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Histopathology 1996, 29, 278–281

CASE REPORT

Metastasising sarcoma of the aorta

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Date of submission: 30 November 1995
Accepted for publication: 3 January 1996

Keywords: aorta, leiomyosarcoma, metastases

Introduction

Primary malignant tumours of the aorta are rare. We report the incidental finding at autopsy of tumour occlusion of the thoracic aorta and the left subclavian artery by a leiomyosarcoma showing osteogenic differentiation.

Case report

A 73-year-old woman was admitted to hospital after a period of persistent vomiting and diarrhoea. On physical examination, a distinct, rough pan-systolic murmur (grade III/VI) was present over the thoracic aorta, radiating to the carotid artery. Abnormalities of the femoral pulse were not noted. The blood pressure was 210/70 mm. An electrocardiogram showed possible left atrial enlargement and left ventricular hypertrophy. The patient had had a biliary sphincterostomy with gallstone extraction in 1989. At that time she had been treated for five years for slight hypertension—150/95 mm.

Thirteen days before death, the patient underwent surgery during which intra-abdominal adhesions, probably caused by a cholecystectomy in 1980, were removed. Post-operatively, the patient deteriorated with diffuse abdominal pains. A few days later a subtotal enterectomy due to mesenteric thrombosis was performed. The operation specimen showed ischaemic lesions consistent with obstruction; no tumour emboli were found. Thereafter, she further deteriorated with coagulation problems, kidney dysfunction and paresis of both distal legs thought to be caused by retroperitoneal vascular disturbances.

PATHOLOGICAL FINDINGS

At autopsy the thoracic aorta was solid and rubbery, the external diameter was normal. The lumen showed almost total obstruction of the thoracic aorta extending from the left subclavian artery to within 10 cm of its origin. The obstruction was caused by friable, pale grey tissue and multiple blood clots (Figure 1).

Microscopically, the aortic occlusion showed a largely necrotic tumour mass. The viable tumour component showed spindle-shaped cells with elongated, blunt-ended nuclei (Figure 2). Pleomorphism and many mitoses were present. In the transition area of the tumour to normal aortic wall, the intima showed endothelial proliferation.

Five centimetres distal to the origin of the left subclavian artery the tumour extended into the adventitia. In this part spindle-shaped cells were arranged haphazardly and in broad intermingling bundles and fascicles, and the tumour showed focal myxoid change and islands of mature differentiated bone (Figure 3a). In addition there was invasion into thin-walled vessels (Figure 3b).

Multiple small infarcts were present in the liver, spleen and kidneys. In relation to these infarcts tumour emboli were found. No lymph node metastases were present.

Both in the aortic wall and in the tumour emboli the cells expressed smooth muscle antigen (SMA-1), actin (HHF) and desmin and, in addition, histiocytic and endothelial differentiation antigens (HAM56, CD68), vimentin, cytokeratin (CAM 5.2) and osteocalcin. There was focal positivity for factor VIII related antigen. Staining for Ulex Europaeus 1, epithelial membrane antigen (EMA), keratin and S-100 protein were negative. Electronmicroscopy was not performed.

Discussion

Primary malignant tumours arising in major blood vessels are rare; approximately 200 cases have been published. Malignant tumours arising in large veins account for approximately 80% of all reported cases and most (70%) are leiomyosarcomas. Reviewing the literature of all 31 primary malignant tumours of the aorta, the ratio of male to female patients is about 2:1 and the mean age of the patients is 55 years (range 3.5 months to 75 years). Anatomically, the sites of origin of primary aortic tumours are evenly distributed between the various aortic segments. The diagnoses are diverse and have included malignant fibrous histiocytoma, undifferentiated sarcoma, fibromyxosarcoma and, in a majority, fibrosarcoma. This varied diagnosis reflects, in part, the changing nomenclature with time and makes direct case comparison difficult. Most cases are classified on the histopathological pattern alone without further elucidation of histogenesis.

In a few cases, further classification was attempted by newer methods, such as immunohistochemistry. Except for factor VIII-related antigen activity, there is no unique immunohistochemical staining pattern in these...
sarcomas. In our case, cytokeratin reactivity within the tumour does not mitigate against a mesenchymal origin, as cytokeratin expression has also been demonstrated in normal human endothelial cells. A myoepithelial differentiation is supported by positive staining for actin and desmin. Despite the absence of binding with Ulex Europaeus, the focal positive factor VIII antigen reactivity and some atypical endothelial cells on the intimal surface in this case may suggest an endothelial origin. In addition, there was focal co-expression of osteocytic and histiocytic differentiation. On the basis of the immunohistochemical staining pattern, the tumour was classified as a primary leiomyosarcoma with osteogenic differentiation. A primary osteosarcoma is improbable because extra-skeletal osteogenic differentiation is reported in tumours from a variety of sites.

This is the second report of leiomyosarcoma of the aorta. The tumour extended focally into the perivascular tissue. Most aortic tumours reported remain confined to the aortic wall and lumen and there are only single reports of proven perivascular extension. Based on this perivascular extension in our case, a primary perivascular tumour localization was considered. However, this was thought improbable because the bulk of the tumour was in the aortic lumen rather than perivascular. Also, tumour extension from the adjacent organs into the aortic lumen has, to our knowledge, never been reported.

Retrospectively, accurate pre-operative or antemortem diagnosis in aortic tumours is often precluded. In addition to the rarity of the tumour, patients present with diverse, non-specific clinical manifestations, such as vascular insufficiency, as a consequence of arterial stenosis and emboli. Emboli occurred in two-thirds of patients and commonly involved the lower extremities and intestinal tract although, occasionally, the upper extremities and brain were affected. Other symptoms such as thoracic and lower back pain, fever and hypertension were common. The extensive intravascular tumour mass in our patient led to thrombo-emboli in the mesenteric arteries and subsequent bowel infarction.

The best means of diagnosis are offered by radiographic techniques and exploratory surgery. In 15 patients (48%) a malignant aortic tumour was suspected or diagnosed antemortem. This group of patients had a mean survival time of eight months. Most of these
patients underwent partial resection of the aorta following symptoms and signs of aortic obstruction. Microscopic examination of the resected specimen showed the malignant nature of the obstruction. One patient was treated post-operatively with radiotherapy and chemotherapy, two others with chemotherapy alone. Radiotherapy was considered in two cases, but not indicated because of the possibility of causing rupture of the aorta by dissolution of the tumour. In a group of six patients who were operated on for aortic obstruction with a pre-operative suspicion of a malignant tumour only three underwent resection. Despite the pre-operative diagnosis, five patients died within four months (one during operation, the others with vascular insufficiency), and the sixth died two years later. Taking all cases of primary aortic tumours into account, the majority of the patients die within a year following the onset of symptoms, although survival as long as three years has been reported.

Because the clinical features that characterize these neoplasms are most often associated with atherosclerotic disease, it is useful for clinicians to add primary aortic neoplasms to their differential diagnoses, particularly in patients with an intraluminal obstructive, potentially resectable, tumour.

Acknowledgements

The authors would like to acknowledge Dr A.C. van der Wal for his comments and suggestions in the preparation of this manuscript.

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