Intimal angiosarcoma of the descending aorta presenting as atheroembolism: case report and description of intravascular biopsy

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ABSTRACT

Aortic sarcoma is a rare entity. In most cases, the diagnosis is established late, owing to the course of the disease, with a median survival time of only a few months. We report the case of a 58-year-old patient with ischemic lesions in the lower limb. The lesions after several investigations were diagnosed by imaging studies (eg, magnetic resonance angiography, contrast-enhanced computed tomography [CT], CT angiography, or positron emission tomography-CT with fluorine-18 fluorodeoxyglucose) as possible primary angiosarcoma of the aorta. It was decided to perform endovascular aortic repair and endovascular biopsy of the lesion. This was chosen as a "palliative" treatment to avoid embolic events, given the presence of metastases, instead of open surgery, which would otherwise have been the treatment of choice. Subsequently, histologic examination confirmed aortic intimal angiosarcoma, and adjuvant chemotherapy was initiated. In the present report, we discuss the clinical presentation, diagnosis, and classification of primary aortic sarcomas. We also critically review the diagnostic and therapeutic management of these patients in previous series of studies to improve their treatment in subsequent cases. (J Vasc Surg Cases Innov Tech 2023;9:101230.)

Keywords: Aorta; Endovascular aortic repair; Intimal angiosarcoma; Malignant aortic tumor

Angiosarcomas of the great vessels are rare entities with a poor prognosis.[1] They affect more men than women (ratio, 2:1 to 5:1) at a mean age of 60 years.[1-4] The abdominal and thoracic aortas are equally affected. Angiosarcomas can be classified according to their location in the arterial wall (mural, 20%; intimal, 70%; or mixed) or their histologic type (sarcoma, 29%; malignant fibrous histiocytoma, 17%; angiosarcoma, 11%; leiomyosarcoma).[1,5,6] As a clinical manifestation, it most often debuts with embolic phenomena (eg, cerebral, lower limb ischemia, visceral) or renovascular arterial hypertension and, rarely, presents as spontaneous rupture of the aorta.

The diagnosis is challenging (both clinically and via imaging studies), because, given that the mean age at diagnosis is ~60 years, most are initially considered to be thrombosis or atherosclerosis.[1] The most useful imaging tests for its evaluation include computed tomography (CT) angiography (CTA), magnetic resonance angiography, and fluorine-18 fluorodeoxyglucose positron emission tomography (PET)-CT.[7,8]

CASE REPORT

A 58-year-old patient with a medical history of atrial fibrillation (with anticoagulation therapy), chronic obstructive pulmonary disease, and dilated cardiomyopathy presented to the emergency department because of infected necrotic lesions on the toes of his left foot, together with a 1-month history of erythrosis. The patient provided written informed consent for the report of his case details and imaging studies. He had no history of claudication or the use of toxic substances. Physical examination showed only femoral pulses, with monophasic distal flows in the left leg and biphasic waves in the right leg (the latter without lesions). Complete laboratory test results revealed elevation of inflammatory markers (ie, erythrocyte sedimentation rate, polymerase chain reaction) and tumor markers (ie, neuronal-specific enolase). CTA revealed a possible myxoid tumor, primary intimal sarcoma of the aorta, or other sarcomatous tumors (Fig 1). Magnetic resonance angiography showed tumor enhancement in the lesion (Fig 2), and PET-CT showed significant metabolic uptake and the presence of multiple metastases (spleen and ribs). Endovascular surgical intervention was chosen as a palliative solution via thoracic endovascular aortic repair with intraoperative biopsy of the lesion. Access was achieved via the bilateral femoral arteries with ultrasound-guided

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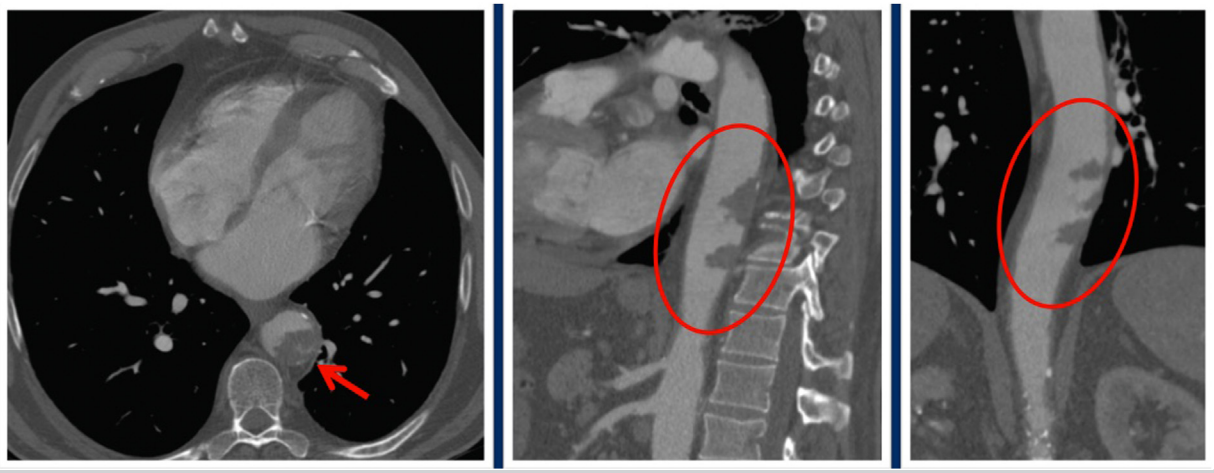

