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Author(s) Alternative	Shimogawara, T; Ono, S; Kobayashi, K; Sasaki, A; Shimizu, H; Matsui, J
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Description	

# Aortic sarcoma mimicking a mycotic aneurysm in the thoracoabdominal aorta

Tatsuya Shimogawara, MD,<sup>a</sup> Shigeshi Ono, MD, PhD,<sup>a</sup> Kanako Kobayashi, MD,<sup>b</sup> Aya Sasaki, MD, PhD,<sup>c</sup> Hideyuki Shimizu, MD, PhD,<sup>b</sup> and Junichi Matsui, MD, PhD,<sup>a</sup> Chiba and Tokyo, Japan

## ABSTRACT

Aortic sarcoma is a rare primary tumor with dismal prognosis. Here, we report a case involving a 74-year-old female patient with aortic sarcoma masquerading as a mycotic aneurysm in the thoracoabdominal aorta. She underwent aortic resection with Dacron prosthetic graft replacement because of rapid growth. The postoperative pathological findings of the resected specimen confirmed the diagnosis of aortic mural sarcoma, which was an unexpected result based on repeat computed tomography angiography performed within 2 months preoperatively. The preoperative diagnosis of aortic sarcoma is often difficult because of its rarity, and this case demonstrates some of the diagnostic pitfalls. (*J Vasc Surg Cases and Innovative Techniques* 2019;5:593-6.)

**Key words:** Aortic mural sarcoma; Aortic intimal sarcoma; Gastric sarcoma; Gastrointestinal metastasis; Pseudoaneurysm

Aortic sarcoma is a rare primary tumor. Although accurate preoperative diagnosis is essential to completely resect the diseased aorta, the rarity of this tumor makes clinical differentiation difficult. We report a case of aortic sarcoma mimicking a saccular mycotic aneurysm on preoperative radiological images and clinical presentation.

The patient's family consented to the publication of the case details and images.

## CASE REPORT

A 74-year-old woman with a history of hypertension and hyperlipidemia presented to a hospital with a primary complaint of upper abdominal pain. Upper endoscopy revealed a 3- × 3-cm mass in the gastric greater curvature with ulceration, pathologically defined as necrotic tissue with nonspecific inflammation. Abdominal plain computed tomography (CT) revealed mild thickening of the gastric wall, slight enlargement of the thoracoabdominal aorta, and celiac trunk occlusion (Fig 1, a). Figure 1 shows the morphological change of the thoracoabdominal aorta in time series. No other remarkable findings were detected. Her symptoms initially resolved spontaneously.

Two months later, she returned to our hospital complaining of persistent upper abdominal pain, general fatigue, and appetite loss. She had low-grade fever and mild tenderness of the upper abdomen. Physical examination was otherwise unremarkable. Laboratory tests revealed elevated white blood cell count and serum C-reactive protein level ( $16.6 \times 10^4/\mu\text{L}$  and 21.7 mg/dL, respectively); the procalcitonin level was normal. Repeat blood cultures taken on admission were negative. Enhanced CT showed a 29-mm saccular thoracoabdominal aortic aneurysm and occlusion of the celiac trunk (Fig 1, b and d). No contrast enhancement surrounding the aneurysm wall was found. Upper endoscopy demonstrated a 7- × 4-cm submucosal tumor on the gastric greater curvature, with necrotic ulceration (Fig 2, a and b). The gastric sarcoma was diagnosed pathologically. A broad-spectrum antibiotic was administered intravenously for a presumptive diagnosis of mycotic aortic aneurysm, but her fever persisted despite intensive therapy.

Two weeks later, repeat CT showed rapid enlargement of the aneurysm and disruption of the anterior wall (Fig 1, c and e), leading to the confident diagnosis of impending aortic aneurysm rupture. She underwent emergent repair through a left-flank thoracoretroperitoneal approach. The aneurysm showed severe adhesions to the surrounding retroperitoneal soft tissue, without any fluid collection. The celiac trunk was occluded with dense intimal thickening, reflecting the preoperative images. The saccular aneurysm was excised along with the adjacent inflammatory soft tissue, and in situ thoracoabdominal reconstruction with a 20-mm rifampicin-soaked Dacron graft was performed. The abdominal branches, including the celiac artery, superior mesenteric artery, and bilateral renal arteries, were all reconstructed, and the prosthetic graft was wrapped with the greater omentum.

