

## Intimal Sarcoma of the Thoracic Aorta; Clinical-course and Autopsy Finding

Kenji Akiyama, MD,<sup>1</sup> Kin-ichi Nakata, MD,<sup>1</sup> Nanao Negishi, MD,<sup>1</sup>  
and Akihiro Henmi, MD<sup>2</sup>

**We report a primary malignant tumor of the thoracic aorta with clinical-course from onset to death. A 63-year-old male was admitted to our hospital with intermittent claudication and bilateral lower extremity pain. The diagnosis was established after an adominal operation. The tumor was subsequently resected including the thoracic aorta and replaced with a dacron graft. The pathological finding was of a primary intimal sarcoma. The patient lived for more than two years excluding the hospitalized period after the diagnosis. (Ann Thorac Cardiovasc Surg 2005; 11: 135–8)**

**Key words:** aortic sarcoma, tumor embolism, pathological finding

### Introduction

Sarcomas of the aorta are extremely rare, and to our knowledge, 145 cases have been reported.<sup>1-10</sup> We report here a case of intimal sarcoma of the thoracic aorta with the pathological and autopsy findings.

### Case Report

A 63-year-old male patient was admitted to our hospital with intermittent claudication for 6 months and acute onset of bilateral lower extremity pain. Magnetic resonance imaging (MRI) showed obstruction in the area of the lower abdominal aorta and bilateral common iliac artery. Echocardiography of the chest showed no evidence of an intracardiac source of embolism. Therefore, we diagnosed the acute change as arteriosclerosis obliterans. We resected this lesion and replaced a Y-shaped prosthetic graft. After the operation, pathological examination revealed atypical cells in the thrombus. A transesophageal

echocardiogram, computed tomography (CT) scan (Fig. 1a) and MRI (Fig. 1b) revealed an intraluminal aortic mass in the proximal descending aorta. No other sources of embolism were discovered and a diagnosis of intraluminal tumor was made. Through a left thoracotomy, the proximal descending aorta was resected and reconstructed with a 6-cm-long prosthetic graft. The limits of resection were free of tumor on frozen section. The patient was discharged from hospital two weeks after the second operation.

One year after the second operation, he was re-admitted because of coldness of the bilateral lower extremity. MRI showed obstruction in the area of the lower abdominal aorta, but the area of the thoracic aorta was free from tumor. Technecium scintigraphy showed no bone metastasis.

We undertook replacement of the abdominal aortic lesion with a Y-shaped prosthetic graft. One year after the third operation, he was admitted again due to weakness of the bilateral lower extremity, tarry stool and renal dysfunction. Sarcoma was noted in the thoracic aorta and abdominal aorta by MRI. Chest X-ray showed right 7th rib metastasis.

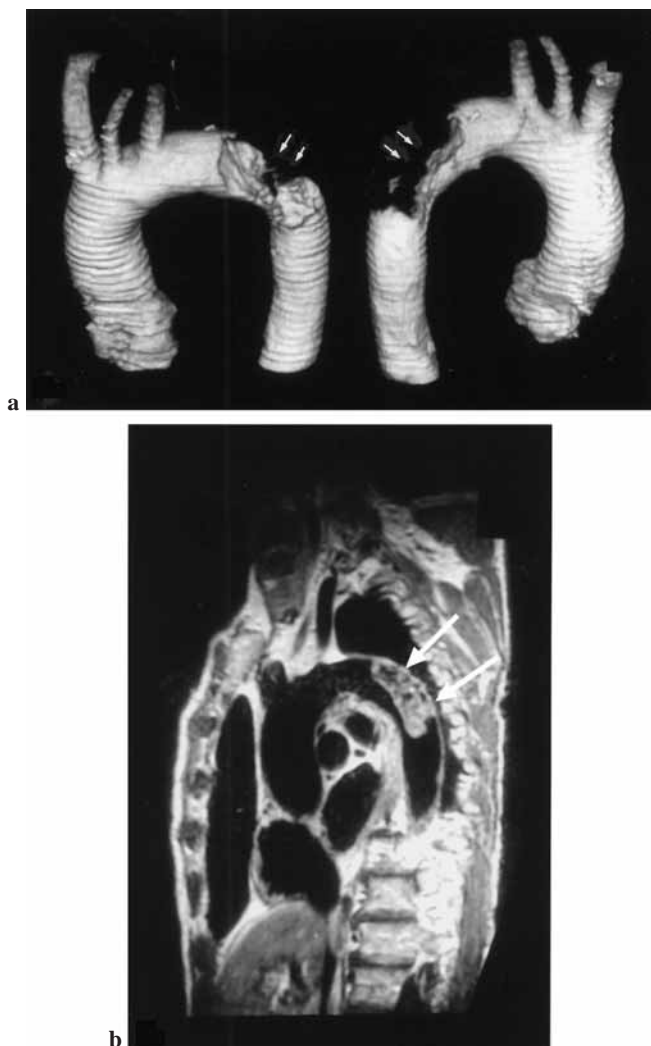
We tried to remove these tumors under extracorporeal circulation but we could not remove enough.

