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CASE REPORT

Endovascular Treatment of an Infrarenal Aortic Epithelioid Angiosarcoma Causing Critical Limb Ischemia in an 80-year-old Patient: A Case Report

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Abstract
Aortic angiosarcomas (AAS) occur in approximately one case per million people annually and are often difficult to diagnose due to their nonspecific clinical and radiological symptoms. An 80-year-old female presented with the absence of pulses of both lower limbs and feet tissue loss. A contrast-enhanced computed tomography (CT) scan showed the occlusion of the infrarenal abdominal aorta extending to the aortic bifurcation, involving both common and external iliac arteries up to the superficial femoral artery bilaterally. A Fogarty embolectomy of both side femoral axes was performed followed by a covered endovascular reconstruction of the aortic bifurcation (CERAB) with a covered stent graft and two covered iliac stents placed in a kissing stent configuration. Histological examination of the Fogarty embolectomy samples revealed an aortic localization of epithelioid angiosarcoma. Healing of feet trophic lesions was observed 2 months after surgery, regardless of physical and rehabilitation team intervention progressive cachexia and physical deterioration led the patient to death 5 months postoperatively. Due to their nonspecific clinical presentation, AAS are typically late diagnosed with poor prognosis despite treatment. Endovascular treatment was chosen as the only feasible option, given the high risk of limb loss and poor general health conditions. Moreover, it led to significant improvement in patient's quality of life with no postoperative complications. Endovascular treatment associated with a physical and rehabilitation program may be advised as a less invasive alternative, for palliative or emergent clinical presentation in order to avoid short-term complications with poor impact on survival and quality of life.

Keywords Angiosarcoma · Aortic diseases · Neoplasms · Chronic limb-threatening ischemia · Case report

Introduction

Primary aortic tumors are generally rare and often misdiagnosed pathologies.

The incidence of AAS is around one case every million people per year [1]. Because of its nonspecific clinical presentation, which often consists of fatigue, unintentional weight loss, and malaise, AAS can be difficult to diagnose.

Acute limb ischemia (ALI), back pain, or chronic limb ischemia (CLI) are some of the most frequent clinical conditions that refer patients who seek medical attention, even without a history of atherosclerosis. In the case of late-stage disease, the previously reported general symptoms and those related to metastatic activity are dominating [2].

Furthermore, radiological findings can be misleading [3], leading to a delay in diagnosis and of their subsequent treatment. In many cases [3], an aortic sarcoma is mistakenly diagnosed as a focal dissection, an inflammatory process of the aortic wall, or an atherosclerotic lesion.

We are reporting the case of a patient affected by Leriche syndrome derived from primary AAS. The patient provided

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written informed consent for the report of her case details and imaging studies.

Case Presentation

An 80-year-old woman presented to our outpatient department after months of progressively worsening lower limb rest pain and tissue loss on both feet (Fig. 1).

The patient's previous medical history included major depression and progressive weight loss of 8 kg in the previous 4 months, dyslipidemia, and arterial hypertension. At admission, the patient was on the following medication regimen: sertraline 50 mg daily, atorvastatin 20 mg daily, and lisinopril 10 mg daily.

Smoking habits or drug abuse were not reported.

Significant swelling, pallor, and absence of peripheral pulses of both lower limbs were noticed. Therefore, a duplex ultrasound exam (DUS) was performed showing a postocclusive signal in common femoral arteries associated with focal stenosis of the profunda femoral artery, bilateral occlusion of the superficial femoral artery, and monophasic venous-like doppler wave in popliteal and tibial arteries.

Subsequently, a contrast-enhanced CT scan was ordered, and it showed the complete occlusion of the infrarenal abdominal aorta extending to the aortic bifurcation, involving both common iliac arteries with no periaortic lymphadenomegaly. The external iliac arteries were stenotic with a low-attenuation filling defect, a finding consistent with a fresh thrombus [4].

A sub-occlusion of the middle third of the superficial femoral artery bilaterally and the right profunda femoral artery was also observed (Fig. 2).

