ELSEVIER

Case Report

Aortic Epithelioid Angiosarcoma after Endovascular Aneurysm Repair

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We report a case of epithelioid angiosarcoma of the abdominal aortic wall after endovascular treatment for abdominal aortic aneurysm (EVAR). A 60-year-old male, treated 7 years before with EVAR, presented with abdominal back pain, general fatigue, and fever. It was assumed to be a graft infection with periaortic tissue compatible with an inflammatory reaction. The endograft was therefore completely removed and a Dacron silver aorto-bisiliac graft was implanted. After a few days the patient worsened, the angio-computed tomography scan showed a progressive increase of the periaortic mass and numerous small nodules in the abdomen were also detected. The patient was again brought to surgery, an axillo-bifemoral bypass was performed, and the aorto-bisiliac graft was removed but the patient died after surgery. The histological examination showed an aortic epithelioid angiosarcoma with peritoneal metastasis.

Angiosarcoma is a primary malignant vessel tumor. It represents 1−2% of all soft tissue sarcomas[1] and very rarely originates from the aorta.

In a review by Seelig et al.[2] of 87 malignant tumors of the aorta, angiosarcoma was present in 10 cases (17%).

The rarity of this tumor makes clinical diagnosis very difficult. By the time of diagnosis, a great number of patients already have metastasis and thus a poor prognosis.

Epithelioid angiosarcoma is a particular morphological subgroup of angiosarcomas in which there is a predominance of epithelioid character among the neoplastic endothelial cells; its incidence is very low and only rarely develops from large arteries.[3]

An association between angiosarcoma and Dacron graft has been rarely reported and any connection has never been demostrated.[4,5]

To our knowledge, this is the first report of epithelioid angiosarcoma after polytetrafluoroethylene (PTFE) aortic endograft.

CASE REPORT

A 60-year-old male presented to our department with abdominal back pain, general fatigue, and fever (37.6°C).

His medical history showed no evidence of cardiac disease, diabetes, or hypertension; medical therapy was only acetylsalicylic acid 325 mg/die.

Seven years before he underwent EVAR with an Excluder stent graft (W. L. Gore & Associates, Flagstaff, AZ) formed by nitinol stents covered with PTFE. The procedure was uneventful but at follow-up a type II endoleak was detected and it was treated 3 times the following years by translumbar embolization with intrasac injection of coils and glue.

Physical examination was negative.

Initially, hemoglobin, leukocytes, and C-reactive protein were 11.6 g/dL, 18.6 × 10^9/L, and 17.60 mg/dL, respectively.

Specimens for emocultures were negative.

An angio-CT (computed tomography) scan showed an abdominal aortic aneurysm sac of 6 cm in diameter with

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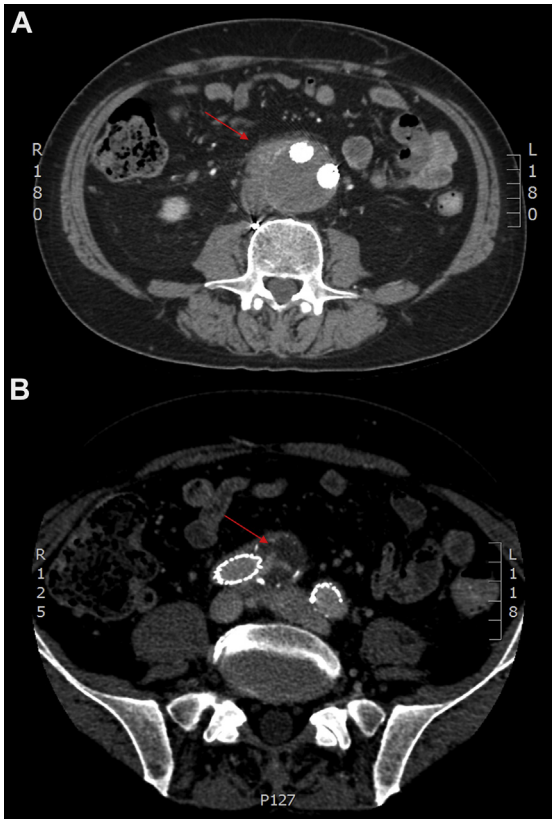

