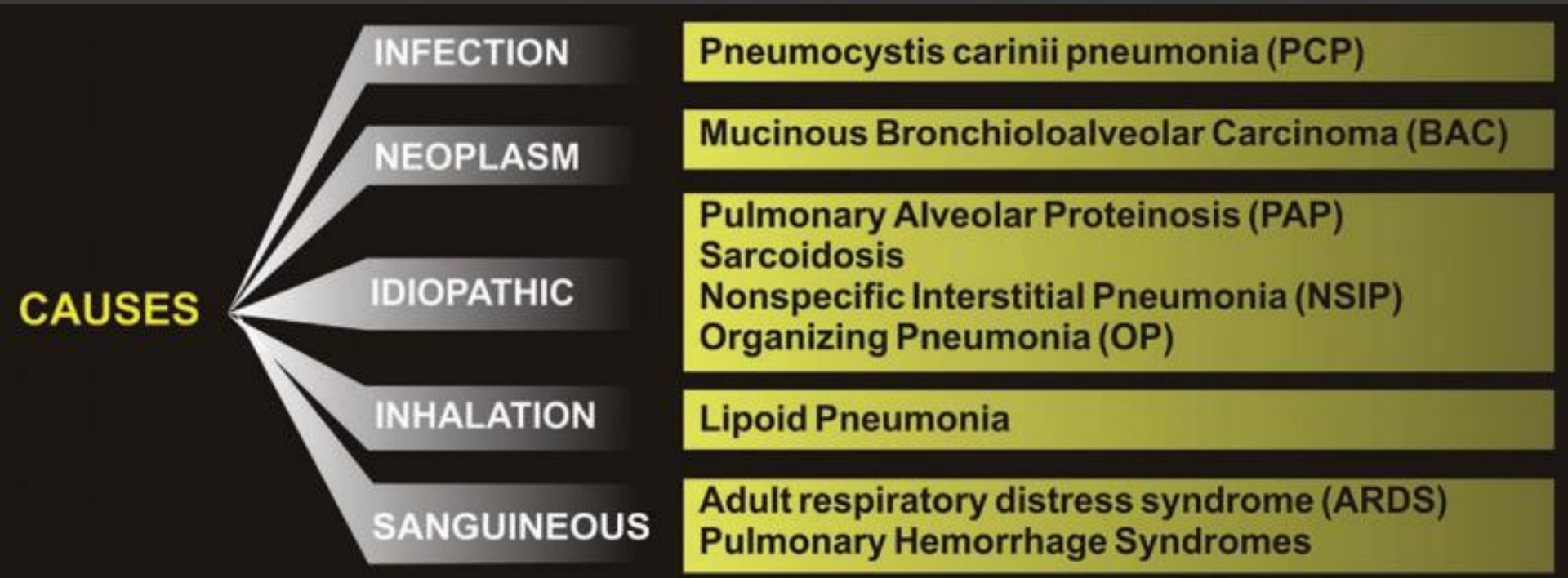
June

pathology
BS-17-41279





Crazy paving: differential diagnosis



Radiographics. 2003 Nov-Dec;23(6):1509-19.

Take Home

- Know your mediastinal mass differential
- Infections can be “mass-like”
- Primary pulmonary lymphoma can present variably as nodules/masses, consolidation or GGO
- Don’t forget about metastasis to the chest

Thank you to

- Dr. Angela Giardino
- Dr. Mark Hammer

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