Radpath Conference Vascular Diseases

6/9/2014

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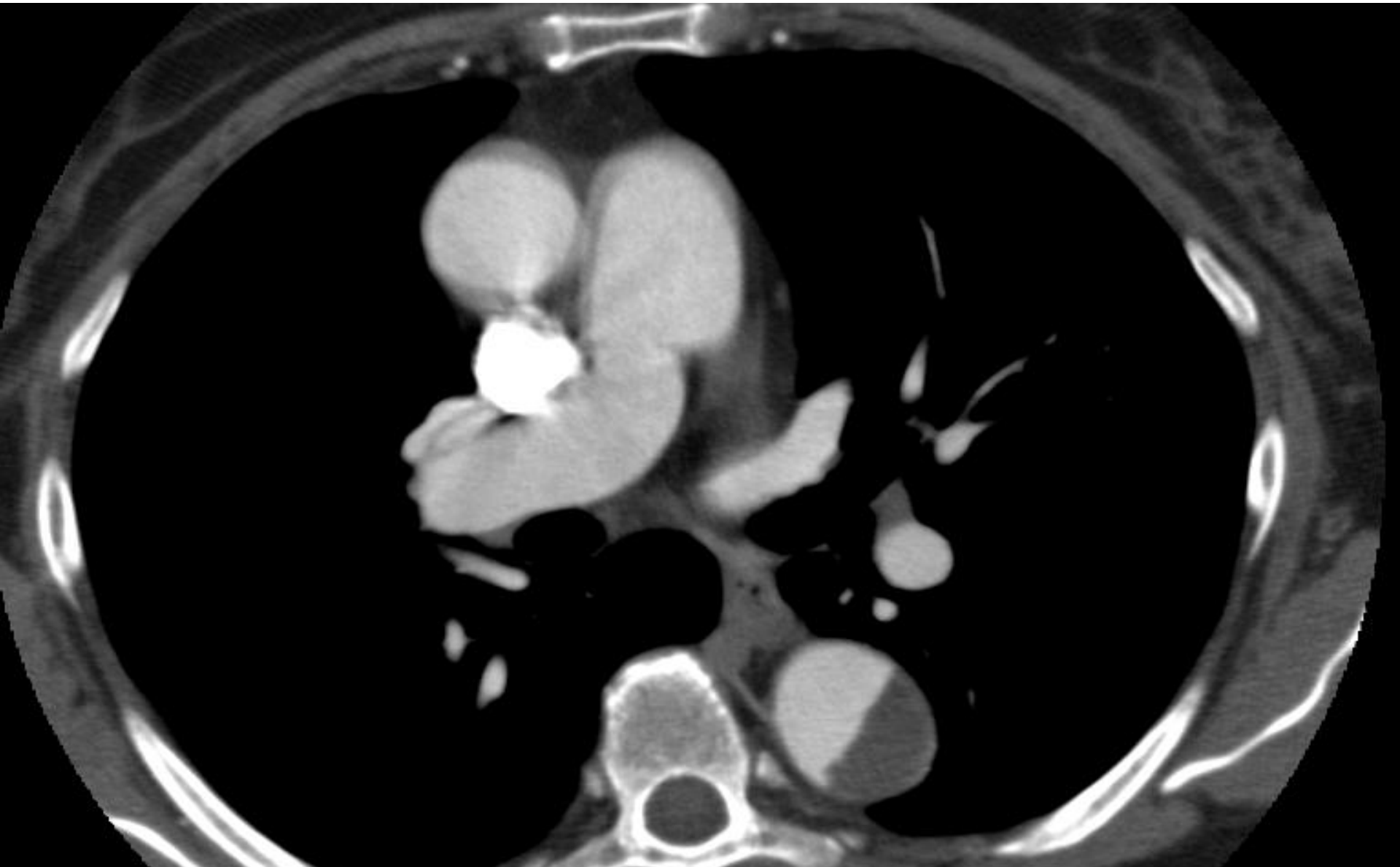
Clinical Condition: **Nontraumatic Aortic Disease**

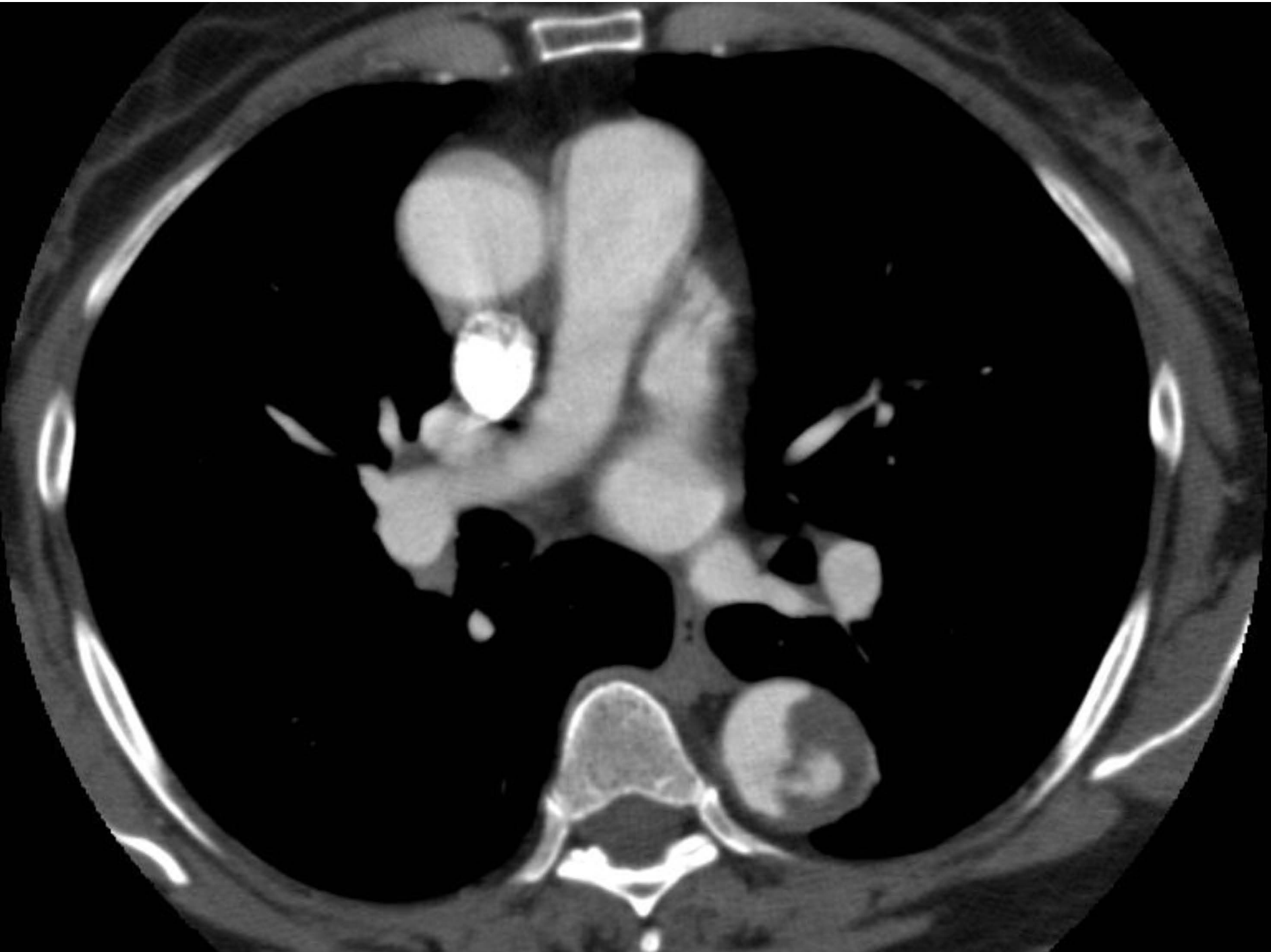
Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	9		☢
CT chest with contrast	8		☢ ☢ ☢
CT chest and abdomen without contrast	8		☢ ☢ ☢ ☢
CT chest and abdomen without and with contrast	8		☢ ☢ ☢ ☢
CTA chest with contrast	8		☢ ☢ ☢
CTA chest and abdomen with contrast	8		☢ ☢ ☢ ☢
MRA chest without and with contrast	8	See statement regarding contrast in text under "Anticipated Exceptions."	O
MRA chest and abdomen without and with contrast	8	See statement regarding contrast in text under "Anticipated Exceptions."	O
US echocardiography transesophageal	7		O
CT chest without contrast	7		☢ ☢ ☢
CT chest without and with contrast	7		☢ ☢ ☢
CT chest and abdomen with contrast	7		☢ ☢ ☢ ☢
MRA chest without contrast	7		O
MRA chest and abdomen without contrast	7	See statement regarding contrast in text under "Anticipated Exceptions."	O
US echocardiography transthoracic resting	6		O
Aortography chest and abdomen	6		☢ ☢ ☢ ☢
FDG-PET/CT chest and abdomen	5		☢ ☢ ☢ ☢
In-111 WBC scan	5		☢ ☢ ☢ ☢
US intravascular aorta	4		O

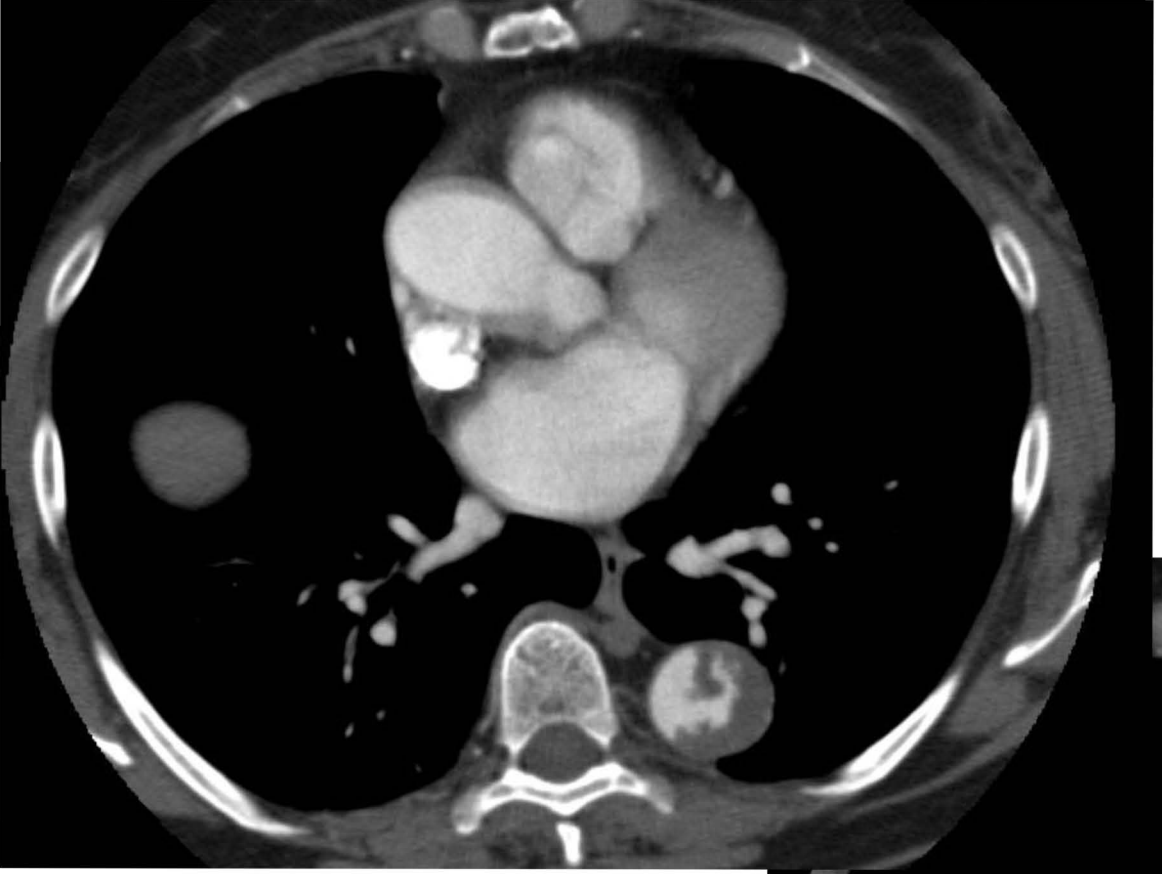
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**

