Case Report

Abdominal Aorta Angiosarcoma after Endovascular Aneurysm Repair

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Abstract: Primary tumors originating within the wall of the arteries are rare and they frequently manifest late, making effective treatment a challenge. We describe here a case of Abdominal Aorta AngioSarcoma masqueraded as an infected EndoVascular Aortic Repair. The knowledge of this pathology from vascular surgeons and radiologist is crucial, because a prompt diagnosis and treatment can improve the prognosis.

Primary tumors originating within the wall of the arteries are rare and they frequently manifest late, making effective treatment a challenge. The most common artery involved is the aorta; moreover, the rarity of this kind of tumor makes clinical diagnosis very difficult. The prognosis is poor.[1] Clinical symptoms of malignant aortic tumors include locally occlusive aortic disease or peripheral emboli, often mesenteric. General symptoms such as fatigue, weight loss or fever can be present, too. Angiosarcoma is an aggressive, malignant neoplasia with significant metastatic potential, but, in view of its rarity, it is sometimes mistaken with other pathologies.[1] We describe here a case of Abdominal Aorta AngioSarcoma (AAAS) masqueraded as an infected EndoVascular Aortic Repair (EVAR).

CASE REPORT

The local IRB approval was obtained. A 72-year-old man presented to our department with fever and back pain. His medical history was consistent for arterial hypertension, atrial fibrillation, chronic renal failure, chronic obstructive pulmonary disease, and 8 years previous EVAR with a bifurcated stentgraft (W. L. Gore and Associated, Flagstaff, AZ) with successful result (stable dimension of aneurysm sac and no evidence of endoleak or collection) at the follow-up. The computed tomography (CT) scan showed a thickened wall of the abdominal aorta in close contact with duodenum including some signs of inflammation and an endoleak without clear

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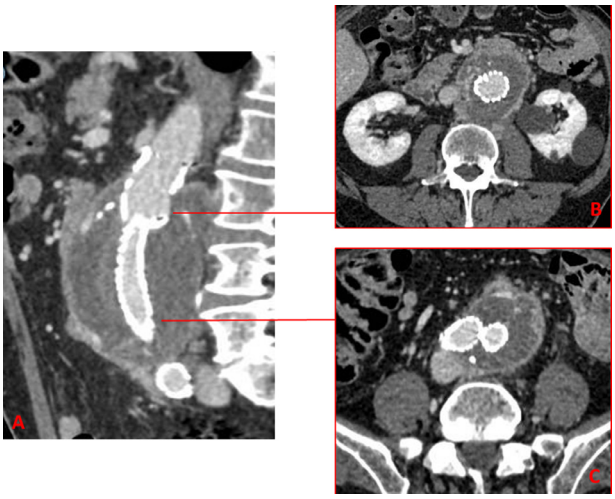

Fig. 1. Preoperative CT scan show some signs of aortic inflammation and an endoleak without clear origin.

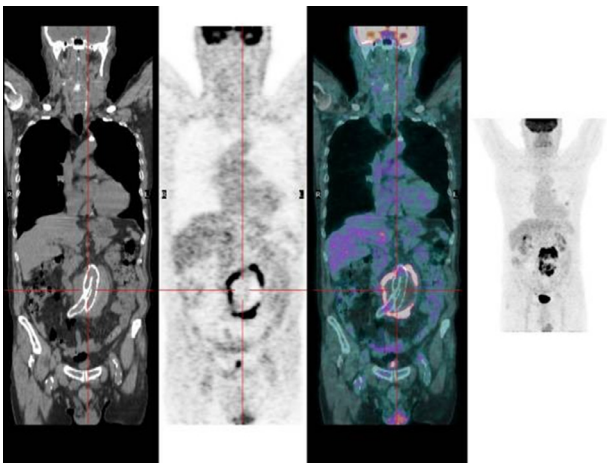

Fig. 2. Preoperative FDG-PET/CT scan showed an abnormal tracer uptake in the proximal neck of endoprosthesis, extending to the aneurysmal sac up to the left iliac branch.

