Intimal angiosarcoma of the thoracic aorta

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ABSTRACT

Introduction: Sarcomas of the great vessels are uncommon, with aortic being the rarest. Only 30 cases of true intimal aortic sarcomas (IAS) are documented. They tend to occur in the abdominal aorta, with less common occurrences in the thoracic aorta. Their growth patterns, predispose them to a propensity for metastases and cause embolic phenomenon. Case Report: A 58-year-old male presented with chest pain and dyspnea and was evaluated for pulmonary embolus and coronary artery disease. Computed tomography angiography (CTA) demonstrated no pulmonary emboli; however, there was severe atherosclerosis/thrombosis of the aortic arch. The process extended centrally, nearly filling the entire lumen. The surgery consultant advised anticoagulation and strict blood pressure control, recommending that the patient come to the outpatient department for surgery. Due to personal reasons, the patient failed to return at the recommended time. Three months after initial presentation the patient was admitted for surgical replacement of the aorta. The surgeon reported the aorta as “chock-full of fibro-fatty material nearly obstructing its course”. The pathology report was aortic sarcoma of intimal origin. Conclusion: Aortic sarcomas are rare tumors, with the intimal subtype in the thoracic aorta being even rarer.

Delay in diagnosis of these tumors often occurs, since the imaging features are nearly identical to atherosclerotic disease. Since atherosclerotic disease is clearly more frequent than intimal sarcoma of the aorta, it is not difficult to understand that this diagnosis is not usually made until after surgical resection or at autopsy. At presentation, nearly all of the patients have metastatic disease.

Keywords: Aorta, Sarcoma, Computed tomography

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doi:10.5348/ijcri-2013-01-263-CII-17

INTRODUCTION

Primary sarcomas of the great vessels (aorta, pulmonary artery and vena cava) are extremely uncommon tumors, with aortic sarcomas being the rarest type; about 26% occurring in aorta compared to about 37% each for pulmonary and vena caval sites. They are sub-classified as either mural or intimal tumors, with intimal tumors characterized as poorly differentiated on histology [1, 2]. Latest reviews, using the strict histological definition of intimal sarcoma reported that only about 21–30 cases of true intimal aortic sarcomas (IAS) have been documented, with the mean age of presentation being 62.2 years [3, 4].

Here, we present a case of undifferentiated intimal aortic sarcoma which is unusual both in location and presentation. This patient’s tumor was located within the thoracic aortic arch, and he presented with chest...
pain, instead of the more typical secondary symptoms of embolic phenomena.

CASE REPORT

The patient was a 58-year-old African-American male with a past medical history significant for type two diabetes mellitus. He presented with chest pain and dyspnea on June 4, 2010 and was subsequently evaluated for a pulmonary embolus as well as coronary artery disease. Cardiac enzymes were negative for indication of ischemic changes. Computed tomographic angiography (CTA) of the chest demonstrated no pulmonary emboli; however, the imaging revealed severe suprarenal atherosclerosis/thrombosis of the aortic arch extending to the descending thoracic aorta, stopping at the level of the diaphragm. The process extended centrally, nearly filling the entire aortic lumen. The adrenal glands were free of involvement (Figures 1–2).

The vascular surgery consultant placed the patient on anti-coagulation therapy (warfarin 5 mg) and strict blood pressure control (metoprolol 50 mg and lisinopril 20 mg), recommending that he return as an outpatient for surgery on the aortic arch and descending thoracic aorta. Due to personal and other preoperative medical reasons (full mouth dental extraction), the patient’s presumed elective aortic surgery was delayed until September 2010.

As a result of chest pain, he visited our emergency department (ED) twice in the month of August 2010 and each time was ruled out for cardiac ischemia. Both times he was continued on his regimen of anti-coagulation and blood pressure control. A computed tomography (CT) scan was performed on both ED visits, demonstrating a new right adrenal mass (2×2 cm) that was not present on initial CT scan, in addition to the previously identified severely diseased aorta (Figure 3). The second time he presented to the ED, he was admitted for one week.

Three months after initial presentation, on September 23, 2010 the patient was admitted for surgical replacement of the descending thoracic aorta with a tube graft utilizing a right axillary artery cannulation site and left femoral artery and vein cannulation site. The surgeon reported the thoracic aorta as “chock-full of fibro-fatty material nearly obstructing its course, blending to a more normal appearing aorta at the diaphragm and in the proximal aortic arch”.

The pathology report described the specimen as “multiple fragments of opaque, yellow-tan to pink-gray friable soft tissue in aggregate measuring 6.2x5.2x3.4 cm. Sectioning revealed markedly friable, partially laminated, yellow-white to gray-brown cut surfaces”. The final pathology report unexpectedly revealed undifferentiated aortic pleomorphic sarcoma of intimal origin (Figure 4).

