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SHORT REPORT

Intimal Sarcoma of the Infrarenal Aorta: Report of a Rare Single Case and Review of the Literature

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Primary malignant tumours of the abdominal aorta are rare and they are usually associated with a poor one-year survival rate. The histological classification of aortic sarcomas (angiosarcoma/intimal sarcoma) is complex and does not appear to affect the prognosis. We present a case of a 68-year-old man with an infrarenal intimal sarcoma with peripheral embolization. The world literature on primary intimal malignancies of the abdominal aorta is reviewed.

Key Words: Abdominal aorta; Intimal sarcoma.

Introduction

Primary malignant tumours of the aorta are aggressive mesenchymal neoplasms. Histologically, angiosarcomas are characterized by a diffuse rather than a lobular or nodular growth pattern as seen in benign vascular lesions.[11] Since the initial report by Brodowski[1] in 1873 only about 100 cases of aortic sarcomas have been published,[2] and only about 40 cases belong to the abdominal region. Intraluminal tumours commonly present with embolization to the peripheral or mesenteric arteries.[2,4] Therefore, the clinical symptoms of malignant aortic disease are usually non-specific. The patient may present with claudication, weight loss, abdominal pain or necrotic skin lesions.

Aortic neoplasms are classified as intimal or mural according to their relation to the aortic wall.[3,4] This classification is relevant because of the wide range of clinical symptoms and prognosis. Intimal sarcomas and angiosarcomas represent the commonest types of primary intraluminal malignant aortic tumours. Intimal sarcomas derive from intimal smooth muscle cells or fibroblasts and angiosarcomas from intimal endothelial cells.[3] Intraluminal tumours present with peripheral arterial embolization, mesenteric ischæmia[2] or multiple skin infarctions.[4] Histologically, most angiosarcomas are found to be myofibroplastic sarcomas and a minority of these tumours produce endothelial antigens.[2] Immunohistological differentiation of histological subtypes is usually difficult. We report a case of an intimal sarcoma of the abdominal aorta with embolization and multiple skin infarctions.

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Case Report

A male patient aged 68 was admitted with symptoms of chronic lower limb ischæmia. Intermittent claudication was present at a walking distance of 15 m, predominately in the left calf and foot. Apart from non-insulin-dependent diabetes mellitus, no major medical problems or cardiovascular risk factors were evident when the medical history was taken. On clinical examination, lower limb pulses were absent and local inspection of both feet was unremarkable. No ulceration or gangrene could be observed. In contrast, erythematous lesions and dark spots were found in the skin of the left thigh and buttock (Fig. 1). A Duplex ultrasound scan and subsequent angiography revealed a high grade stenosis of the infrarenal aorta (Fig. 2) with an embolic occlusion of the left popliteal artery. The aortic stenosis was of functional relevance with a significant reduction of the ABPI (<0.5 at rest). Since the claudication was disabling a

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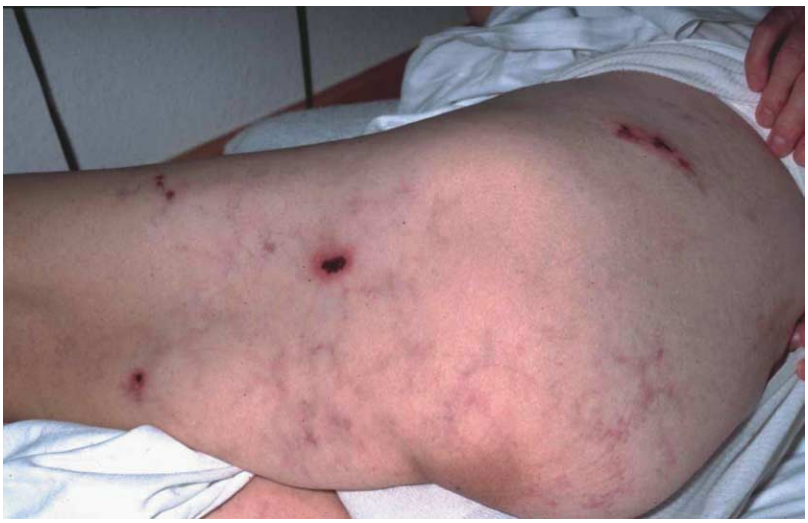

Fig. 1. Skin infarction.

decision was made to operate (Fig. 3). Thromboendarterectomy of the abdominal aorta with implantation of a 16 mm polyester (Dacron) graft was performed. The occlusion of the left popliteal artery was treated by an open embolectomy of the popliteal segment with local arterial reconstruction using a polyester patch. Apart from a postoperative retroperitoneal haematoma, the patient recovered well from this major surgical intervention. Toe necrosis in the left foot developed within the first postoperative week.