Fig. 1. Angio-CT scan shows the contrast enhancement of the periaortic tissue like an inflammatory reaction **(A)** and type II endoleak **(B)**. Red *arrow* shows the the contrast enhancement of the periaortic tissue in Fig. 1A and the type II endoleak in Fig. 1B.

periaortic tissue contrast uptake compatible with an inflammatory reaction and type II endoleak (Fig. 1).

The fluorodeoxyglucose-positron emission tomography/computed tomography (FDG-PET/TC) scan showed abnormal tracer uptake in the aneurysmal sac and in the tissue between the aortic wall and the vena cava (Fig. 2). It was assumed to be a graft infection.

He was placed on large spectrum antibiotic therapy for 3 weeks without any benefit.

A surgical conversion was therefore performed and it was surprising that there was no evidence of macroscopic infection, nor of abscesses. The periaortic tissue was friable and hypervascularized. The endograft was totally removed and a Dacron silver aorto-bisiliac graft (Maquet Getinge Group, Rastatt, Germany) was implanted and covered with omentum.

The intraoperative microbiological samples were negative for bacterial colonization. Unfortunately, we did not perform any histologic screening.

The early postoperative period was uneventful but after a few days the general conditions of the patient worsened: he developed fever, increased white blood cells (WBC), progressive weakness, and chronic anemia requiring blood transfusions every 3–4 days. Angio-CT scans were performed at 8, 18, and 30 days postsurgery (Fig. 3) and showed a progressive and rapid increase of the periaortic mass from a diameter of 63–102 mm. In the last angio-CT scan, numerous small nodules in the abdomen were also detected.

Progressively, the patient's conditions worsened: the inflammatory markers increased. The patient had short breath and a crisis of acute pulmonary edema treated with diuretic therapy. We theorized a reinfection of the graft, so repeat surgery was scheduled: an axillo-bifemoral bypass was first performed; then on re-laparotomy, we found multiple mesenteric nodules and in retroperitoneum a large friable and hypervascularized mass, extremely difficult to separate from the intestine. The Dacron silver aorto-bisiliac graft was removed and the infrarenal aorta was closed by a patch of femoral artery. The patient however died after surgery due to his generally weakened condition.

The final diagnosis was made by histological examination that showed an angiosarcoma consisting of hemorrhagic multinodular masses composed of epithelioid endothelial cells of high malignancy and multinodal peritoneal metastasis (Figs. 4 and 5).

DISCUSSION

In the last years, EVAR has become the most common technique used in abdominal aortic aneurysm repair. At the same time, a steady increase in complications such as endoleaks has been seen.

Moreover, an incidence of infection after EVAR is reported between 0.2% and 0.7%[6] and percutaneous procedures to treat endoleaks increase that risk.

The diagnosis of graft infection is based on the measurement of inflammatory markers and on the CT-scan and FDG-PET/TC findings. The gold standard treatment of aortic endograft infection is the total removal of the prosthesis and on aortic regrafting in situ or extra-anatomic.

Angiosarcoma involving primarily the aorta is rare; its diagnosis is difficult[7] and normally is not suspected clinically.[8] In some cases, aortic angiosarcoma masquerades as an infectious aortitis[9] or an inflammatory aortic aneurysm.[10] Generally, this tumor grows asymptomatically and often the first sign is the occlusion of the aorta or iliac arteries with possible distal embolism.[2,11,12] At the time of diagnosis, the tumor is very large and metastasis is already present. The treatment is only surgical with very poor long-term prognosis.[13,14]

In this patient, the symptoms were nonspecific (weight loss, fatigue, fever); claudication was not present probably because the tumor grew in the aortic sac, outside the endograft, thus maintaining the aorta and iliac arteries patent. The angio-CT