Malignant Fibrous Histiocytoma Arising From the Aortic Wall Mimicking a Pseudoaneurysm With Ulceration

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Malignant fibrous histiocytoma of the thoracic aorta associated with ulcer-like projection has not been previously reported. The hypovascular tumor of the thoracic aorta involved the mural layer, which showed intra- and extra-mural growth patterns and no significant filling defect within the aortic lumen, and mimicked ulcer-like projection with secondary pseudoaneurysm formation. Aortic tumor, although rare, should be included among the causes of an ulcer-like projection. (Circ J 2007; 71: 1659–1661)

Key Words: Aorta; Computed tomography (CT); Tumor

P rimary malignant lesions of the aorta occur rarely and are associated with a very low survival rate. Common presentations are distal embolic phenomena, aortic dissection; or even occlusion of the aortic lumen. This case report describes a patient with a primary malignant fibrous histiocytoma (MFH) of the thoracic aorta associated with ulcer-like projection (ULP). The aortic mural tumor invaded into the surrounding tissue, not into the aortic lumen, and the radiological appearance mimicked ULP with aortic pseudoaneurysm formation. To our knowledge, a primary aortic tumor with ULP has not been previously reported and knowledge of the characteristic imaging findings is clinically important.

Case Report

A 57-year-old woman with a 1-month history of heartburn and who suffered from progressive back pain was admitted to hospital. Her past medical history was rheumatoid arthritis. Physical examination revealed: blood pressure, 130/90 mmHg; pulse, 96 beats/min; respiration, 16 breaths/min; and body temperature, 36.8°C. Lung and cardiac sounds were normal. Blood count revealed slight anemia (hemoglobin level: 10.5 g/dl). Other laboratory data, including blood chemistry, coagulation studies, and cardiac enzymes, were not abnormal. Precontrast and contrast-enhanced dynamic computed tomographic (CT) imaging was performed using a 64-row multislice scanner (Aquilion-64, Toshiba). Contrast agent (100 ml) at an iodine concentration of 300 mg/ml (Iomeron, Eisai) was injected at a rate of 4 ml/s, and arterial phase imaging was performed 30 s later. The delay time was 3 min for delayed phase imaging. CT images in the arterial phase showed a 6.5×4.3×6.7 cm aortic lesion with ULP in the thoracic descending aorta (Fig 1). The attenuation values (in Hounsfield units (HU)) were measured at regions of interest (ROIs) in the aortic lesion in each phase. Attenuation of each ROI was 53 HU, 59 HU, and 62 HU on the precontrast CT, arterial-phase, and delayed-phase CT images, respectively. Because the case was initially diagnosed as a pseudoaneurysm with ULP and possible impending rupture, emergency endovascular stent-graft placement was selected as the treatment, using a tailored 100-mm stent-graft device (Gianturco Z-stent with UBE woven Dacron graft material). The immediate postinterventional result showed complete exclusion and thrombosis of the ULP (Fig 2). One month after the stent-graft treatment, the ULP was completely excluded but the aortic mass lesion showed no regression in size on CT imaging. Two months later, the aortic mass lesion had increased in size and there was distinct enhancement on contrast-enhanced CT imaging (Fig 3). The patient was referred for surgery during which it was found that a tumor of the thoracic aorta had solidly invaded the pulmonary hilus and was considered to be nonresectable. Surgical biopsy was performed. On histological examination, the tumor comprised a haphazardly arranged mixture of pleomorphic spindle and giant cells with frequent mitosis. Immunohistochemistry detected a positive reaction to vimentine and α-1-antitrypsin, and negative reactions to desmin and actin. The tumor was diagnosed as a MFH of the aorta.

Subsequently, radiation therapy (a dose of 2 Gy per fraction; total dose of 66 Gy) was selected and resulted in partial remission. Unfortunately, surgical-site disseminated metastasis in the chest wall occurred 3 months later.