Thus, the patient was diagnosed with Leriche syndrome associated with peripheral embolizations and scheduled for surgery in the coming days.

Because of the weight loss and the general condition of the patient, an underlying oncologic pathology was suspected. Therefore, a brain CT scan and a complete biochemical oncologic biomarker panel with carcinoembryonic antigen (CEA), cancer antigen 125 (CA125), cancer antigen 19–9 (CA19-9), and cancer antigen 15–3 (CA15-3) were requested. However, all tests resulted in being negative.

Under general anesthesia, after exposing both common femoral arteries and positioning two short 6 Fr introducer sheaths, the lesion was crossed, using Terumo 0.035″ guidewires and then exchanged with super stiff 0.035″ guidewires, and finally, an over-the-wire Fogarty embolectomy was performed bilaterally, with the removal of a large amount of white material with fleshy consistency. Several samples of the thrombus were prepared for histopathological and cultural examination.

Covered endovascular reconstruction of the aortic bifurcation (CERAB) was carried out with a balloon-expandable Bentley BeGraft (BeGraft Aortic, Bentley InnoMed, Hechingen, Germany) covered aortic stent measuring 18 × 48 mm, approximately 15 mm above the aortic bifurcation.

Post-dilatation was performed with an Atlas Gold PTA Dilatation balloon (BARD Peripheral Vascular Inc., Tempe, AZ)—20 × 20 mm—in order to achieve a funnel-shaped form of the Bentley BeGraft stent.

Then, two balloon-expandable covered iliac stents, 10 × 38 mm Atrium Advanta (Atrium Medical Corp., Hudson, NH, USA) and 10 × 59 mm Atrium Advanta, were placed in kissing stent configuration and simultaneously inflated, on the right and left side respectively.

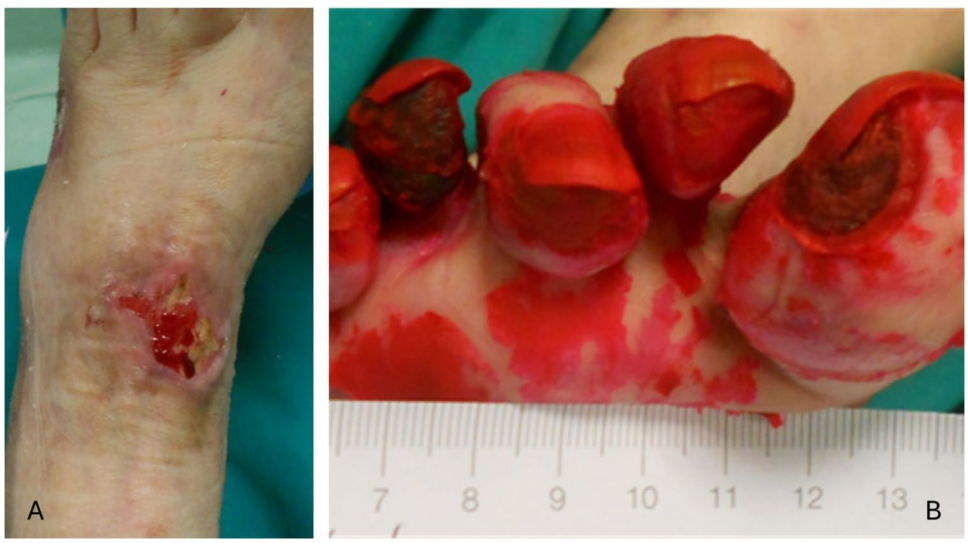

Fig. 1 **A** Dorsal left foot ulcer and **B** right gangrenous changes of the first, second, and fourth right digit

