
Case Report

Intimal Sarcoma of the Descending Aorta

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Primary intimal angiosarcoma of the aorta (i.e., mostly intraluminal sarcomas with evidence of endothelial differentiation) is extraordinarily rare. We report a case in which the diagnosis was accurately made using immunohistochemistry in an embolectomy specimen. The patient was a 78-year-old man with a two-month history of bilateral claudication. Doppler ultrasound proved an embolus in both popliteal arteries, which was removed. The highly atypical cells comprising these emboli were positive immunohistochemically for CD68, vimentin, and CD31. Magnetic resonance imaging also showed an irregular tumor (invasion to the left main bronchus). This case emphasizes the need for a wide panel of immunohistochemical studies in tumor emboli of unknown origin.

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Introduction

Intimal sarcoma of the aorta is a very rare, but aggressive tumor. It is mostly accompanied with embolic phenomena. En-bloc resection with postoperative chemo-radiotherapy are its treatments, but in most cases, the prognosis is still dismal. Herein, we report a case of aortic angiosarcoma presented with bilateral popliteal thromboemboli.

Case Report

A 78-year-old heavy-smoker man was admitted to our center for further evaluation of cardiovascular malignancy. He had a history of bilateral claudication in the past two months. He had cold lower extremities with absence of pulses in the popliteal, tibialis posterior, and dorsalis pedis arteries two days before his first hospital

admission. Doppler ultrasound of the lower extremities revealed bilateral popliteal artery thrombosis. Bilateral popliteal embolectomy was performed. The pathological study of the embolectomized material showed some spindle to ovoid cells with severe nuclear pleomorphism and hyperchromasia accompanied by multinucleated giant cells, with myxoid background. The immunohistochemical study was positive for CD68, vimentin, and CD31 and negative for CD34 and CKAE1/3 markers. All these findings were compatible with a diagnosis of intimal angiosarcoma. In our center, echocardiography revealed a mild diastolic dysfunction with aortic valve calcification. There was no evidence of tumor in the cardiac chambers. An irregular tumoral mass was found in the descending aorta by magnetic resonance imaging (MRI) (Figure 1A). The mass had invasion to the left bronchus (Figure 1B) with heterogeneous enhancement after contrast injection (Figure 2). Considering the extensive tumoral involvement, chemo-radiotherapy was started. The patient refused to receive the treatment and died of multi-organ failure after two months.

Discussion

There are two types of aortic sarcomas; intimal and mural sarcomas. Intimal sarcoma may cause

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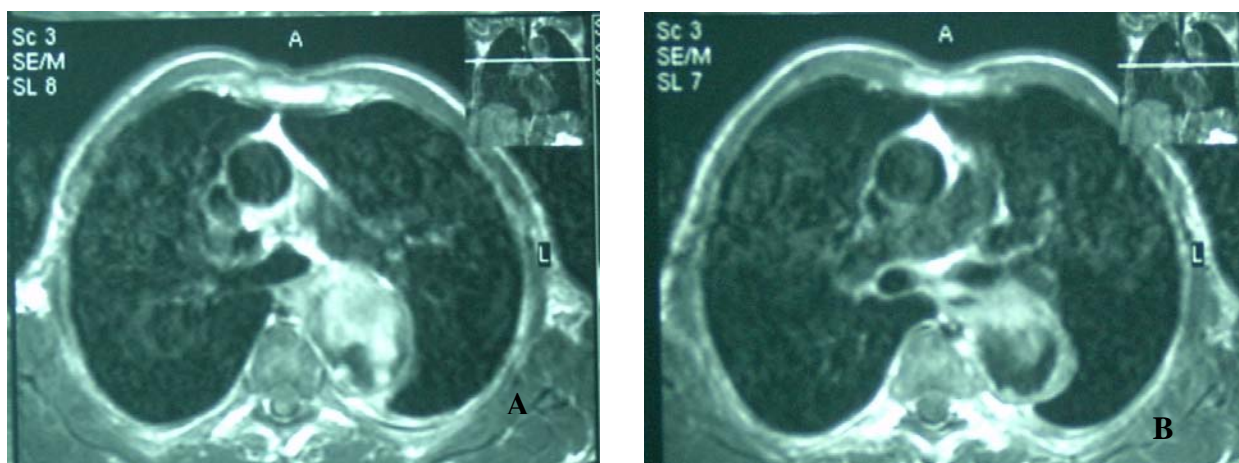


Figure 1. Axial T1-weighted image revealing irregular tumoral mass in the descending aorta (A). The arrow shows invasion to the left main bronchus (B).

peripheral emboli or grow along the lumen. The mural type originates from media or adventitia and extends to paraaortic tissues.¹ In most cases, the intimal subtype originates from the descending thoracic or abdominal aorta.² Most cases are diagnosed at autopsy. The symptomatic patients usually have signs of tumor embolus in lower extremities or mesenteric arteries.² The disease is more prevalent in men with a mean age of 60 years. The work-up of a patient presenting with peripheral emboli, begins with echocardiography.

Once a cardiac source has been excluded, magnetic resonance angiography (MRA) of the aorta is the most sensitive imaging modality for detection of the tumor. MRI study can differentiate tumor from atheromatous plaque by enhancement and reveals the extension of tumor to adjacent

structures. Compared with conventional angiography, there is no risk of catheter-induced embolization or contrast-induced nephrotoxicity with MR study.³ When malignant tumor of aorta is suspected in MR, bone scintigraphy should be carried out, since the rate of bone metastasis is high.³ If bone metastases are confirmed, major surgical intervention is not indicated. Immunohistochemistry is important for making a definite diagnosis of intravascular malignancy. Positivity for CD31, von-Willebrand factor, and Ulex Europeans with negative result for CD34 are in favor of intimal angiosarcoma.⁴ In patients without bone metastasis, treatment consists of en-bloc resection with chemo-radiotherapy, but prognosis is still poor with a life expectancy at diagnosis of almost 14 – 27 months.⁵

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Figure 2. Axial T1-weighted image after contrast injection. The tumor shows heterogeneous enhancement in favor of malignancy.