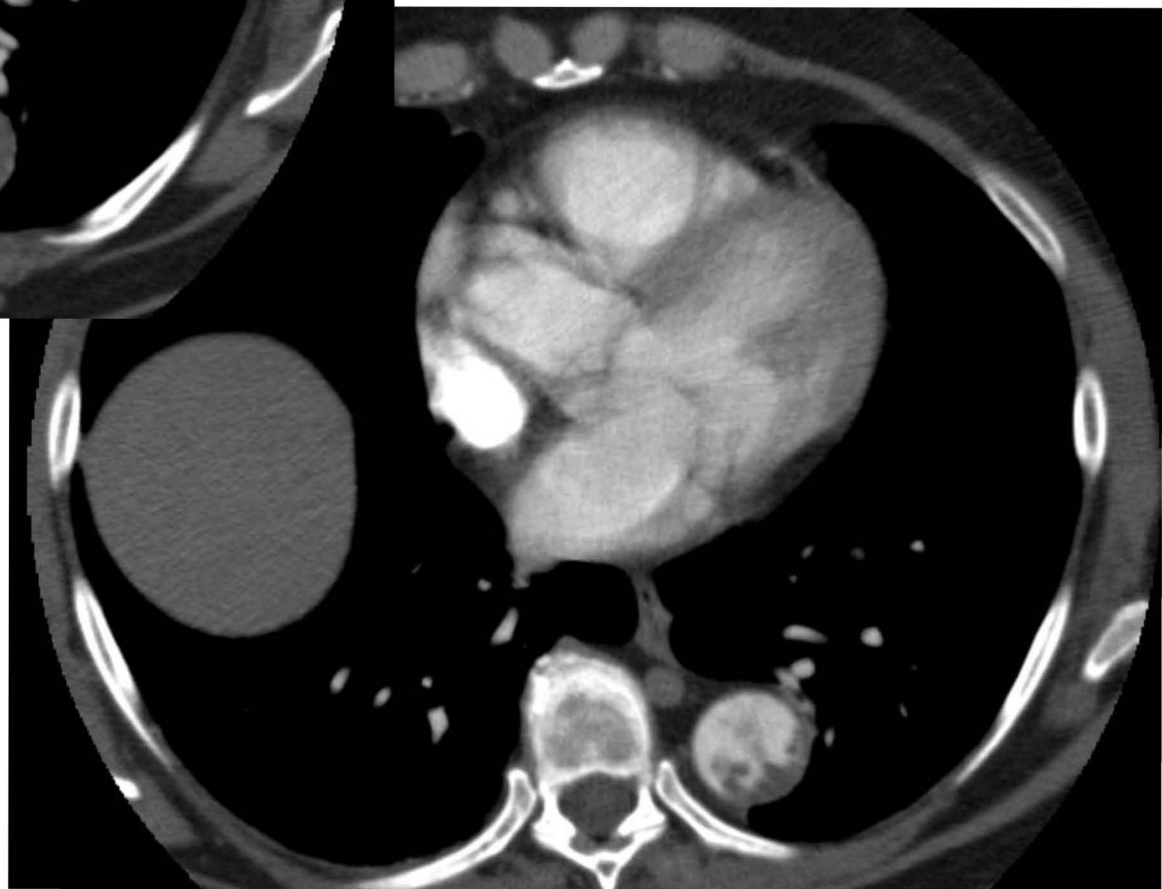
***Relative Radiation Level**

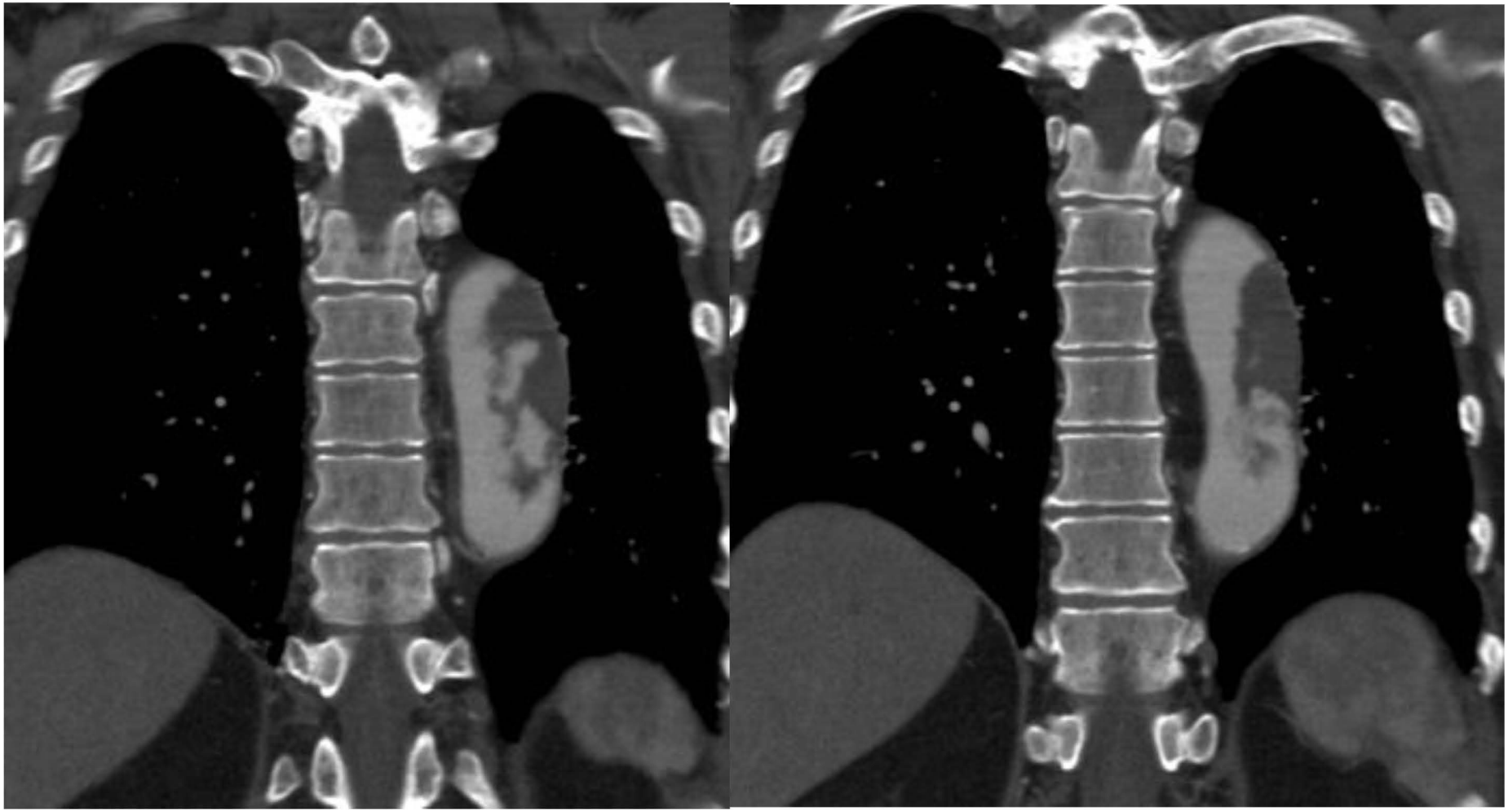
Case 1





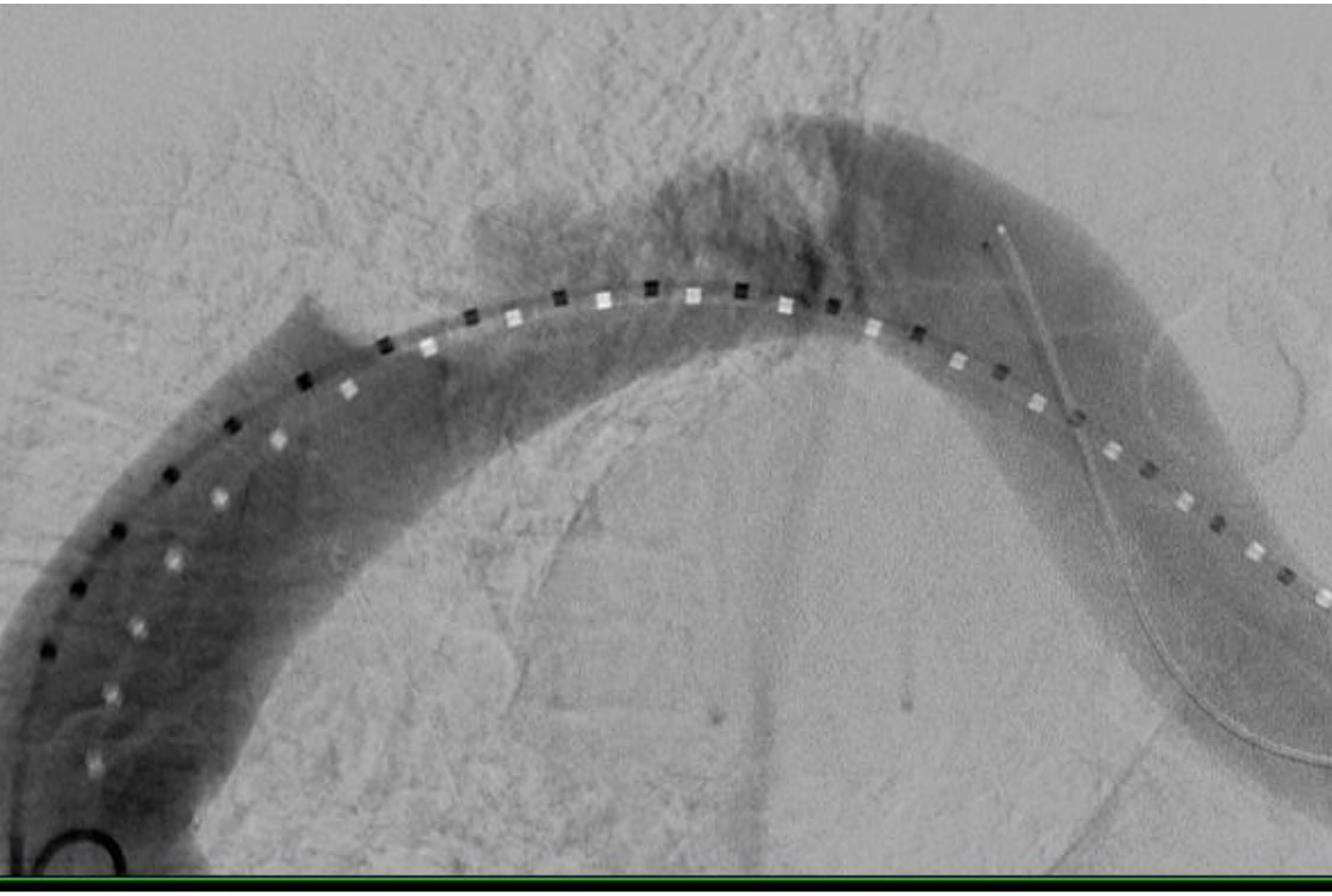






Clinical Course

- Presented to OSH with chest tightness and left arm numbness
- CTA chest and abdomen read as mural thrombus in the descending thoracic aorta spanning from the level of main pulmonary artery extending into the inferior aspect of left atrium
- Transferred to another OSH: ECHO demonstrated no evidence of intracardiac thrombus and eventually had a thoracic aortic angiogram followed by deployment of a covered stent while on AC (ASA/heparin)
  - Based on clinic note, wall stent was deployed in the distal aorta to prevent distal embolization during stent graft procedure which was removed and incidentally found to have soft tissue material within the wallstent → sent to pathology for review



Differential diagnoses

- Mural thrombus
- Severe atherosclerosis
- Primary malignant tumors of the aorta (PMTA)
- Vasculitis

Primary Malignant Tumors of the Aorta (PMTA)

- Aggressive class of sarcomas arising from aortic wall or intima
  - Intima with intraluminal extension (80%): intimal sarcoma, angiosarcoma, malignant fibrous histiocytoma, fibrosarcoma, osteosarcoma, chondrosarcoma
    - More likely to obstruct lumen
    - Often poorly differentiated
  - Mural with extraluminal extension (20%): leiomyosarcoma, angiosarcoma, fibrosarcoma, osteosarcoma, chondrosarcoma
    - Often well differentiated
    - May obstruct lumen

- Presentation: Progressive chest or abdominal pain
  - May develop sx related to embolic disease or occlusion (mesenteric ischemia/ claudication, HTN, etc)
  - Male >female (2:1)
  - Mean age 60