Fig 1. Computed tomography angiographic images showing, in the descending aorta, prominent, irregular, vegetating, dense soft tissue repletion defects (30-40 HU; marked in *red*), extending from L8 to L11 (62 mm long), and seated in the posterior and external wall of the aorta.

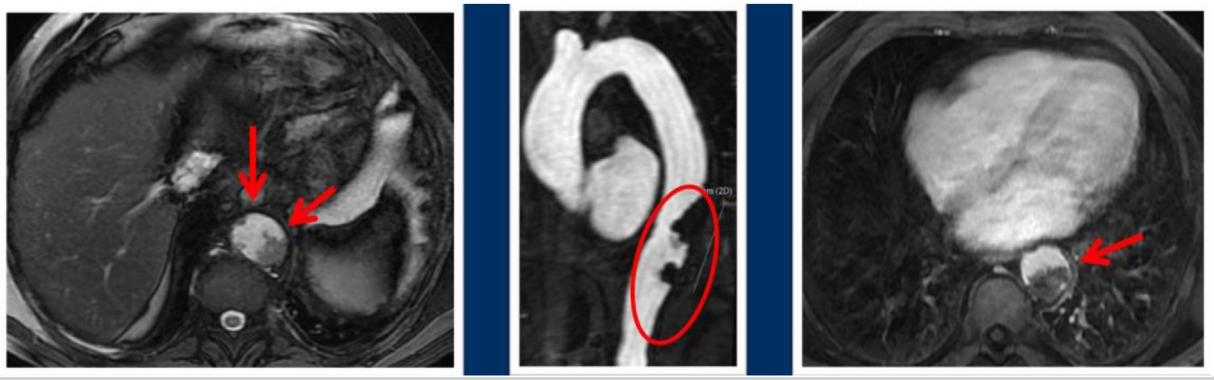

Fig 2. Magnetic resonance angiographic images showing a voluminous filling defect in the descending thoracic aorta dependent on the posterior aortic wall, with irregular contours and a low resonance signal, without significant enhancement and causing stenosis of >50% of the cross-sectional area (marked in *red*).

puncture and 6F introducers, and preclosure devices were placed (Perclose ProGlide; Abbott Vascular) in standard fashion. On the left side, a 45-cm, 12F sheath was delivered at the level of the tumor over a rigid guidewire, ensuring that it remained toward the greater curvature of the aorta and guided with fusion imaging. On the right side, a 22F (65-cm) DrySeal sheath (W.L. Gore & Associates) was placed in the aortic arch. After insertion of both sheaths, systemic heparin was administered (1 mg/kg). Angiography was performed, which showed the tumor imprint in the lumen. The Cydar image fusion system was adjusted to avoid the use of excess contrast and radiation. A 34 × 34 × 150-mm cTAG prosthesis (W.L. Gore & Associates) was deployed (Fig 3, *A*), leaving the 12F sheath between the endograft and aortic wall. A 6F Fogarty catheter was pushed over the 12F sheath and, under fluoroscopic guidance, two sweeps were performed, introducing whitish tumor material into the sheath (Fig 3, *B*). This material was simultaneously aspirated with a 50-mL syringe and sent for analysis. The distal part of the prosthesis was dilated with a balloon simultaneously with removal of the 12F sheath, aspirating through it to avoid embolization.

Completion angiography was performed to visualize correct implantation of the endoprosthesis and patency of all the visceral, iliac, and femoral branches (Fig 3, *C*). The femoral punctures were closed, and the patient was taken to the intensive care unit. Subsequently, pathologic examination of the anatomy confirmed the diagnosis of primary intimal angiosarcoma of the aorta.

The patient was discharged with anticoagulation therapy (previously indicated for atrial fibrillation) and aspirin. Subsequently, he refused chemotherapy. He was admitted 5 months after surgery with cerebral metastases and died in hospital.

