

Persistent Aortic Sac Expansion following EVAR Secondary to Malignancy

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Disclosures

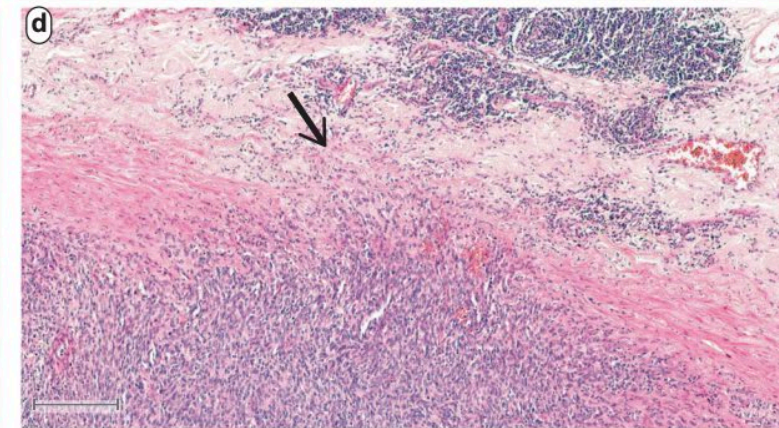
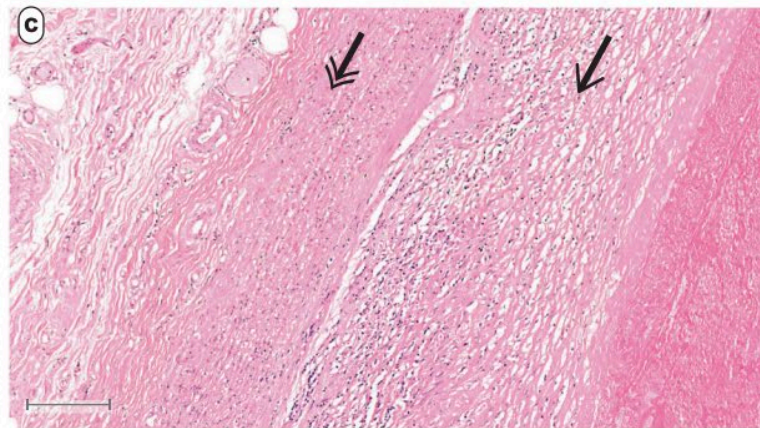
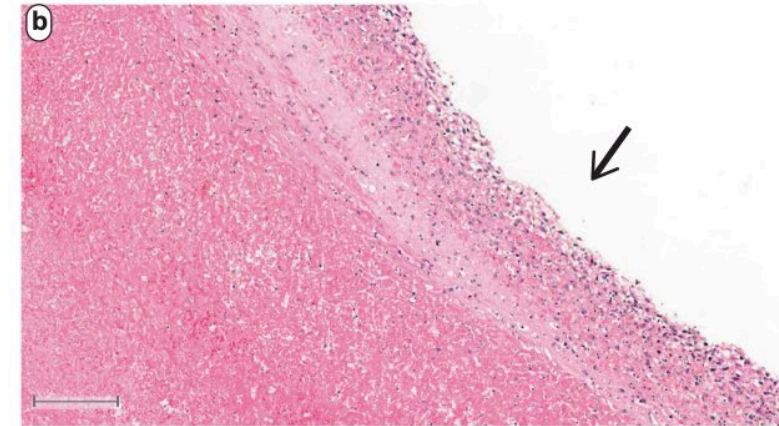
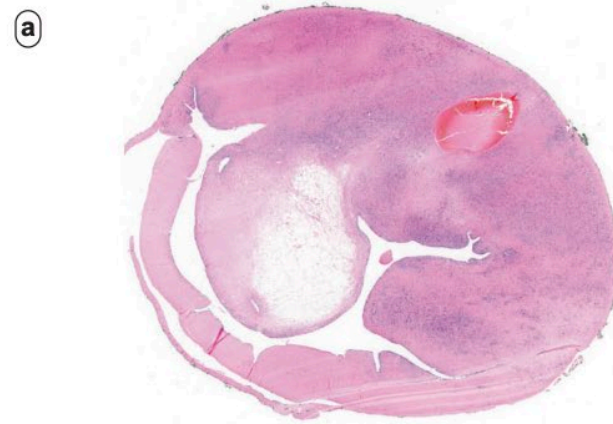
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- We have no disclosures otherwise