Discussion

Since its initial description in 1964, MFH has been recognized as a distinct form of sarcoma. Fewer than 100 aortic sarcomas have been reported in the world literature. They occur throughout the aorta and are most common in middle-aged patients, with mean age 54 years. There is a male-to-female ratio of 2:1, with an average survival of 1.5 years after diagnosis. Aortic tumors are classified by their
Fig 1. Transaxial precontrast and contrast-enhanced dynamic computed tomography (CT). [(A) Precontrast CT image; (B) arterial-phase CT image; (C) delayed-phase CT image] and oblique sagittal multiplanar reconstruction (D) images show the large mass lesion of the descending aorta, but no significant contrast enhancement. The transaxial and oblique sagittal multiplanar reconstruction CT images in the arterial phase show the aortic ulcer-like projection (arrow) and circumferential extent of the tumor (arrowheads).

Fig 2. Transaxial contrast-enhanced computed tomography image shows complete exclusion and thrombosis of the aortic ulcer-like projection immediately after stent-graft placement.

Fig 3. Transaxial contrast-enhanced computed tomography image shows enlargement of the aortic mass and its enhancement characteristics (arrows) 2 months after stent-graft placement.
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Histologic type and location within the aortic wall. Those found most often are sarcomas (25), MFHs (18), angiosarcomas (13), leiomyosarcomas (9), and fibrosarcomas (9). Aortic sarcomas are also described as intimal or mural tumors, depending on their origin within the aortic wall. Intimal-based aortic sarcomas form polypoid masses projecting in the aortic lumen and grow along the intima. These lesions often obstruct branches of the aorta, causing ischemia of the organs by embolism, thrombosis, and/or mechanical obstruction. Mural-based aortic sarcomas, such as the present case, originate in the media or adventitia and invade the surrounding tissue with limited metastasis.

Because of their rarity, primary aortic tumors are seldom suspected and diagnosed in clinical practice. Radiology plays an important role in their diagnosis, but it can be difficult to differentiate between an aortic hypovascular tumor and atherosclerotic disease, both in clinical manifestation and radiological appearance. In the present case, the aortic tumor involved the mural layer, which showed intra- and extra-mural growth patterns, and no significant filling defect within the aortic lumen. An irregular aortic surface localized to the tumor and the circumferential extent of the tumor were seen on CT imaging (Fig 1), but the aortic tumor showed only little enhancement on dynamic CT imaging, and thus the differentiation between ULP with aneurysmal formation and aortic sarcoma was difficult. Initially, we considered that the aortic lesion was ULP with aortic pseudoaneurysm formation, and we selected endovascular stent-graft placement as the treatment because it is effective. In this case, however, the ULP was actually tumor erosion, not an atheromatous ulcer. We suggest that the irregularity of the aortic lumen and circumferential extent of tumor may be characteristic CT findings of a mural-based aortic tumor. Clinicians should be highly alert to the possibility of a primary aortic tumor in the differential diagnosis, and consider other investigations to aid the final diagnosis. Magnetic resonance imaging (MRI) of a MFH usually shows a mass with nonspecific T1 and T2 signal characteristics. Contrast-enhanced MRI, however, is possibly useful in differentiating tumor from atherosclerotic disease because of the higher contrast resolution. Also, we believe positron emission tomography may be helpful for diagnosis of the aortic tumor.

The appropriate therapeutic protocol of primary malignant aortic tumors has not been clearly established and, unfortunately, the prognosis is poor, with metastatic disease causing death in most patients. Autopsy studies show that distant metastasis to bone, kidneys, liver, the adrenal glands, and lungs occurs in 80% of patients. Operative treatment consists of node sampling and en-bloc resection with an interposition graft. Thromboembolectomy is often used if thrombus has propagated distal to the aortic bifurcation. The identification of tumor cells in the retrieved thromboembolic material indicates the need for systemic chemotherapy to treat metastatic disease. Adjuvant therapy is used if there is embolic, metastatic, or nonresectable disease. The recommended chemotherapy guidelines for treatment of aortic sarcomas is doxorubicin based. Radiation therapy can be used to treat nonresectable disease: 50–60 Gy delivered over 5–6 weeks in 2 Gy fractions.

In summary, we describe a 57-year-old woman with a primary MFH arising from the wall of the aorta, which, to our knowledge, has not been described before. Differentiation of hypovascular aortic tumor and atherosclerotic disease can be difficult, so careful examination of radiological findings, together with the medical history, is important for diagnosis of rare aortic tumors.

References

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