  - Rare: ~150 cases of sarcomas are reported worldwide since 1873
  - Predominantly descending thoracic (35%)> abdominal (27%) > thoracoabdominal > arch (11%)
  - Mean survival: 16 months
- Metastasis to liver, lungs, bone, brain, and skin

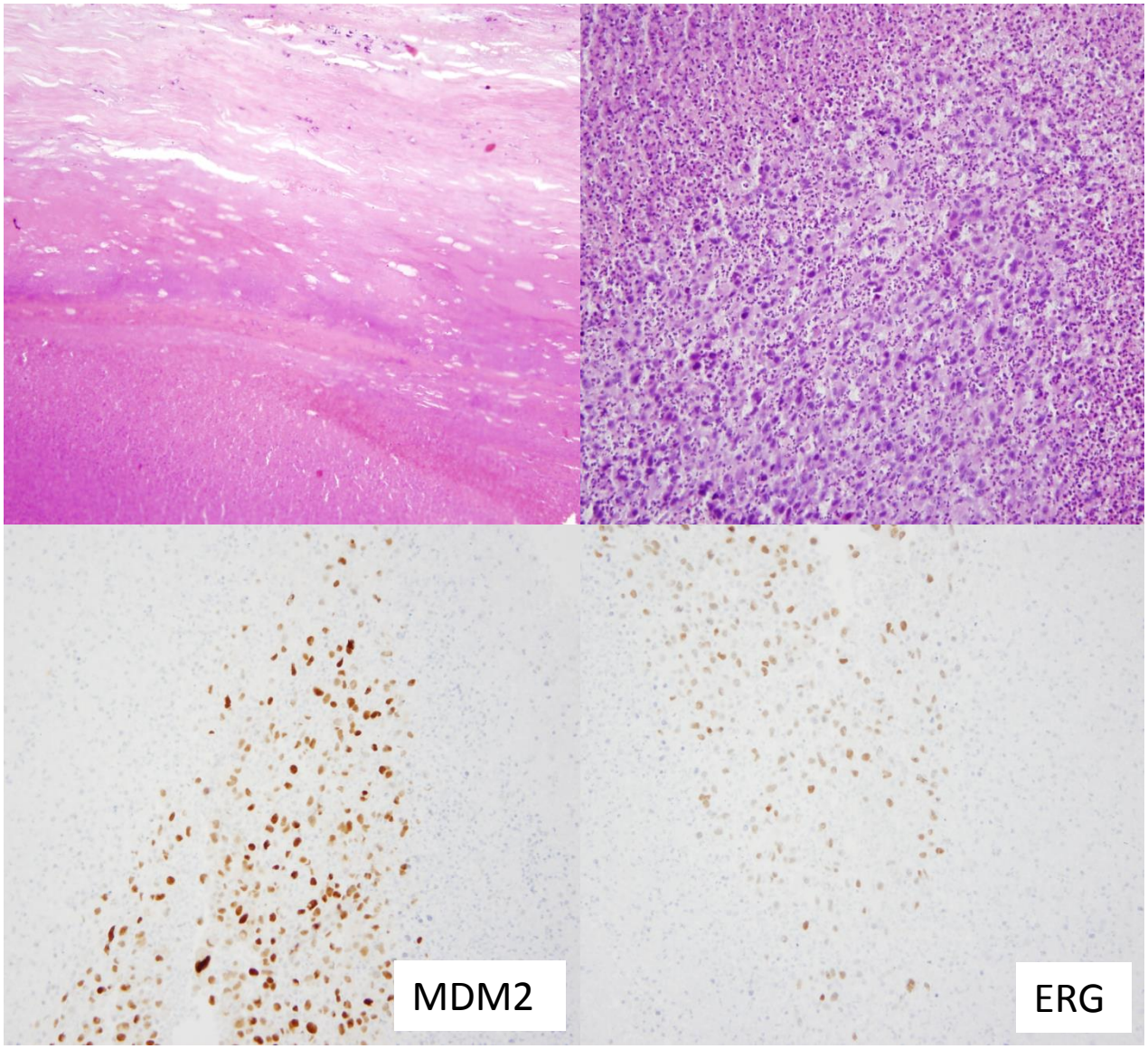
- Image findings:
  - Radiograph: mediastinal widening/lobulated contour
  - CT: Polypoid mass within, along, or surrounding aortic wall with narrowing/occlusion of lumen secondary to tumor emboli
  - MR: Similar to CT with wall enhancement
    - MRA is test of choice as it is easier to see enhancement on MR for differentiation of thrombus/plaque from PMTA
  - PET: Focus of FDG uptake in or around aortic wall
    - Useful for staging/met search
  - Although aggressive cases may appear mass like, some may only demonstrate mural thickening with minimal or no enhancement
  - Can be confused with vasculitis (consider if steroid resistant)
- Complications: Aneurysm formation secondary to wall weakening
- Treatment: Resection, chemoradiation, stent graft
- Worse prognosis with intimal sarcomas, involvement of ascending aorta or arch, and incomplete resection
- Extraluminal/medial aortic sarcomas have better prognosis
- Similarly, sarcomas can also arise from pulmonary artery, IVC, SVC, and heart

INTIMAL SARCOMA

- Malignant tumor arising in tunica intima of large blood vessels, usually aorta

- Pathologic findings:
  - Tumors are typically largely necrotic, with poorly differentiated epithelioid and pleomorphic cells associated with tunica intima

  - Positive for MDM2 (amplified by FISH), ERG, CD31, FLI1



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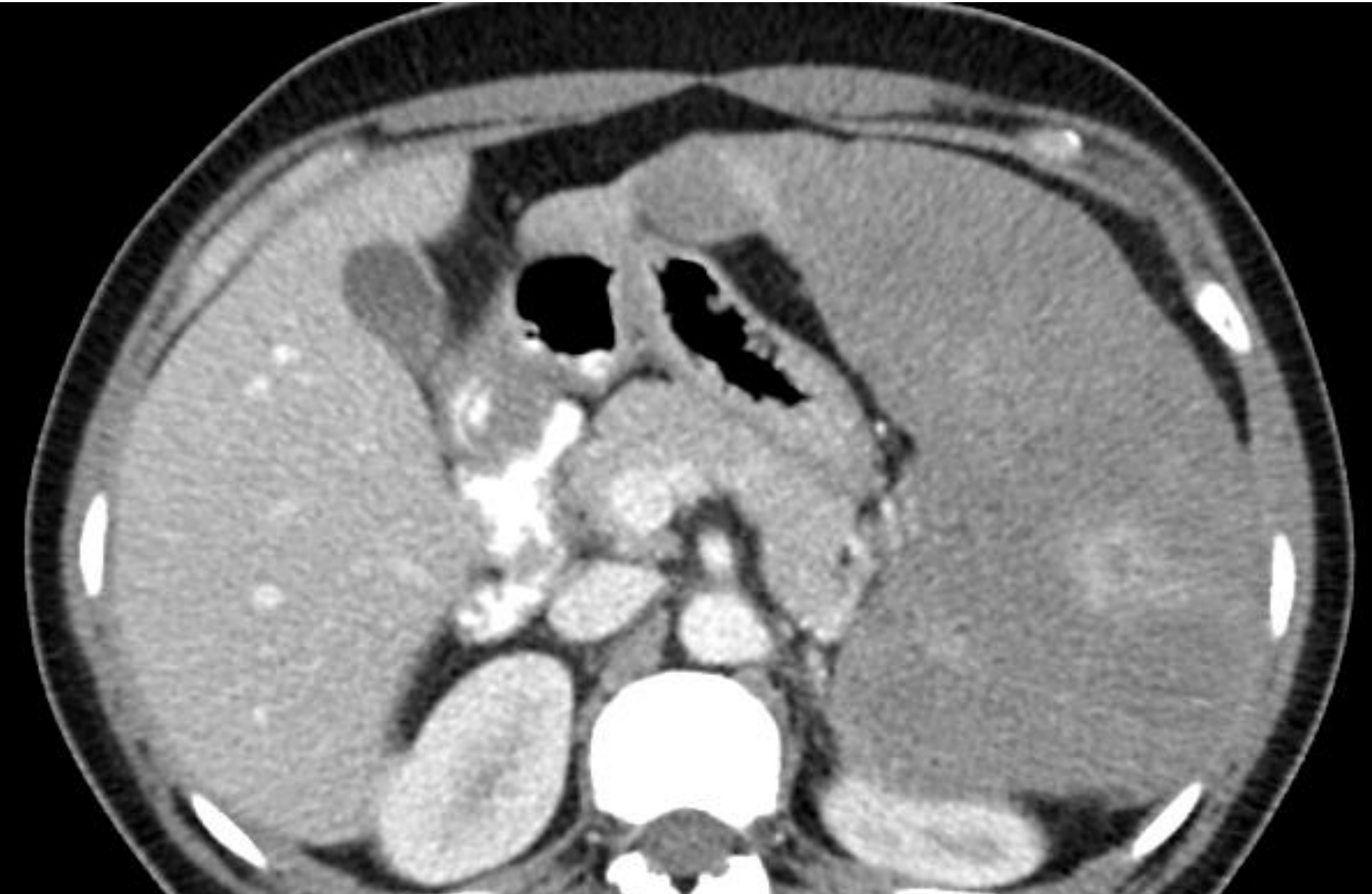