The patient was discharged from the hospital two weeks later. However, he returned to the ED after one week, complaining of neurologic symptoms (confusion, aphasia) and was admitted on October 13, 2010. CT scan and magnetic resonance imaging (MRI) of the brain demonstrated two intra-axial masses (4×3 cm in left temporal region and 2×2 cm in right occipital region). CT scan of the chest, abdomen and pelvis showed increased size of the right adrenal mass (3×2 cm), a new left adrenal mass (1×1 cm), indication of tumor thrombus in the IVC, and a peripherally enhancing paravertebral soft tissue mass involving the musculature which, in hindsight, was present on the initial CT of the chest from June 2010 (Figures 5–8). A positron emission tomography (PET) performed a few days later showed increased metabolic activity in the above lesions as well as in the T3 and T4 vertebral bodies. After consultations with medical oncology and radiation oncology specialists, a decision was made to first treat the brain lesions with external beam radiation therapy (XRT). A course of systemic chemotherapy was planned to follow. The brain lesions were treated with 6 mV photon therapy for a total of 30 gray in 10 fractions. Two weeks after completing the initial XRT, because of worsening pain, the patient had his paraspinous lesion treated with 6 mV photon therapy for a total of 25 gray in 5 fractions. Finally in mid December 2010, he was able to begin three cycles of chemotheraphy with gemcitabine and taxotere.

With complaints of pain, nausea and vomiting from the metastatic disease and subsequent treatment, he was admitted multiple times in the following months. November 2010, January 2011, February 2011, and March 2011 admissions involved multiple follow-up imaging studies, all showing progression of the bilateral adrenal masses, invasion of the inferior vena cava (IVC), paravertebral soft tissue mass, and vertebral body involvement (Figures 9–10). New findings included right lower lobe pulmonary emboli. A follow-up MRI of the spine in March 2011 showed epidural metastases at T3, where he also had bony disease. His MRI of the brain in March 2011 showed marked decrease in size of the brain metastases and no new foci of metastasis.

Early in presentation, the patient already demonstrated metastatic disease. Metastatic disease was confirmed in a soft tissue lesion on the patient's back, again with pathology of metastatic undifferentiated pleomorphic sarcoma/malignant fibrous histiocytoma.

As of March 2011, the patient’s diagnosis was stage IV metastatic, undifferentiated, pleomorphic sarcoma. He received three cycles of the chemotherapy regimen of gemcitabine and taxotere from December 2010 through February 2011, as well as supportive treatment for the neurological symptoms from the temporal and occipital lobe brain metastasis but died in July 2011 at hospice care.

DISCUSSION

Aortic sarcomas are rare tumors, with the intimal subtype occurring in the thoracic aorta, as the case reported here, being even more uncommon. Delay in diagnosis for these tumors often occurs as the imaging
features appear nearly identical to atherosclerotic disease. Seeing that atherosclerotic disease is clearly a more frequent diagnosis than intimal sarcoma of the aorta, it is not difficult to understand that this diagnosis is not usually made until after surgical resection or at autopsy.

Sarcomas of the aorta tend to occur mostly in the abdominal aorta, with less common occurrences in the thoracic aorta. For example, in one case series, four out of 21 cases of sarcoma presented in the chest. By definition, in contrast to the mural aortic sarcomas, intimal aortic sarcomas actually grow within the vascular lumen. Because of this growth pattern, they have a greater propensity for fragments to dislodge and be carried in the blood stream and metastasize [5]. Additionally, these tumors commonly present with sequelae from embolic phenomenon with symptoms ranging from absent peripheral pulses to mesenteric occlusion [1–4, 6, 7]. In one case series of 11 patients, all the patients with aortic sarcoma, died within 16 months of diagnosis [1].

Due to the rarity of this disease, there have been no randomized trials for the definitive treatment of IAS, with the therapies that are currently used being based solely on observational studies. Definitive treatment methods for IAS have not been delineated. The literature suggests that resection of the affected portion
Figure 5: Increased size of right adrenal mass, new small left adrenal mass, and new tumor thrombus in the inferior vena cava (October 2010).

Figure 6: After surgical repair of the aorta, a more conspicuous peripherally enhancing right posterior paravertebral soft tissue mass is seen which was subsequently removed (arrow) (October 2010).

Figure 7: (A, B) Nonenhanced computed tomography scan of the brain demonstrating heterogeneous left temporal mass and right occipital mass with surrounding edema (October 2010).

Figure 8: Fluid attenuated inversion recovery (FLAIR) magnetic resonance imaging demonstrating both left temporal and right occipital masses with surrounding edema (October 2010).

Figure 9: Increased size of right and left adrenal masses with expansion and involvement of the inferior vena cava (November 2010).

of the aorta with placement of a graft and subsequent chemotherapy and radiation is the best approach [2]. Endarterectomy is also a reported treatment choice [4]. At presentation, nearly all of the cases of IAS have evidence of metastatic disease.
CONCLUSION

Our case of intimal aortic sarcoma illustrated a number of interesting facts. First, the tumor may occur in the thoracic aorta, more specifically at the aortic arch, which is an uncommon location. Second, the clinical presentation may not be the classic one of symptoms from embolic phenomenon; instead patient may present with symptoms of chest pain likely from the primary tumor. Third, patients with intimal sarcoma of the aorta tend to die shortly after presentation. However, despite the evidence of metastatic disease early in presentation, patient may lived for many months after diagnosis and treatment.

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Author Contributions
Michelle Forman – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Michael Mulligan – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES