Primary aortic sarcomas are rare; only 37 cases have been reported world-wide, and most of these have been diagnosed after death.\textsuperscript{1-3} Postirradiation sarcomas are also infrequent, occurring in approximately 0.03\% to 0.05\% of patients who undergo radiotherapy.\textsuperscript{4} Here we report an unusual case of an aortic sarcoma that was diagnosed before death in a patient who had been treated previously with radiotherapy for testicular seminoma.

**CASE REPORT**

A 46-year-old man was admitted in September 1996 as a medical emergency through the Accident and Emergency Department with sudden onset left-sided chest pain. There was no history of breathlessness or hemoptysis. The only medical history was of a left testicular seminoma that had been treated with a left inguinal orchidectomy and radiotherapy to the para-aortic and left iliac lymph nodes in 1988. The latter consisted of 15 fractions of 8 MeV photons, through anterior and posterior "dog leg" fields, to a total dose of 30 Gy. The upper level of this field was the T10/T11 vertebral disc space.

The patient was in pain, tachycardic, and mildly pyrexial on admission. Clinical examination was generally unremarkable except for dullness and reduced breath sounds at the left lung base.

The admission chest radiograph revealed a left-sided pleural effusion but normal mediastinal contours and otherwise clear lung fields. A pleural tap was performed that yielded a blood-stained exudate with a high lactate dehydrogenase content. Arterial gases on air at the same time showed a marked hypoxia. The working clinical diagnosis at this stage was pulmonary embolus.

A ventilation/perfusion scintogram did not show a ventilation/perfusion mismatch, but in view of the high clinical suspicion, a pulmonary angiogram was performed. This confirmed the pleural effusion and lack of pulmonary emboli but demonstrated a right paravertebral soft tissue mass. As part of a comparative study of thromboembolic disease, chest magnetic resonance imaging (MRI) using dark blood imaging (two-dimensional magnetization prepared rapid gradient echo: TR11, T4.2, flip angle 40, TI15; field of view, 330 × 440; matrix size, 128 × 256; time of acquisition, 16 seconds) and contrast-enhanced magnetic resonance angiography (Fast Imaging with Steady State Precession [FISP] three-dimensional sequence: TR5, TE2, flip angle 40; field of view, 400 × 450; matrix, 150 × 256; time of acquisition, 25 seconds) were performed with a 1.5T Siemens Vision Scanner (Siemens, Erlangen, Germany). During the contrast-enhanced sequence, breathholding was used. Fifteen milliliters of gadolinium DTPA (Magnevist; Schering AG, Berlin, Germany) was injected at 1 mL/s, and scanning was commenced after the injection of 5 mL of contrast. A single acquisition with the FISP three-dimensional sequence was obtained.

On black blood images (Fig 1), a large left-sided pleural effusion containing areas of high-signal intensity adjacent to the diaphragm were seen, suggesting recent hemorrhage. Coronal images revealed an abnormal structure straddling the diaphragm in the position of the descending aorta at the T10-T12 level. On axial sections, this appeared as a large circumferential soft tissue mass surrounding an aneurysm of the descending thoracic aorta. The gadolinium-enhanced angiographic images (Fig 2) demonstrated the aneurysm again, plus an anterior outpouching above this arising from the thoracic aorta. No pulmonary emboli were seen.

Postirradiation aortic sarcoma demonstrated by magnetic resonance angiography

J. Graham Pollock, FRCR, Alan R. Moody, FRCR, Catherine N. Ludman, FRCR, Michael Sokal, FRCR, and William G. Tennant, FRCS, Nottingham, United Kingdom

This is the first ever reported case of a radiation-induced aortic sarcoma. This patient had symptoms and signs initially interpreted as a pulmonary embolus. The extent of the disease was demonstrated with magnetic resonance imaging and magnetic resonance angiography, in particular, allowing rapid surgical intervention. (J Vasc Surg 2000;31:798-801.)
In view of these images, a thoracic aortic aneurysm leaking into the left pleural space was diagnosed. Abnormal soft tissue around the aorta was noted and felt to represent an aortitis. Urgent vascular intervention was advised.

