

# PRIMARY LEIOMYOSARCOMA OF THE THORACIC AORTA MIMICKING AORTIC DISSECTION

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Primary malignancies of the aorta are extremely rare. Leiomyosarcomas arise from the smooth muscle of the aortic wall, and may have intraluminal and extravascular components. Only six cases have been previously reported.<sup>1-3</sup> Clinical presentations are nonspecific and related to pattern of tumor growth. Our case is one of a leiomyosarcoma of the descending thoracic aorta masquerading as aortic dissection on clinical and computed tomography (CT) features. The magnetic resonance imaging (MRI) suggested the diagnosis of mediastinal mass with aortic and vertebral extension. To our knowledge, no similar case has previously been reported in the English literature.<sup>2</sup>

## Case Report

A 65-year-old Caucasian female was admitted with acute chest pain. On examination, she appeared in good general health. Her blood pressure was normal and there were no cardiac or pulmonary abnormalities. Laboratory analyses revealed anemia. ECG was normal, but chest radiography showed enlargement of the descending aorta associated with a retrocardiac mass. Thoracic and transesophageal echocardiography were not performed. CT demonstrated a thickening of the luminal narrowing (Figure 1). A heterogeneous periaortic mass 7 cm in diameter with peripheral enhancement after contrast media injection was visualized. Clinical signs and CT findings supported the diagnosis of aortic dissection with fissuration and periaortic hematoma extending paravertebrally.

MRI was performed with a 1.0-T system (Magnetom, Siemens). MR images were obtained from the aortic arch to the renal vessels in axial, coronal, and sagittal planes, with spin-echo (SE) and gradient-echo (GRE) sequences (Figure 2). On SE T2-weighted images, we noted an heterogenous and laterovertebral mass associated with a marked luminal narrowing of the aorta with intraluminal nodules. Post-

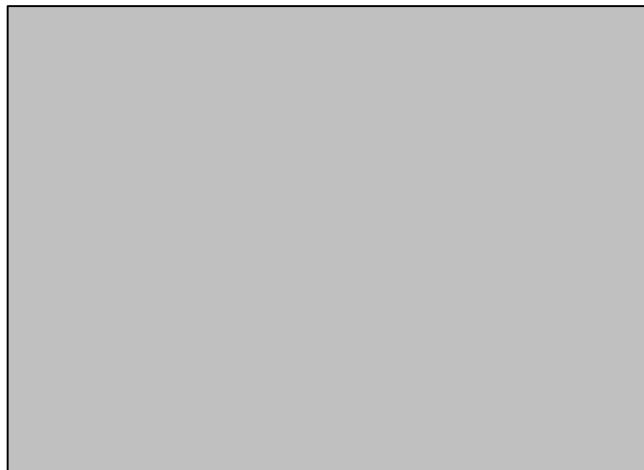


FIGURE 1. Axial contrast-enhanced CT image at the level of the primary bronchi showing pseudo-intimal flap (arrow) with thickening of aortic wall (arrowheads).

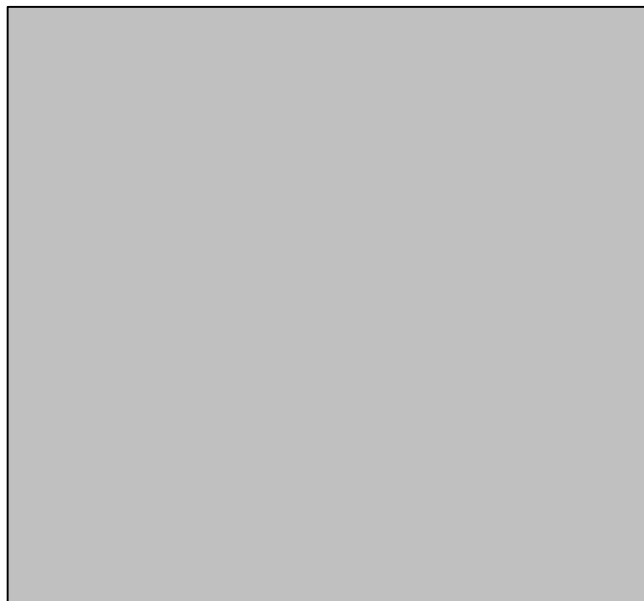


FIGURE 2. Coronal T1-weighted spin-echo magnetic resonance image after injection of gadolinium showing irregular and nodular narrowing of the aortic lumen (arrowheads). Note the moderate enhancement of the periaortic mass (arrow).

gadolinium SE, T1-weighted images and contrast-enhanced turbo-FLASH (fast low angle shot) images demonstrated enhancement of the mass and improved visualization of the aortic nodular lesions, which were 3 to 6 mm in diameter,

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and located the vertebral extension to the bodies of T9 and T10. Preoperative diagnosis was mediastinal tumor with aortic and vertebral extension.

