

Beating Heart, Aggressive Sarc: case-study of an atrial angiosarcoma

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Background:

One of the rarest types of cancers are cardiac tumors, with only 20-30% of the cardiac neoplasms being malignant. Of the most common cardiac malignant mass is an angiosarcoma, a mass arising in the endothelial lining of blood and lymph vessels. Angiosarcomas are rare, encompassing only 2% of the soft tissue sarcomas and particularly affecting elderly white men. The tumor is particularly aggressive with difficulty of obtaining clear margins due to its infiltrative nature and early systemic metastasis. Considering this, cardiac angiosarcomas are especially difficult due to the presence of the mass, typically arising in the right atrium (RA). Epidemiological manifestations of the cardiac angiosarcomas consists of pericardial effusion, heart failure, arrhythmias, obstruction of the vena cava, and cardiac tamponade. The location of the tumor can also lead to invasion into the pericardium and other surrounding heart structures (right ventricle, superior vena cava, and tricuspid valve). Patients with history of radiation treatment or chronic lymphedema have a higher risk factor angiosarcomas. While the survival rate of angiosarcoma is approximately 30%, the location of the mass along, early metastases, and late detection can all lead to worse prognosis. Typically, a multidisciplinary approach with radical resection along with potential chemotherapy and radiotherapy is considered for treatment.

Clinical history:

A 32-year-old with no history of cancer or radiation treatment presented to the Emergency Department (ED) in November 2025 with weeks of progressing flu-like symptoms, including shortness of breath, fever, dry cough, and fatigue, complaining that his chest felt heavy. The patient is a non-smoker, who rarely drinks alcohol, with a significant history of lipoma removal (arms, torso, and scrotum). COVID, flu, and RSV tests were negative. An electrocardiogram (ECG) proved the heart was non-ischemic. The positron emission tomography/computed tomography (PET-CT) scan showed the right ventricle was normal in size with a non-mobile electron density on the right atrial (RA) free wall. A computed tomography angiography (CTA) of the chest revealed a 4.1 x 3.5 cm lobulated intermediate mass-like structure in the lumen of the RA, associated with the anterior wall and abutting the tricuspid valve. Additionally, moderate pericardial effusion was identified. Concern for potential neoplasm and possible invasion into the adjacent pericardium motivated the cardiac team to proceed with surgical intervention.