Clinical course

- Incidentally found to have HCT 19.5 → received 2 units of pRBCs
- Abdominal and pelvic US demonstrated splenomegaly (17.6 cm)
- CT and PET demonstrated no other findings; FDG uptake in the spleen
- BM biopsy→erythroid hyperplasia, but no malignancy
- Splenectomy

Differential Dx

- Normal: ≤ 13 cm
- Causes/Differentials:
  - Congestive: Right heart failure, portal HTN, splenic or portal vein thrombosis, sickle cell (splenic sequestration)
  - Hematologic: Polycythemia vera, leukemia, myelofibrosis, acute infarction
  - Malignant: Lymphoma, metastatic disease, primary splenic tumors
    - Primary splenic tumors are rare: most common is splenic angiosarcoma
  - Infectious: Mononucleosis, abscess
  - Infiltrative and storage disorders: Amyloidosis, hemochromatosis, gauchers disease (glycogen storage disease)
  - Trauma (intrasplenic or subcapsular hematoma)

Splenic angiosarcoma

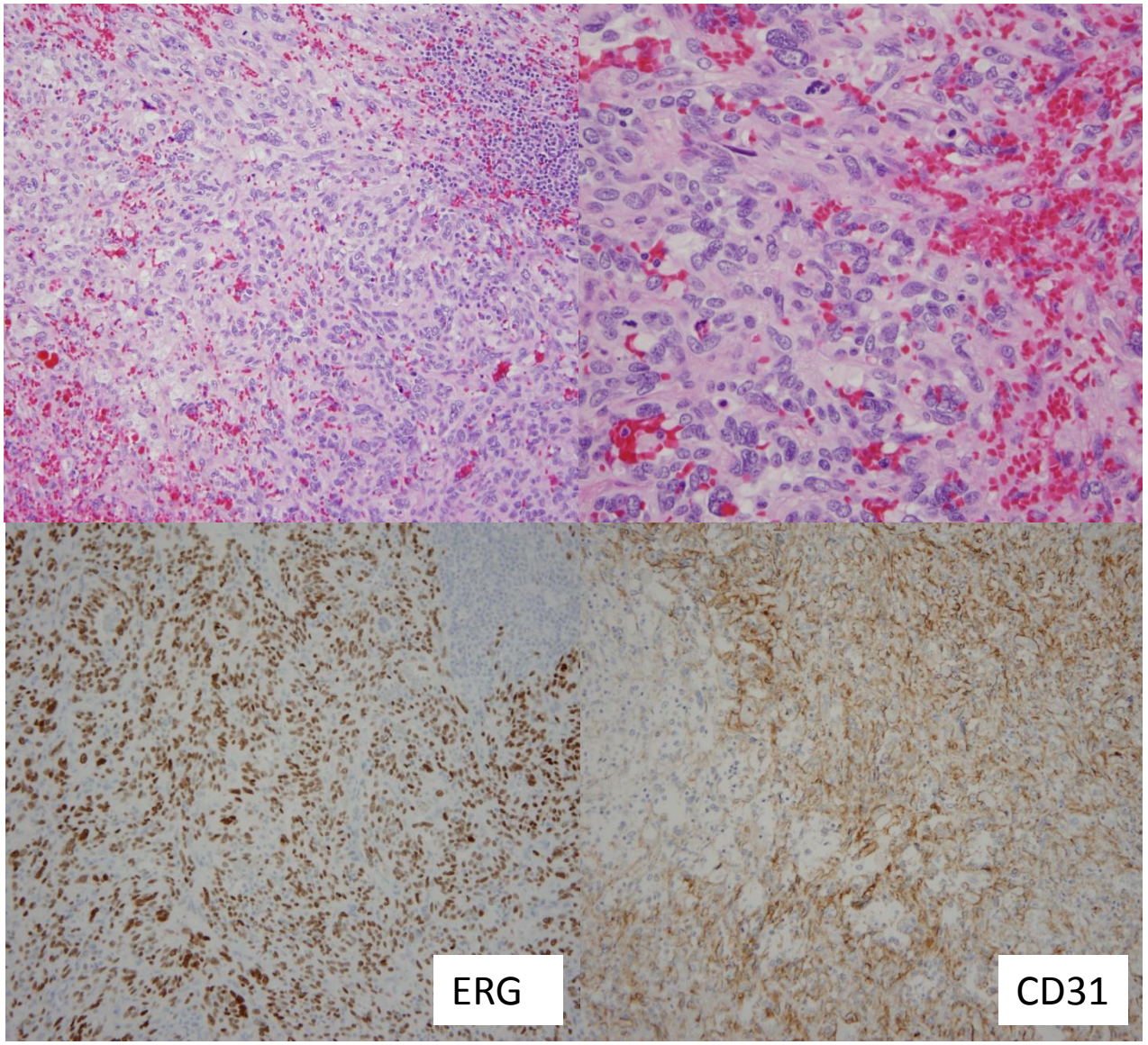
- Very rare but most common, extremely aggressive primary malignancy of the spleen
  - Neoplasm of endothelial-type cells lining the vessel
  - <1% of all sarcomas
  - Mean age 47 (ranges from 14months to 84 years)
  - Presents with LUQ pain, anemia, weight loss, fatigue, dyspnea
  - Median survival 24-36 months or less (some report 10.3-14.4 months)
  - Ambiguous taxonomy in literature: angioma, hemangioma, angioblastoma, hemangioendothelioma of the spleen
- No known association with vinyl chloride, arsenic, or thorium dioxide as is with hepatic angiosarcoma

- Image characteristics are non-specific
  - US: Splenomegaly, mixed echogenic masses may be seen
  - CT: Splenomegaly although at times can have cystic and solid low attenuating masses or areas of necrosis. Occasionally can demonstrate acute hemorrhage or subcapsular hematoma.
    - With contrast, can demonstrate peripheral enhancement like hepatic hemangiomas
  - MR: T1 and T2 hypointense nodular masses
    - With subacute hemorrhage or tumor necrosis: Increased T1/T2 intensities
    - With gadolinium can demonstrate multinodular enhancement with focal areas of non-enhancement (hemorrhage/necrosis)
- Complications: May cause splenic rupture in up to 30% of cases
- Early metastasis to liver, lung, bone, LN, and brain
- Treatment: Resection, chemoradiation (?)

SPLENIC ANGIOSARCOMA

- Most common malignant nonlymphoid tumor of spleen
- Associated: microangiopathic anemia, thrombocytopenia, consumptive coagulopathy
- May develop years after insertion of foreign body
- Pathologic findings:
  - Solid, papillary or freely anastomosing vascular channels (variable even within same case), lined by atypical (occasionally epithelioid), hyperchromatic cells

  - Positive stains: Endothelial markers (CD31, CD34, ERG) and histiocytic markers (CD68, lysozyme)



