A case of primary aortic sarcoma with tumor infarction after stent graft placement

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Hiroki Nakamura, MD[1], Akihiko Kanki, MD[1], Hiroyuki Watanabe, MD[1], Kentarou Ono, MD[1], Noriaki Kuwada, MD[2], Shinsuke Saisho, MD[3], Hirotake Nishimura, MD[4], Akira Yamamoto, MD[1] and Tsutomu Tamada, MD[1]

Abstract

Primary aortic sarcoma is a very rare disease, and most primary aortic tumors are malignant mesenchymal tumors. We present the case of a 62-year-old man with sudden epigastric and back pain. Contrast-enhanced computed tomography (CT) revealed a mass lesion about 33.8 mm in diameter, in contact with the left side of the abdominal aorta. Impending rupture of an abdominal aortic aneurysm was suspected, so cardiovascular surgery for stent graft placement was performed the same day. Symptoms immediately improved and CT at 3 months postoperatively showed a marked decrease in lesion size, but the lesion subsequently grew again. Fluorodeoxyglucose (FDG)-positron emission tomography/CT was performed due to the possibility of malignant solid tumor, revealing markedly increased FDG accumulation (maximum standardized uptake value, 36.95) in the mass lesion. Primary aortic sarcoma was diagnosed from thoracoscopic biopsy. Here, we report a primary aortic sarcoma that shrank due to tumor infarction after stent graft placement, followed by tumor regrowth.

Keywords

Primary aortic sarcoma, tumor infarction, stent graft placement, computed tomography, magnetic resonance imaging

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Introduction

Primary aortic sarcoma is a very rare disease, and most primary aortic tumors are malignant mesenchymal tumors. The pathology was first reported by Brodowski in 1873,[1] and 170 cases have since been reported.[2] These tumors are reportedly more common in men, with a male-to-female ratio of 2:1, predominantly occurring around 60 years old.[3] They occur most often in the descending thoracic aorta, followed by the abdominal and thoracoabdominal aorta.[4] Aortic sarcomas are classified into intimal or mural types based on their location.[5] Among aortic sarcomas, 80% are intimal type and the remaining 20% are mural type.[1] The intimal type grows in the lumen along the intima, while the mural type grows outward, with the main seat being the media and adventitia. Vascular occlusion is unlikely without intimal infiltration. In contrast, in the intimal type, vascular occlusion is the main symptom, and intermittent claudication, mesenteric artery occlusion, renal artery stenosis, occlusion-related hypertension, and sexual dysfunction might also be observed.[1] Mural-type lesions, on the other hand, are not associated with vascular occlusion and can be asymptomatic. Symptoms that do appear are typically non-

[1]Departments of Radiology, Kawasaki Medical School, Kurashiki, Japan
[2]Department of Cardiovascular Surgery, Kawasaki Medical School, Kurashiki, Japan
[3]Department of General Thoracic Surgery, Kawasaki Medical School, Kurashiki, Japan
[4]Department of Pathology, Kawasaki Medical School, Kurashiki, Japan

Corresponding author:
Hiroki Nakamura, Departments of Radiology, Kawasaki Medical School, 577 Matsushima, Kurashiki 701-0192, Japan.
Email: h.nakamura@med.kawasaki-m.ac.jp

specific, including dyspnea, dysphagia, low back pain, weight loss, and malaise.[1] Most cases have developed metastases by the time of diagnosis.[6] Bone, lungs, liver, and the skin are considered the most common sites of metastasis, in that order.[7,8] Generally, the prognosis of aortic sarcoma is poor, with a 5-year survival rate of 2.6% for intimal-type and 29.8% for mural-type tumors.[1] Computed tomography (CT) and magnetic resonance imaging (MRI) are important for differentiation from wall thrombosis and atherosclerosis for intimal-type lesions,[2] and from aortic aneurysm and infectious aortic aneurysm for mural-type lesions.[9–11]

Here, we report the case of a 62-year-old man who underwent aortic stent grafting for suspected impending rupture of an abdominal aortic aneurysm that was later diagnosed as primary aortic sarcoma.

