

Late Diagnosis of Undifferentiated Grade 2 Pleomorphic Sarcoma Involving the Spine and Encasing the Aorta. Delayed Diagnosis of Primary Aortic Pleomorphic Sarcoma Initially Misdiagnosed as a Mycotic Aneurysm

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ABSTRACT

Summary

Background: Primary aortic sarcomas are exceptionally rare malignancies and are frequently diagnosed late because of their non-specific clinical and radiological presentation.

Case presentation: A 56-year-old man initially investigated for a presumed mycotic thoracic aortic aneurysm underwent emergency thoracic endovascular aortic repair. Despite prolonged antimicrobial therapy, his clinical and radiological findings progressed. Subsequent imaging and tissue biopsy confirmed an undifferentiated pleomorphic sarcoma involving the thoracic aorta and adjacent vertebrae.

Outcome and follow-up: The patient was treated with palliative radiotherapy followed by single-agent doxorubicin chemotherapy, achieving a partial response before later disease progression.

Conclusion: This case highlights the diagnostic challenge of distinguishing primary aortic sarcoma from infective aortic pathology and emphasizes the importance of reconsidering the diagnosis when expected clinical response is not achieved.

Keywords: Primary aortic sarcomas; Palliative radiotherapy; Aortic pathology

Background

Primary aortic sarcomas are exceptionally rare and highly aggressive malignant tumors arising from the aortic wall, accounting for less than 1% of all sarcomas¹⁻⁵. Because of its rarity and non-specific clinical presentation, diagnosis can be

delayed or mistaken for more common vascular conditions such as mycotic aneurysm, thrombus or severe atherosclerotic disease⁶⁻¹².

Multimodal imaging, including CT angiography, MRI and FDG PET-CT, may help to identify suspicious

features of malignancy¹³⁻¹⁵. However, definitive diagnosis relies on histopathological examination with appropriate immunohistochemical evaluation¹⁶⁻²⁰.

Due to the aggressive behavior of aortic sarcoma and frequent delay in diagnosis, prognosis remains poor in many reported cases.²¹⁻²⁴ Early recognition and prompt tissue diagnosis are therefore essential for timely management²⁵⁻²⁸.

We report a diagnostically challenging case of primary aortic undifferentiated pleomorphic sarcoma initially managed as a mycotic aneurysm because of overlapping clinical and radiological features.

Case Presentation

A 56-year-old man presented in January 2023 with acute chest and back pain and was found to have a ruptured thoracic aortic aneurysm. CT angiography demonstrated erosion of the posterior wall of the supradiaphragmatic aorta, 98 mm in diameter, with tracking in the paravertebral space, with Rt sided pleural effusion, features thought to be consistent with a mycotic thoracic aortic aneurysm. Bloods on admission showed C-RP level of 32/mg/dL (0-5 mg/dL), Albumin 26 g/L (35-50 g/L), Ca 2.64 mmol/L (2.2-2.6 mmol/L), ALT 63 U/L (0-41 U/L), WBC 11.8X10⁹ (4.3-11.2X10⁹/l), Platelets 539x10⁹/L (150-400 x 10⁹/L). Blood cultures did not show any growth, pleural fluid aspiration was done and showed a protein level of 46 g/L (more than 35 g/L indicates exudate) and cytology did not show any malignant cells. He underwent emergency thoracic endovascular aortic repair (TEVAR) and insertion of a Hickman line for prolonged intravenous antimicrobial therapy (Flucloxacillin and Gentamicin). FDG- PET CT arranged 3 weeks post TEVAR, which confirmed that the picture is more consistent with a mycotic aneurysm rather than malignancy.

Following TEVAR, the patient experienced persistent breathlessness, progressive thoracic back pain, significant unintentional weight loss and night sweats. Serial imaging showed a persistent right-sided pleural effusion and no improvement in aortic morphology. Despite prolonged broad-spectrum antibiotics, his condition deteriorated, with treatment complicated by deranged liver function tests (ALT elevated to 322 U/L) with newly developed non-malignant left-sided effusion.

A repeat CT in April 2023 showed heterogeneous attenuation surrounding the distal part of the thoracic aorta with a few patchy areas of enhancement, with an increase in the diameter of the aortic lesion to 10.6 mm. A decision to proceed with a biopsy was taken.

ACT-guided biopsy showed a microscopically focally necrotic neoplasm composed of large epithelioid cells to polygonal to spindle cells with moderate to abundant uniform eosinophilic cytoplasm and bizarrely shaped, enlarged, irregularly contoured hyperchromatic nuclei. Many multinucleate giant cells are scattered with bizarre nuclei. Possible osteoid seen focally. Mitotic count was up to 3 per 10 hpf.