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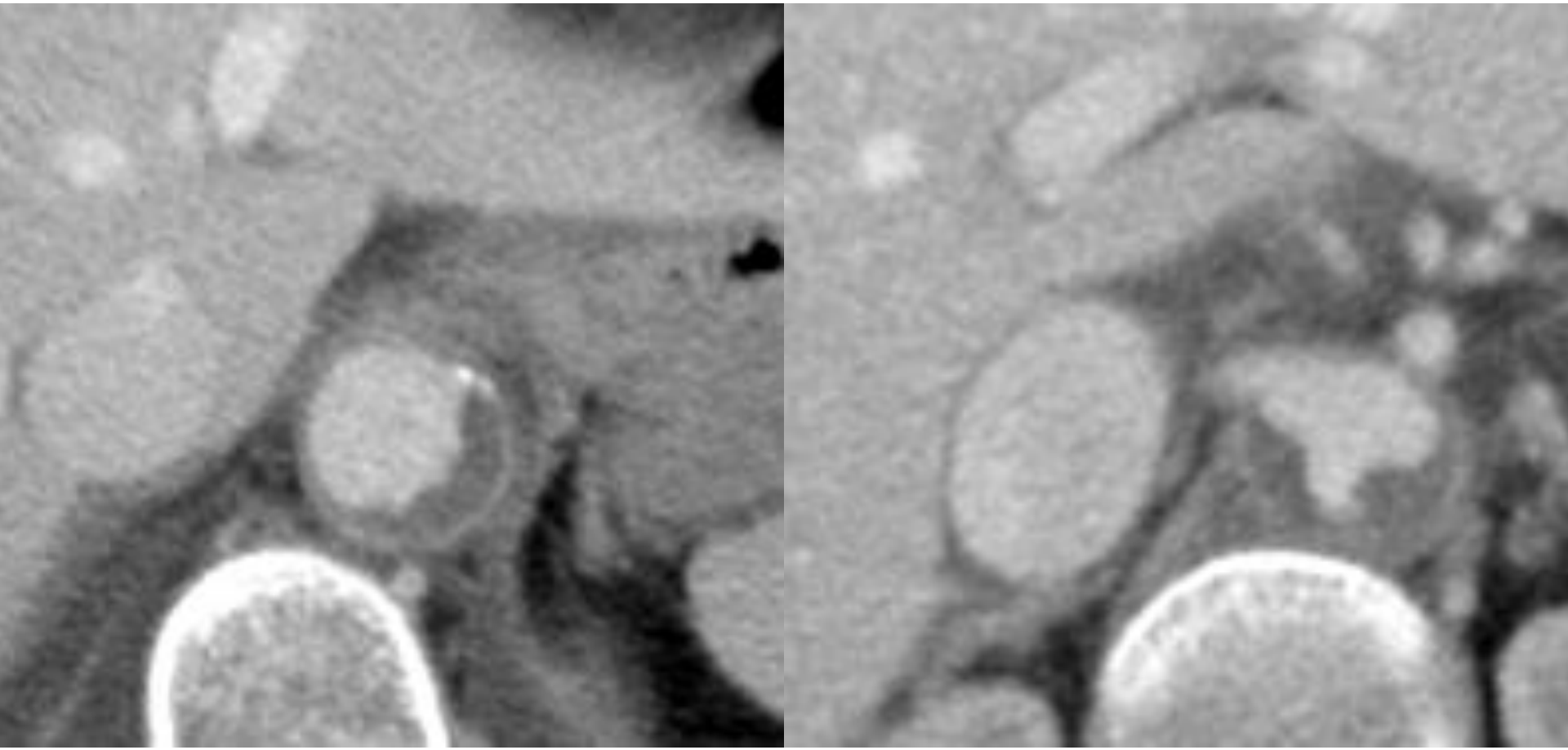
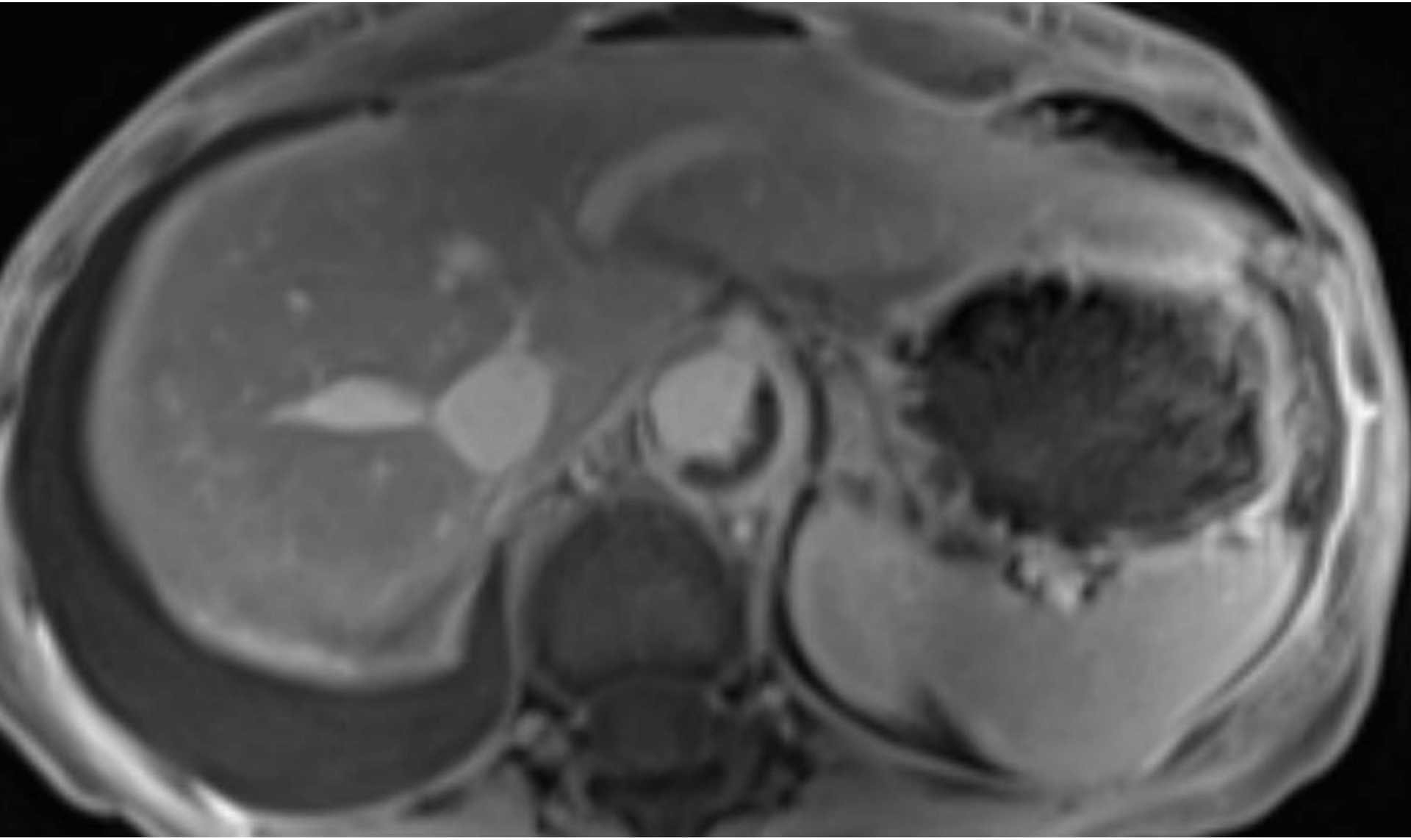
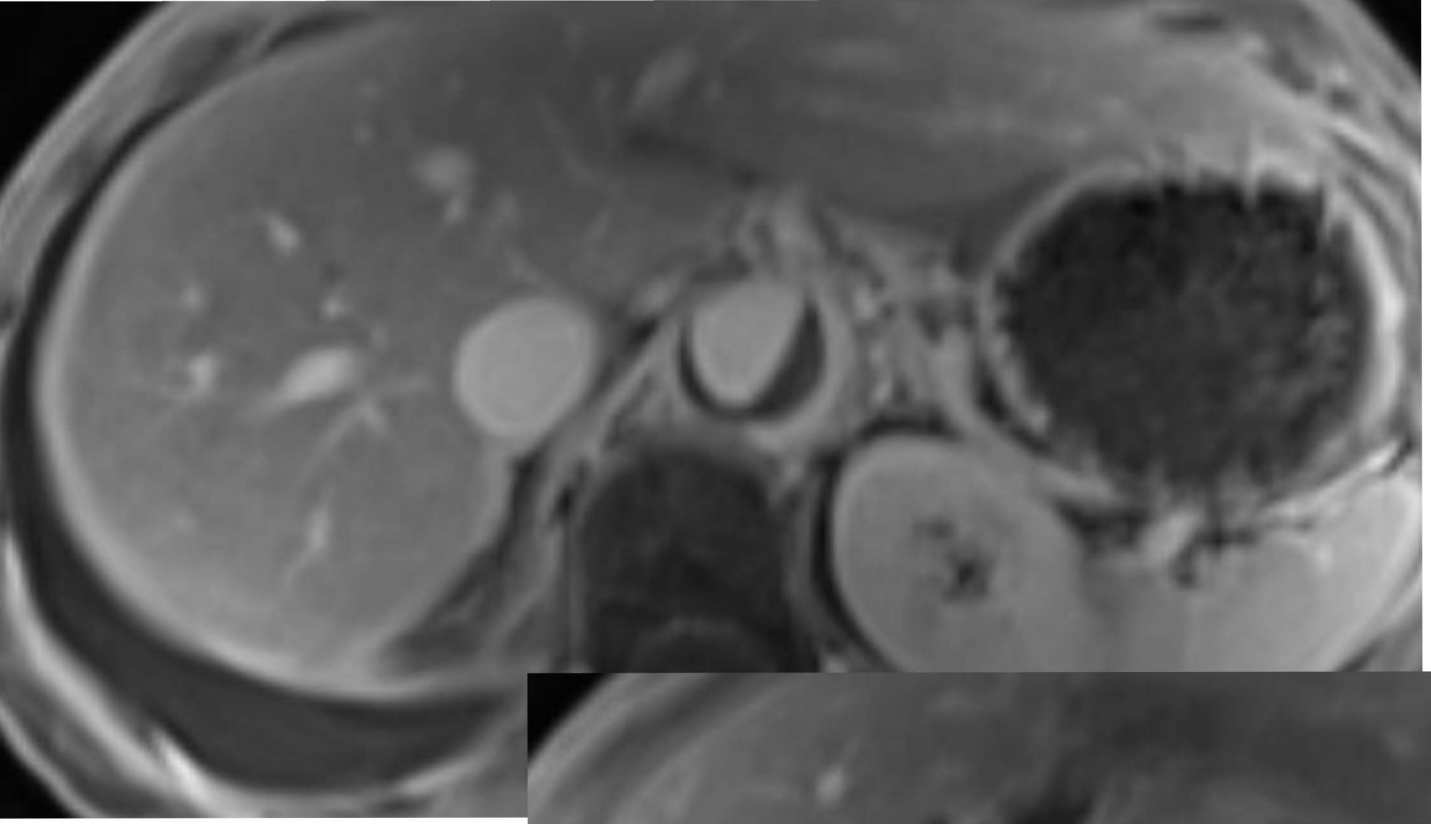
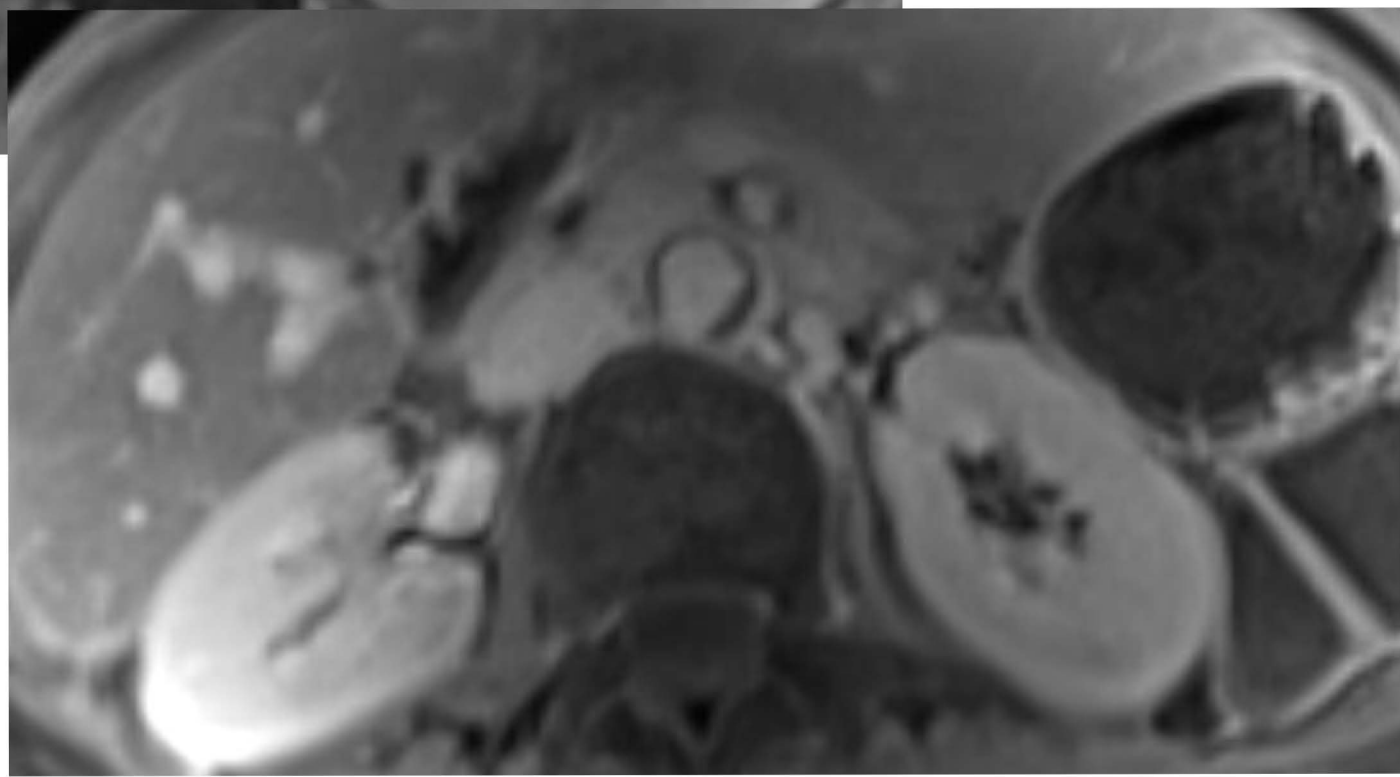