Case

A 62-year-old man presented with sudden epigastric and back pain, for which abdominal contrast-enhanced CT was performed. CT showed a mass-like lesion about 33.8 mm in diameter, in contact with the left side of the abdominal aorta and showing poor contrast effect (Figure 1). On suspicion of impending rupture of an abdominal aortic aneurysm, the patient underwent cardiovascular surgery for stent graft placement on the same day. At that time, intraoperative digital subtraction angiography did not reveal any clear contrast findings in the tumor. Symptoms improved after the stent graft was placed. MRI performed 3 days after stent graft placement showed a rim-like high-intensity signal on T1-weighted imaging (T1WI) and a low-signal area at the tumor margin on T2-weighted imaging (T2WI) (Figure 2). The tumor displayed a marked reduction in size (to about 18.5 mm) on CT performed 3 months after stent graft placement, but had then regrown to about 41.7 mm on contrast-enhanced CT performed 7 months after stent graft placement. Moreover, while the lesion had previously appeared hypovascular, an internal non-uniform contrast effect was also observed at this time (Figure 3). In addition, multiple nodular shadows appeared in the lung fields on CT. The patient did not report any symptoms. Fluorodeoxyglucose (FDG)-positron emission tomography (PET)/CT was performed on suspicion of primary malignant tumor of the retroperitoneum with lung metastases, revealing marked

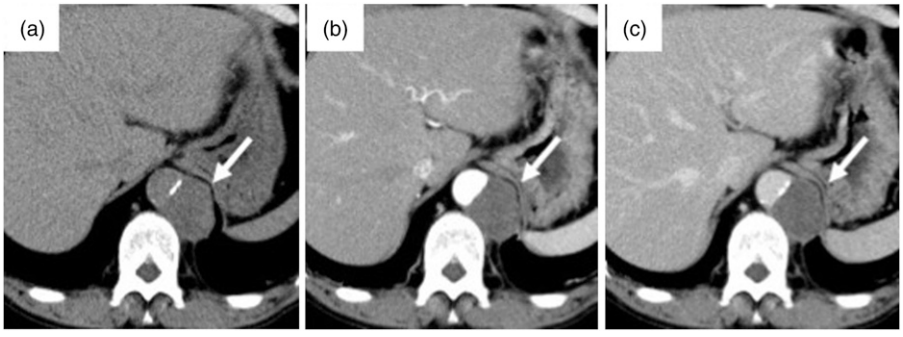

Figure 1. Contrast-enhanced computed tomography (CT) of the abdomen from the first scan. (a) Plain CT shows a mass-like lesion about 33.8 mm in diameter (arrow) in contact with the left side of the abdominal aorta. (b) and (c) contrast effect is poor in the arterial phase (b) and equilibrium phase (c).

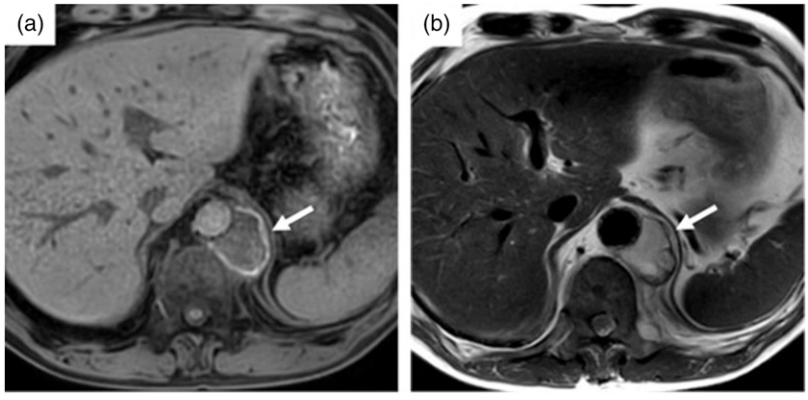

Figure 2. Magnetic resonance imaging performed 3 days after stent graft placement shows a rim-like, high-intensity signal on T1-weighted imaging (a) (arrow) and a low-signal area on T2-weighted imaging at the tumor margin (b) (arrow).

