Pleomorphic undifferentiated aortic sarcoma presenting as persistent endoleak after endovascular aneurysm repair

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ABSTRACT

Sarcomas of the aorta are rare tumors with an unknown incidence and wide variety of clinical presentations. These malignant neoplasms are often manifested in an advanced state and with symptoms of nonmalignant vascular disease owing to a delay in diagnosis. We present the case of a 78-year-old man in whom workup was initially performed for a persistently enlarging abdominal aortic aneurysm after endovascular aortic repair but who was subsequently found to have a pleomorphic undifferentiated sarcoma of the aorta. (J Vasc Surg Cases and Innovative Techniques 2019;5:294-7.)

Keywords: Angiosarcoma; Endoleak; EVAR; Aneurysm

Repair of abdominal aortic aneurysms (AAAs) is primarily performed with an endovascular rather than an open approach because of improved morbidity and faster recovery without inferiority in repair.[1,2] Primary malignant sarcomas of the aorta are often manifested as nonmalignant vascular disease, such as aneurysms, endoleaks after endovascular repair, and embolic phenomena. We present the case of a pleomorphic undifferentiated sarcoma discovered during an attempted open abdominal aorta repair with explantation of an aortic stent graft. The patient's wife provided written consent for publication of this report and images.

CASE REPORT

A 78-year-old man was referred to a tertiary care medical center for an enlarging infrarenal AAA with endoleak after initial endovascular repair in 2007 with a Gore Excluder stent graft (W. L. Gore & Associates, Flagstaff, Ariz). He had a type II endoleak with persistent dilation of the aneurysm and underwent lumbar artery coiling in 2009. He subsequently underwent proximal aortic cuff and distal bilateral iliac stent placement, but the aneurysm continued to enlarge. He complained of mild abdominal pain radiating to his back, which had been present since his initial stent graft placement. He took prednisone 5 mg daily for polymyalgia rheumatica and had a >50-pack-year smoking history. His family history was not significant for malignant disease. On examination, he was a thin, elderly man with a palpable midabdominal mass extending to the left of the abdomen. He was not cachectic and denied any unexpected weight loss leading up to his preoperative evaluation. Computed tomography angiography (CTA) revealed an AAA measuring 8.4 × 8.2 cm (Fig 1) and several liver lesions identified as cysts (Fig 2).

He presented in February 2017 for elective open repair of the infrarenal AAA with explantation of all previously placed endovascular aneurysm repair stents. On gaining of entry into the abdomen and exposure of the infrarenal aorta, there was an 8.4-cm mass extending from the aorta into the retroperitoneum with multiple areas of nodularity (Fig 3). Intraoperative biopsy specimens were taken of this mass and sent for pathologic examination, which revealed pleomorphic undifferentiated sarcoma. The surgical oncology team was consulted, and a biopsy specimen was taken from one of many liver lesions, revealing sarcoma. With the diagnosis of metastatic sarcoma, the decision was made to abort the operation and to notify the patient of the new diagnosis.

The patient recovered from surgery, and the medical oncology service was consulted; outpatient staging with positron emission tomography/computed tomography and chemotherapy were recommended. The patient declined further treatment, was discharged home on hospice care, and died several months later. Final pathologic examination revealed high-grade spindle and pleomorphic sarcoma with lymphovascular invasion (Fig 4) and positive CD68 staining (Fig 5) obtained from both the aortic and the liver mass biopsy specimens. CD68 staining identifies cells in the monocyte lineage and has been associated with intimal vascular sarcoma.[3]

DISCUSSION

Aortic sarcomas are rare; 165 cases have been reported in the literature,[3] with the first case reported by Brodowski[4] in 1873. Aortic sarcomas often are manifested with symptoms of nonmalignant vascular disease, such as aortic aneurysm, acute arterial embolization, abdominal pain, nausea and vomiting, dissection, and aortic rupture,[3] which may explain the frequent delay in diagnosis. The distribution of primary aortic sarcoma evolves with increasing reports of aortic disease. Current data suggest that the thoracic aorta makes up 46% of cases; abdominal

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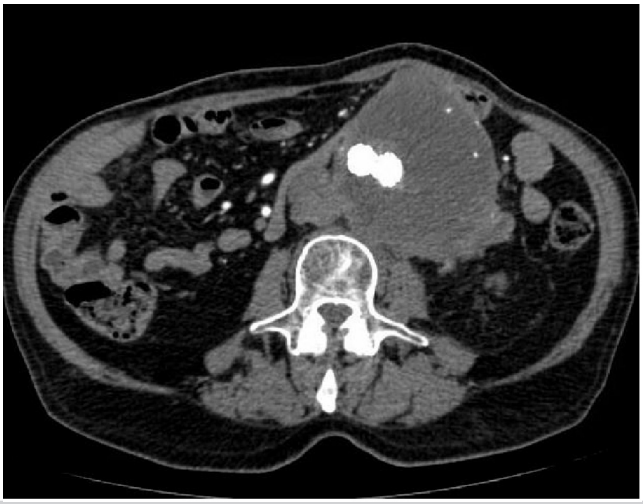

Fig 1. Preoperative computed tomography angiography (CTA) image of infrarenal aortic aneurysm with aortic sarcoma.

