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Aortic angiosarcoma masquerading as a thoracic aortic aneurysm

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Aortic angiosarcoma is an exceedingly rare clinical entity. Significant delay in diagnosis can occur due to a low index of suspicion on the part of the clinician. We report a case of aortic angiosarcoma masquerading as a descending thoracic aneurysm arising from a penetrating ulcer. The patient was initially treated with an endovascular stent graft for rapid growth, but the lesion continued to enlarge despite angiographic exclusion. FDG-PET CT scan and biopsy ultimately confirmed the diagnosis of aortic angiosarcoma. This case highlights some of the difficulties of making the early diagnosis of aortic angiosarcoma. (J Vasc Surg 2009;50:1477-80.)

Aortic sarcomas are an extremely rare, malignant entity with fewer than 145 cases reported in the literature to date. The clinical presentation and imaging studies frequently mimic aneurysmal disease or aortic occlusive disease. As such, there is often a delay in diagnosis. Despite aggressive management, patients with aortic angiosarcomas have a median survival time of only a few months. We report a case of a primary mural aortic angiosarcoma of the descending thoracic aorta presenting as an enlarging aortic aneurysm in the setting of a penetrating ulcer.

CASE PRESENTATION

A 68-year-old male presented to the hospital with intermittent abdominal pain radiating to the left chest for two months and no other complaints. Physical examination was unremarkable. Of note, the patient experienced postprandial abdominal pain, as well as weight loss of approximately 20 pounds over the last ten months. His past medical history included hypertension, hypercholesterolemia, asthma, and a significant smoking history of 45 pack-years.

Given the patient's presentation and history, workup was completed to rule out occult gastrointestinal (GI) malignancy with an upper endoscopy, ultrasound, and abdominal computed tomography (CT) scan. Although the endoscopy and ultrasound were unremarkable, the CT scan with contrast revealed what was presumed to be a descending thoracic aortic aneurysm secondary to a penetrating ulcer (Fig 1). There was noted to be an eccentric outpouching of contrast associated with an intramural hematoma and degeneration of the aorta just beyond the ulceration. The aneurysm was found to be above the level of the diaphragm, terminating 2 cm proximal to the origin of the celiac axis. Given the multiple comorbidities of this patient in the setting of focal disease, endovascular repair was recommended and electively scheduled. During this interval period, however, the patient returned with worsening abdominal pain. At this visit, the patient had a repeat abdominal CT demonstrating a 5-mm increase in diameter of the lesion (Fig 2). In light of the rapid growth and the worsening epigastric pain, urgent repair was recommended. The aneurysm was treated with endovascular placement of an aortic stent-graft with no postoperative complications (Fig 3). Two months following stent graft repair of the aneurysm, the patient returned complaining of new-onset back pain. A chest CT with contrast was obtained showing a 2 cm interval increase in the size of the soft tissue between the thoracic spine and the thoracic aorta (Fig 4). Of note, there was no demonstrated endoleak or device migration. The differential diagnosis at the time included possible mycotic aneurysm versus a primary aortic malignancy. Of note, the patient's white blood cell count (WBC) was always normal, but he had an elevated C-reactive protein (46.7 mg/L, nml 0-8) and erythrocyte sedimentation rate (ESR; 114 mm/Hr, nml 0-15). A fluorodeoxyglucose-positron emission tomography (FDG-PET) scan demonstrated hypermetabolic activity in the rim of the lesion, as well

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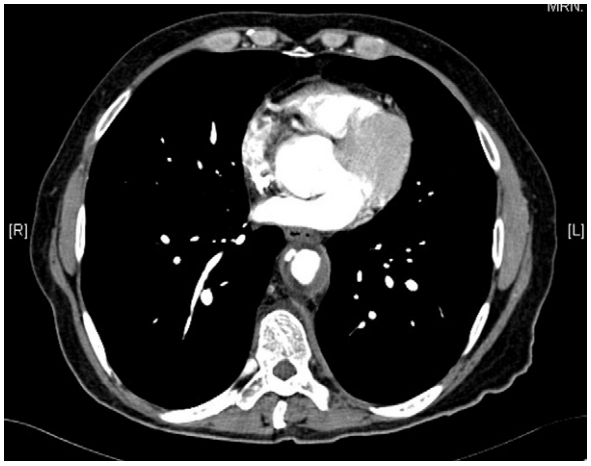

Fig 1. A computed tomography (CT) scan with contrast prior to graft repair of patient's presumed aneurysm. Note the eccentric outpouching of contrast and wall thickening.