Immunohistochemistry of the majority of tumor cells showed staining with SMA. There was no staining with Alk-1, Desmin, H-Caldesmon, SOX10, Pan-cytokeratin, Calretinin, D2-40, DOG-1, MyoD1, Myogenin, ERG, P63, SMM, Melan A and TTF-1. There was no loss of INI-1. FISH showed loss of CDKN2a, an atypical FUS rearrangement and no evidence of MDM2 amplification.

The features are suggestive of undifferentiated pleomorphic sarcoma FNCLCC grade 2; the possible osteoid could be interpreted as undifferentiated sarcoma with osteosarcomatous differentiation. The main differential was Leiomyosarcoma due to strong staining with SMA, although it is unlikely due to the absence of staining of other smooth muscle markers. The absence of MDM2 amplification excluded dedifferentiated liposarcoma. SMARCA4-deficient undifferentiated thoracic sarcoma was a possibility, but that was not tested. Sarcoma MDT recommended that the disease be treated as pleomorphic sarcoma.

Treatment

The patient was managed under the care of the oncology team. He was admitted to the hospital for further support of his general condition, Rt sided chest drain was inserted for his Rt sided large effusion, followed by pleurodesis. Also, the patient commenced on a therapeutic dose of enoxaparin. Given the risk of impending spinal cord compression, palliative radiotherapy was delivered to T10–T11 (8 Gy / one fraction) mainly for symptom control in June 2023.

Systemic treatment options were discussed within the sarcoma multidisciplinary team. Combination chemotherapy was deemed unsuitable due to poor performance status and cardiovascular comorbidity. Single-agent weekly doxorubicin (20 mg/m² weekly, 3 weekly cycles) was initiated as palliative chemotherapy, in line with accepted management strategies for advanced soft tissue sarcoma. Treatment was initiated in July 2023^{8,9}.

Outcome and Follow-Up

The patient tolerated treatment well, experiencing only mild nausea. Interim CT imaging after three cycles of chemotherapy demonstrated a partial radiological response with a reduction in tumor size and pleural effusion.

Following completion of six cycles of chemotherapy in November 2023, imaging confirmed continued disease control. Cardiac monitoring revealed a reduction in left ventricular ejection fraction consistent with possible anthracycline-related cardiotoxicity.

Serial 3-monthly CT scans showed stable disease from November 2023 till August 2024. Disease progression was noted on surveillance imaging in August 2024 and second-line combination chemotherapy with gemcitabine (540 mg/m² D1, D8) and docetaxel (60 mg/m², D1) 3 weekly was commenced. He finished 6 cycles (last cycle in February 2025) and is currently the patient on follow-up with 3 monthly scans and the CT scans are still showing stable disease (**Figures 1-5**).

January-February 2023	Initial presentation with symptoms suggestive of mycotic aneurysm. Antibiotics started as a management.
April 2023	Progression despite antibiotic treatment. Decision to proceed with biopsy
May 2023	CT guided biopsy confirmed the diagnosis of Pleomorphic soft tissue sarcoma
July-November 2023	Systemic 1 st line treatment given with partial radiological response achieved. Then patient remain on surveillance with 3 monthly scans.
August 2024	Surveillance CT scans showed progression of the disease
August 2024- February 2025	Second line chemotherapy given with CT scans showing stable disease and patient on surveillance



Figure 1: CT scans from 12/2022 which diagnosed as mycotic aneurysm.

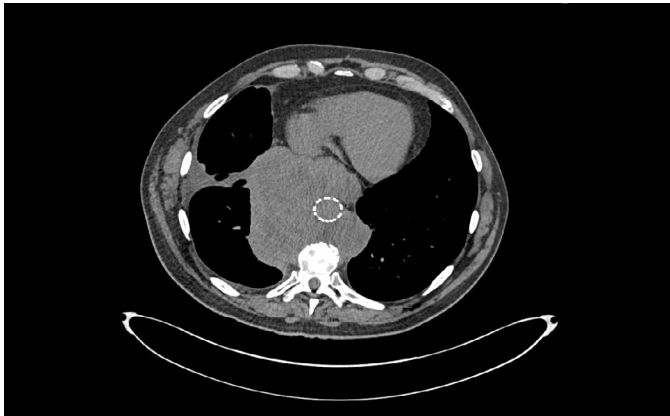


Figure 2: CT scans from 5/2023 showed progression of the aortic mass size.



Figure 3: CT scan in 11/2023 showed partial response to chemotherapy single agent Doxorubicin.



Figure 4: CT scans from 5/2024 showed stable appearance regarding the aortic mass.

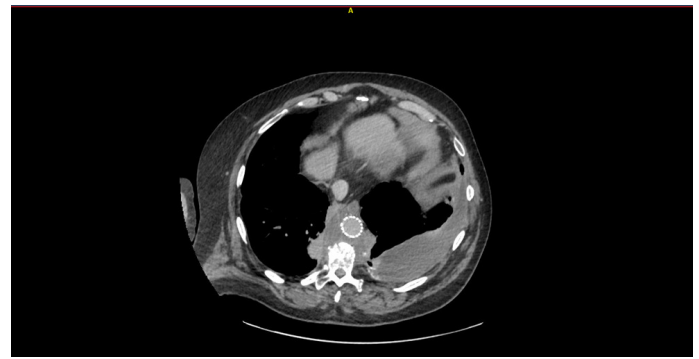


Figure 5: CT scans in 8/2024 showed progression regarding soft tissue component of the aortic mass.