At the operation, the patient was placed in a half-lateral position with the shoulders vertical and left side up. The pelvis was allowed to fall into the supine position, exposing the abdomen. A thoracoabdominal incision was made extending along the sixth intercostal space to the midline of the abdomen then taken vertically to the umbilicus. The retroperitoneal plane was developed, and the thorax was opened. The diaphragm was incised to the aortic hiatus, and the crus was divided to allow access to the entire thoracic and upper abdominal aorta. A ruptured 7-cm thoracic aortic aneurysm was found, extending superiorly from a point 2 cm above the origin of the coeliac axis. There was an obvious breach in the left lateral wall of the aneurysm that was plugged by thrombus. The appearance was of an "inflammatory aneurysm" with marked perianeurysmal fibrosis. A small saccular aneurysm was also found about 5 cm above the first aneurysm. The mediastinal pleura was divided, and normal aorta above and below both aneurysms was clamped. The patient was not given heparin. The aneurysm was opened. There was no lumbar back bleeding. An 18-mm diameter woven Dacron graft was inserted in the standard inlay fashion with 2/0 polypropylene for both anastomoses. A biopsy specimen was taken from the aortic wall because of the inflammatory appearances, and the sac was closed over the prosthesis. On declamping, the femoral pulses appeared immediately. The incisions were closed over two chest drains, and the patient was transferred to the intensive care unit.

The histologic features of the aortic aneurysm wall, however, identified a moderately differentiated spindle cell sarcoma resembling a malignant fibrous histiocytoma with no evidence of testicular seminoma.

The patient made an uneventful postoperative recovery, but further imaging revealed rapidly progressive local disease with para-aortic, subcutaneous, and pleural metastases. He received four cycles of single agent doxorubicin 75 mg/m² chemotherapy over 3 months with no durable response and died in April 1997, just 7 months after diagnosis.
DISCUSSION

Primary aortic sarcomas are rare; in over 120 years, only 37 cases have been documented. The most common site is the abdominal aorta, and most of these are now histologically classified as malignant fibrous histiocytomas (MFHs). They tend to have one of three morphologic appearances: intraluminal (polypoidal mass is seen with embolic episodes), intimal (spread along the intima and are seen as vascular occlusions), adventitial/medial (nonspecific, seen as aneurysms or masses).

Because of their late presentation, critical location and serious complications (eg, rupture, mesenteric embolization, or aortic occlusion) most of these sarcomas are diagnosed after death. Ultrasound scanning, conventional angiography, computed tomography, and MRI have all been used to diagnose aortic sarcomas in the literature; but this is only the second case of a diagnosis before death with the use of magnetic resonance angiography (MRA). Aortic obstruction has been demonstrated with the use of MRA in the one other patient to date; the obstruction was diagnosed as an abdominal aortic leiomyosarcoma. Interestingly, our patient’s diagnosis initially masqueraded as a pulmonary embolus in view of the sudden chest pain, hypoxia, and bloody pleural effusion. This is a common presentation of pulmonary artery sarcomas, where embolization of a tumor frequently occurs. Magnetic resonance has been used to diagnose several of these pulmonary artery sarcomas, typically showing low T1 and high T2 signals and gadolinium enhancement of the sarcoma mass.

Our investigative algorithm for pulmonary embolism at that time used MRI/MRA in a comparative study with conventional pulmonary angiography. Computed tomographic pulmonary angiography was not our standard practice, and the limited field of view and scanning range may not have included the aortic lesion. The information on the MRA was sufficient to achieve a fast and minimally invasive diagnosis of a leaking aneurysm. The multiplanar images were particularly helpful, and our vascular surgeons were able to operate on the basis of the MRA alone. Further imaging was not necessary.

The role of ionizing radiation in the pathogenesis of soft tissue sarcomas has been well established since the turn of the century. Radiation-induced sarcomas (RISs) are, however, rare; and recent estimates suggest an incidence of 0.03% to 0.05% of patients undergoing radiotherapy. However, with the increasing use and improved prognosis of patients who receive radiotherapy, the long-term complications such as RIS are of increasing concern. Definite cases of RIS are diagnosed with criteria proposed by Cahan et al and later modified by Arlen et al. Women tend to have a greater incidence of RIS than men, but this is probably due to the fact that radiotherapy is used to treat those common malignancies predominantly affecting women (ie, breast and cervical carcinomas). The most common histologic type is MFH. The most frequent sites reported are the chest, chest wall, pelvis, and skeleton, reflecting the irradiated sites. The mean latency period between irradiation and sarcoma is around 10 years, but a large range is seen. Our patient had a latent period of 8 years and histologically had an MFH. Associations between aortic sarcomas and synthetic vascular grafts have been described, but to date there have been no reports of a direct link to radiotherapy.

In conclusion this is the first reported case of a radiotherapy-induced aortic sarcoma that has been demonstrated with MRA. MRI was able to reveal both the mural component and intraluminal abnormality caused by this rare tumor, allowing rapid management of the condition of this patient with confusing clinical signs. The bitter irony of this case is that prophylactic radiotherapy to eliminate the possibility of subclinical nodal spread caused this lethal sarcoma.

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