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Aortic pseudoaneurysm after endarterectomy for small aorta syndrome

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ABSTRACT

INTRODUCTION: Small Aorta Syndrome (SAS) or hypoplastic aorto-iliac syndrome is a rare pathology of the aorta that affects almost exclusively young or middle-aged women and is characterized by smaller dimension of the aorta and iliac axes. Etiopathogenesis is unclear and many factors have been invoked. The smaller caliber of the aorta and iliac arteries may predispose to aorto-iliac occlusive disease development.

In the past aorto-iliac endarterectomy (AE) with patch closure was utilized as an alternative to surgical bypass in order to correct steno-obstructive syndromes affecting carriers of SAS. Little is known about long term outcomes of this type of surgery.

PRESENTATION OF THE CASE: During investigations for acute cholecystitis, an aortic pseudoaneurysm (PA) was diagnosed by ultrasound in a 73 old year woman. She was submitted twenty-two years ago for SAS with disabling claudication to aortic endarterectomy (AE) with patch graft insertion. Considering all the vascular options available she was submitted to open surgery with replacement of the aortic bifurcation.

DISCUSSION: Aortic PA is a relatively common complication after bypass surgery but is rarely observed after AE. It requires prompt intervention to prevent subsequent complications such as rupture, thrombosis, distal embolism or aorto-enteric fistula.

CONCLUSION: Endovascular treatment for aortic PA should be always considered the treatment of choice but the open surgical option was preferred in this particular case because of the small diameters of the iliac accesses, making them unsuitable for an endovascular approach.

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1. Introduction

The present work has been reported in line with the SCARE criteria [1].

Small Aorta Syndrome (SAS), also called hypoplastic aorto-iliac syndrome, is a congenital anatomical entity characterized by a smaller caliber of the aorta and iliac arteries. It may predispose to aorto-iliac occlusive pathology that can be corrected with open or endovascular surgery [2–4]. Aortic endarterectomy (AE) was a widely adopted surgical option for treatment of SAS in last decades. Adjunctive patch angioplasty was frequently performed to repair the arteriotomy and to enlarge the aortic lumen. A possible complication of AE is the development of a pseudoaneurysm (PA) in the patch site. We present a case of post AE aortic PA in a woman with a history of SAS, discussing pathogenesis, diagnosis and treatment options of this unusual complication.

2. Case report

A 73-year-old woman, with a history of essential arterial hypertension, underwent AE for treatment of SAS when she presented with disabling claudication 22 years prior. The aortotomy was closed with a Dacron patch graft using a 4-0 monofilament polypropylene suture. At discharge all peripheral tibial pulses were present. The patient, who lived in a rural area, was lost to follow-up.

As part of a work-up for cholecystitis, she underwent an ultrasound scan (US) of the abdomen which revealed a pulsating mass, extending from the level of the aortic bifurcation, measuring 43 × 45 mm. The patient was completely asymptomatic and afebrile. Blood tests showed no evidence of leukocytosis as might be expected in the setting of infected PA. The aorto-iliac digital subtraction angiography confirmed the presence of an aortic PA of the aortic bifurcation, with very narrow iliac arteries (Fig. 1).

Computed tomographic angiography of the abdomen demonstrated a 37 mm length aortic neck, 43 mm antero-posterior aortic diameter and 40 mm antero-lateral diameter, 5.5 mm bilateral common iliac and 5.0 mm bilateral external iliac diameters. Angio-CT scan failed to show any evidence of aortic inflammation or other

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Fig. 1. Preoperative aortography of the aortic pseudoaneurysm (PA) after aortic endarterectomy (AE).

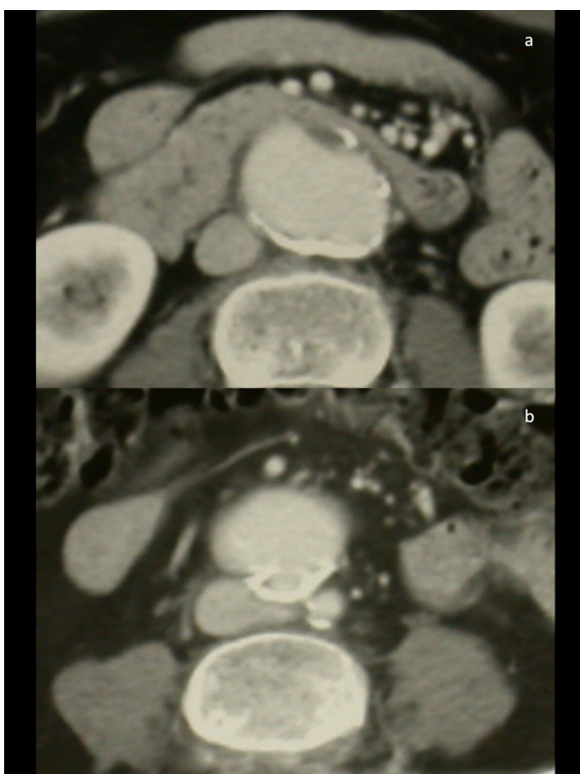


Fig. 2. a–b; CT scan of the aortic PA.

signs of periaortic infection (absence of gas bubbles, fat stranding, phlegmon or abscess) (Fig. 2a and b).