Clinical presentation

- 57 year old Caucasian female with complicated PMHx including discoid lupus and stage III rectal CA s/p multiple bowel resections presenting with progressive weight loss of 60 lbs and failure to thrive
  - Due to extensive surgical history and with question of short bowel syndrome in the setting of copious ostomy output→ CT abdomen and pelvis
  - CT demonstrated thrombus throughout the aorta (nonocclusive)→ f/u MRA
  - MRA demonstrated intraluminal irregularity consistent with mural thrombus in the ascending thoracic aorta descending thoracic aorta and abdominal aorta with concentric wall enhancement in the suprarenal abdominal aorta consistent with aortitis
  - Patient later developed nonspecific transient waxing and waning headaches
  - No visual changes or neurologic symptoms
  - Temporal artery biopsy

Differential diagnoses

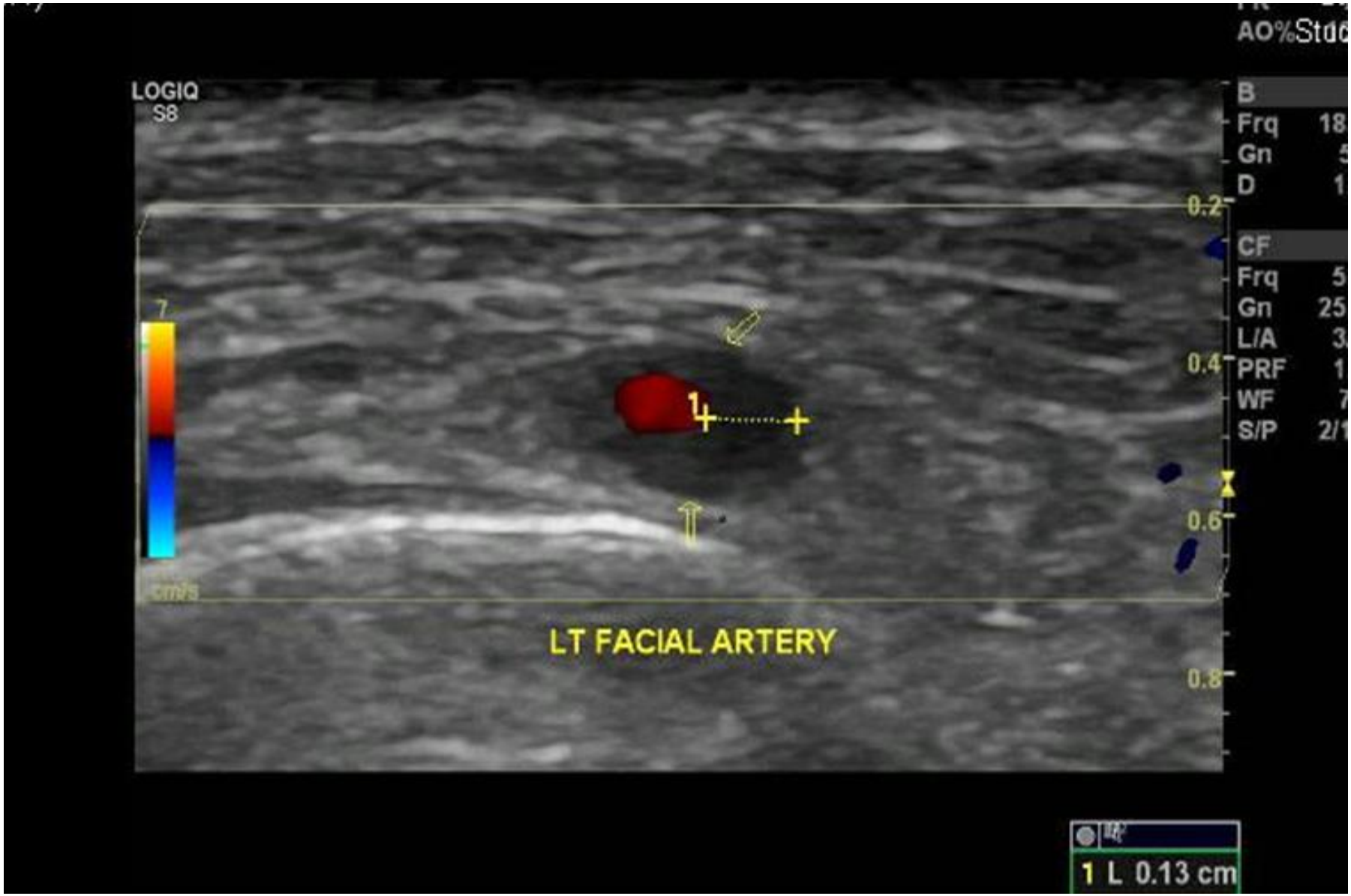
- Giant cell/temporal arteritis (>50s)
  - Predominantly Northern Europeans/ F>M
- Takayasu arteritis (<30s)
  - Predominantly in Asians/ aortic occlusion highly specific
- Atherosclerotic disease/penetrating atherosclerotic ulcer/thrombus
- Other systemic vasculidities: Polyarteritis nodosa, syphilitic aortitis
  - Often small and medium vessel
- FMD
- PMTA

Giant cell arteritis

- Medium and large vessel inflammatory disease of the tunica media
- Clinical presentation: low grade fever, weight loss, transient neurologic symptoms, jaw claudication, headache, temporal tenderness, visual disturbance
- Gold standard Dx: pathologic diagnosis
- As name implies, typically involves extracranial carotid vessels (superficial temporal) and rarely involves the thoracic/abdominal aorta, coronary, vertebral, subclavian/axillary, and other medium/large vessels resulting in stenosis/occlusion
- Up to 18% demonstrate aortic involvement

- Imaging:
  - CTA: Circumferential mural thickening with stenosis/occlusion; may not demonstrate wall enhancement
  - MRA: Similar findings to CTA with irregularities within the lumen, mural thickening, varying stenosis/occlusion, collateralization
    - T1: Can make mural thickening more conspicuous
    - T2: Edema=T2 intense= active inflammation
    - STIR can be more sensitive for detection of mural edema than T2
    - T1 C+: wall thickening with mural enhancement
      - More enhancement= increased inflammation
  - US: grayscale US of the superficial temporal artery may demonstrate halo sign (edema within inflamed vessel) or color Doppler US demonstrate luminal stenosis
  - Echo: Can have AV involvement so can assess for cardiac involvement
  - Angio: Multisegmental smooth luminal narrowing
  - PET: Can demonstrate FDG uptake if with active inflammation
    - Potential to assess for extravascular involvement as in polymyalgia rheumatic
      - 50% develop PMR
    - Evaluation for response to therapy
- MRA preferred over CTA however can still be used to exclude acute complications (dissection, aneurysm, occlusion)

- Complications: 17 fold risk thoracic aortic aneurysm, 2 fold risk AAA, aortic dissection, and MI
  - Use CTA to exclude such complications as aneurysm, dissection, intramural hematoma, acute luminal occlusion, or coronary stenosis
- Treatment: Steroids



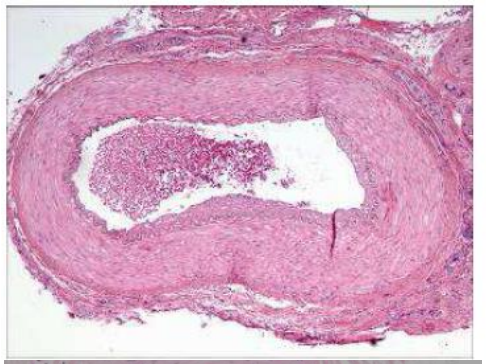
Courtesy of Magda Abdou

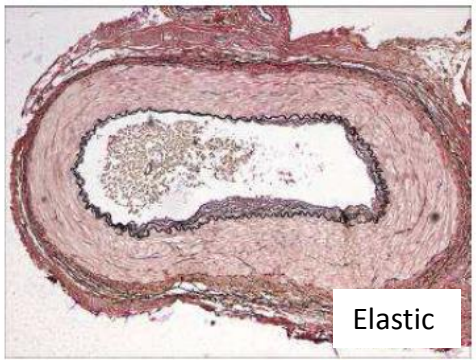
TEMPORAL ARTERITIS

- Pathologic findings:
  - Granulomatous inflammation of arteries: *cells are mainly chronic inflammatory cells including lymphocytes, macrophages, and foreign body giant cells*

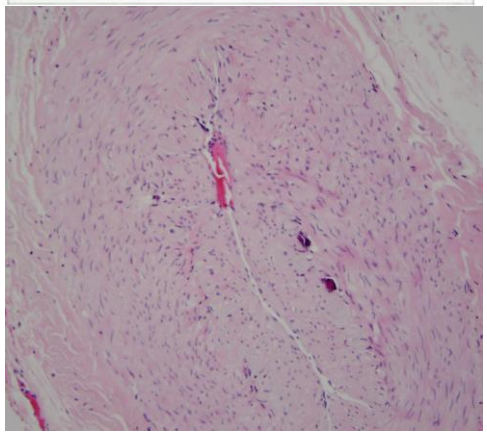
- Discontinuous and fragmented internal elastic lamina: *the inflammatory infiltrate in this disease seems to center on the IEL after making its way through the media*