The patient died of multiple organ failure, one month after the last operation and 29 months after the initial embolic event.

From <sup>1</sup>Departments of Cardiovascular Surgery, and <sup>2</sup>Pathology, Nihon University School of Medicine, Tokyo, Japan

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Address reprint requests to Kenji Akiyama, MD: Department of Cardiovascular Surgery, Nihon University School of Medicine, 30-1 Oyaguchikami-machi, Itabashi-ku, Tokyo 173-0032, Japan.



**Fig. 1.** 3D-CT (a) and MRI (b) showing an intraluminal aortic mass in the proximal descending thoracic aorta. Clear arrow points to an intraluminal aortic mass.

### Histopathologic features

In the excised specimen, a tumor mass measuring 4×1.5 cm protruded into the lumen and filled the aorta, measuring 4.5×2.2 cm (Fig. 2a).

The tumor protruded from the tunica intima into the lumen and most of the tumor was necrotized. Tumor cells were observed only in the surface layer under a microscope (Fig. 2b). Electron microscopic examination showed microvilli, lumen formation and pinocytic vesicle in the tumor cells (Fig. 2c). Tumor cells were positive for vimentin and CD34, whereas all other immunohistochemical markers were negative.

From these findings, a diagnosis of intimal sarcoma was made.

### Autopsy finding

Autopsy was performed one hour after death. The procedure involved a detailed examination of all the thoracic and abdominal organs and the brain. Tumors occupied the thoracic and abdominal prosthetic grafts and large tumor emboli were found in the superior mesenteric artery and renal arteries. Microscopic emboli were found in all the organs examined except the heart, the lung and the brain. However, no evidence was seen of metastasis in the thoracic and abdominal organs.

