Epithelioid Angiosarcoma With Metastatic Disease After Endovascular Therapy of Abdominal Aortic Aneurysm

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Abstract Malignancies of the aortic wall represent a rare condition, and only a few reports have covered cases of sarcomas arising at the site of a prosthesis made of Dacron. A coincidence with endovascular repair has only been reported in one case to date. We report a patient with epithelioid angiosarcoma and metastatic disease, which was found in an aneurysmal sac after endovascular aortic repair for abdominal aortic aneurysm.

Keywords Endovascular aortic repair · Abdominal aortic aneurysm

Introduction

Primary malignancies of the aorta represent a rare but aggressive tumor entity and constitute approximately 1–2% of soft-tissue sarcomas [1]. As a subtype of angiosarcoma, epithelioid angiosarcomas have a low incidence and only rarely arise from the larger blood vessels themselves [2]. By the time of diagnosis, a great number of patients already have metastatic disease and thus have a poor prognosis. They are most often diagnosed by chance on autopsy or

from surgical specimens [3]. The clinical presentation of these patients can imitate symptoms of claudication or thrombembolism. There are several reports on the disease in connection to foreign-body reaction due to the implantation of Dacron material [4].

Here we report a case of a high-grade epithelioid angiosarcoma of the aortic wall with metastatic bone disease and involvement of regional lymph nodules several years after endovascular aortic repair (EVAR).

Case Report

A 77-year-old patient with an abdominal aortic aneurysm (AAA) was successfully treated with an aortic endograft (Talent Endoprosthesis; Medtronic, Santa Rosa, CA, USA), which consists of nitinol springs covered with woven Dacron. The patient had a history of diabetes, cerebral ischemia, thrombosis of the basilar artery, and coronary vascular disease. There was no evidence of malignancy of any kind with respect to clinical and diagnostic parameters.

The patient initially had suspected type II endoleak during the first 3 months after EVAR, presumably arising from the inferior mesenteric artery, but further follow-up showed rapid shrinkage of the aneurysm sac and no further endoleak. Seven years after EVAR, when the patient was 85 years old, increased aneurysmal size was observed along with perfusion, presumably arising from a lumbar artery, which was successfully embolized (Fig. 1). However, radiological controls demonstrated persistent perfusion with noticeable circular contrast enhancement of the vessel wall but no evidence of a mural mass (Fig. 2).

On further follow-up, the patient presented with symptoms of general fatigue, weight loss, and diffuse abdominal discomfort. There had been no clinical improvement with

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Fig. 1 Given suspected endoleak on follow-up CT scan, intra-arterial examination was performed. Angiogram performed before embolization of a lumbar artery demonstrated normal placement at the proximal end of the prosthesis

Fig. 2 CT scan after embolization of a lumbar artery (portal venous phase) showing increased circular enhancement of the aortic wall in the proximal treated segment. In addition, an enlarged lymph node (arrow) was found in proximity to the aorta

steroid treatment for the suspected diagnosis of arteritis, and there were increased blood inflammatory levels and blood sedimentation rate. Therefore, fluorodeoxyglucose-positron emission tomography/computed tomography (FDG-PET/CT) scan was initiated, which showed pathologic tracer uptake in the aneurysm sac and aortic wall as well as a progressive regional lymph node (Fig. 3A). In

addition, the scan showed an osteolytic lesion located in a lumbar vertebra (Fig. 3B). The changes in the aortic vessel wall were assumed to represent an infection of the prosthesis, and fluoroscopically guided biopsy was performed to rule out malignant disease affecting the lumbar vertebra. Histologic evaluation of the material showed increased signs of bone remodelling and residual signs of bleeding with few inflammatory cells but no signs of malignancy. Repeat PET/CT scan after an interval of 3 weeks still showed markedly increased FDG uptake of the aortic wall, the aneurysm, and the adjacent lymph node; however, there was a new vertebral lesion.