Introduction

- Intimal sarcomas
 - Incidence of 0.0014 per 100,000
 - Composes 0.9% of malignant sarcomas of the aorta
 - Most Common:
Angiosarcomas at 67.7%
- Other malignant aortic tumors include: metastases, leiomyosarcomas, fibromyxosarcomas, myxosarcomas, fusiform cell sarcomas, fibrosarcomas, malignant fibrous histiocytomas
- Benign aortic tumors include: myxomas, fibromyxomas

Introduction- Presentation

- Symptoms of intimal sarcoma depend on the location of the lesion
 - Cardiac may mimic heart failure
 - Pulmonary artery may mimic pulmonary emboli
- Computed tomography (CT) may be suggestive of sarcoma
- Pathology is critical to the diagnosis as imaging findings often are usually not confirmatory



Introduction- Management

- Surgery, radiation, and chemotherapy have all been attempted interventions with no superior regimen
 - Surgical options include:
 - Endarterectomy
 - Replacement
 - Excision and extra-anatomic reconstruction
 - Chemotherapy regimens include alkylating agents, anthracyclines, tyrosine kinase inhibitors
- Median survival < 2 years

Case Presentation

- 65 year old Male
- CC: Fever and back pain
- PSH: EVAR
- PMH: Hypertension, insulin dependent diabetes, sleep apnea, COPD, tobacco use