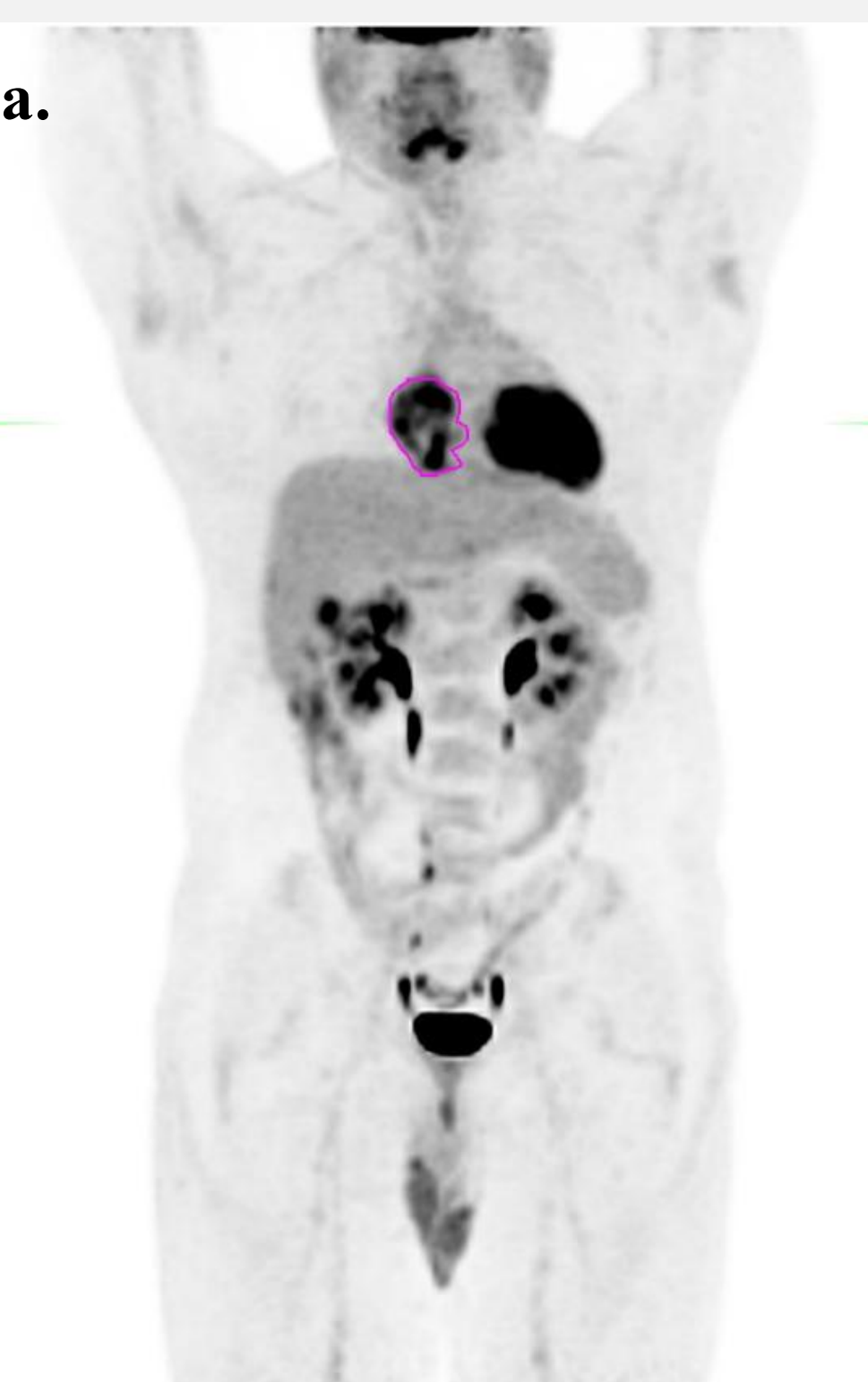


Figure 1 PET-CT with atrial mass outlined in pink **1a.** coronal plane **Figure 1b.** axial plane.

Methodology:

The Cleveland Clinic cardiac surgical team removed a portion of the RA with the mass as well as fragments of the pericardium, epicardial rind, and a chest lipoma. The team then repaired the tricuspid aortic valve. In Surgical Pathology, the specimen was differed from the cardiac to the soft tissue subspecialty due to the possibility of a mass being an angiosarcoma. After reviewing orientation with the Soft Tissue team, the Pathologists' Assistant (PA) performed the gross examination after overnight fixation in 10% neutral buffered formalin.

Results:

Gross dictation:

Received in formalin labeled as "right atrial tumor" is a segment of the right atrium measuring 7.7 x 4.7 x 4.5 cm. Per the specimen requisition "stitch is marked atrial border". The epicardial surface is tan-red and roughened. Abundant epicardial fat is present and the myocardium is pale, measuring 0.3 cm in thickness. A roughened possible soft tissue resection margin is appreciated and inked blue. The "atrial border" is inked orange. The pericardial soft tissue is inked black. Within the endocardium is a blue-gray multinodular bulging mass with underlying firmness, overall measuring 7.1 x 4 x 3.8 cm. There are two bulging areas on the atrial surface measuring 3.3 x 3.2 x 2.4 cm and 3 x 2 x 2 cm. The mass approaches within 0.1 cm of the nearest possible soft tissue margin, 0.2 cm from the pericardial surface, and 1 cm of the nearest atrial border. The cut surfaces of this lesion is red, hemorrhagic, and slightly cystic. Photographs are attached to the case. This case is reviewed with Dr. Smith. Representative sections are submitted as follows:

- D1 lesion to atrial border, perpendicular section;
- D2 remainder of atrial border, shaved;
- D3-D5 lesion to possible soft tissue margin to include epicardium, perpendicular sections;
- D6 lesion to uninvolved atrium with epicardium;
- D7 larger bulging site of lesion to soft tissue margin, perpendicular section;
- D8 smaller bulging site of lesion to soft tissue margin, perpendicular section;
- D9 lesion to pericardium;
- D10 lesion to pericardium with uninvolved atrium.

