ELSEVIER

Angiosarcoma of the Thoracoabdominal Aorta Presenting with Systemic Hypertension, Anemia, and Visceral Ischemia

Tara Karamlou,[1] Melissa K. Li,[2] W. Kent Williamson,[3] Lloyd Heller,[4] and John W. Wiest,[3] Portland, Oregon

Aortic angiosarcomas, one of the three major types of primary aortic tumors, are exceedingly rare, with only 25 cases reported in the literature. Peripheral thromboembolic complications are the most frequently described presenting feature, and therefore, these tumors can be mistaken for aortoiliac occlusive disease. We describe a rare case of an extensive thoracoabdominal angiosarcoma that manifested with hypertension, profound anemia, and visceral ischemia in a young woman.

Brodowski first described primary aortic tumors in 1873.[1] Aortic angiosarcomas, one of the three major types of primary aortic tumors, are exceedingly rare, with only 25 cases reported in the literature.[2-4] Peripheral thromboembolic complications are the most frequently described presenting feature, and therefore, these tumors can be mistaken for aortoiliac occlusive disease.[2,5,6] We describe a rare case of an extensive thoracoabdominal angiosarcoma that manifested with hypertension, profound anemia, and visceral ischemia in a young woman.

CASE REPORT

A woman of age 46 years was referred from another hospital. She presented with hypertension, profound anemia, acute renal failure, and an abdominal bruit. One year prior to admission, the patient was evaluated in her primary-care clinic with mild dysuria, hypertension, and fatigue. Physical examination revealed a midabdominal bruit, with no other abnormalities. Abdominal ultrasound was unremarkable. Computed tomography angiogram (CTA) and subsequent magnetic resonance angiogram (MRA) revealed mild orificial stenosis of the right renal artery and 75% stenosis of the celiac artery at the origin. The superior mesenteric and inferior mesenteric arteries were widely patent. Mild atherosclerotic plaque was noted in the abdominal aorta, without any other abnormality (Fig. 1). Several months later, the patient re-presented complaining of lower back pain with radiation to both lower extremities. Her hematocrit was 37 mg/dL and serum creatinine was 1.3 mg/dL, without other significant abnormality. Magnetic resonance imaging (MRI) of the thoracic and lumbar spine demonstrated degenerative changes with mild herniation of the nucleosis pulposis at L4-L5. She was diagnosed with radiculopathy, and no further evaluation was undertaken at that time (Fig. 2).

Four months later, she was seen in the emergency room of a referring hospital with severe hypertension and headache. She was afebrile and weighed 118 lb. Her vital signs at that time were notable for a blood pressure of 205/81 mm Hg without tachycardia, tachypnea, or respiratory distress. Physical examination revealed bilateral carotid bruits, an abdominal bruit, mild peripheral edema, and a normal peripheral arterial exam without pulse deficit. Laboratory studies demonstrated profound normocytic anemia, with a hematorit of 17.9 mg/dL, elevated serum glucose of 141 mg/dL, and elevated creatinine of 2.6 mg/dL. Lumbar puncture was normal. Erythrocyte sedimentation rate was only mildly elevated at 12 mg/dL, but C-reactive protein was high at 7.6 mm/hr. White blood count and liver enzymes were normal, and chest radiograph showed only increased pulmonary vascularity.

[1]*Department of Surgery, Oregon Health & Science University, Portland, OR.*

[2]*Department of Pathology, Providence St. Vincent's Medical Center, Portland, OR.*

[3]*Department of Vascular Surgery, Providence St. Vincent's Medical Center, Portland, OR.*

[4]*Department of Radiology, Providence St. Vincent's Medical Center, Portland, OR.*

Correspondence to: Tara Karamlou, Department of Surgery, Oregon Health & Science University, 3181 SW Sam Jackson Park Road, Mail Code L223, Portland, OR, USA, E-mail: karamlou@ohsu.edu

Ann Vasc Surg 2008; 22: 459-464
DOI: 10.1016/j.avsg.2007.09.010

Published online: March 25, 2008

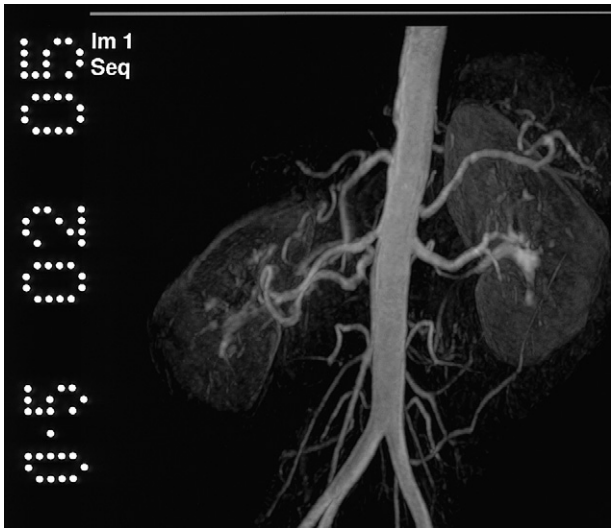

Fig. 1. MRA on initial evaluation revealed mild right renal stenosis. Not seen as well on this view, but noted on the saggital reconstructions was 75% celiac artery stenosis at origin.