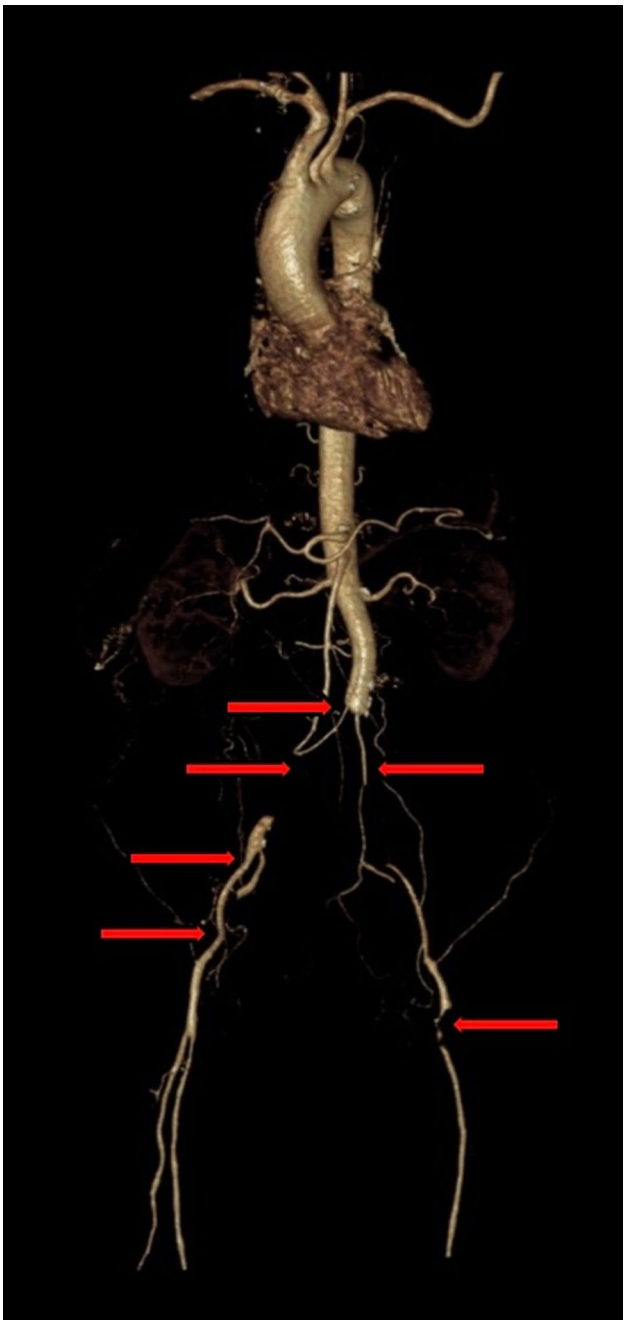

Fig. 2 3D reconstruction of contrast-enhanced CT scan at first emergency department