Histopathology revealed a primary aortic intimal sarcoma. Radiotherapy was offered but the patient declined this and died nine months later from widespread malignant disease.

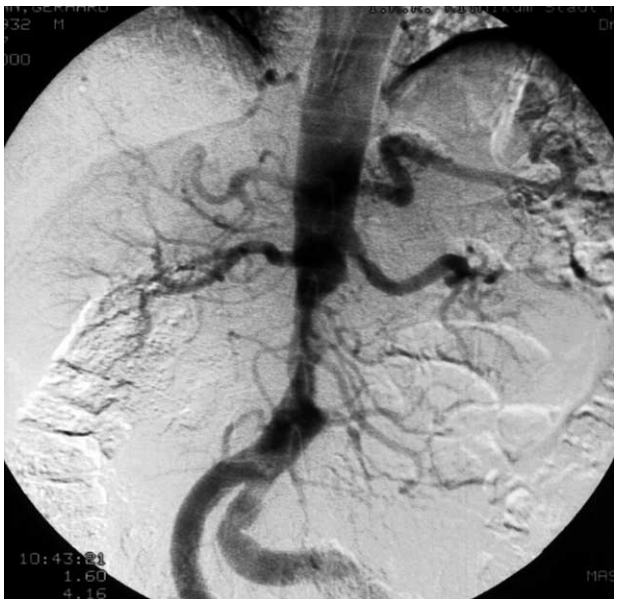

Fig. 2. Abdominal arteriography.

Pathology findings

A segment of the abdominal aorta measuring 5 cm in length and a fragmented section (1.5 cm) of the popliteal artery were examined. The specimen showed a yellowish polypoid surface of soft particles (each 2–3 mm in diameter) intruding into the vessel lumen. Light microscopy of this necrotic material revealed tissue infiltrated with inflammatory and necrotic cells with hyperchromatic nuclei, with increased and atypical mitosis. Immunohistological examination revealed numerous positive reactions to vimentin-antibodies and weak positive reactions to CD34 antibodies. Antibodies against common leukocyte antigen, pancytoceration, S100 and desmin were negative. The immunohistological constellation of the results was consistent with an intimal sarcoma at both locations.

Autopsy findings

The patient had a massively reduced body weight (53 kg/190 cm) due to tumour cachexia. He showed several signs of multiple wide spread embolization and paraneoplastic activated coagulation with fresh and old thromboembolic particles in the lungs. Massive proliferation of tumour cells at the aortic graft anastomosis was found. Disseminated endovascular spread of the tumour caused peripheral arterial occlusion in both legs. The patient suffered from chronic heart failure with concentric hypertrophy of the myocardium. The cause of death was right heart failure.

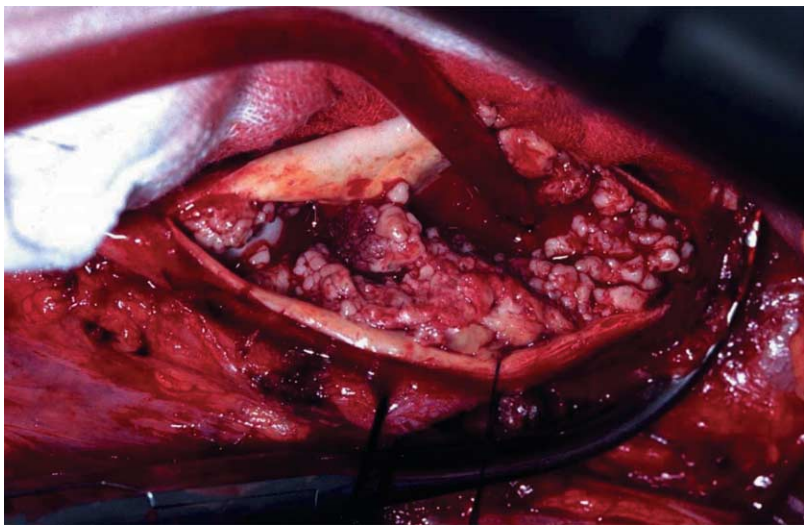

Fig. 3. Intraoperative.