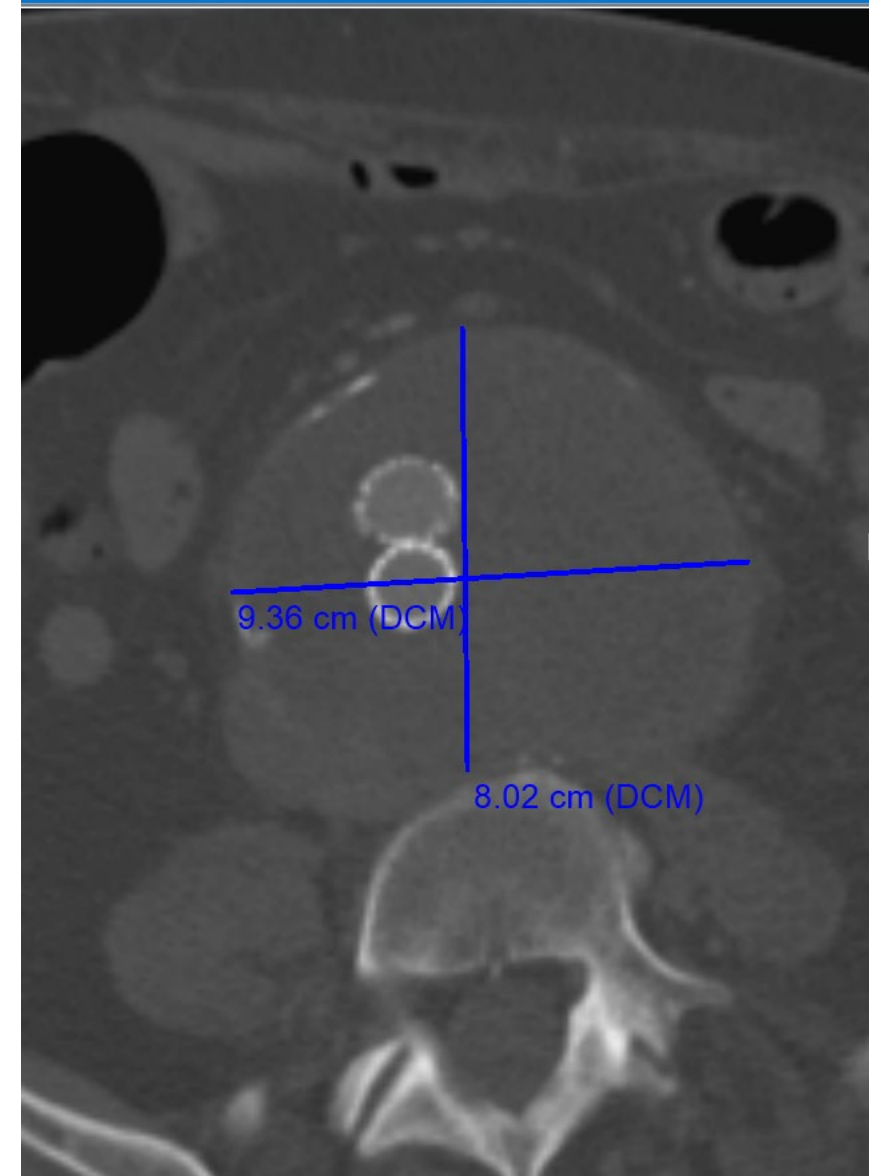
Case Presentation

- Blood Cultures: Bacillus
- Infectious Disease consult
 - 6 weeks of Ceftriaxone
- Interventional Radiology consult
 - Translumbar IMA embolization



Case Presentation

- ED: abdominal pain
 - Unchanged exam
- CTA: aneurysm sac expansion to $>9\text{cm}$
 - No clearly visible endoleak
 - Perianeurysmal stranding concerning for infection

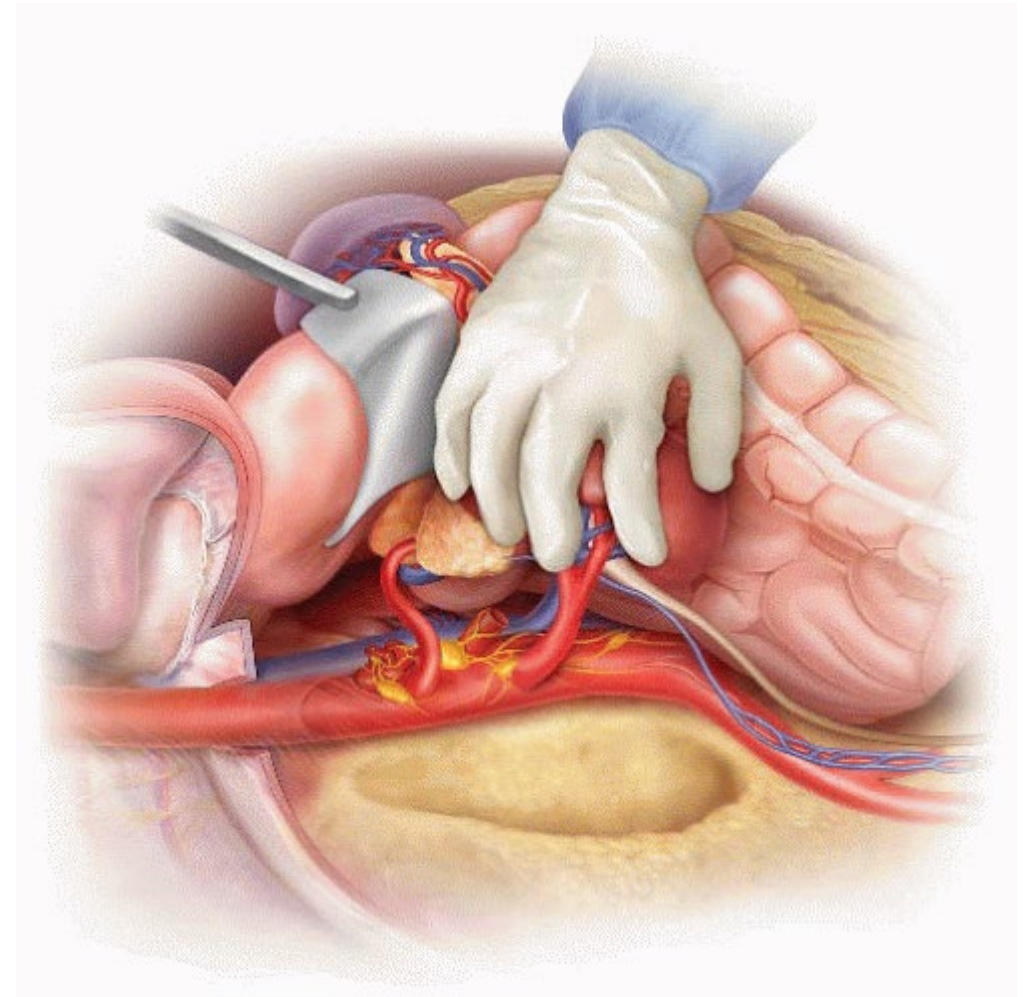


Case Presentation

- Blood cultures negative
- ID: continue Ceftriaxone
- IR: aspiration of aneurysm sac
- Culture (-) for anaerobic/aerobic/fungal culture