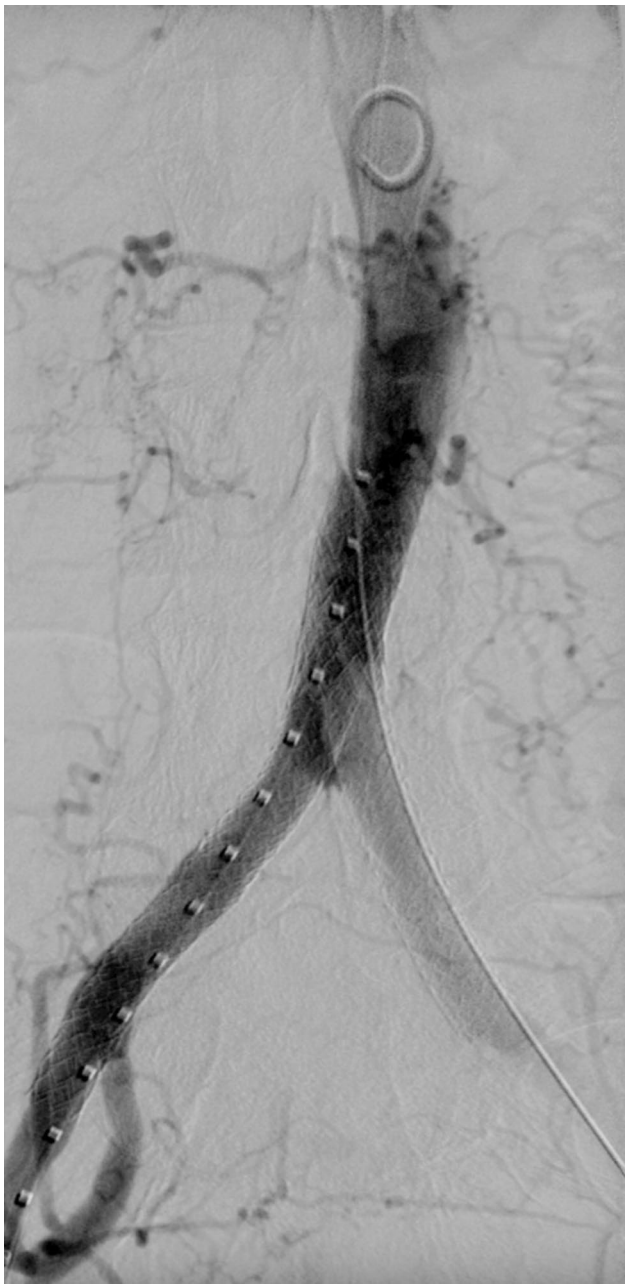

Fig. 3 Final angiogram showing the complete revascularization of the aortic bifurcation

Aortic lumen area was calculated in correlation with the dimension of the stent grafts for the iliac arteries in order to achieve the best CERAB configuration.

The final angiogram showed a complete revascularization of the aortic bifurcation and of the iliac limbs (Fig. 3). The surgical procedure was then completed with a superficial femoral artery and a profunda femoris artery embolectomy performed on both sides, with peripheral pulse recovery of both lower limbs.

Intraoperative sample analysis tested negative for bacterial infection, but the histological examination revealed the presence of aortic localization of epithelioid angiosarcoma.

The immunohistochemical study showed positivity for CD31, ERG, Fli-1 (weak), pooled cytokeratins, and CD34 (focal) and negativity for protein S-100, CD20, and CD3, with a proliferation index (ki67) of 40% confirming the histological diagnosis (Fig. 4).

A positron emission tomography (PET) and a CT scan were performed postoperatively after the histological

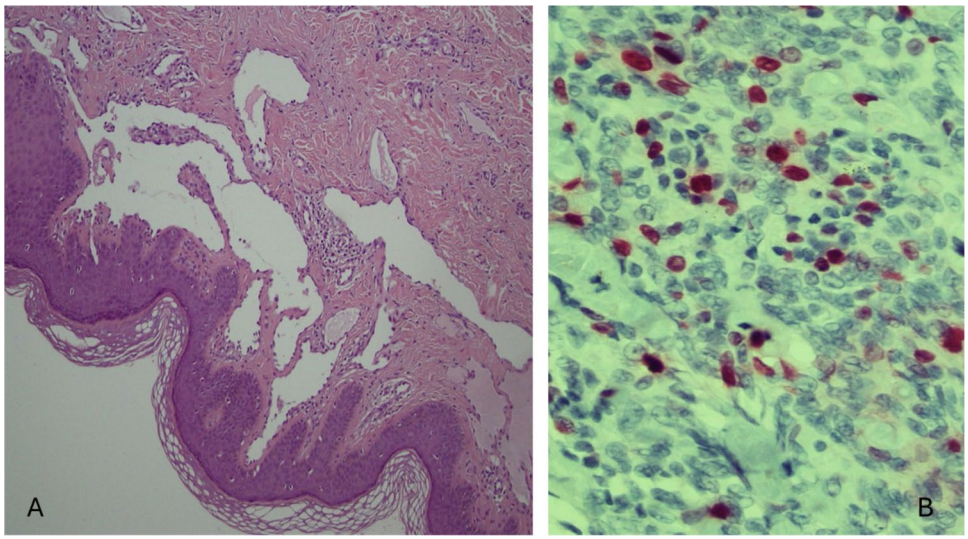

Fig. 4 **A**, **B** Hematoxylin and eosin staining showing cohesive and pleomorphic tumor cells with dense chromatin