Discussion

Malignant neoplasms involving the aorta most commonly occur as spread of a local tumour. Metastases to the aorta from distant primary neoplasms are unusual.[12] Primary aortic sarcomas are rare and only 40 cases have been described arising from the abdominal aorta. Common clinical symptoms include abdominal pain and claudication.[5] Claudication is caused by aortic occlusion, aortic stenosis or peripheral embolism.[12] The differential diagnosis includes thromboembolism from a cardiac source, Takayatsu arteritis, atherosclerotic thrombosis and left atrial myxoma with systemic embolization, the last cause being almost as rare as a tumour itself. Aortic sarcomas have been described at sites of vascular surgery such as arterio-venous fistula (haemodialysis), or graft interposition.[9]

Aortic sarcomas are mainly found at post mortem or during reconstructive surgery.[7] Kattus[6] reported the first ante-mortem case of a primary aortic tumour in 1960. The prognosis for primary aortic neoplasm is poor and death is usually caused by peripheral metastasis. Autopsy of 42 patients with aortic tumours (thoracic and abdominal) showed peripheral metastasis in 71.2% of cases. The metastases are located primarily in bone (28.8%), kidneys (27.1%), liver (23.7%), adrenal glands (20.3%) and lungs (15.3%). The mean survival rate is 14 months.[4] Only two cases of long term survival after primary malignant tumour have been described. Majeski[12] presented a patient with 8 years survival after resection of an intimal sarcoma of the endothelial cell type and recently Shuster reported of a case of abdominal aortic intimal sarcoma with a survival time of 48 months after adjuvant therapy.[8]

Malignancies of the aorta are classified by site as mural (primarily in the aortic wall) or intimal (predominantly luminal).[3] Sarcomas of the media or adventitia produce intramural or extramural extension with vague clinical signs such as abdominal and/or back pain.[12] The tumour can induce aneurysmal change. The clinical signs become obvious only if external compression of the tumour produces stenosis or results in thrombosis of the vessel lumen. Sarcomas of mural origin show a more protracted course and have a slightly longer survival time.[2] Tumours of intimal origin grow into the aortic lumen, extend along the aortic wall and produce a polypoid mass. Embolization of these polypoid structures produces peripheral occlusion and metastasis, becoming obvious as claudication or skin infarction.

Aortic sarcomas rely for their diagnosis solely on morphologic features identified using light or electron microscopy. A review of the sarcoma literature by Seelig[4] found a wide spread of histological diagnoses in 39 primary malignant tumours of the abdominal aorta (15 cases of sarcoma, four malignant fibrous histiocytoma, six angiosarcoma, five leiomyosarcoma, five fibrosarcoma, one hemangioendothelioma, one myxoid chondrosarcoma, one myxomatous endothelioma, and one fibromyxosarcoma).

Table 1 represents the currently published intimal tumours of the abdominal aorta according to their subtype, symptoms, location of metastasis and time of follow-up. Fourteen cases (58.3%) belong to the intimal type and 10 (41%) to angiosarcomas. There is a male-to-female ratio of 3:1 with an average survival rate of 11.5 months (1 day to 72 months). According to the

Table 1. Clinical and pathologic features of intimal and angiosarcomas

Histology	Reference	Year	Age	Sex	Symptoms/signs	Follow-up (+ = death)	Metastasis
1. Intimal sarcoma	Sladden[10]	1964	64	m	Lumbago, sciatia	5 months +	Pleura/skin
2. Intimal sarcoma	Sladden[10]	1964	59	m	Leg pain, pulslessness	18 months +	None
3. Angiosarcoma	Winkelmann[16]	1971	56	m	Pain feet and toes	8 months +	Intestinal, skin
4. Intimal sarcoma	Steffelaar[21]	1975	70	m	Claudication	5 days +	None
5. Angiosarcoma	Fehrenbacher[17]	1981	67	m	Pulslessness peripheral	7 months +	Liver, lungs
6. Angiosarcoma	Mason[18]	1982	70	m	Abdominal pain, anorexia	6 days +	Spleen
7. Intimal sarcoma	Wright[13]	1985	46	f	Bowel infarction	3 days +	Liver, pancreas
8. Angiosarcoma	Becquermin[19]	1988	67	m	Claudication, cutan. nodules	8 months	Skin
9. Angiosarcoma	Weiss[20]	1991	59	m	Abdominal and back pain	11 months	Liver
10. Intimal sarcoma	Burke[5]	1993	63	f	Abdominal pain, leg pain	1 months +	Bone multiple
11. Intimal sarcoma	Burke[5]	1993	68	m	Leg embolism	2 months +	None
12. Intimal sarcoma	Burke[5]	1993	64	m	Mesenteric occlusion	1 month +	None
13. Intimal sarcoma	Burke[5]	1993	64	m	Ruptured aneurysm	No follow-up	None
14. Intimal sarcoma	Burke[5]	1993	68	f	Obstructed AAA-graft	5 months +	Bilateral femurs
15. Angiosarcoma	Burke[5]	1993	58	m	Leg pain	7 months +	Peritoneal, liver
16. Intimal sarcoma	Daniel[23]	1997	73	f	Peripeheral embolisation	10 months +	Bone
17. Intimal sarcoma	Daniel[23]	1997	56	m	Back pain	7 months +	Bone, lung
18. Intimal sarcoma	Majeski[12]	1998	69	m	Claudication	72 months +	Femur, pelvin
19. Angoisarcoma	Hottentott[11]	1999	69	f	Claudication	12 months	Spinel, right foot
20. Angiosarcoma	Hottentott[11]	1999	68	m	Claudication	5 months	None
21. Intimal sarcoma	Nishida[15]	2000	73	m	Dyspnea, general fatigue	14 months +	Lung, adrenal
22. Angiosarcoma	Santonja[2]	2001	64	f	Abdominal pain, vomiting	11 months +	Spleen, liver
23. Intimal sarcoma	Shuster[8]	2002	47	m	Hip pain	48 months	No
24. Angiosarcoma	Rudd[22]	2002	59	m	Claudication, gangrenous toe	8 months	Skin

