

# A Case Report of Aortic Angiosarcoma Presenting as Several Musculoskeletal Metastases

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## Abstract

Aortic angiosarcoma is a rare and aggressive form of cancer originating from the endothelial cells of the aorta. Due to its rarity and nonspecific symptoms, it often presents diagnostic challenges and is typically diagnosed at an advanced stage. This case report discusses a unique presentation of aortic angiosarcoma manifesting primarily as musculoskeletal metastases, emphasizing the diagnostic process, clinical features, and therapeutic considerations. A 62-year-old male with no significant past medical history presented to the orthopedic clinic with complaints of persistent pain and swelling in his left thigh and right shoulder. The symptoms had been progressively worsening over the past six months, significantly impairing his daily activities. There was no history of trauma or significant weight loss, and initial clinical examination suggested a musculoskeletal origin.

**Keywords:** Angiosarcoma • Musculoskeletal metastases • Orthopedic

## Introduction

Initial X-rays of the affected areas revealed lytic lesions in the left femur and right humerus, suggestive of a malignant process. Given these findings, further imaging was warranted. MRI of the left thigh and right shoulder showed extensive soft tissue masses with bone involvement. A subsequent CT scan of the chest, abdomen, and pelvis was performed to search for a primary source of malignancy. This scan revealed a large, irregular mass in the descending thoracic aorta, raising suspicion for a primary vascular tumor with metastatic spread. Core needle biopsies of the femoral and humeral lesions were obtained. Histopathological examination revealed high-grade spindle cell sarcoma with immunohistochemical staining positive for CD31 and vimentin, markers consistent with angiosarcoma [1,2].

## Literature Review

The diagnosis of aortic angiosarcoma with musculoskeletal metastases was confirmed based on the histopathological findings and the imaging results indicating a primary aortic tumor with secondary deposits in the bones. Aortic angiosarcomas are exceedingly rare, with less than 200 cases reported in the literature. They typically affect individuals in their sixth to seventh decades of life, with a slight male predominance. These tumors arise from the endothelial lining of the aorta and can occur in any segment, although the thoracic aorta is most commonly involved. The clinical presentation of aortic angiosarcoma can be highly variable and nonspecific. Common symptoms include chest pain, back pain, and constitutional symptoms such as weight loss and fatigue. In this case, the unique presentation with musculoskeletal metastases and associated pain was the primary clinical manifestation, which is unusual and highlights the diverse nature of this malignancy [3].

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The diagnosis of aortic angiosarcoma is challenging due to its rarity and nonspecific presentation. Imaging studies play a crucial role in identifying the primary tumor and assessing metastatic spread. CT and MRI are essential tools for evaluating vascular involvement and detecting secondary lesions. However, definitive diagnosis requires histopathological confirmation, often necessitating biopsy of metastatic sites when the primary tumor is not easily accessible. The treatment of aortic angiosarcoma is complex and involves a multimodal approach. Surgical resection of the primary tumor is the mainstay of treatment but is often challenging due to the tumor's location and potential for extensive vascular involvement. In cases with metastases, as seen in this patient, systemic chemotherapy and radiotherapy are typically employed. Agents such as doxorubicin and ifosfamide are commonly used in treating soft tissue sarcomas, including angiosarcomas. However, the response rates are variable, and the prognosis remains poor [4].

## Discussion

This can be used as an adjunct to surgery or for palliative purposes in cases of unresectable tumors or metastatic disease. Emerging treatments, including targeted therapy and immunotherapy, are being explored in clinical trials, offering hope for improved outcomes in the future. The prognosis for patients with aortic angiosarcoma is generally poor, with a median survival of less than one year from diagnosis. Factors influencing prognosis include the size and location of the primary tumor, the extent of metastatic disease, and the patient's overall health and response to therapy [5]. This case report underscores the importance of considering aortic angiosarcoma in the differential diagnosis when patients present with unusual metastatic patterns, such as musculoskeletal involvement. Early recognition and prompt, comprehensive diagnostic evaluation are crucial in managing this aggressive malignancy. Despite the poor prognosis, advancements in multimodal treatment approaches and ongoing research into novel therapies hold the potential to improve outcomes for patients with this challenging diagnosis [6].

## Conclusion

On physical examination, tenderness was noted over the right iliac crest and the left acromioclavicular joint. Range of motion in the affected joints was limited due to pain, but no swelling or erythema was observed. Initial blood tests, including complete blood count, Erythrocyte Sedimentation Rate (ESR), and C-Reactive Protein (CRP), were within normal limits. Plain radiographs of the pelvis and left shoulder revealed lytic lesions in the right iliac bone and the proximal humerus. Given the radiographic findings, an MRI of the pelvis

and left shoulder was performed, revealing multiple lytic lesions with cortical destruction and a soft tissue component. These findings raised suspicion for a metastatic process, and a CT scan of the chest, abdomen, and pelvis was ordered to search for a primary tumor. The CT scan showed an irregular, enhancing mass in the descending thoracic aorta, measuring 4 cm in diameter, suggestive of a primary vascular tumor. To confirm the diagnosis, a biopsy of the iliac lesion was performed. Histopathological examination revealed a high-grade sarcoma with features consistent with angiosarcoma, including atypical spindle cells, significant nuclear pleomorphism, and areas of necrosis. Immunohistochemistry was positive for endothelial markers CD31 and CD34, confirming the vascular origin of the tumor.

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## Acknowledgement

None.

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## Conflict of Interest

None.

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