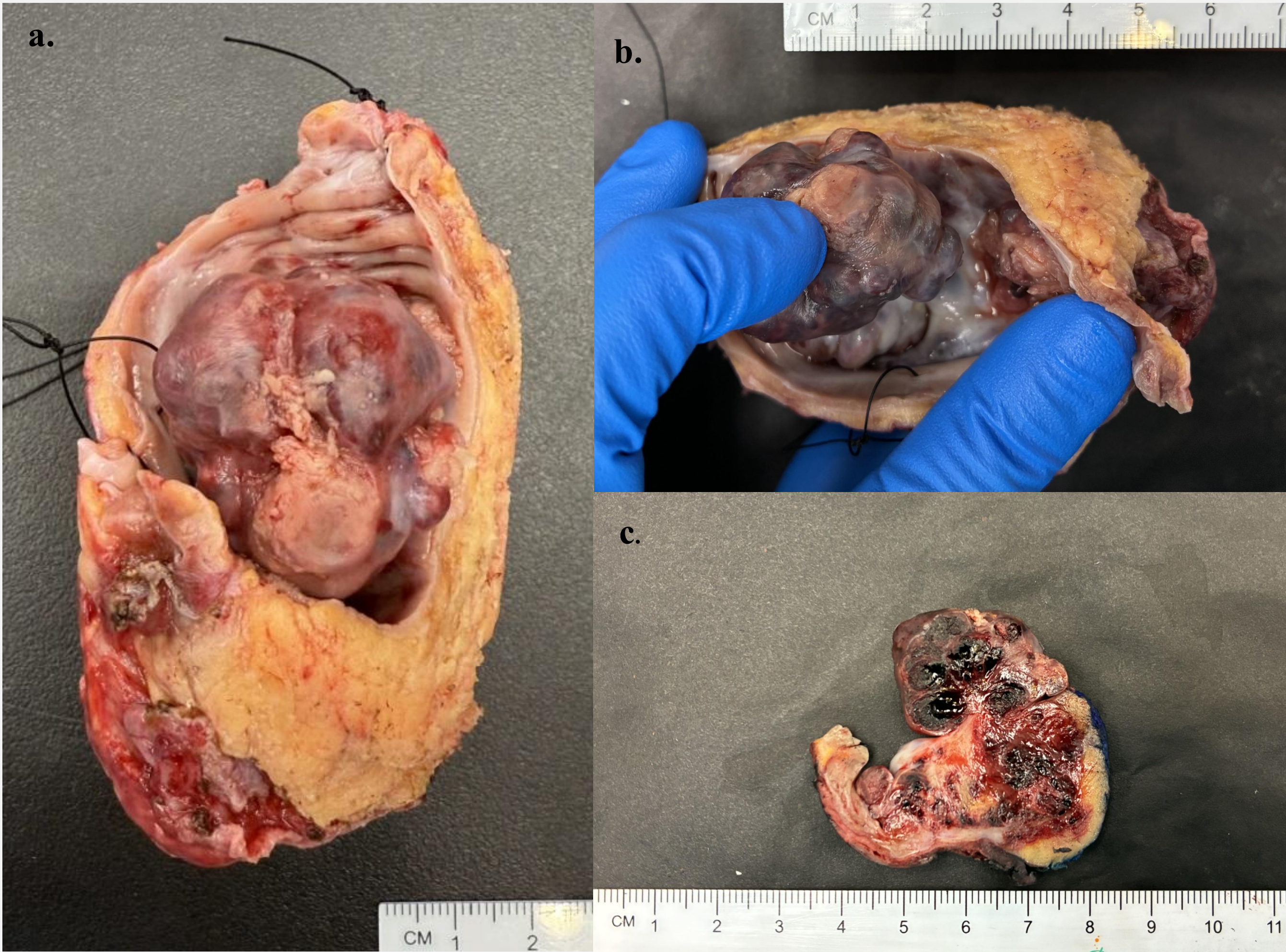


Figure 2. Gross images **2a.** overall specimen **2b.** Atrial surface **2c.** Cross-section

The final diagnosis of this case is angiosarcoma, with the tumor extending to the inked epicardial soft tissue resection edge. Additional tumor infiltration is identified in both the pericardium and the epicardial rind excision of the case.

References:

- Kim, E.J. and Kim, H. K. (2023) Current understanding of angiosarcoma: disease biology and evolving treatment. *Archives of Craniofacial Surgery*. 24(5):203-210. doi: 10.7181/acfc.2023.00409
- Guo Y, Liu Q and Wu H (2023) Primary cardiac tumor: a case report of right atrial angiosarcoma and review of the literature. *Frontiers in Oncology*. 13:1164153. doi: 10.3389/fonc.2023.1164153
- Liu, C., Zhao, Y., Yin, Z., et al. (2018) Right atrial epithelioid angiosarcoma with multiple pulmonary metastasis confirmed by multimodality imaging-guided pulmonary biopsy: a case report and literature review. *Medicine*. 97(30):p e11588. doi: 10.1097/MD.00000000000011588
- Luong, HTT, Vercammen, S, de Marco, A., de Rooster, H. and Cosma, A. (2025) Angiosarcoma: a systematic review of biomarkers in diagnosis, prognosis, and therapeutic strategies. *Frontiers in Oncology*. 15:1623327. doi: 10.3389/fonc.2025.1623327

Discussion:

The case was signed out with a pT2, M1 following the American Joint Committee on Cancer (AJCC) Sarcomas of the Abdomen and Thoracic Visceral Organs staging. The pT2 staging was due to the mass measuring >5cm and ≤10cm. In this case, the pT2a or pT2b staging was not utilized since the serosa or visceral peritoneum (pT2a) or mesentery (pT2b) were not applicable for this specimen. The mass extension to the pericardium and soft tissue indicated metastasis. Atypical cells with hyperchromatic features and cell layering were appreciated in the slides, typical of aggressive angiosarcoma. The slides also included 20 mitoses per 10 high-power fields with 40% necrosis, further supporting the mass was of a vascular origin. A clean line of distinction between the tumor and normal tissue was not identified, indicating the mass to be infiltrative. Immunohistochemistry (IHC) stains ERG, CD31, and CD34 were all positive for the mass. All three of these markers are highly expressed in angiosarcoma cases, however, to confirm a mass is an angiosarcoma, a positive stain of only two out of the three is sufficient.