Cultures obtained from the resected specimens, including the aortic wall and surrounding retroperitoneal inflammatory tissues, showed negative results. Microscopic examination of the resected aortic segments showed large, poorly differentiated atypical cells with a palisading arrangement (Fig 3, a and b). Although the cells mainly occupied the outer layers of the aorta,

From the Department of Surgery<sup>a</sup> and Department of Pathology,<sup>c</sup> Tokyo Dental College, Ichikawa General Hospital, Chiba; and the Department of Cardiovascular Surgery, Keio University School of Medicine, Tokyo.<sup>b</sup>

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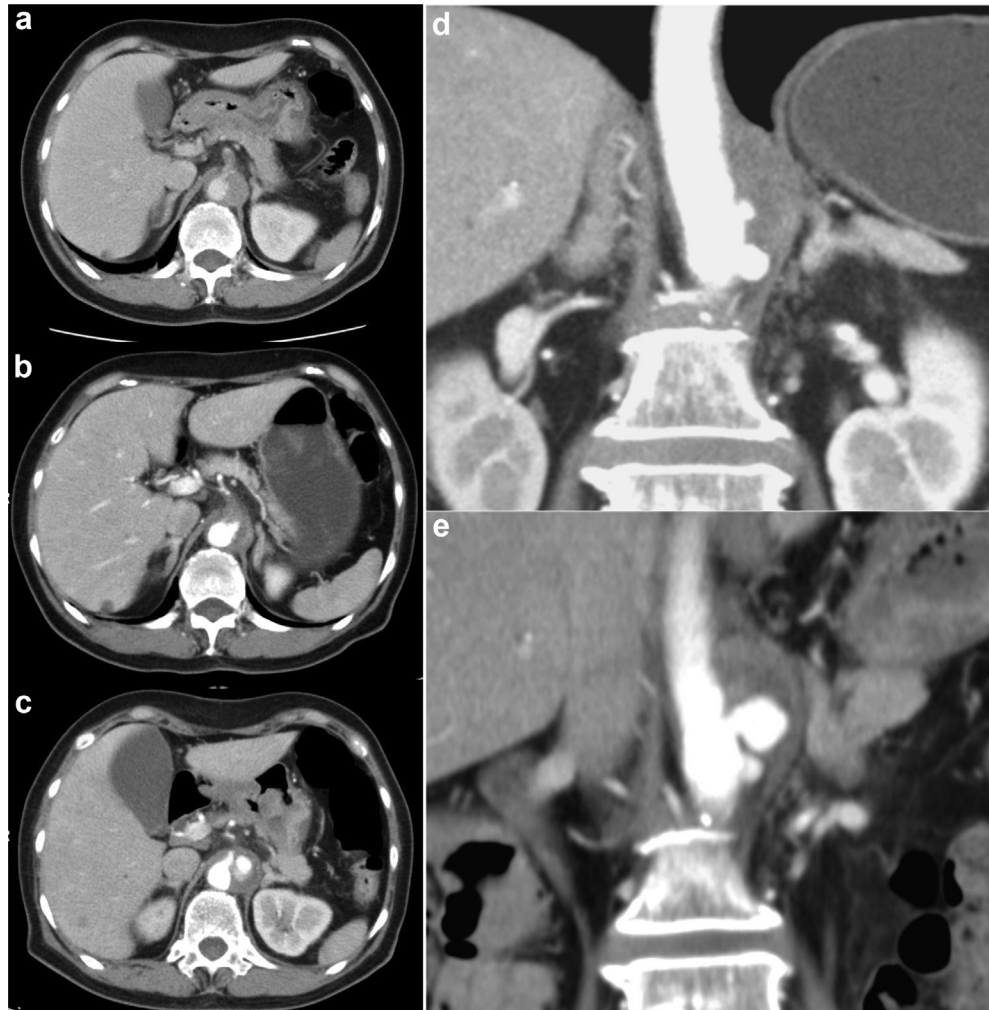
Correspondence: Shigeshi Ono, MD, PhD, Department of Surgery, Tokyo Dental College, Ichikawa General Hospital, 5-11-13 Sugano, Ichikawa, Chiba 272-8513, Japan (e-mail: [sono@tdc.ac.jp](mailto:sono@tdc.ac.jp)).