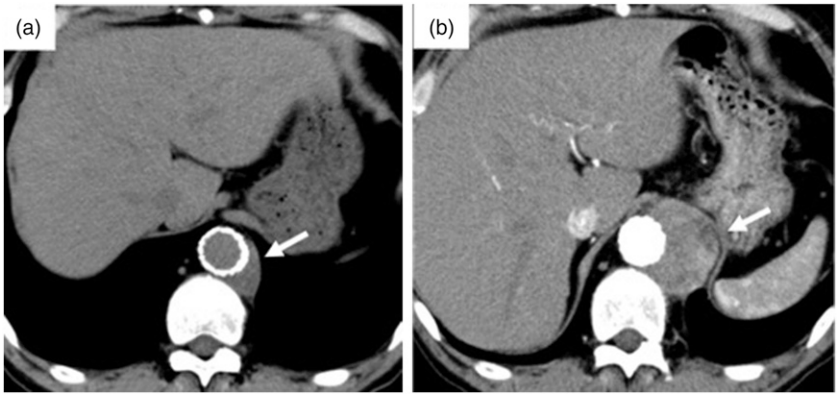

Figure 3. The tumor shows a marked reduction in size (to about 18.5 mm) on plain computed tomography (CT) 3 months after stent graft placement (a), but has increased to about 41.7 mm in diameter on arterial-phase contrast-enhanced CT 7 months after stent graft placement (b). Furthermore, the mass appears hypovascular at 3 months (a) (arrow), but displays an internal, non-uniform contrast effect at 7 months (b) (arrow).

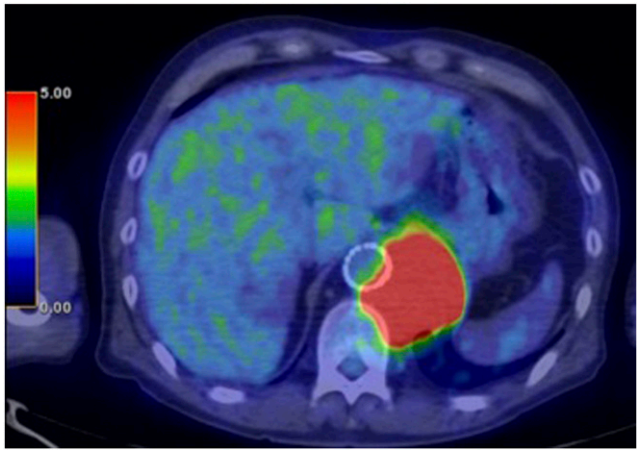

Figure 4. Fluorodeoxyglucose (FDG) positron emission tomography/computed tomography performed 7 months after stent graft placement shows a marked increase in FDG accumulation (maximum standardized uptake value = 36.95) in the mass lesion (arrow).

uptake of FDG (maximum standardized uptake value = 36.95) in the mass lesion (Figure 4). Thoracoscopic biopsy was conducted to confirm the diagnosis, as a malignant solid tumor was suspected. Since the biopsy report suggested the possibility of aortic sarcoma, partial resection of the middle and lower lobes of the right lung, resection of the tumor involving the aorta and ascending aorta–abdominal aorta bypass surgery were performed. Macroscopic examination of the resected aortic tumor showed a solid, whitish mass continuous with the aorta, along with some dark red blood clots inside the tumor (Figure 5). Microscopic evaluation showed dense proliferation of cells with a high degree of nuclear atypia, with spindle-shaped cells growing in bundles apparent in some areas. The tumor mainly occupied the media and adventitia, with partial infiltration of the intima. Coagulation components and hemosiderin deposits were observed along with post-bleeding changes in the lesion. Immunostaining showed positive results for the endothelial markers avian v-ets erythroblastosis virus E26 oncogene homolog, α-smooth muscle actin, and podoplanin, but negative results for CD31 and CD34. Based on these observations, high-grade, mural-type primary aortic sarcoma was diagnosed. As of 6 months postoperatively, the patient showed no sign of local recurrence or metastasis.

Discussion

Aortic sarcoma is a rare disease with no specific imaging findings, making preoperative diagnosis difficult.[1,2] This entity is classified into intimal and mural types, each with different imaging findings and differential diagnoses. Intimal-type aortic sarcoma needs to be distinguished from mural thrombus, while mural-type sarcoma must be differentiated from aortic aneurysm, closed-type aortic dissection, infectious aortic aneurysm, and type II endoleak after stent graft placement. Although contrast-enhanced CT and MRI can aid in the diagnosis, contrast effects are often poor. Increased FDG accumulation on FDG-PET/CT is also useful in reaching the correct diagnosis.[3] Ultrasonography is not very useful in the diagnosis since distinguishing between tumors and thrombus on ultrasonography is difficult.[3] Contrast-enhanced CT/MRI in cases of aortic sarcoma often shows an irregular, lobulated surface, with an appearance referred to as "shaggy aorta."[2] Imaging can also reveal other tumors in cases with extramural extension, absence of significant atheromatous disease (e.g., lack of intimal calcification) in the lesion, internal heterogeneous imaging effects, neovascularization, increased accumulation on FDG-PET/CT, lymph node metastasis, and distant metastasis.[2]