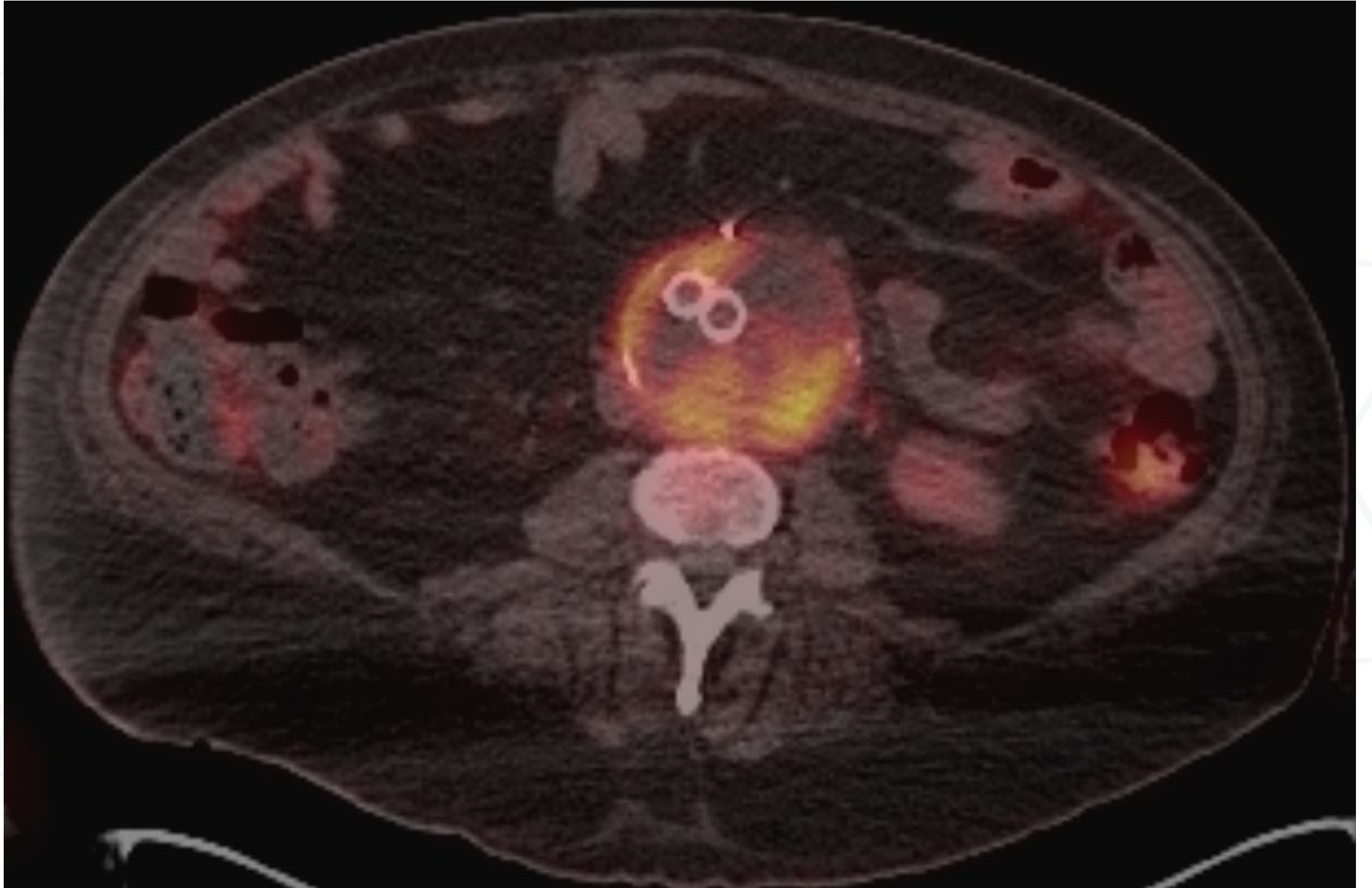
Case Presentation

- Offered an open exploration and possible explantation
- Left retroperitoneal exposure performed without gross signs of infection evident
- Well-organized, firm thrombus was removed, and sent for culture and pathology
- IMA suture ligated, and aneurysm sac closed



