

Angiosarcoma of the abdominal aorta after endovascular aneurysm repair

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ABSTRACT

Primary aortic sarcoma is a rare diagnosis that carries a poor prognosis. This case report features a 68-year-old man, treated 4 years earlier with an endovascular aortic aneurysm repair, who presented with fever, low back discomfort, and abdominal pain. Given the concern for an infected endograft, the patient underwent explantation and replacement with a cadaveric aortoiliac cryograft. Ultimately, the pathology returned as an angiosarcoma. Although endovascular aortic aneurysm repair is the gold standard for abdominal aortic aneurysm repair in patients with suitable anatomy, there are trade-offs associated with less invasive approaches compared with open approaches. (*J Vasc Surg Cases and Innovative Techniques* 2019;5:506-8.)

Keywords: Angiosarcoma; EVAR; Aortic sarcoma

Angiosarcoma of the abdominal aorta is an extremely rare malignancy that carries a poor prognosis. Even rarer are those diagnosed in the setting of an endovascular aortic aneurysm repair (EVAR). Soft tissue sarcomas represent less than 1% of all cancer types and angiosarcomas compose 1% to 2% of those soft tissue sarcomas.¹ A poor prognosis is thus affected by the rarity of diagnosis, an insidious presentation, and progression of disease with metastatic potential. Endovascular repair of abdominal aortic aneurysm has become increasingly more common. Lost in this technique, versus an open approach, is the ability to make an earlier diagnosis, as well as possible curative resection. Moreover, in the setting of a possible endograft infection, we suggest sending specimen(s) for pathologic examination in a comprehensive workup. We present a case of angiosarcoma masquerading as an infected EVAR. The patient agreed to have his case details and images published in this report.

CASE REPORT

A 68-year-old man with a history of chronic lymphocytic leukemia (CLL) presented to the emergency department with

a 2-week history of fever, low back discomfort, and abdominal pain. Past medical history was significant for an infrarenal abdominal aortic aneurysm that was repaired four years prior with a Gore Excluder EVAR (W. L. Gore & Associates, Flagstaff, Ariz). His postoperative course was unremarkable. One year after the repair, he was noted to have a type II endoleak on imaging surveillance (*Fig 1*). This endoleak was managed nonoperatively with serial imaging over the subsequent four years. Of note, 4 months before his presentation, imaging surveillance was also notable for a new right adrenal gland nodule. Given his history of CLL, the patient underwent a transgastric fine needle aspiration of the suspicious nodule. Pathology was inconclusive for disease recurrence and both the patient and oncologist agreed on close observation.

Admission laboratory evaluation at the emergency department revealed leukocytosis (white blood cell count of $23 \times 10^9/L$) with a left shift, an elevated C-reactive protein (72 mg/L) and an elevated erythrocyte sedimentation rate (90 mm/h). Multiple sets of blood cultures were negative. Computed tomography (CT) findings (*Fig 2*) were notable for aortic wall thickening with surrounding lymphadenopathy concerning for infection. The aneurysm sac with the known type II endoleak was slightly larger compared with outpatient surveillance scans. A tagged white blood cell scan was negative for infection. A positron emission tomography (PET) scan was unable to be obtained secondary to insurance logistics. Our working differential diagnosis included endograft infection, worsening endoleak, recurrence of CLL, or aortitis.

Despite the institution of empiric broad-spectrum antibiotics, the patient continued to have a leukocytosis and abdominal pain. Given the concern for endograft infection, the patient underwent explantation of his EVAR, reconstruction with cadaveric aortoiliac cryograft (CryoLife, Kennesaw, Ga), and wide local debridement of the aortic wall including adjacent necrotic lymph nodes. Significant periaortic inflammation with lymphadenopathy was noted at the time of operation without frank purulence. In addition, there was mild

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Fig 1. Computed tomography (CT) surveillance scan 4 months before presentation with a type II endoleak.



Fig 2. Computed tomography (CT) scan at the time of presentation with an increase in the size of the aneurysm sac and significant periaortic thickening and lymphadenopathy.

bilious staining, likely from his previous transgastric biopsy of his right adrenal nodule. Intraoperative cultures were negative.

Given his history of CLL and concern for disease recurrence, the aortic wall was sent as specimen for pathologic examination. The aortic wall pathology was significant for a 4.2-cm epithelioid angiosarcoma, present at the margin, with foci of lymphovascular invasion (Fig 3). The patient recovered well from the operation. He has since been seen by us as an outpatient as well as hematology-oncology and radiation oncology.

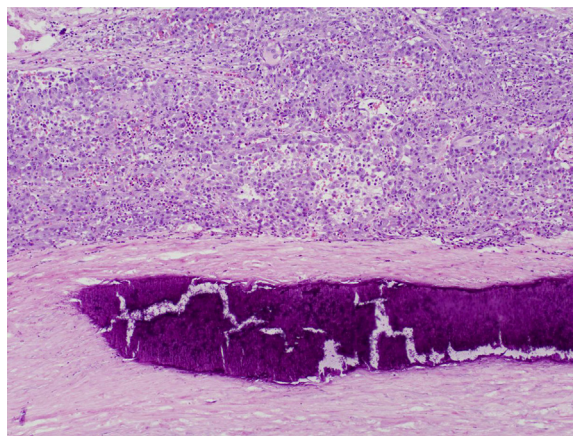


Fig 3. Abnormal cancer cells next to native calcific aortic wall (original magnification $\times 100$).

Recommendations included chemotherapy, but no radiation therapy or discussion of further oncologic resection. Postoperative imaging at 2 months was notable for rapid progression of disease with invasion of his lumbar vertebrae and emergence of hepatic lesions. He is considered to have a very poor prognosis from the angiosarcoma and has been initiated under various chemotherapy regimens, currently on gemcitabine.

DISCUSSION

Soft tissue sarcomas, let alone angiosarcomas, are an extremely rare form a malignancy. More common examples include a gastrointestinal stromal tumor, liposarcoma, or leiomyosarcoma. Angiosarcomas most commonly manifest as cutaneous lesions of the head and neck in elderly white men. Most arise spontaneously, and risk factors include radiation, chronic lymphedema, exogenous toxins, and familial syndromes. Hallmark pathologic findings include abnormal, pleomorphic, and malignant endothelial cells.² Staging is based on the Tumor-Node-Metastasis (TNM) system. However, unlike soft tissue sarcomas in general, angiosarcomas are considered high-grade tumors by definition and therefore do not include histological grading. Angiosarcomas have an overall 5-year survival rate of about 35% with a median survival of 7 months. Given the rarity of the disease and subsequent paucity of published data in the literature, treatment options lack uniformity. R0 resection is recommended at initial resection to improve overall survival rate in patients with angiosarcoma. However, more than 50% of patients with primary angiosarcoma have metastasis at the time of presentation which is why the 5-year survival is so poor.^{1,2} The National Comprehensive Cancer Network guidelines address angiosarcoma indirectly within the broader category of sarcoma treatment.³ Recommendations are R0 resection with no specified margin size other than a tumor-free microscopic margin. Resection should involve all structures seeming to be involved in the primary tumor

with preservation if vital structures seem to not be invaded. This should be confirmed by a pathologist to confirm disease-free microscopic margins, which is often after completion of the operation. Leaving surgical clips in the resection bed can aid in adjuvant radiation therapy. Consideration of repeat resection to obtain disease-free margins is strongly recommended in the event of R1 or R2 resection.² Finally, patients with metastatic disease should be offered palliative treatment with chemotherapy and radiation therapy.⁴ For metastatic angiosarcoma, recommended cytotoxic chemotherapy regimens include anthracyclines, ifosfamide, and taxanes.² Biological therapies have shown some progress, but require further studies.

Milite et al¹ described a 60-year-old male with a similar presentation of abdominal pain and an EVAR 7 years prior. Given the concern for infection, that patient also underwent an extra-anatomic bypass and eventual explant of the endograft, but ended up expiring postoperatively owing to his complicated hospital course.¹ Fenton et al⁵ reported on a 66-year-old man with back pain and weight loss in the setting of an EVAR 6 years prior. Imaging demonstrated a lytic lesions on the spine and biopsy confirmed an angiosarcoma. That patient underwent palliative spine surgery and outpatient chemoradiation, and expired 1 month after diagnosis.⁵ Fatima et al⁴ described a case series of 13 patients, over the course of 26 years, with a primary angiosarcoma of the aorta, great vessels and the heart. These investigators included a 57-year-old patient with an EVAR 6 years earlier who presented with back pain and, fevers, and weight loss. That patient underwent an endograft explantation and replacement with a rifampin-soaked Dacron graft for presumed infection. His pathology came back as metastatic angiosarcoma and he expired 2 months postoperatively. Fatima et al⁴ go on to quote a median survival of 14 months (range, 1-75 months). Kamran et al⁶ also noted mean overall duration of survival at 14 ± 2.4 months and 3- and 5-year survival rates of 11.2% and 8.0%, respectively.

With an atypical presentation of an infected endograft, one should have a heightened degree of suspicion when evaluating the CT imaging. Kamran et al⁶ noted the following imaging characteristics to consider: protrusive vegetations or nodular soft tissue components, lack of atherosclerosis in the area of suspicion, heterogeneous thrombus, evidence of enhancement and neovascularity,

persistent enlargement of the excluded sac, and avid fluorodeoxyglucose uptake on the PET study. They went on suggest multimodal imaging, such as magnetic resonance angiography or PET, to aid in the diagnosis. Furthermore, when possible, a tissue biopsy would definitively change the treatment options.

CONCLUSIONS

Although EVAR is considered by many to be the gold standard for abdominal aortic aneurysm repair in patients with suitable anatomy, there are trade-offs associated with less invasive approaches compared with open repair. This case highlights the missed opportunity for aortic tissue biopsy and possible curative resection in a patient with a rare vascular malignancy. It is crucial that vascular surgeons pay close attention to the noncontrast CT images of the aorta on surveillance to recognize any abnormal lymphadenopathy or wall enhancement.

In the era of duplex imaging for EVAR surveillance to avoid contrast and radiation, it is possible that opportunities may be missed to evaluate the aortic wall. In a possibly infected endograft, one should also consider sending the specimen for pathology during an explantation. The late but vague clinical presentation delayed diagnosis and potentially allowed disease progression, leading to a poor prognosis.

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