Surgical exploration showed a mediastinal mass arising from the thoracic descending aorta with paravertebral extension. Identification of intraluminal nodules after the opening of the aortic lumen ruled out the diagnosis of aortic dissection. A large part of the lesion was resected, and a vascular prosthesis was placed. The diagnosis of leiomyosarcoma arising from the aortic wall was based on histological analysis and confirmed immunohistochemical findings. The patient died three months after surgery.

### Discussion

Leiomyosarcomas of vessels are rare.<sup>1,2</sup> They occur in the veins five times more frequently than in the arteries.<sup>3,4</sup> In the pulmonary arteries, they are twice as frequent as those found in other systemic arteries.<sup>4,5</sup> A review of the literature disclosed only six cases of primary leiomyosarcoma of the thoracic aorta.<sup>2,3</sup>

The value of MRI and transesophageal echography in the diagnosis of aortic leiomyosarcoma has been previously reported in only two cases.<sup>2,3</sup> Aortic tumors can be classified into two categories, according to the site of occurrence in the aortic wall.<sup>6</sup> The first group involves the intima, and grows in the direction of the aortic lumen. In these cases, symptoms are due to luminal obstruction, and it may be difficult to distinguish them from atherosclerosis. In this pattern, hematogenous metastasis is very frequent, mainly involving the spine, liver, adrenal glands, pancreas, lung and soft tissues.<sup>1,2,5-7</sup> The second group of aortic tumors arises in the media and adventitia, and the growth pattern is extravascular.<sup>2,6</sup> Clinical signs are nonspecific and are similar to those of aortic dissection, ruptured or dissecting aneurysm.<sup>1,5-8</sup>

Symptoms of previously reported cases of primary malignant neoplasms of the thoracic aorta typically included back pain, hypertension, fever, dyspnea and paresthesia.<sup>2,3,8</sup> The episode of sudden chest and back pain may be explained by intratumoral bleeding or fissuration, which are the more frequent complications. Because of these diverse clinical manifestations, aortic tumors are almost always identified during operation or at autopsy.<sup>8</sup>

In our case, clinical and CT findings suggested the diagnosis of aortic dissection with periaortic hematoma. There were no abnormalities of spine at chest radiography and CT scans. MRI was very helpful for the diagnosis of tumoral origin, and demonstrated wall nodules, periaortic

and vertebral extensions. These lesions were enhanced after gadolinium injection, which is also useful in detecting residual tumor.<sup>9</sup> MRI enabled us to rule out the diagnosis of aortic dissection. In effect, no intimal flaps in different sequences were found.

Transesophageal echography may be useful for the diagnosis and identification of wall thickness and nodules, but it is less informative than MRI on periaortic extension.<sup>2</sup> The management and optimal treatment of aortic tumors is difficult to determine even in the cases in which the diagnosis is established before surgery.<sup>1,2,8</sup> As described in this report, the treatment may be surgical, combining primary resection with reconstruction. The longest period of survival from an aortic tumor is estimated to be 4.2 years after surgery.<sup>10</sup> The other reported cases, which were treated with radiotherapy and chemotherapy with or without surgical resection, also indicated poor results, with an average survival period of 18 months.<sup>1,2,6,8</sup>

Aortic leiomyosarcomas are very rare, and usually have unfavorable prognosis. Clinical awareness, with the help of MRI findings, can lead to early diagnosis. Our data suggest that MRI may be very useful in preoperative diagnosis of aortic tumors, indicated by irregular lumen and identification of gadolinium-enhanced intraluminal nodules.<sup>1,9</sup> As well, MRI is more appropriate than CT to detect leiomyosarcoma metastasis, particularly in the spine and mediastinum and to rule out aortic dissection.<sup>1,2,10</sup>

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