origin (Fig. 1), not identified by the contrast-enhanced ultrasound. High reactive C-protein (RCP, 12 mg/dL) and intermediate Procalcitonin (0.9) ng/mL) were present in blood tests; no leukocytosis was present and blood cultures were negative. The fluorodeoxyglucose-positron emission tomography/computed tomography (FDG-PET/CT) scan showed an abnormal tracer uptake in the proximal neck of endoprosthesis, extending to the aneurysmal sac up to the left iliac branch (Fig. 2) without involvement of duodenum or presence of any remote foci. Due to the presence of probable high flow type II endoleak and the suspicion of prosthetic infection, after written informed consent, a surgical conversion was

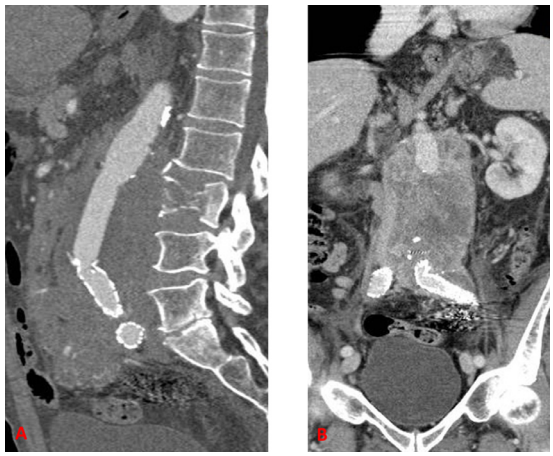

Fig. 3. A 3-week CT scan showed an aortic wall thickened with inflammatory tissue around the spinal column and the ureters, causing ureteronephrosis.

performed with partial removal of the prosthetic body (a complete explant of the prosthesis was avoided due to the absence of intraoperative signs of infection and the resistant incorporation of the iliac legs) and straight silver Dacron graft (Maquet Getting Group, Rastatt, Germany) interposition. Intraoperatively significant periaortic inflammation with lymphadenopathy was noted without liquid collection or aorto-enteric fistula. The aortic wall and lymph nodes specimens were sent for pathologic examination. When aorta was opened, a periprosthetic thrombus with different grading of clotting was found. The thrombus was removed and it was observed a chronic rupture of the posterior wall of the aorta, with a type II endoleak by a lumbar artery. Postoperatively the patient recovered well and blood tests improved. He was discharged at home on the seventh postoperative day. Three weeks after the intervention, the patient was readmitted to the infectious disease ward of our hospital, due to the recurrence of back pain and increased markers of inflammation (ESR 132 mm/H, CRP 16.15 mg/dL, WBC 12120/mmc, NEU 77.5%) without fever. Several samples for blood culture were collected without any results. A CT scan showed an aortic wall thickened with inflammatory tissue around the spinal column and the ureters, causing ureteronephrosis (Fig. 3). It was assumed to be a recurrent infection and an antibiotic therapy was prescribed, but the results of the histological analysis confirmed the diagnosis of aortic angiosarcoma few days after the admission. In consideration of the malignancy of the histotype, the aggressiveness of the pathology and the poor condition of the patient all therapeutic procedures

were suspended and the patient was transferred to a hospice and died 1 month later.

