Primary Intimal Sarcoma of the Aorta
—Role of Transesophageal Echocardiography—

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Diagnosis of primary tumors of the aorta is difficult. A patient who had a primary intimal sarcoma of the aorta with metastasis presented with obstructive symptoms and computed tomography showed a thrombus-like mass in the aorta. However, transesophageal echocardiography revealed an inhomogeneous and echo-dense mass with an outer membrane, unlike a thrombus, and suggestive of a primary aortic tumor. Pathologic examination of specimens from exploration and autopsy revealed a primary intimal sarcoma. (Circ J 2002; 66: 111–113)

Key Words: Aortic tumor; Primary intimal sarcoma; Transesophageal echocardiography

A ccurate preoperative or antemortem diagnosis of primary tumors of the aorta is difficult because of their rarity and varying presentation. Most of the diagnoses have been made postoperatively or at postmortem examination. Angiography and magnetic resonance imaging may assist in more accurate evaluation and diagnosis of primary aortic tumors. Recently we managed a patient with a primary aortic intimal sarcoma that presented as vascular occlusive symptoms, but was preoperatively considered to be a primary tumor of aorta based on the transesophageal echocardiography findings.

Case Report

A 42-year-old woman was referred from another hospital. Her chief complaint was claudication that had become progressively worse during the past 3 months. Fifteen days before admission, cramping abdominal pain had occurred in the periumbilical area and it was of an intermittent nature and short duration of about 4 or 5 min. The pain was followed by nausea, vomiting and bloody watery diarrhea. There was no fever, but 4 kg of body weight had been lost over a 6-month period. She did not smoke and was not taking any medications other than a herbal preparation for their rarity and varying presentation. Most of the diagnoses have been made postoperatively or at postmortem examination. Angiography and magnetic resonance imaging may assist in more accurate evaluation and diagnosis of primary aortic tumors. Recently we managed a patient with a primary aortic intimal sarcoma that presented as vascular occlusive symptoms, but was preoperatively considered to be a primary tumor of aorta based on the transesophageal echocardiography findings.

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Laboratory tests revealed anemia (hemoglobin: 10 mg/dl) and the erythrocyte sedimentation rate was 19. Electrolyte levels and liver function tests were within normal limits. Fibrinogen was within normal limits, but fibrinogen degradation product was positive. Antithrombin III was mildly decreased to 73%. The protein C and S levels were in the normal range. Antiphospholipid antibody was negative, but lupus anticoagulant was positive. Chest radiography showed normal heart and lungs.

Computed tomography (CT) with contrast enhancement revealed near total occlusion of the abdominal aorta and both common iliac arteries by an intraaortic mass, which was attached to the anterior wall of the abdominal aorta at the level of the upper abdomen and extended both upward and downward (Fig 1). The mass in the thoracic aorta appeared to have a rod-like structure. The left renal artery was occluded and the mass extended to the celiac axis and superior mesenteric artery. There were also small masses in the aortic arch.

Transesophageal echocardiography showed no abnormal findings, but transesophageal echocardiography (TEE) revealed highly mobile masses attached to the wall of the descending thoracic aorta, just after the origin of the left subclavian artery, which protruded into the aortic lumen. At 30 cm from the incisor, the intraaortic mass was attached to the thoracic aorta and extended into the distal aorta. Unlike a thrombus, it had a rod-like structure with a central echo-dense area and an outer membrane (Fig 2). After 1 month, TEE was repeated and showed that the intraaortic mass had increased in size. In the aortic arch, 2 masses had newly formed and were huge in size and highly mobile (Fig 3).

With the presumptive diagnosis of a primary aortic tumor, exploratory surgery was undertaken for tumor removal and bypass graft if needed. The masses in the abdominal aorta and both iliac arteries were removed, but not those in the thoracic aorta. After removal, both femoral arterial pulses were palpable. A 15×10×5 cm small bowel mass adhering to the uterus was resected because it was protruding into the intestinal lumen and nearly obstructing it. It had the appearance of a submucosal tumor of the intestine and histologic sections revealed sarcoma cells.

Postoperatively, the patient developed acute renal failure and 7 days after the operation, she became stuporous and died of massive upper gastrointestinal bleeding.

At autopsy, a long tract of the aorta from the descending portions disclosed diffuse intimal thickening
with a mass-forming appearance. The cut surface was grayish yellow, solid, rubbery and myxoid. There were multiple tumor emboli at the branches of the thoracic aorta, celiac artery, left common carotid artery and left subclavian artery. The left renal artery had mixed thromboembolic and neoplastic lesions, and the kidney was infarcted from the multiple tumor metastases and thromboembolic lesions. Histologic sections of the aortic intimal mass disclosed spindle-shaped sarcoma cells with large hyperchromatic pleomorphic nuclei in abundant myxoid stroma, which is

![Abdominal CT scan](image1)

Fig 1. Abdominal CT scan. (A) Mass within the aorta (arrowhead) shows as a round and low-density filling defect. (B) Left kidney shows areas of low density, which is caused by multiple embolic infarcts. The aorta is filled with a low-density mass (arrowheads). (C) Sagittal reformatted image in the plane through the abdominal aorta shows an intraaortic mass that is attached to the anterior wall of abdominal aorta with upward and downward extensions (arrowheads).

![Transesophageal echocardiography](image2)

Fig 2. Transesophageal echocardiography. (A) Inhomogeneous echo-dense mass is attached to the aorta at 30cm from the incisor. (B,C) Rod-like mass with a central echo-dense area and outer membrane (arrowheads) in the descending thoracic aorta.
compatible with the histologic diagnosis of sarcoma (Fig 4).

**Discussion**

Since the first case described by Brodowski in 1873, 50–100 cases of primary tumors of the aorta have been reported in the English literature. These tumors are of various histologic types and the clinical signs and symptoms are nonspecific, such as headache, weakness or poorly localized chest and/or abdominal pain, because they are determined by the location and growth pattern of the tumors. The most common symptom is referable to vascular occlusion, as in the present case. As described by Wright et al, tumors primarily involving the intima tend to grow in the direction of the aortic lumen, forming polypoid masses. The clinical signs and symptoms are referable to the luminal obstruction and these tumors frequently give rise to thromboembolic metastases.

The diagnosis of primary aortic tumor is difficult because of its diverse clinical manifestation and rarity. Recently, with the advance of various diagnostic modalities, preoperative and antemortem presumptive diagnosis has become possible. In the present case, CT with contrast enhancement had a limited role in the evaluation because the findings were suggestive of thrombus. However, TEE revealed the inhomogeneous and echo-dense mass with an outer membrane, unlike a thrombus and suggestive of a primary aortic tumor.

Treatment of aortic tumors is difficult, because these tumors are rare and usually noticed late, and is based on the presenting symptoms. Recently, some cases were successfully treated by resection of the tumor mass and patch replacement. The preoperative presumptive diagnosis was aortic tumor, so it is important to do the appropriate diagnostic workup, such as magnetic resonance imaging and TEE. To our knowledge, this is the second case that has been diagnosed with TEE and we believe that it is the most valuable diagnostic modality because it can differentiate aortic tumor from thrombus and atheromatous plaque.

**References**