CASE REPORT

Intimal Sarcoma of the Thoracic Aorta: a Case Report

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Introduction

Sarcomas of the aorta are extremely rare, most of the cases being discovered at postmortem autopsy.1-5 We report a case that presented with bilateral peripheral tumour emboli.

Case Report

A 78-year-old female patient without any significant past medical history underwent a bilateral embolectomy of both femoral arteries because of sudden bilateral lower limb ischaemia. The removed material was unusually white, friable, with an “encephaloid” aspect, and microscopic examination revealed sarcoma cells. Physical examination was normal. There were no intracardiac abnormalities on transoesophageal echocardiography. CT scan revealed an intraluminal lesion, approximately 2.5 cm in length, located at the left posterior wall of the descending thoracic aorta (Fig. 1). No other sources of embolism were discovered and a diagnosis of an intraluminal sarcoma with peripheral tumour embolisation was made. Through a left thoracotomy, the diseased aorta was resected and reconstructed with a 4-cm long prosthetic graft. The intimal and mural segments were examined histologically. There were no signs of embolism in the aortic arch. Microscopic examination showed an intraluminal aortic sarcoma, sessile and polypoid, that measured 3 cm × 2 cm, reducing the cross-section of various sites throughout the body. Intimal sarcoma of the aorta (Fig. 2). The media was normal, and a necrotic polypoid intraluminal lesion surrounded by a rim of large neoplastic cells arose from the intima. These cells were positive to vimentin and to CD 31, negative to CD 34. Less than 10% of cells were positively stained with KL1, whereas all other immunohistochemical markers (EMA, CLA, PS 100, HMB 45, AFP, HCG, ACE, CD15) were negative. These various elements were consistent with a haemangioendotheliosarcoma.

Postoperatively a CT scan was performed because of abdominal distention and this showed occlusion of the mesenteric superior artery. An open embolectomy was successfully performed and intestinal resection was not necessary. Microscopy showed a similar picture to the aortic primary sarcoma. Postoperative recovery was thereafter uneventful. No complementary treatment was administered. Five months later, the patient died of profound cachexia, with clinical evidence of generalised metastases. No autopsy was performed.

Discussion

Since the initial report made by Brodowski in 1873,1 Seelig et al.2 have reported 87 cases of aortic sarcomas, almost 60% of them only discovered at autopsy. Wright et al.3 distinguished two types of aortic sarcomas, intimal and mural forms. Intraluminal sarcomas give signs of peripheral emboli as in our patient, and/or vascular signs of aortic obstruction. Because of embolisation, distant metastases may occur early in various sites throughout the body. Intimal sarcoma may also grow along the lumen, obstructing all branches of the aorta.2,4 Mural aortic sarcoma originates from the media or from the adventitia,3,4 and has a tendency to extend extramurally, involving para-aortic tissues and lymph nodes. It has a lower risk of producing emboli and early distant metastases. As in
Fig. 2. Macroscopic aspect of the intraluminal aortic sarcoma.

with adherent thrombus. With various immunohistochemical stains, it is often possible to define the subtype of sarcoma, a F VIII positive presentation suggesting an endothelial origin. Aortic sarcomas generally have a poor prognosis. The 1-year survival rate has been reported as only 13%, and Seelig et al. calculated that the median survival of patients was 7 months (0.25 to 168 months); 12 months for mural forms and 6 months for intimal forms.

References


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