Case Presentation

- Cultures negative but invasion into the aorta present
 - Grade 3 sarcoma without any positive stains but fibrin deposition and necrosis consistent with an intimal sarcoma
- Oncology consulted
 - Started on pazopanib, a tyrosine kinase inhibitor
- PET-CT 6 weeks postoperatively



Discussion

- Given the scarcity of this process (0.0014 per 100,000), risk factors have not yet been determined
- Previous cases have occurred in the setting of endografts, but this does not necessarily determine causation
 - More frequent cross-sectional imaging
- Aortic intimal sarcomas are most commonly abdominal consistent with our patient

Discussion

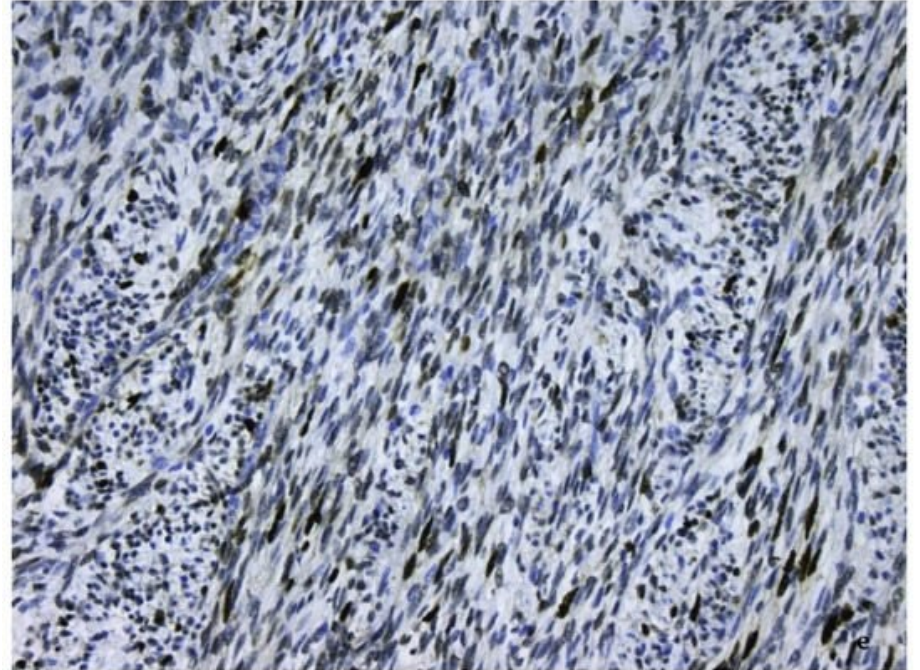
- Location determines symptoms
 - Arch may present with stroke
 - Mesenteric vessels may present with ischemic bowel
 - Distal (near iliac arteries) may present limb ischemia
 - Others have nonspecific symptoms similar to our patient
- Maintaining endoleak, infection, and rheumatologic etiologies in the differential for sac expansion and inflammation is prudent until sarcoma is pathologically confirmed

Discussion

- Blood cultures, CBCs, and endovascular sampling are beneficial to first rule out infection
- CTA is the best imaging modality for possible malignancies of the abdominal aorta, but is not confirmatory
- MRI may be useful in thoracic aorta with potential cardiac involvement, and PET-CT may be useful for staging

Discussion

- Pathologic differential includes other sarcomas, myxomas, and metastases and this is predominately determined by staining including MDM2, which is present in 70% of cases
- Our patient differs in that MDM2 was negative, but he was very poorly differentiated.



Discussion

- Surgical options include: endarterectomy, aortic replacement, or excision and extra-anatomic reconstruction
- Unclear if neoadjuvant or adjuvant chemotherapy is more beneficial as most are diagnosed postoperatively
- Chemotherapy regimens include alkylating agents, anthracyclines, or tyrosine kinase inhibitors
- Utility of radiation has yet to be determined

Discussion

- Patients with embolic disease often receive anticoagulation, but it is unclear if this is beneficial
- Prognosis is generally poor as is seen in our patient



Questions?

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