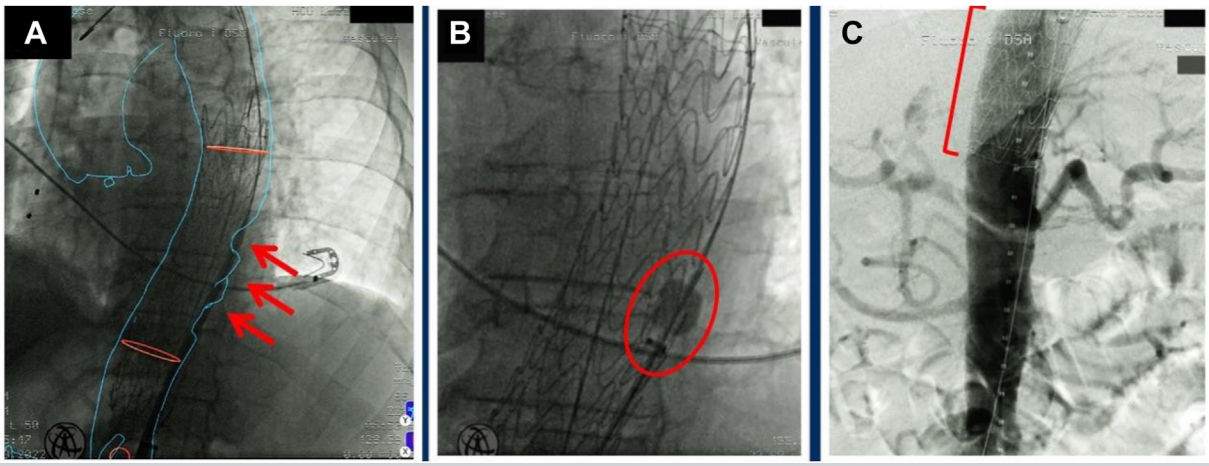

Fig 3. A, cTAG (W.L. Gore & Associates) 34 × 34 × 150-mm stent implant (*arrow*). **B,** Sweeping performed with a 6F Fogarty catheter over the 12F sheath, drawing tumor material with a whitish aspect into the sheath (*circle*). **C,** Completion angiography (*bracket*).

DISCUSSION

Primary aortic tumors are extremely rare and frequently correspond with malignant mesenchymal tumors.[1-10] Few studies have been reported (<100 cases since 1998), with the latest a case series with a review of the literature by Böhner et al.[4,9] Because of its deep location, silent growth, and clinical and imaging findings that mimic thrombosis and atherosclerosis, its diagnosis can be difficult.[1,2] Doppler ultrasound and intravascular ultrasound are not useful for tissue characterization or differentiation between tumor and thrombus.[7,8] CTA usually shows aortic wall irregularity, polypoid intraluminal mass, and complete occlusion of the aorta or aortic branch, without significant enhancement. The presence of concomitant atherosclerosis calcification in the wall and associated thrombus adds confusion to the diagnosis. Aneurysmal dilatation and rupture of the aortic wall is rare.

The most specific tests for differentiating between tumor and thrombus and/or atherosclerosis are gadolinium-enhanced MRA and fluorine-18 fluorodeoxyglucose PET-CT, which show tumor enhancement (not always present) and significant metabolic uptake.[7,8] The gold standard for diagnosis continues to be pathologic examination of the anatomy,[1,6] with an antemortem diagnosis made in 73%. Most specimens are obtained by surgical resection. Intra-arterial biopsy is rarely performed because of possible embolic complications.

The most effective treatment is radical surgical resection; however, that is rarely possible.[10] Studies have described cases of endarterectomy without resection and reported that it could be an adequate treatment, because the malignant cells in intimal angiosarcomas are limited to the luminal surface of the thrombus. However, studies to support this are still lacking.

As palliative surgery, implantation of an endoprosthesis is indicated to avoid distal embolization that worsens patients' quality of life. In our patient, palliative surgery was the intention of the intervention (no curative surgery was possible), in addition to obtaining a pathologic specimen for histologic analysis of the tumor and the possibility of performing subsequent targeted chemotherapy. We decided to implant the cTAG endograft because it is deployed from distally to proximally; thus, the distal exit is sealed first, and embolization is also avoided.

Several studies have shown that even metastatic aortic sarcomas can be controlled for some time with radiotherapy or chemotherapy (doxorubicin and ifosfamide) if the metastasis is limited, if the tumor is not resectable, and in embolic situations.[4-9] Even with adjuvant treatment, these entities have a very poor prognosis (worse with thoracic aortic involvement), with a median survival of 14 to 16 months. The cause of death for 80% of these patients is metastatic disease (ie, bones, kidneys, liver, adrenal glands, lungs).[1]

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