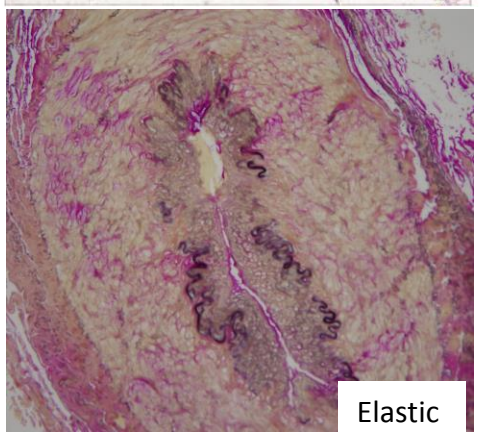
Normal

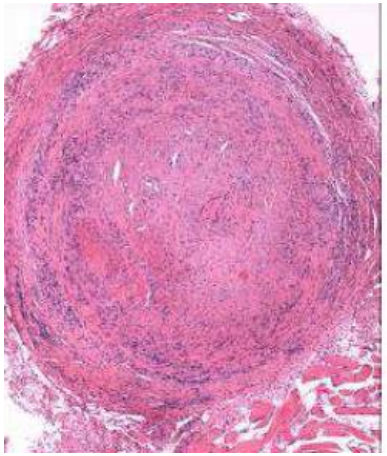


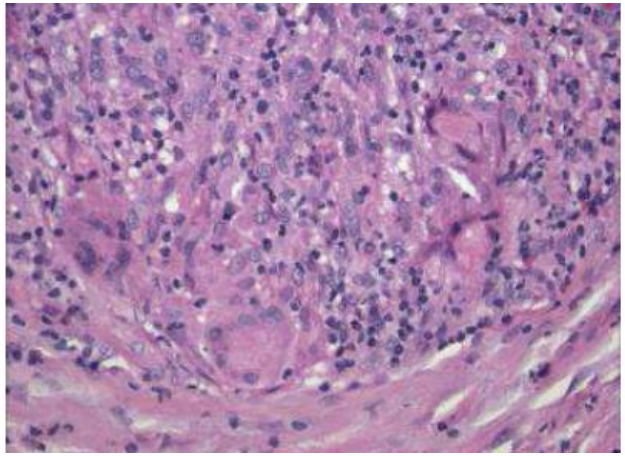


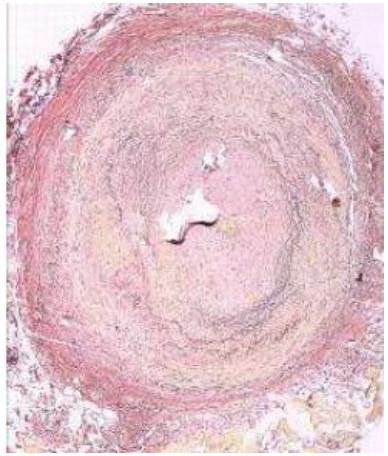
Patient



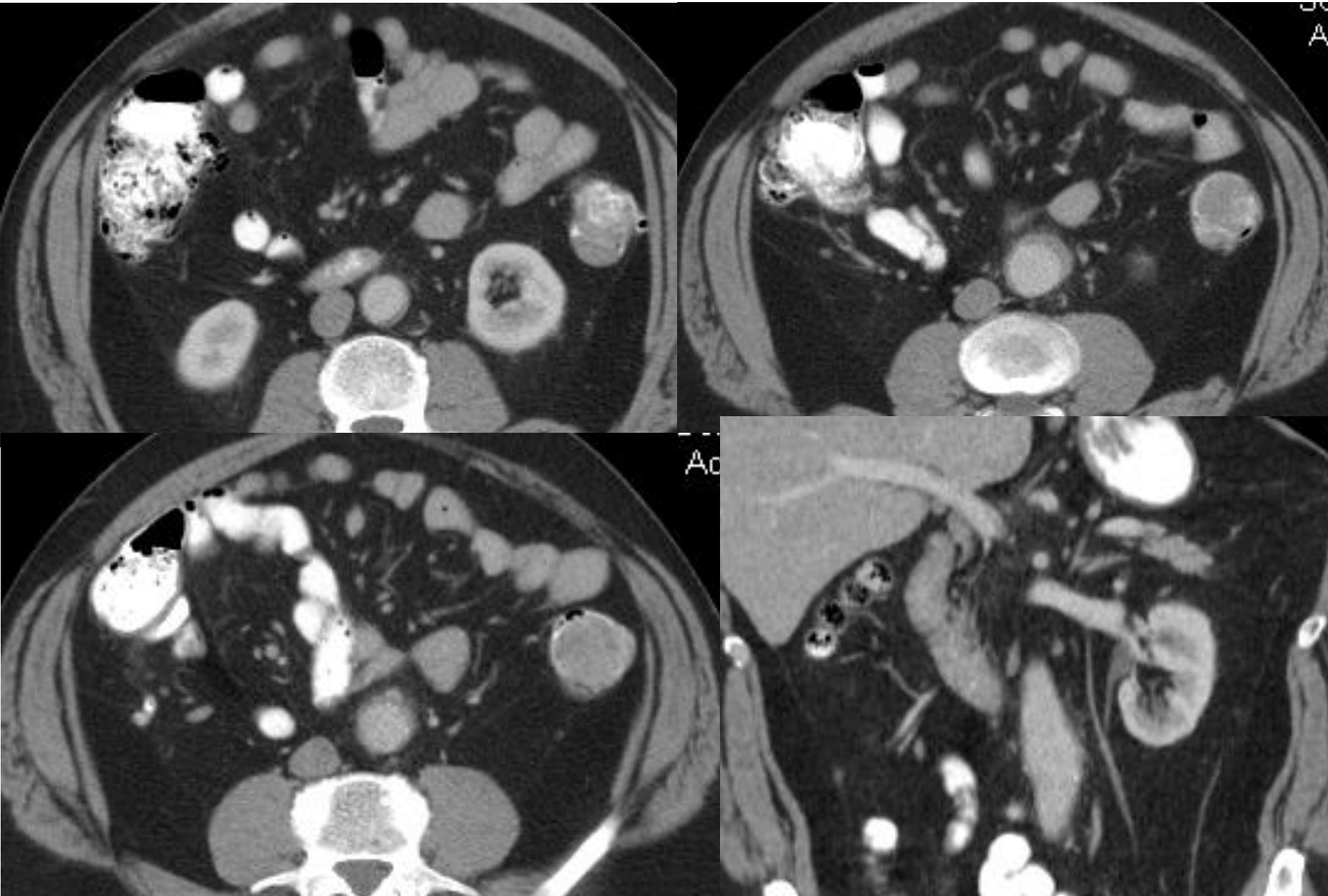


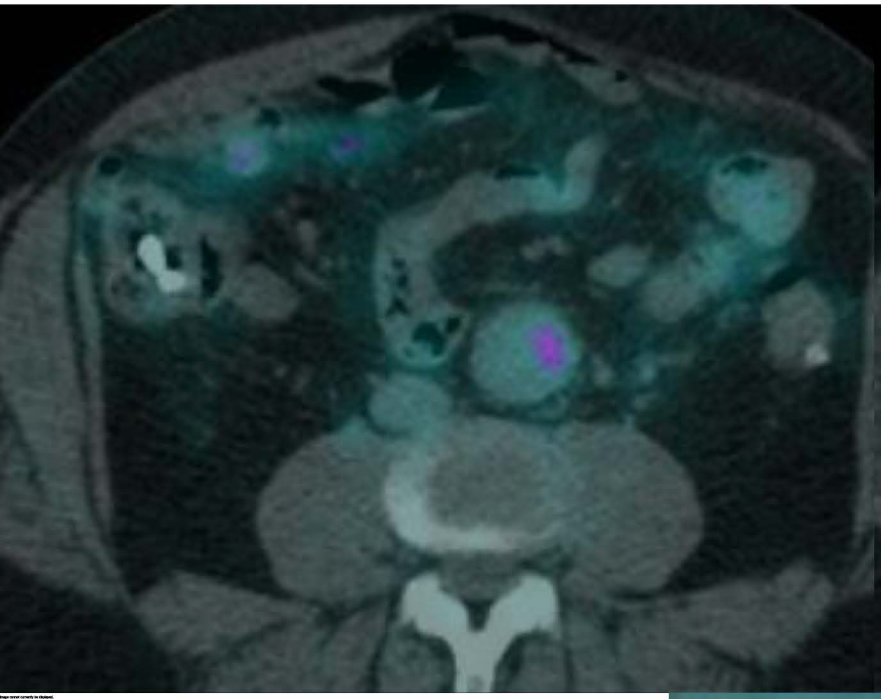


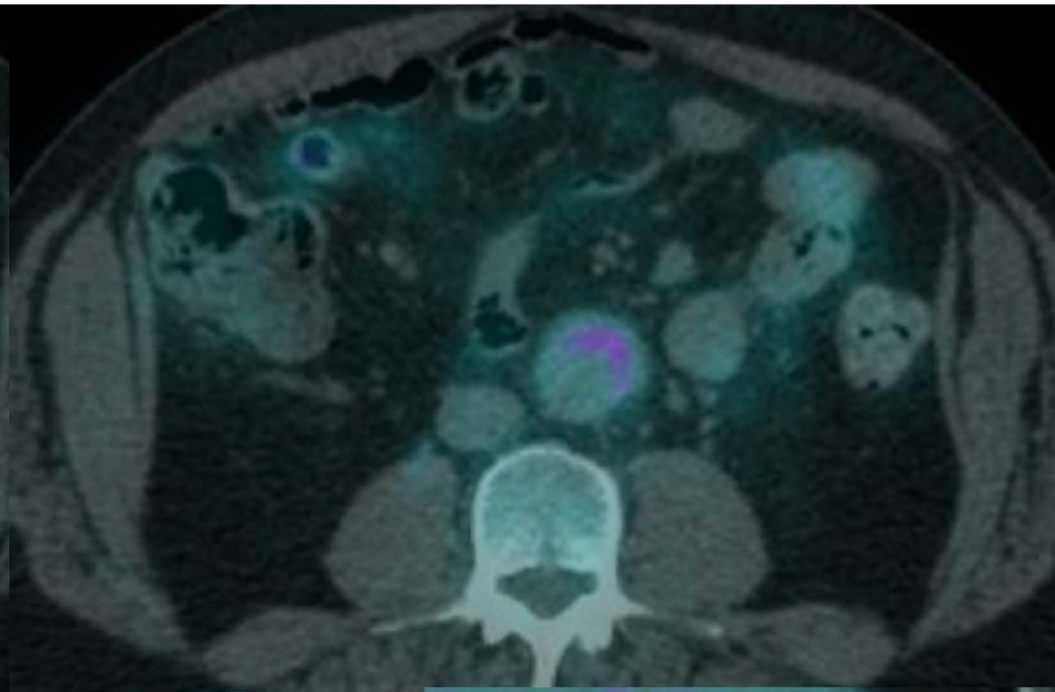




Case 4







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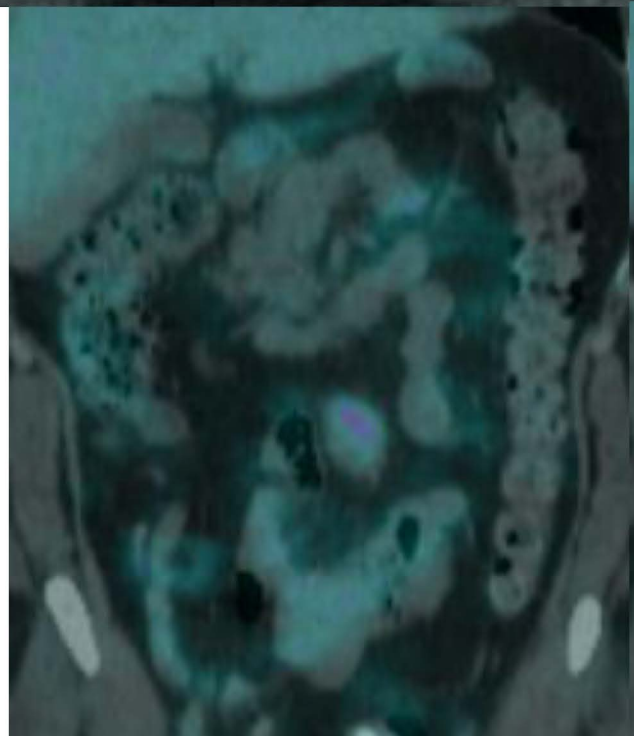
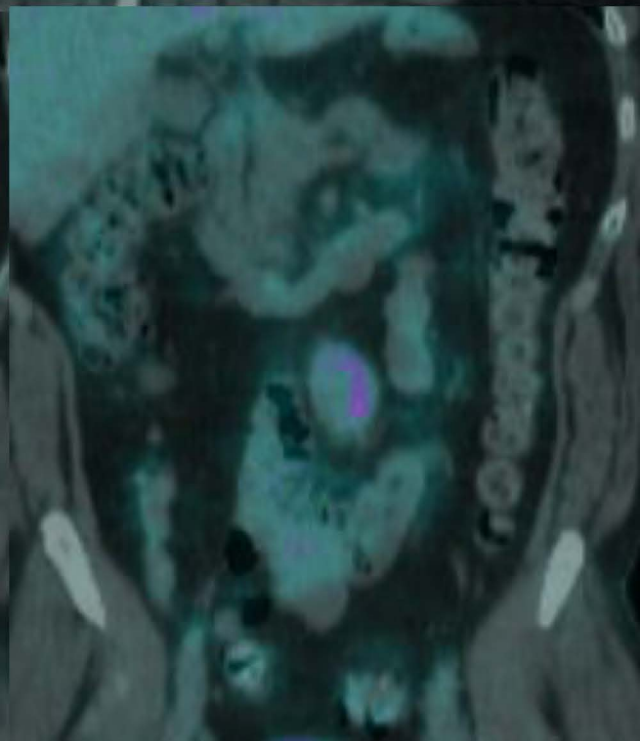