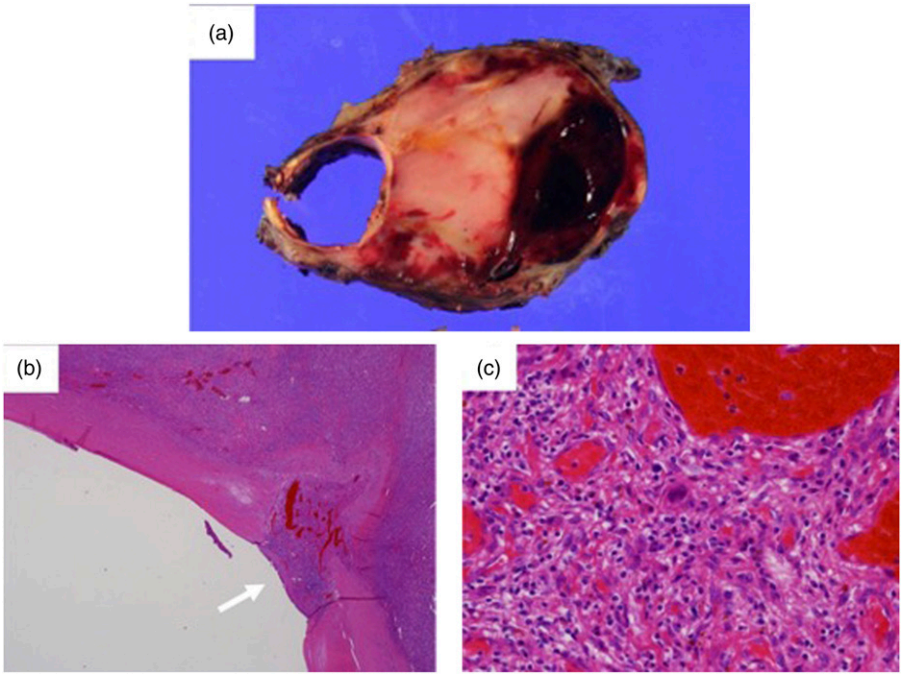

Figure 5. Gross evaluation of the specimen shows a solid, whitish mass continuous with the aorta, along with some dark red blood clots (a). Microscopic evaluation of hematoxylin and eosin-stained images show-s the tumor mainly occupies the media and adventitia, with partial infiltration of the intima (arrow) (b). Tumor cells show dense proliferation with a high degree of nuclear atypia (c). b, ×40; c, ×400.

In the present case of mural-type primary aortic sarcoma, the contrast effect on the first contrast-enhanced CT was very poor, and the diagnosis of solid tumor was missed. Therefore, the patient received endovascular stent graft placement under a misdiagnosis of impending rupture of an abdominal aortic aneurysm. Several cases of aortic sarcoma treated by endovascular stent graft placement due to suspected thoracic aortic aneurysm have been reported.[9,10,13] In those cases, PET/CT and biopsy were performed due to increases in lesion size despite an absence of endoleak after stent graft placement, leading to the correct diagnosis of aortic sarcoma. Those studies made no mention of temporary reductions in lesion size after stent placement or changes in contrast effects over time. In our case, MRI after placement of the stent graft showed a rim-like high-intensity signal at the tumor margin on T1WI and a low-signal area on T2WI. The T1WI hyperintensity in this case was attributed to the presence of methemoglobin, and the T2WI hypointensity was due to deoxyhemoglobin in the lesion, both of which were considered to represent tumor infarction.[14] In the present case, tumor infarction probably occurred following placement of the stent graft, leading to a temporary decrease in tumor diameter due to necrosis of the tumor tissue. Further, contrast effects were enhanced during the course of tumor regrowth after placement of the stent graft. Aortic sarcomas often show a poor contrast effect, although tumor ischemia and surgical invasion reportedly promote tumor growth, new blood vessel growth and metastasis.[15] Likewise in this case, angiogenic hyperplasia may have developed due to the infarction following stent graft placement. This report describes a very rare course of aortic sarcoma, consisting of lesion shrinkage due to tumor infarction after placement of the aortic stent graft, followed by tumor regrowth.

In conclusion, primary aortic sarcoma is a rare disease that lacks specific imaging findings. However, a diagnosis of primary aortic sarcoma should be considered in patients showing an atypical presentation of aortic aneurysmal disease such as absence of significant atheromatous disease—and internal heterogeneous imaging effects in the lesion on contrast-enhanced CT/MRI.

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Declaration of conflicting interests

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ORCID iD

Akihiko Kanki https://orcid.org/0000-0002-2458-5598

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