DISCUSSION

Aortic sarcoma is a rare disease, with only approximately 200 cases being reported in the medical literature[1]; AAAS represents a minute proportion with only 35 cases reported and only 4 after EVAR. Mean age at the diagnosis is 60 ± 10 years (M:F, 2:1) with metastasis present in more than 80% of the cases at the time of diagnosis. These tumors traditionally have a very poor prognosis, with a survival rates of 11.2% and 8% at 3 and 5 years, respectively.[2,3] Most AAAS arise spontaneously, but several well described risk factors exist. Radiotherapy is an independent risk factor. Although the association between radiotherapy and angiosarcoma is best described for breast cancer therapy, it is not exclusive of breast lesions. Various chemicals are associated with the development of angiosarcoma, particularly within the liver. Reports of angiosarcoma associated with foreign bodies include accidentally retained surgical gauzes or vascular prosthesis.[4] The development of sarcomas, including AAAS, has been well established in experimental animal models of tumor-genesis. Due the rarity of the AAAS related to the high number of EVAR, an animal model cannot be realistically used.[2] The most common location of this lesion is the abdominal aorta (38.5%) and the clinical presentation depends on the involved structures.[5] The clinical presentation is variable and specific symptoms do not exist, so the diagnosis is challenging. There can be general symptoms like fever, fatigue, and weight loss. Most patients present claudication, abdominal or back pain due to the compression of retroperitoneal structure, signs and symptoms of distal vascular compromise and organ ischemia. Diagnosis with imaging studies is likewise not easy; ultrasounds are highly sensitive for the detection of mobile intra-aortic components, but they are operator dependent. CT is the most commonly used imaging modality. Heterogeneous thrombus, protrusive vegetation and lack of atherosclerosis have been proposed as differentiating features.[6–10] Magnetic resonance imaging (MRI) is considered the most sensitive diagnostic tool for the detection of an aortic tumor, because it provides precise anatomic details that are comparable with that of conventional angiography, without the risk of embolization. In addition, MRI may provide excellent contrast between the vessel lumen and the aortic wall and the surrounding tissues. Finally, MRI may distinguish intraluminal tumor from atheromatous material.[11–14] FDG-PET, that shows an avid uptake of FDG, can be useful for the confirmation of the diagnosis, for staging and for monitoring treatment response. In a patient previously treated with EVAR, an AAAS can be confused with atypical endoleak, infection or thrombus of the graft. The association between angiosarcoma and EVAR has only been described in a few cases in the literature (ref). In all cases, the suspicion was prosthetic infection and the patients presented with fever, weight loss and posterior pain, and had contrast enhancement of the aortic wall at CTA. The median survival was 2 months after vascular open surgery.[15] Kamran et al.[3] proposed as nonspecific signs to CTA: (1) protrusive vegetations or nodular soft tissue components, (2) lack of atherosclerosis in the area of suspicion, (3) heterogeneous thrombus, (4) evidence of enhancement and neovascularity, (5) persistent enlargement of the excluded sac. Due to the rarity of this pathology, a standardized protocol for the treatment does not exist. If an aortic tumor is suspected during the surgery, it would be appropriate to perform a frozen biopsy for immediate examination and if positive it is recommended to perform a radical excision of the mass and of the aorta. Resection should involve all structures, seeming to be involved in the primary tumor with preservation if vital structures seem not to be invaded. If the diagnosis is made in the postoperative period, a repetition of the resection should be taken into consideration and it is strongly recommended, in order to obtain disease-free margins. Finally, patients with metastatic disease should be offered palliative treatment with chemotherapy and radiation therapy.

CONCLUSIONS

In the last few years, EVAR has become the most common technique used in abdominal aortic aneurysm repair. This technique reduces perioperative mortality and morbidity avoiding the invasiveness of open surgery. On the other hand, it does not allow radicality on the aortic pathology and the opportunity for aortic tissue biopsy. Diagnosis of AAAS must be considered in a patient with atypical clinical presentation and much attention must be paid to the observation and interpretation of the CTA images. In addition, it is mandatory in case of surgical conversion for endoleak or suspected infection to carry out samples of the aortic wall and periaortic lymph node tissue to allow a prompt diagnosis even of rare pathologies such as vascular neoplastic ones. The knowledge of this pathology

from vascular surgeons and radiologist is crucial, because a prompt diagnosis and treatment can improve the prognosis.

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