Clinical history

- 56-year-old male with history of diverticulitis s/p Abx presenting with worsening LLQ pain of 3 week duration
- On repeat CT: found to have fat stranding and wall thickening in the abdominal aorta with focal area of ectasia concerning for aortitis→ PET which demonstrated focal uptake
- PE essentially normal
- ESR 31 and CRP 30
- Resection/graft repair

Differential Diagnoses

- Infectious aortitis
  - Bacterial
  - TB
  - Syphillis
  - Mycobacterial
  - Viral (HIV)
  - Aortic graft infections
- Noninfectious aortitis
  - Rheumatic disease
    - Takayasu
    - Giant cell
    - Ankylosing spondylitis
    - RA
    - SLE
    - Behcet
  - Idiopathic
    - Idiopathic aortitis
    - Inflammatory
    - RP fibrosis/fibrosing mediastinitis
  - Radiation induced aortitis
- Atherosclerotic aneurysm
  - Fusiform
- Inflammatory aneurysm
  - Fusiform
- Infectious aneurysm
  - Saccular
- Intramural hematoma
- Contained rupture
- Aortoenteric Fistula

Mycotic aneurysm

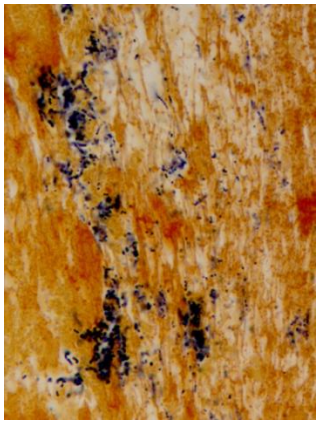
- AKA infectious aneurysm: focal eccentric outpouching secondary to infection/inflammation
  - Represents 0.7-2.6% all aortic aneurysms
  - Infectious aortitis leads to weakening of walls with contained rupture and pseudoaneurysm formation
  - Most commonly bacterial (staph aureus and salmonella)
  - Predisposing conditions: Atherosclerosis, DM, immunosuppression, grafts, IV catheter, alcoholism, and neoplasms
  - syphillis- usually affects ascending aorta but spares sinuses of Valsalva
  - Primary: arise from distant unknown or remote source of infection
  - Secondary: arise from specific source of infection (endocarditis/TB)
  - Infrarenal abdominal aorta most common followed by descending thoracic aorta
  - Mortality ~67%
- Clinical features: Fever, pain, nonspecific symptoms
  - Blood cultures are negative in 25% of cases and acute rupture is seen in 75% of cases

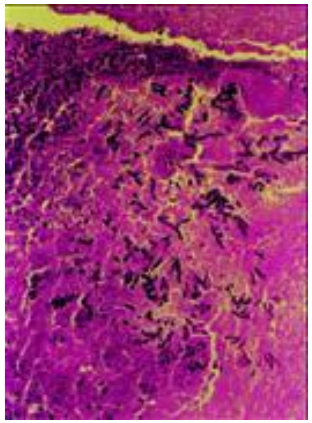
- Imaging
  - Predominantly in branch points
  - Rapidly growing focal saccular outpoutching
  - Surrounding findings: inflammation, abscess, gas, vertebral body abnormalities
  - CT:
    - Noncon: periaortic soft tissue stranding/edema, gas, vertebral body abnormalities
      - Syphillis: calcifications
    - Con-enhanced: Focal saccular aneurysm with lobular contours, periaortic enhancement
  - MR
    - Periaortic T1 hypo and T2 hyperintensities
    - Aortic and periaortic enhancement with contrast
- Complications: Acute rupture in 75%
- Treatment: Resection and grafting, antibiotics

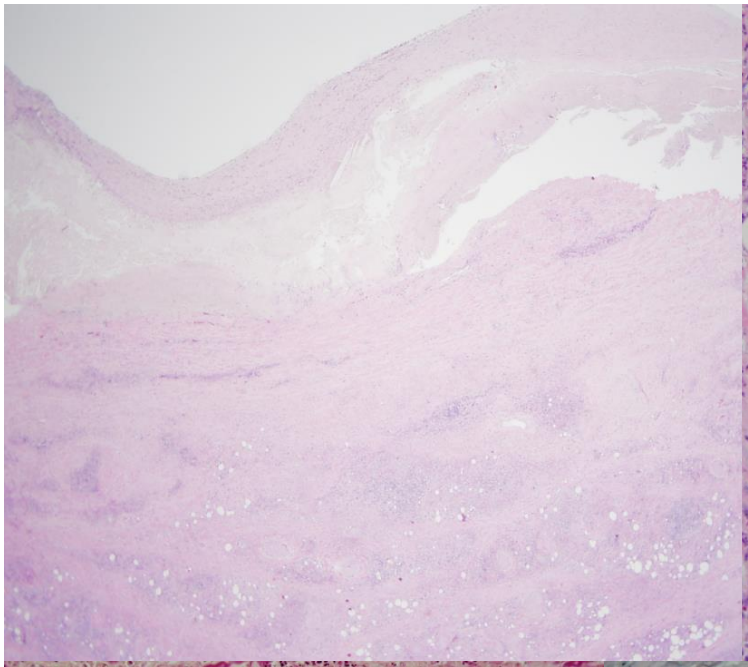
MYCOTIC ANEURYSM

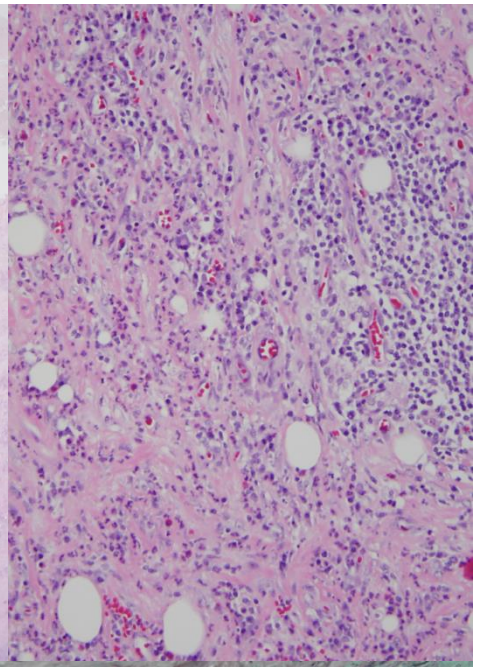
- Septic embolus lodges at branch point of an artery and infects surrounding tissue which causes weakness (and aneurysm)
- Infectious: Fungi not the usual culprits (though infection with aspergillus & mucormycosis can occur)

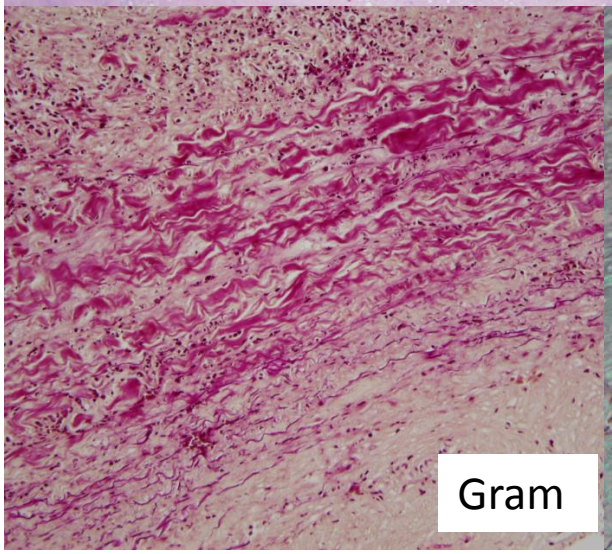
- Pathologic findings:
  - Often seeds atherosclerotic plaques
  - Neutrophilic infiltrate involving media with extension into adventitia

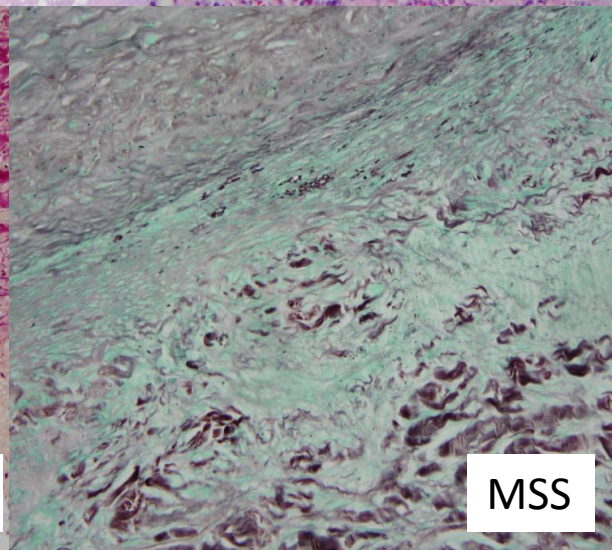












Take home points

- Although rare, primary malignancies of the aorta exist and should be considered in the differential
  - Thrombus may not be all thrombus! Think of intimal sarcoma!
  - Look for evidence of perivascular inflammation or enhancement
  - If concerns for malignancy, consider MRA or PET to differentiate between thrombus and soft tissue mass
- With idiopathic splenomegaly or anemia/pancytopenia without evidence of lymphoma, leukemia, or myelofibrosis consider splenic angiosarcoma
- PMTA and vasculitis may demonstrate periaortic enhancement
- Mycotic aneurysms are more often bacterial in origin

Special thanks to

- Dr. Angela Giardino
- Julia Seol
- Magda Abdou

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