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**Fig 1.** Representative preoperative computed tomography angiography images of the aorta performed (a) 60 days, (b and d) 14 days, and (c and e) 1 day before the operation. The thoracoabdominal aorta showed rapid expansion accompanied by celiac trunk occlusion.

the internal lamina was also partially destroyed, with consequent spread of the tumor cells from the adventitia to the intima. Immunohistochemical studies showed positive staining for CD68 only, representing nonspecific differentiation; this finding matched that of the gastric sarcoma; thus, aortic mural sarcoma with gastric metastasis was diagnosed clinicopathologically.

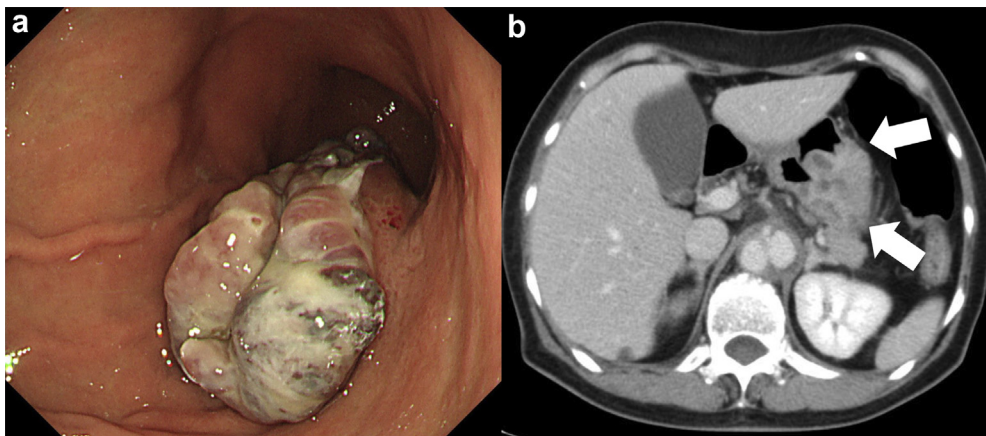
Intravenous broad-spectrum antibiotic therapy was continued postoperatively until the pathological findings revealed the existence of aortic sarcoma. The patient showed continued general fatigue and low-grade fever postoperatively. She developed antibiotic-related encephalopathy and acute renal failure requiring temporary hemodialysis. CT performed 2 months later demonstrated regrowth of the aortic sarcoma and multiple new lesions of the lungs, liver, skin, and sacral spine, interpreted as systemic metastasis. Her general condition rapidly deteriorated while awaiting adjuvant chemoradiotherapy, and she was deemed ineligible for these treatments. She died of multiple organ failure 86 days after the operation.

Autopsy revealed local tumor recurrence of the anastomosed aorta, with intimal thickening of the anastomosed aortic wall demonstrating dense proliferation of spindle cells, presumed to be the main cause of the distant metastases.

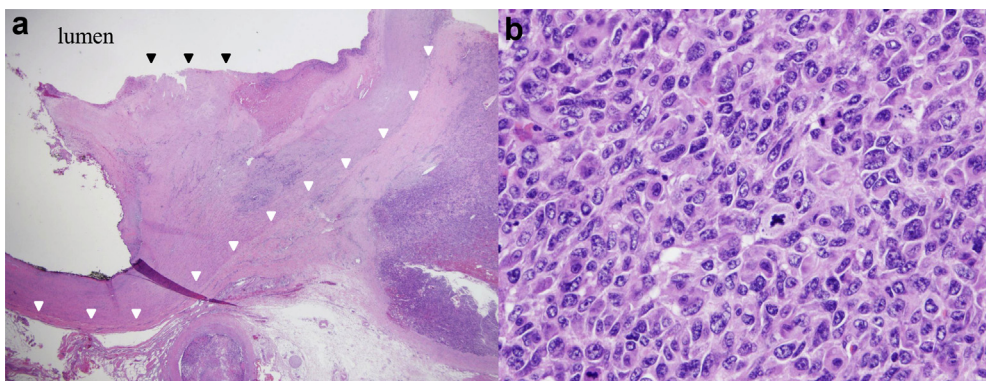
## DISCUSSION

Primary malignant aortic sarcoma is extremely rare, with only 140 cases reported.<sup>1</sup> In 1873, Brodowski documented the first case of aortic sarcoma. This tumor shows a slight male predominance, and occurs at a mean age of 60 years.<sup>2</sup> The descending thoracic aorta is the most common site, followed by the abdominal aorta and thoracoabdominal aorta.<sup>2</sup>