With the diagnostic procedures giving evidence for graft infection, explorative laparotomy was performed, and the graft was removed and replaced by a silver-coated prosthesis. At surgery, there were signs of chronic inflammation with tissue adhesions. Comparable with the CT scan, an enlarged lymph node was seen firmly attached to the aortic wall. The initial pathological examination showed thrombotic material with signs of chronic inflammatory

Fig. 3 **A** On follow-up imaging, progression of the periaortic lymph nodes was observed (arrow and arrowhead). **B** CT scan also showed an osteolytic lesion in a lumbar vertebra

reaction. The further clinical course was complicated by acute retroperitoneal haemorrhage, which required another surgical procedure to remove. CT scans were performed to locate the bleeding source as well as determine the progression of the lesions in the lumbar spine as described in previous text.

The final diagnosis of aortic malignancy was made by examination of the completely explanted vessel section surrounding the graft. The therapeutic plan consisted of palliative measures, including chemotherapy and possible laminectomy with ventral stabilization in case of myelon compression. However, the patient did not recover from the surgical procedures due to his generally diminished condition.

Histological examination of the paraffin-embedded tissue showed infiltrates of a high-grade epithelioid neoplasm that was composed of moderately sized tumor cells with rudimentary vascular channel formation (Fig. 4A). Immunohistochemical stains showed strong positive staining for CD31 (Fig. 4B) and focal positive staining for pancytokeratin, whereas staining for CD30, EMA, and especially CD34 was negative. Mib-1 staining as an indicator for proliferation was positive in approximately 40% of the epithelioid cells and positive for P53 in approximately 10% of epithelioid cells.

Discussion

The diagnosis of angiosarcoma with primary intimal aortic origin is rare [5]. Immunohistochemical staining shows positivity for CD31, CD34, factor VIII-associated protein, S100, or HMB 45, whereas sarcomas deriving from intimal smooth muscle cells or fibroblasts express mesenchymal antigens [6].

Depending on the growth pattern of the tumor, the focal tissue alterations can lead to intraluminal occlusion or aneurysmatic changes in the vessel wall. Therefore, the clinical presentation of patients with primary malignancies of the aortic wall may vary from symptoms of aortoiliac occlusive disease with possible embolic complications to symptoms of vasculitis.

Signs of claudication were not present in this case assumably due to the growth inside the aneurysm sac and the thrombotic material were obscured from the blood flow by the endovascular stent-graft. Therefore, the patients' general symptoms, such as weight loss and fever, were the only symptom of inflammatory malignant disease, whereas signs of vasculitis may appear in cases of epithelioid angiosarcoma [7]. Due to the aggressive nature of the tumor, prognosis is poor, and there is high rate of metastatic disease at the time of diagnosis; the most common site of metastasis is the bone.

Fig. 4 **A** Angiosarcoma grade 3 with solid growth and rudimentary vascular channel formation (hematoxylin-eosin stain, scale bars 50 μ m). **B** CD31 immunohistochemistry showed strong positive immunoreactivity in nearly all tumor cells

Because the presence of primary malignant disease of the aorta is an unexpected diagnosis, differential diagnosis includes, amongst others, chronic inflammation with granulation tissue formation and papillary endothelial hyperplasia (Masson's tumor), both of which can be excluded by the absence of atypia and invasive growth. In the presented case, the radiological findings as well as pathological analysis showed signs of chronic inflammation; however, graft infection after endovascular repair of AAA is a rare condition itself [8].

Sarcomas resulting from foreign-body reaction have a latency period of decades. Most cases of epithelioid angiosarcoma reported in the literature have described an association with the foreign material Dacron in surgically implanted grafts [7, 9]. However, in the only case published so far, Umscheid et al. [10] reported a high-grade epithelioid angiosarcoma arising at the abdominal aortic

wall after implantation of a Vanguard endograft. Even if the existence of a similar case does not prove an association between the occurrence of this malignancy and the presence of Dacron material, more cases might report on comparable conditions with longer follow-up periods after EVAR.

Conclusion

In conclusion, the epithelioid angiosarcoma in this case became noticeable in terms of unspecific symptoms in combination with increased markers of inflammation. Occlusive vascular disease was not evident, presumably due to endovascular repair of the AAA. The presence of foreign material, as well as the presentation of clinical symptoms in accordance with the diagnostic procedures, lead to the suspected diagnosis of graft infection. However, consistent with other reports in the literature, symptoms appeared late during the course of the disease and metastatic disease that was already evident. Even if there is no definite proof of an association between the material and the tumor in our case, it emphasizes the rare but possible differential diagnosis of malignancies in patients having the described symptoms after EVAR.

Conflict of interest None.

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