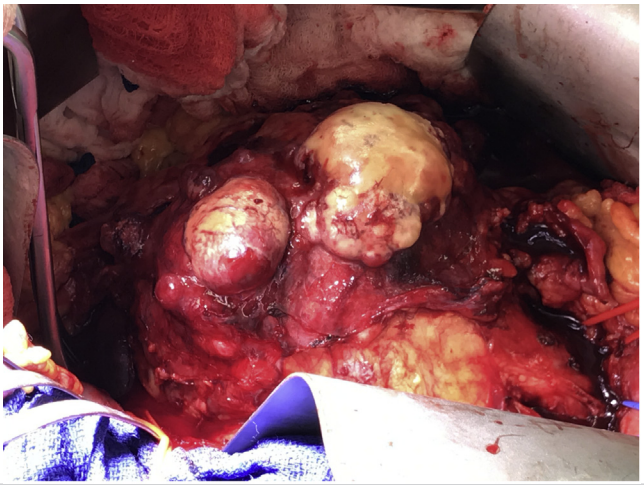

Fig 3. Large exophytic neoplastic mass involving the retroperitoneum identified as an aortic sarcoma. The *right* of the image is the cranial direction. The *red vessel loop* is around the left renal vein.

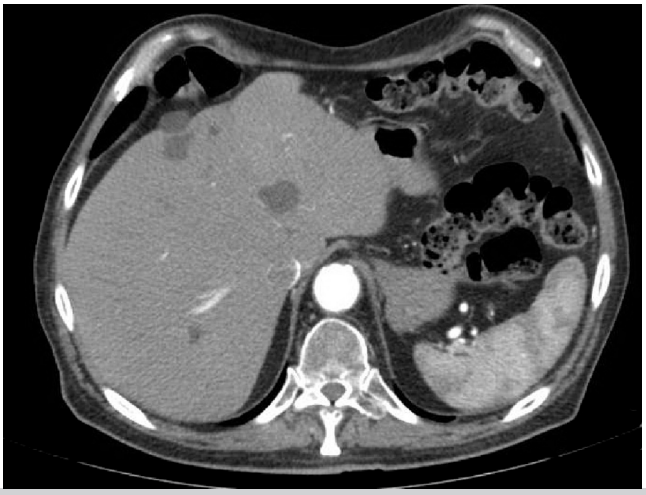

Fig 2. Preoperative computed tomography angiography (CTA) image showing liver lesions reported as hepatic cysts.

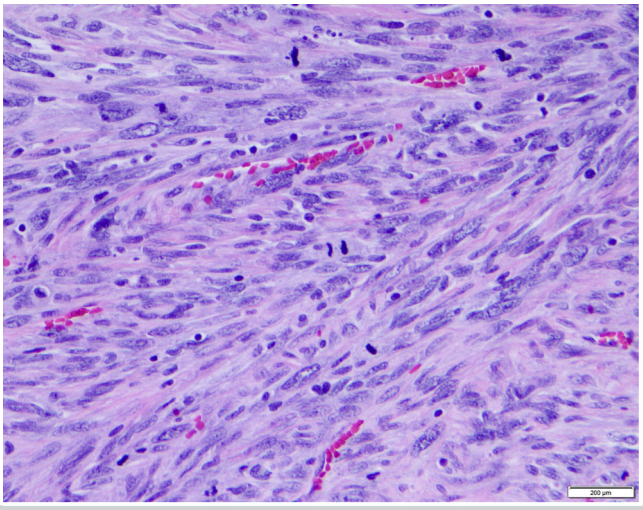

Fig 4. Intraoperative aortic wall/retroperitoneal mass biopsy specimen showing pleomorphic sarcoma with spindle cell morphology, nuclear pleomorphism, and numerous mitoses (hematoxylin and eosin stain, magnification ×200).

aorta, 27% to 42%; and thoracoabdominal aorta, 12% to 25%. The aortic arch was involved in 26% of cases.[5,6]

The malignant neoplasms can arise from the adventitia, media, or intima. Primary intimal lesions are the most common and primarily are manifested with embolic events or aortic obstruction.[3,5,7] Tumors arising from the adventitia or media tend to form mural tumors with extravascular growth and aortic wall compromise.[3] Aortic arch sarcomas of intimal origin are uniquely associated with cerebral emboli and early metastasis to the brain.[3] Undifferentiated histology is the most common at 39%, followed by angiosarcomatous at 37%, with leiomyosarcomas and fibrosarcomas significantly less common.[3]