Thalheimer et al. differentiated aortic sarcomas clinicopathologically into intimal and mural subtypes.<sup>3</sup> Sarcomas of the intimal subtype originate from the intima and express endothelium-specific immunohistochemical markers, such as CD31 and/or factor VIII.<sup>4</sup> It commonly forms a luminal mass, causing arterial



**Fig 2.** Gastric metastasis on the greater curvature was detected on upper endoscopy and enhanced computed tomography. **a**, Representative endoscopy image; **b**, dense thickening of the gastric wall is evident (*white arrows*: lesion).



**Fig 3.** Hematoxylin and eosin-stained images of the resected aorta. Large, poorly differentiated cells mainly occupied the adventitia, and malignant cell invasion from the adventitia to the intima was present (*white arrows*: external lamina). The internal lamina was partially damaged (*black arrows*). **a**,  $\times 40$ ; **b**,  $\times 400$ .

obstruction and distal embolization. The mural subtype is less frequent, develops from the media or adventitia, and involves the extra-aortic soft tissues. This subtype rarely interrupts the aortic circulation. It commonly lacks the immunohistochemical expression of endothelial markers.<sup>4</sup> In our case, the primary tumor showed extraluminal growth and no aortic obstruction, clinically similar to the mural subtype.

Although preoperative diagnosis of aortic sarcoma is important for performing adequate resection, it is difficult to distinguish sarcoma from a mycotic aneurysm using common radiological modalities. CT and ultrasonography are performed routinely to evaluate aortic diseases; however, these techniques lack specific imaging characteristics. The characteristic features of aortic sarcoma, such as pseudoaneurysm-like tumor formation and rapid aortic expansion, are remarkably similar to those of saccular mycotic aneurysms. Although aortic sarcoma is rarely diagnosed preoperatively owing

to this difficulty, some reports on the efficacy of fluorodeoxyglucose-positron emission tomography have been published.<sup>5-7</sup> Fluorodeoxyglucose-positron emission tomography could be helpful for distinguishing aortic sarcomas from mycotic aneurysms because the standardized uptake value of a sarcoma tends to be higher.<sup>5-9</sup>

In the current case, the aortic and gastric lesions were preoperatively considered to be mutually independent diseases. However, the postoperative pathologies revealed the simultaneous existence of aortic and gastric sarcomas—one as the primary tumor, whereas the other was a result of metastasis. The aortic lesion was distantly separated from the greater curvature, suggesting that direct invasion by the gastric sarcoma was unlikely. Additionally, tumors resulting from metastases to the aorta are rare in comparison to primary tumors forming in the aorta. Consequently, the aortic sarcoma was diagnosed as the primary tumor. The primary aortic sarcoma

extending from the adventitia to the intima destroyed the lamina intima, and the hematogenous metastatic pathway from the celiac trunk to the gastroepiploic artery presumably led to gastrointestinal metastasis. As demonstrated in this case, the greater gastric curvature is reported as the main gastrointestinal metastatic site for soft-tissue sarcomas.<sup>10</sup>

There is no well-established management for aortic sarcomas. Complete surgical resection of the tumor and surrounding tissues may prolong survival<sup>11</sup>; however, in the majority of reported cases, aortic sarcomas were unexpected diagnoses resulting in insufficient resection of the diseased aortic lesions and adjacent tissue.<sup>1-3,6,11-13</sup> These facts also highlight the difficulty in making a preoperative diagnosis of aortic sarcoma. The efficacy of multidisciplinary treatment combining complete surgical resection, adjuvant chemotherapy, and radiation therapy has been reported,<sup>3,6,11,12,14,15</sup> but long-term survival is extremely poor because of widespread metastatic potential.<sup>15</sup> The 5-year survival rate is approximately 10%,<sup>1</sup> with a mean survival of 12.8 months.<sup>3</sup>

In conclusion, aortic sarcoma is a very rare entity with a poor prognosis, which may mimic mycotic aneurysm. Accurate preoperative diagnosis is of great importance to determine an appropriate treatment option.

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