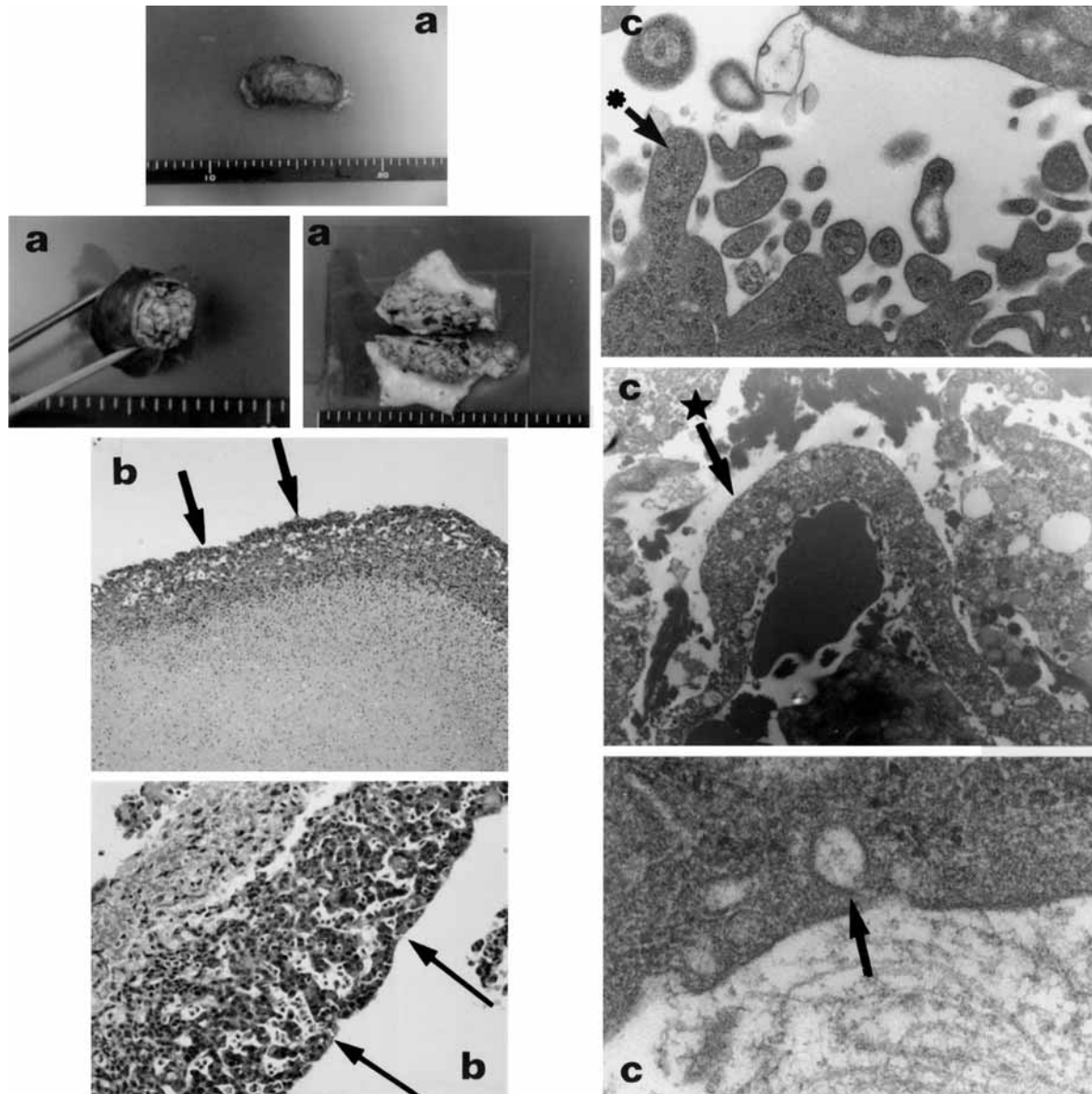
### Discussion

Primary tumor of the aorta is very rare. In the analyzable information of 145 patients including our patient from 142 reports,<sup>1-10)</sup> the male/female ratio was 9:5, and the age ranged from 3.5 months to 85 years old with a mean of 59.5 years old. The site of the origin was the thoracic aorta in 67, thoracoabdominal aorta in 36, and abdominal aorta in 39 patients. The tumor morphology was divided into two types,<sup>9)</sup> and was the intimal type in 90, mural type in 27, mixed in 14, and unknown in 11 patients.<sup>3)</sup>

The most frequent symptoms were due to embolic events, and the primary lesion was determined upon the pathological examination of the embolus in relatively many cases. Since the intimal type develops in the vascular lumen, the tumor is likely to metastasized via blood flow and cause embolic symptoms.

Surgical cases have recently been reported. In contrast, the mural type manifests few symptoms because it develops in the arterial wall, lacking a characteristic feature of imaging, and in many cases is not diagnosed before surgery, and is incidentally discovered during surgery. Many cases were autopsied. In both types, few symptoms were derived from the primary lesion, avoiding preoperative diagnosis. Although this patient developed embolic symptoms, only echocardiography of the chest was performed without consideration of this disease.<sup>3,9)</sup>

The only effective therapy was excision of the lesion with the arterial wall, and the prognosis was poor. The stump after excision of the primary lesion was negative for tumor in intraoperative rapid pathological examination, but tumor recurred in the stump of the excised abdominal aortic region one year after surgery, and in the stump of the primary lesion of the thoracic aorta one year later. The tumor recurred in the manner of tumor cells settling in the injured intimal region, similar to platelet thrombus formation, at the site of intimal damage. Bone



**Fig. 2.** Histological features.

In the excised specimen (a), a tumor protruded into the lumen and filled the aorta. The microscopic (b) and electron microscopic (c) appearance of the aortic tumor in the surgical specimen. Most of the tumor was necrotized and tumor cells were observed only in the surface layer (b: arrow).

Microvilli (c: \*), lumen formation (c: ★) and pinocytotic vesicles (c: →) were observed in the tumor cells.

metastasis observed 2 years after excision of primary lesion was also found in the region of the posterior intercostal artery that corresponded to the site of thoracic local recurrence. These findings suggested that take in the normal intimal region was not easy for the tumor cells due to the speed of aortic blood flow. Individual tumor cells were not readily released in blood flow, and when tumor cells grew to some extent and the central region of the tumor was necrotized, the tumor mass became unable

to resist the blood flow and caused embolism. At autopsy, a number of fresh multiple embolism that may have been caused by the final surgery were observed, but no distant metastatic lesion other than that observed in 7th rib was evident, supporting the hypothesis that the tumor cells were not yet distributed throughout the body. This was not our assumption at the establishment of diagnosis.

The life expectancy was about 8-14 months in many reports,<sup>1,4)</sup> and some physicians refrained from active

therapy.<sup>10)</sup> However, the patient lived normally for more than two years excluding the hospitalized period after the diagnosis, suggesting that excision of the primary lesion and additional excision of the abdominal aortic lesion were sufficient. Aortic tumor rarely develops, and the histologic type varies.<sup>1)</sup> In some types, recurrence occurs at the site of injured arterial intima. Formation of distant metastatic lesion is unlikely, as in this patient, suggesting that course observation by transesophageal echocardiography starting early after surgery, and surgery for prevention of multiple embolisms when local recurrence is found, may prolong the life of some patients. Long-term survival of patients has recently been reported, in which the patients received chemotherapy after surgery and survived without recurrence.<sup>6)</sup>

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