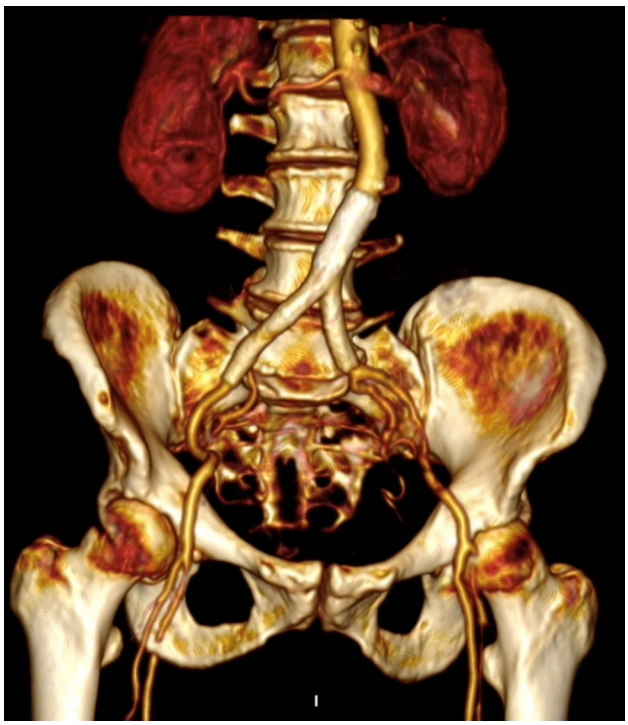

Fig. 5 Postoperative 3D reconstruction of contrast-enhanced CT scan showing the absence of recognizable endoleaks

diagnosis in order to identify any metastatic lesion; however, no evidence of extra aortic tumor localization or metastatic lesions was found as well as no evidence of endoleaks (Fig. 5).

The case underwent a multidisciplinary examination with oncologists and radiologists.

Because of the absence of noticeable extra-aortic pathology and the progressive worsening of the patient general clinical conditions with an Eastern Cooperative Oncology Group (ECOG) score = 4 and due to her age, no chemotherapy treatment was set up; physical and rehabilitation medicine (P&RM) team evaluation was requested, and an individualized rehabilitation program (IRP) drawn up.

The patient was discharged after 15 days in stable conditions with anticoagulant (unfractionated heparin) and aspirin 100 mg daily therapy.

In the first 2 months of follow-up (FUP), we obtained a partial regression of the trophic lesions and a complete regression of lower limb pain.

Although follow-up PET and CT scan were negative and the regular P&RM FUP with IRP adaptations, the patient developed a progressive cachexia and physical deterioration that led to her death 5 months postoperatively.

Discussion

AAS is a very uncommon pathology, described for the first time in 1873 by Brodowski [5]. It typically involves the abdominal and thoracic aorta with a median age of onset of 62.2 years [6, 7].

To date, there are few recognized risk factors that may lead to AAS development mainly represented by radiation, chemical toxins exposure, or foreign bodies (including vascular endoprosthesis) [8].

Diagnosis is often made post-mortem [9], because of its non-specific, and, in the majority of cases, asymptomatic initial clinical presentation may lead to a late diagnosis.

However, contrast-enhanced CT angiography can discriminate with high sensitivity and specificity the tumor growth pattern despite it may be challenging to discriminate between aortic neoplasms and other aortic pathologies such as atherosclerosis plaques, intraluminal thrombotic apposition, or inflammatory vasculitis.

Growth patterns and subsequent different radiological findings may be linked to which vessel part the neoplasm arises from: intima, media, or adventitia.

In case of an intimal origin, the tumor typically invades intraluminal spaces, exhibiting a polypoid-like appearance while, in case of media or adventitia origin, AAS shows an outward growth, involving the periaortic tissues [10].

To further discriminate the possible involvement of the aortic wall or the perivascular soft tissue, aortic contrast-enhanced magnetic resonance imaging (MRI) could be used due to its higher sensitivity when compared to contrast-enhanced CT angiography [11, 12].