histological subtype, we noticed a longer survival rate with 14.2 months (1–72 months) for the intimal sarcoma against 7.8 months (1–12 months) for the angiosarcoma.

Phenotypical evaluation in previous reports of aortic intimal sarcoma varies in the era before immunohistochemistry. With the advantage of immunohistological examination the sub classification of primary malignant tumours of the vessels has become more precise using morphologic and immunohistochemical characteristics.[2] Nevertheless there is a heterogeneity of aortic sarcomas and it seems to be impossible to determine exactly the cell type of origin by histological and immunohistological findings.[3,5] For the purposes of classification it is reasonable to suggest that differentiated luminal angiosarcomas derive from intimal endothelial cells and intimal sarcomas from intimal smooth muscle cells or fibroblasts. The diversity of immunohistological results and the problem of exact classification of histological origin of primarily malignant tumours of the abdominal aorta are summarized in Table 2 (review of the current literature).

The reported case represents one of seven aortic intimal sarcomas in the world literature diagnosed before death. The patient showed typical clinical symptoms for the disease with claudication caused by peripheral embolization associated with multiple skin infarctions. The intraoperative macroscopic pathology (Fig. 2) was characterized by a polypoid mass with soft particles responsible for the popliteal embolization. After reconstructive surgery the

Table 2. Immunohistological results

Histological type	Reference	Factor VIII	Vimentin	Actin	Desmin	Keratin	S-100	QBend/CD34
1. Intimal sarcoma	Present	–	Positive	Negative	Negative	Negative	Negative	Positive
2. Intimal sarcoma	Shuster[9]	Positive	Positive	Positive	–	–	Positive	–
3. Intimal sarcoma	Burke[5]	Negative	Positive	Positive	Negative	Negative	Negative	Negative
4. Intimal sarcoma	Majeski[12]	Positive	Positive	–	Negative	Negative	Negative	Negative
5. Intimal sarcoma	Nishida[15]	Negative	Positive	Negative	Negative	Negative	Negative	Negative
6. Intimal sarcoma	Daniel[23]	Positive	Positive	Positive	Negative	Negative	–	Negative
7. Intimal sarcoma	Daniel[23]	Positive	Positive	Positive	Negative	Negative	–	Negative
8. Angiosarcoma	Burke[5]	Positive	Positive	Negative	Negative	Negative	Negative	Positive
9. Angiosarcoma	Weiss[20]	Positive	Positive	–	–	Negative	–	–
10. Angiosarcoma	Santonja[2]	Positive	Positive	Negative	Negative	–	–	Negative
11. Angiosarcoma	Rudd[22]	Positive	–	–	–	Positive	Negative	Negative
12. Angoisarcoma	Hott.-rott[11]	Positive	Positive	–	–	–	Negative	Positive
13. Angoisarcoma	Hott.-rott[11]	–	Positive	–	–	Positive	Negative	Positive

patient rejected further adjuvant therapy and died 9 months after diagnosis. Post-mortem examination showed beneath no other organ metastasis apart from popliteal and skin tumour embolization. Immunohistological results revealed a positive reaction for vimentin and QBend/CD34. In consideration of the histological characteristics the present case was defined as an intimal sarcoma.

As far as diagnosis and treatment is concerned Higgins suggested an algorithm based on patients with mesenteric tumour embolisation.[14] Magnetic resonance imaging with gadolinium enhancement is considered to be the most sensitive diagnostic tool for aortic tumours. For operative treatment a complete resection of malignant tissue and interposition of an artificial graft is recommended. Best results are obtained after aggressive treatment including surgical resection, radiation and chemotherapy. Majeski[12] reported a survival of 8 years after intimal sarcoma resection (thoracic) followed by combined adjuvant external beam radiation and chemotherapy with doxorubicin hydrochloride. Nevertheless primary aortic tumours are aggressive and the survival rate after resection and treatment is less then 15 months. Surgery seems to be only a palliative procedure because of the presence of metastases at the time of diagnosis.

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