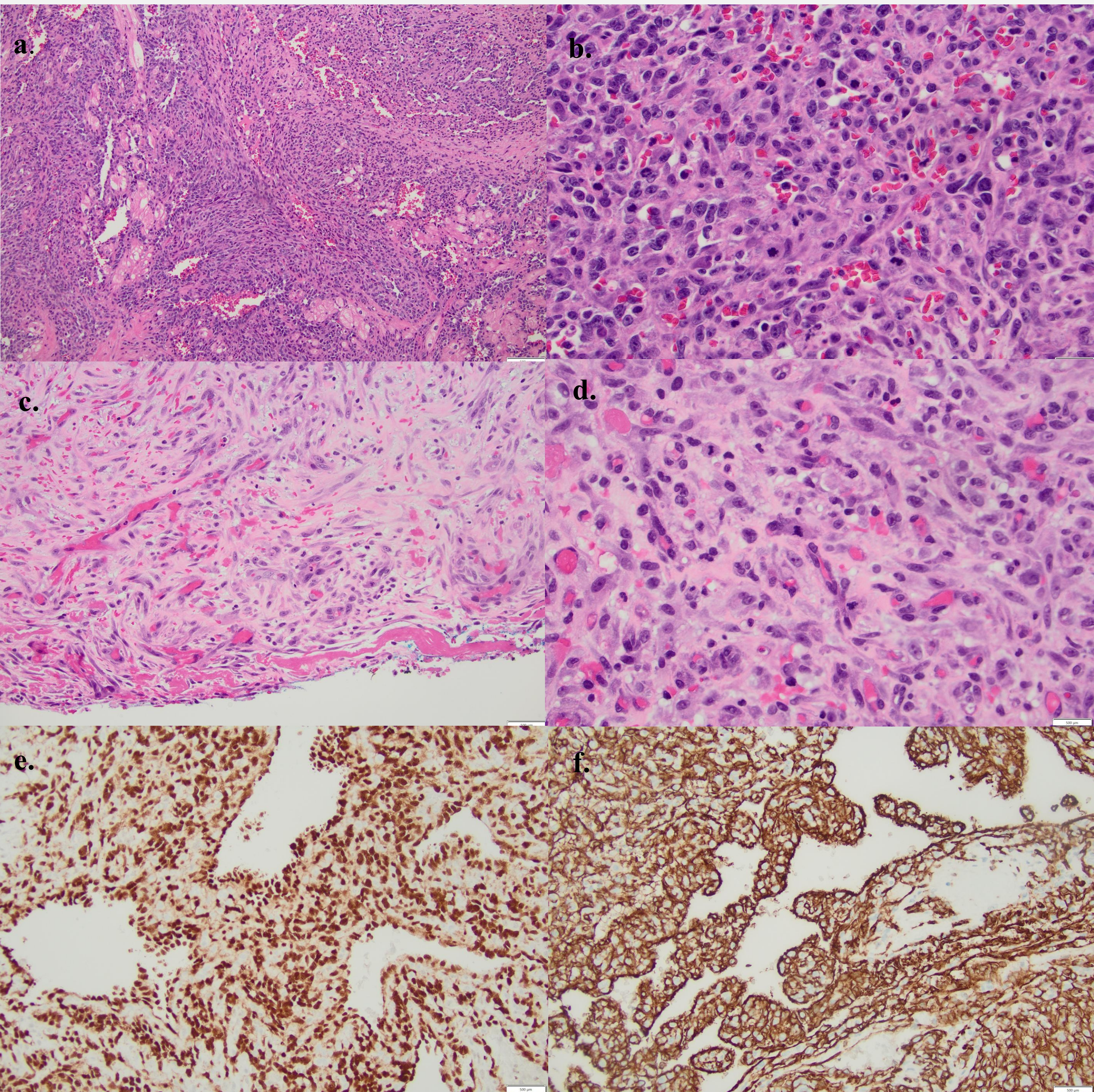


Figure 3. **3a.** Angiosarcoma to include atypical cell patterns with hyperchromatic expression, multiple layering of cells, vascular formations **3b.** Mitotic features and some hobnail architecture with the cells pushing into the lumen **3c.** Extension to blue inked soft tissue margin **3d.** Mass extension in pericardium part of the case **3e.** Positive ERG stain **3f.** Positive CD31 stain

Conclusion:

The aggressive, infiltrative nature of this tumor makes it important for the PA to carefully examen the specimen, the pathologist to confirm the mass is of an angiosarcoma by using IHC, and for the clinical team to consider a multidisciplinary approach. Since the processing of this case, multiple other areas of metastases have been identified, including the liver, brain, lungs, vertebra, and sacrum. The patient had to undergo whole brain radiation and is undergoing chemotherapy.

Acknowledgements

I would like to thank Dr. Josephine Dermawan for revieing this case with me.