In addition, PET scan may be associated with previous imaging methods to evaluate the presence of metastasis and rule out the diagnosis of atherosclerotic lesions or thrombotic apposition.

Anyway, due to its low specificity [13] and the rarity of neoplastic aortic pathology, this methodology has poor cost-effectiveness compared to other imaging methods. Therefore, the PET stand-alone use can be considered controversial, and in common practice, it is often performed once the histopathological diagnosis is achieved.

Indeed, the definitive diagnosis can be achieved only after surgical explantation and histopathological examination.

Surgical treatment can be considered in the case of intraluminal AAS [14], while in the case of metastatic disease, chemotherapy with standard protocols (mitomycin C, doxorubicin (adriamycin), and cisplatin) [15] is considered the gold standard therapy in association with radiotherapy in order to treat localized bone metastases and to achieve optimal pain control, but with low impact on long term survival rate [16].

Since AAS has intrinsic resistance to chemotherapy, its highly aggressive growth pattern, and metastatic potential (in particular to lungs, lymph nodes, liver, and bone) [17], the estimated overall survival rate still ranges between 6 and 16 months [18].

To date, radical surgical treatment with resection of the involved vessel and subsequent reconstruction, combined with multidisciplinary management may increase survival by up to 3 years [19], and it is still considered the gold standard for AAS therapy [15, 20].

In the literature, there are just two reported cases of endovascular approach related to this condition, and in both cases, the angiosarcoma was affecting the thoracic aorta [21, 22].

Considering the patient clinical presentation, an extreme rescue therapy was considered appropriate in order to attempt a limb rescue by combining the CERAB technique and Fogarty embolectomy.

Although the patient poor prognosis, the authors consider the surgical result satisfactory; in fact, no further vascular complications have been recorded and the peripheral lower limb lesions healed with a significant improvement in the patient's quality of life. Despite this, lower extremity embolic skin cutaneous metastases may develop after angioplasty and stenting for AAS endovascular treatment [19].

Retrospectively, in our specific case, the malignant disease could be at least linked as a determinant for the development of the patient's Leriche syndrome.

However, even if suspected, the postoperative diagnosis of aortic angiosarcoma would not in fact have changed the type of treatment performed on the patient.

Since the general clinical conditions and the perioperative risk, it would not have been possible to carry out more demolishing interventions which could have possibly guaranteed greater oncological radicality.

Conclusions

Aortic angiosarcomas are typically late diagnosed, and their prognosis remains poor despite the possible adjuvant or neoadjuvant chemotherapy administration [23, 24].

Up to now, no specific treatment can be advised, and any case should be singularly evaluated as well as no specific diagnostic method may allow an early diagnosis.

Surgical treatment followed by rehabilitation should be advised in case of palliative or emergent clinical presentation in order to avoid short-term complications with poor impact on survival and quality of life.

Moreover, despite the actual lack of solid evidence, endovascular treatment should be taken into consideration to guarantee a less invasive alternative, mostly in those patients with poor general conditions at the time of diagnosis.

Eventually, a neoplastic pathology of the aorta should be considered in differential diagnosis in case of association of vascular problems and constitutional symptoms, especially in the absence of a widespread atherosclerotic pathology or cardiovascular risk factors.

Author Contribution L.G., M.G.V., F.F. and G.H. have contributed to the conception, design, acquisition of data, analysis and interpretation of data, drafting the article and revising it critically, and final approval of the manuscript to be published.

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Data Availability No datasets were generated or analysed during the current study.

Code Availability Not applicable.

Declarations

Ethics Approval No ethics committee evaluation was required for a single case report. All methods were performed in accordance with

the ethical standards as laid down in the Declaration of Helsinki and its later amendments or comparable ethical standards.

Consent to Participate Informed written consent was obtained from all individual participants and/or their legal guardians included in the study.

Consent for Publication Informed written consent for publication was obtained from all individual participants and/or their legal guardians included in the study.

Competing Interests The authors declare no competing interests.

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