Discussion: Include a very brief review of similar published cases

Distinguishing between primary aortic sarcoma (PAS) and mycotic aneurysm (MA) is a well-recognized diagnostic challenge, given their overlapping clinical presentations. Both conditions may manifest with non-specific symptoms such as chest or abdominal pain, fever, elevated inflammatory markers and weight loss. As a result, imaging modalities play a decisive role in early recognition. The most important clinical points that could differentiate between a mycotic aneurysm and a sarcoma are a negative blood culture and a poor response to antibiotics in Sarcoma patients. Sarcoma should be suspected in patients with mycotic aneurysm who do not show clinical improvement after commencing antibiotics.

Accurate radiologic interpretation can significantly alter the patient's management and prognosis. CT angiography (CTA) is often the first-line imaging modality. Aortic sarcomas classically appear as irregular intraluminal or mural soft-tissue masses arising from the intima. The masses typically extend longitudinally and may involve large segments of the aorta or branch vessels. By contrast, mycotic aneurysms usually demonstrate saccular aneurysmal dilatation with focal outpouching, commonly located at arterial branching points or sites of pre-existing vascular injury. The hallmark in MA is destructive wall change rather than a discrete enhancing intraluminal lesion^{1,4,14,15}. PAS often lacks any surrounding inflammatory reaction; instead, it presents with eccentric thickening of the aortic wall due to tumoral infiltration. Therefore, lack of this inflammatory changes in our case patient supported neoplastic rather than infective etiology^{13,17,20}.

MRI can add value in characterization of soft tissue components and identification of tumour heterogeneity, necrosis and mural invasion^{16,18}. FDG PET-CT is increasingly reported in differentiating these entities. PAS exhibits intense, focal and heterogeneous FDG uptake within the intraluminal mass, consistent with malignancy. Although both conditions can demonstrate FDG avidity, the distribution and morphology of uptake provide critical clues. Moreover, PET-CT can detect metastatic disease in sarcoma and systemic septic emboli in MA^{11,12,19}.

Histopathological examination ultimately established the diagnosis in this case. Accurate pathological assessment is essential because undifferentiated pleomorphic sarcoma is largely a diagnosis of exclusion requiring careful immunohistochemical evaluation to exclude vascular, neural and smooth muscle differentiation²⁹.

The published literature consistently demonstrates that PAS is frequently diagnosed late because of its close clinical and radiological resemblance to infective aortic disease. Persistent constitutional symptoms, negative blood cultures, progressive imaging findings and failure to respond to antimicrobial therapy should alert clinicians to the possibility of underlying malignancy. Early consideration of tissue diagnosis and multidisciplinary evaluation is essential to avoid delays in definitive oncologic management.

Rusthoven, et al.¹, in the largest pooled analysis of published cases, demonstrated that most patients present with advanced disease and poor prognosis, with delayed diagnosis being a major cause of mortality. Shimogawara, et al.¹⁷ described a case of aortic sarcoma mimicking mycotic aneurysm in which persistent symptoms and lack of response to prolonged antibiotic therapy raised suspicion for malignancy. Fukunaga, et al.¹² further demonstrated the diagnostic utility of FDG PET-CT in differentiating PAS from infected aneurysm. Kim, et al.² reported an undifferentiated pleomorphic sarcoma of the thoracic aorta presenting with a ruptured saccular aneurysm requiring emergency intervention. Similar to our patient, urgent vascular management preceded histological diagnosis due to the life-threatening presentation. Their report showed that tissue diagnosis is often not feasible before emergency repair, contributing to delayed oncologic referral.

Conclusion

Primary aortic sarcoma is a rare but important differential diagnosis in patients suspected of having a mycotic aortic aneurysm. Failure to respond to appropriate antimicrobial therapy should prompt reconsideration of the initial diagnosis. Imaging features such as enhancing intraluminal masses and invasion of adjacent structures may suggest malignancy. Early multidisciplinary collaboration and tissue diagnosis may reduce diagnostic delay and improve management.

Disclosures

We can confirm that written informed consent was obtained from the patient for writing this case report and for the publication of it, including the publication of his scan images.

Conflicts of interest

The authors declare that they have no conflict of interest.

Data Availability

The data supporting the findings of this case report are available from the corresponding author upon reasonable request.

Authors' Contributions

All authors contributed to the conception, clinical data collection, manuscript drafting, critical revision and final approval of the manuscript. All authors have read and approved the final version of the manuscript.

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