The diagnosis of primary aortic sarcoma is difficult because the presenting symptoms are similar to those of nonmalignant vascular disease. With the trend toward endovascular intervention, there is less opportunity for examination of aortic specimens to determine malignant potential.[2] The diagnosis of aortic sarcoma is commonly made during open aortic repair.[6-8] Many imaging modalities have been evaluated, including CTA, arteriography, and magnetic resonance angiography (MRA). Unfortunately, there appear to be no specific imaging characteristics on CTA; the most common pathologic changes represent dissection, aneurysm, or intraluminal filling defects.[9] MRA appears to offer the

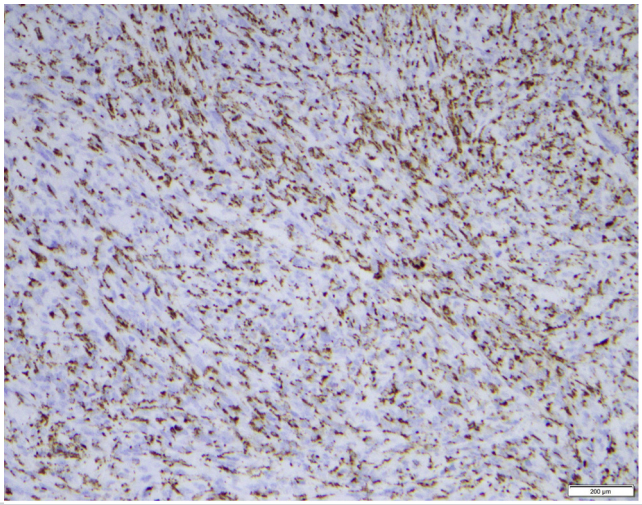

Fig 5. Aortic biopsy specimen tumor cells positive for CD68 as denoted by *brown staining* (CD68 immunostain with hematoxylin counterstain, magnification ×100).

most diagnostic potential, with lobulated heterogeneously enhancing masses involving the aorta being associated with malignant disease.[10-12] However, it remains difficult to identify the populations who benefit from MRA evaluation; some argue for MRA evaluation in those with uncharacteristic disease (ie, aortic thrombus without aneurysm or aortic dissections without risk factors), those with intraluminal masses, or those with lesions in bone or lung and liver as these are common points of sarcoma metastasis.[12] MRA could also be expanded to those with a refractory endoleak as this may be a manifestation of aortic sarcoma masquerading as an endoleak, as noted in our patient and others in the literature.[5]

Treatment of aortic sarcoma remains difficult, with overall 5-year survival at 8% and 50% mortality at 1 year.[2] The most commonly accepted avenue of treatment is en bloc resection of the tumor with bypass grafting, which is believed to offer the greatest possibility of cure.[5,13] Palliative bypass grafting, endarterectomy, and stenting have been proposed for those with metastatic disease. There is some limited evidence for the use of chemotherapy in the treatment of aortic sarcomas in combination with surgery, but the overall survival outcome remains poor.[6] The factor most prognostic for survivability is that of metastatic disease on diagnosis, which appears to affect nearly 80% of known cases; the most common sites of spread include the lung, bone, skin, and liver.[12,14,15]

A phenomenon noted in several case reports and murine models is the association of angiosarcoma at sites of aortic Dacron grafts and polytetrafluoroethylene dialysis grafts.[16-19] The graft can be that of an open replacement or endovascular repair. The patient often presents many months or years after intervention with development of symptoms such as pain and enlarging pseudoaneurysm. Patients often undergo multiple examinations and interventions before an open repair is undertaken whereby the diagnosis is made.[5,14,20,21] The time from graft placement to tumor diagnosis varies (3 months-7 years) but averages 48 months.[3] In the majority of graft-associated aortic sarcoma cases, patients already suffer from metastatic disease and their survival is marginal. As in nongraft-associated aortic sarcomas, MRA may offer higher sensitivity of detection, but this has yet to be formally proven.

CONCLUSIONS

We describe the case of a 78-year-old man diagnosed with metastatic pleomorphic undifferentiated aortic sarcoma at the site of a previously placed Gore Excluder device during attempted explantation for persistent aneurysmal enlargement. Aortic sarcomas are a rare disease with poor outcomes probably related to a delay in diagnosis, allowing disease metastasis. In review of our preoperative imaging, the expanding aortic sac with asymmetric wall morphology and cystic liver lesions were the only findings suggesting a neoplastic process. Recognizing aberrant symptom patterns associated with aortic sarcoma and the use of MRA may allow early diagnosis and improved outcomes.

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