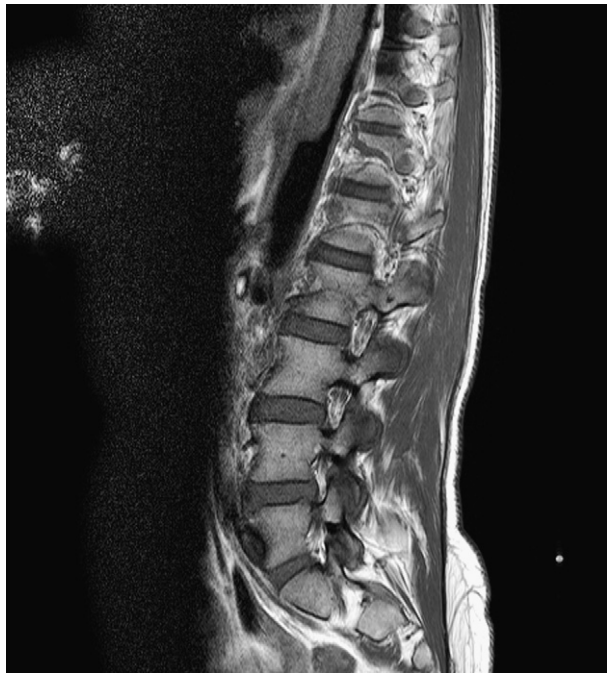

Fig. 2. Sagittal reconstruction MRI of the lower thoracic and lumbar spine revealed a large intraluminal filling defect in the descending aorta (*arrowhead*).

She was transferred to our institution for further evaluation and continued care.

Transthoracic echocardiography revealed normal aortic valve morphology and function, normal aortic root size, and left ventricular ejection fraction of 65%. MRA of the chest and abdomen was interpreted as a descending thoracic aortic dissection associated with severe luminal compromise with intramural hematoma, critical short-segment stenosis of the right renal artery at the origin, occlusion of the celiac artery, and moderate stenosis of the superior mesenteric artery (Figs. 3 and 4). The right kidney was atrophic, and there were bilateral pleural effusions. Mesenteric duplex was then performed, which showed elevated peak systolic velocity in the abdominal aorta of 291 cm/sec.

The patient was admitted to the intensive care unit, intubated, and transfused with two units of blood. Intravenous esmolol and nitroprusside infusions were instituted to reduce her blood pressure. Within 24 hr, however, she became anuric and her serum creatinine rose to 3.4 mg/dL. Arterial blood gas revealed a severe metabolic acidosis with serum bicarbonate of 9.7 mmol/L despite hyperventilation and a pCO_2 of 23 mm Hg.

She was taken to the operating room, where a right axillary artery to right common femoral bypass with a ringed 7 mm Goretex graft was performed to establish retrograde perfusion to the visceral and iliac arteries. A temporary hemodialysis catheter was placed for postoperative hemodialysis. At the conclusion of the case, the patient had Doppler signals in both legs but remained anuric.

During the next 24 hr, however, she became progressively more acidotic with an elevated venous lactate level to 4.2 mmol/L despite ultrafiltration with continuous venovenous hemodialysis. Abdominal duplex revealed a patent abdominal aorta, with flow in the splenic, hepatic, and superior mesenteric arteries. Because of her worsening clinical condition, she was returned to the operating room with a presumptive diagnosis of mesenteric ischemia due to hypoperfusion. At laparotomy, the small bowel had patchy areas of necrosis throughout its length and the entire colon was also ischemic. The stomach and liver appeared normal. The infrarenal aorta was examined and found to be soft, though with a diminished pulse. Similarly, the superior mesenteric artery was soft with an intact Doppler signal. The thoracic aorta immediately below the diaphragm was thickened, though without any extraluminal inflammatory changes. Because of the extensive intestinal necrosis, her condition was thought to be irreversible, and her abdomen was closed without further operative intervention. She expired several hours later under comfort care measures.

Postmortem examination showed a 30 cm occlusive lesion in the descending aorta with associated thrombus formation and thickening of the adjacent aortic wall (Fig. 5A). Gross inspection showed no evidence of aortic dissection or significant atherosclerotic disease. The celiac artery was occluded, and the superior mesenteric artery and right renal artery were severely narrowed, with thickening of the walls and intraluminal thrombus (Fig. 5B). Microscopic examination of the aortic mass revealed a largely necrotic lesion with a thin layer of preserved malignant epithelioid cells with hyperchromatic, pleomorphic nuclei at the intimal surface. The media was uninvolved. The tumor cells involved the visceral branches, with near occlusion of the celiac, superior mesenteric, and right renal arteries. Metastatic tumor also involved the left adrenal