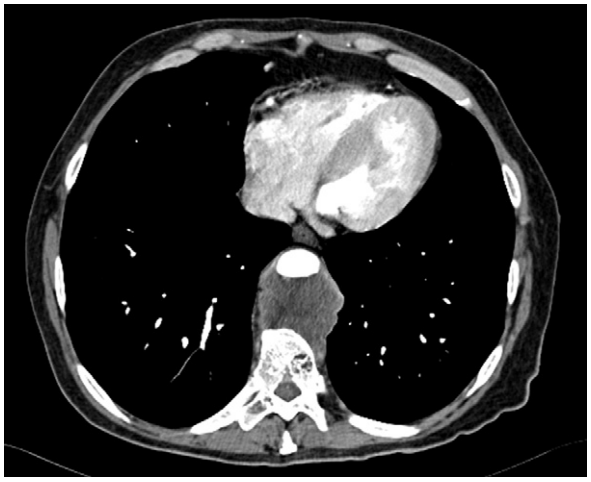

Fig 2. A preoperative computed tomography (CT) scan with contrast one month after initial visit and workup for isolated abdominal pain. The maximal aortic diameter measured from this CT scan was 60 mm, 5 mm larger than before.

as a single focus of metabolic activity in the spleen (Fig 5). The splenic lesion was new when compared to prior CT. A CT-guided biopsy of the mass was performed, which only demonstrated necrotic tissue. A left video-assisted thoracoscopic biopsy of the lesion was completed. Exploration of the chest revealed a large, complex, fungating mass of the distal descending thoracic aorta that was friable. Confirmation with frozen tumor section demonstrated cells consistent with an aortic angiosarcoma invading the lung parenchyma. Stains for cytokeratins AE1/AE3, CAM5.2, CK5/6, CD34, CD31, S100, WT1, TTF-1, actin, and desmin were negative. Palliative radiation and chemotherapy was discussed with the patient, but he declined treatment and ultimately expired four months later.

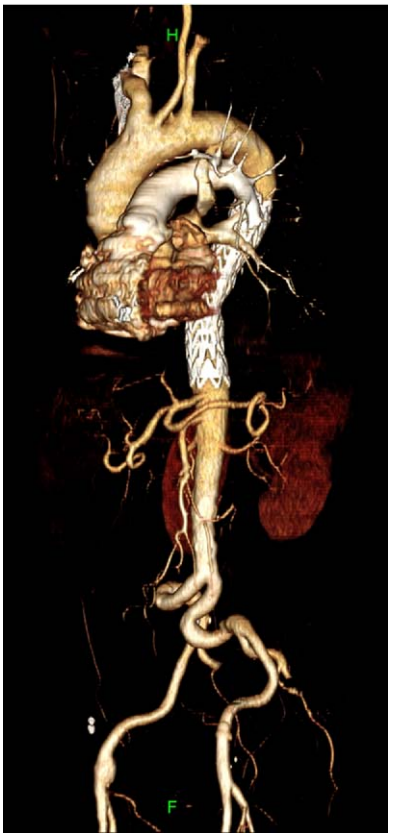

Fig 3. A 3-D reconstruction image status post endograft deployment demonstrating graft location in the thoracic aorta.

DISCUSSION

Primary aortic sarcomas are an extremely rare entity, with fewer than 145 reported in the literature to date.[1-6] Between 24 and 34 of these cases have been specifically identified as aortic angiosarcomas.[6,7] While both thoracic and abdominal aortic sarcomas have the same incidence, aortic angiosarcomas occur more frequently in the descending thoracic aorta.[1,7,8] They also occur more frequently in men, with a male-to-female ratio of 2:1. The average age of those affected is 62 years, with a range of 46 to 85 years.[1,2,5-7] There are no established risk factors for developing aortic angiosarcomas to date.[9,10]

Clinically, most patients with aortic sarcomas present with symptoms that are non-specific. Symptoms are typically a result of either tumor embolization, local mass effect, or luminal obstruction. One review of 135 aortic angiosarcomas found that 25.9% of patients complained of pain on presentation.[2] Rare complications seen with aortic sarcomas are aortic rupture, cerebral infarction, deep vein thrombosis, spontaneous paraplegia, and gastrointestinal hemorrhage secondary to metastasis.[1-7] This patient's initial complaints of abdominal pain and 20-lb weight loss were not felt to be related to the lesion in the descending thoracic aorta at the time of presentation. Part of the delay in therapy was due to the workup to exclude an occult GI malignancy.

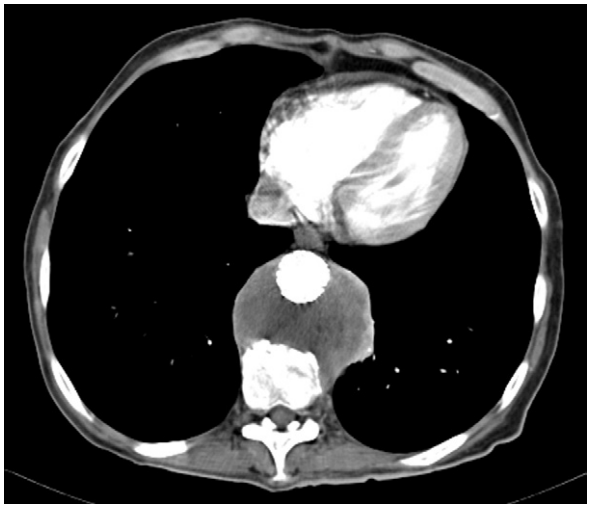

Fig 4. A computed tomography (CT) scan with contrast taken two months postoperatively when the patient returned to the hospital complaining of back pain. The lesion increased significantly in size since the last visit to a maximal diameter of 80 mm. Of note, there is no appreciable extravasation of contrast into the growing lesion.

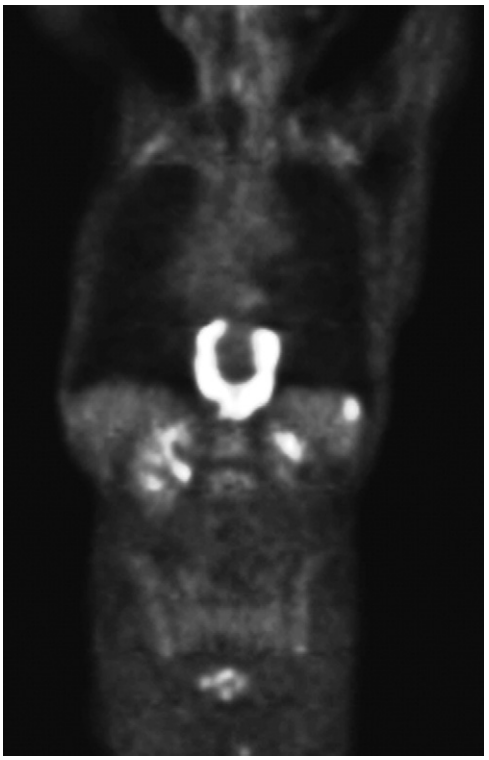

Fig 5. A coronal section of the positron emission tomography (PET) demonstrating hypermetabolic foci in the wall of the aorta and in a single focus of the spleen.

The similarity of presenting symptoms between aneurysmal disease, arteriosclerotic aortoiliac occlusive disease, and aortic sarcomas makes diagnosis challenging.[4,6,7] While most imaging modalities reveal what is presumed to be severe mural disease, magnetic resonance angiography (MRA) may be able to differentiate atheromatous lesions from neoplastic tissue. MRA may be able to distinguish these two entities on the basis of increased enhancement of tumor by gadolinium.[5,7,11-14] To date, no specific features on imaging that would suggest the diagnosis of aortic sarcoma have been reported in the literature. In our patient, the effacement of the surrounding tissues and the rapid growth of the lesion raise the suspicion for a malignancy; however, these findings can also be seen in the setting of a penetrating ulcer or a mycotic aneurysm, both of which are much more common entities. The patient did not manifest any clinical signs of infection, and the lesion grew despite being excluded by the stent graft. These facts led us to consider a primary malignancy as a possibility. Once the diagnosis of a primary malignancy is suspected, FDG-PET CT may be of clinical utility. Increased metabolic activity may be seen in the wall of the aorta, and distant metastases may be identified, as was the case in our patient.

Histological classification of aortic angiosarcomas can be accomplished by staining for distinct tumor markers. Immunohistochemical staining for pankeratins (AE1, AE3, S-100 and CAM5.2), endothelial markers (CD31, Qbend10/CD34, WT-1, von Willebrand factor/Factor VIII-related antigen and FLI-1), vimentin, desmin, and actin are targeted to endothelial and myofibroblastic cells, which are thought to give rise to angiosarcomas.[5,6,15,16] While useful confirmatory tests, many tumor markers have limited utility given their vastly variable sensitivity and specificity in identifying aortic angiosarcomas. CD31 and CD34, for instance, are classically regarded as highly sensitive and specific markers for endothelial angiosarcomas but have demonstrated inconsistent yield with 33%-92% positivity in angiosarcomas.[17,18] As such, a three-reagent immunohistochemical panel of CD31, CD34, and von Willebrand factor is recommended, as it has a higher sensitivity and specificity in identifying endothelial angiosarcomas more than any one individual marker.[16]

Treatment of aortic sarcomas has proven to be, at best, mildly therapeutic. This is likely a combined result of a highly aggressive cancer and nonspecific diagnostic tools lending to late diagnosis. Treatment includes en bloc resection of affected aortic tissue and periaortic soft tissue with graft interposition of the resected aortic segment.[2,4] In patients who are not good surgical candidates or who have already developed distant metastases, chemotherapy and adjuvant radiation may offer some palliation.[2,5,6] The life expectancy at the time of diagnosis is 14 to 27 months, with a mean overall survival length of 16 ± 2.4 months and a range of 0 to 168 months.[1,2] Cumulative overall survival rates for 111 cases of primary aortic tumors at three and five years were 11.2% and 8%, respectively. After surgical intervention, the survival rates at three and five years increased to 16.5% and 11.8%, respectively.[2]

The rarity of aortic angiosarcomas makes establishing a diagnosis challenging. Clinical presentation and imaging

findings are often non-specific. Magnetic resonance angiography (MRA) and PET-CT may help in establishing the diagnosis of aortic sarcoma. Vascular pathology with immunohistochemical stains are important confirmatory tests in effectively identifying and managing this aggressive and rare neoplasm.

AUTHOR CONTRIBUTIONS

Conception and design: SE
Analysis and interpretation: VR, SE
Data collection: VR, SE
Writing the article: VR
Critical revision of the article: VR, SE
Final approval of the article: SE
Statistical analysis: N/A
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