Open surgical correction was performed under general anesthesia through a mid-line abdominal incision; the aortic PA was

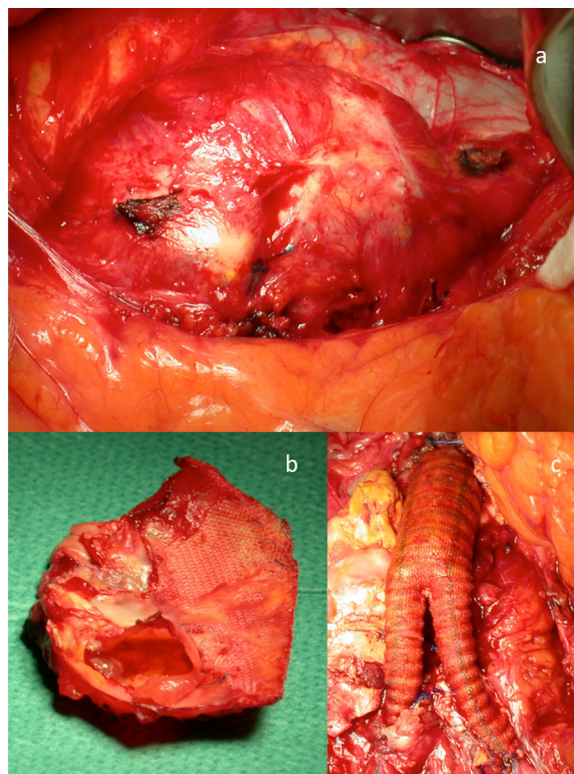


Fig. 3. a. Intraoperative image of the PA; b. Specimen of the aortic wall and Dacron patch showing the dehiscence of the suture line; c. Aortobiliac bypass graft to correct the aortic PA.

isolated (Fig. 3a) and, after 2500 I.U. of heparin with aortic cross clamping, then resected. The PA specimen showed a 5 cm in length dehiscence of the suture line (Fig. 3b). A 14 × 7 mm diameter aortobiliac knitted, double velour Gelatine-coated vascular graft (Uni-Graft® K DV, B. Braun Melsungen AG, Tuttlingen, Germany) was interposed (Fig. 3c). No purulent material was observed near or in contact with the aorta. Bacterial cultures of the specimen resulted negative for growth.

The patient had an unremarkable postoperative course and was discharged eight days later. On three years follow-up, the patient remained asymptomatic and had no evidence of recurrent anastomotic dilatation or pseudoaneurysm on ultrasound imaging.

3. Discussion

SAS was originally described in 1847 by Quain [5] and later, in 1969, by Johnson who named it “Small Blood Vessel Syndrome” [6]. This particular clinical picture is actually also known as “Hypoplastic Aortoiliac Syndrome” [7]. Some authors prefer the more descriptive term of “premature aorto-iliac steno-occlusion in women” [3].

This rare arterial pathology is characterized by a tapered terminal aorta with narrow iliac arteries, affecting young or middle-age women with mild obesity and aggressive atherosclerosis. Absence of predisposing risk factors such as smoking or hyperlipidemia is frequently observed. Etiopathogenesis is unclear and many factors have been invoked. Arnot et al. observed in cadavers the presence of a unique origin for the lowest pair of lumbar arteries and hypothesized that a congenital defect, due to overfusion of the two embryonic dorsal aortas, could cause hypoplasia of the terminal aorta [8]. High level of antiphospholipid antibodies, local inflammation, radiation, infection, aortic haemodynamics or aggressive atherosclerosis have been also considered, but the most accredited

ited theory suggests a mix of congenital aortic hypoplasia and atherosclerosis-induced lesions [2,9–12].

When symptomatic, aortic reconstruction is often challenging, considering the small size of the vessels. In the past, open surgery was the preferred treatment for SAS. Some authors supported closing the aortotomy with a Dacron patch, preferably “Y” shaped, while others preferred aorto-iliac bypass revascularization. Conservative therapy and lumbar sympathectomy are also described [12]. Recently, endovascular techniques have been proposed with good results [13].

Complications of surgery include late postoperative failure with results ranging from 46 to 79% for bypass surgery [14,15] and 71.4% for endovascular treatment [13]. In small series, AE has good long term results, with patients remaining free from symptoms for several years [7].

Aortic PA is a relatively common complication after bypass surgery but is rarely observed after AE [16]. PA following AE can be multifactorial. Etiologies include: infection, anastomotic tension, tensile stress unevenly distributed on the different parts of the anastomosis, inadequate suture technique, excessive space between stitches, defective suture materials, increase in peripheral resistance and intrinsic weakness of the artery wall [16]. Patch graft angioplasty has been commonly used to repair the aortotomy in cases of SAS treated with AE because a primary closure risks narrowing the vessel. Patch angioplasty was performed with synthetic or autologous vein material. Weakness of the residual arterial wall could be largely responsible for PA, due to interruption of the vasa-vasorum and altering district blood flow, especially in presence of extensive artery preparation [16].

Diagnosis of non-infected aortic PA is difficult because of their deep localization and their slow and silent development. An abdominal painful mass may be sometimes felt by the patient and/or by the physician. Abdominal pain radiating to the back, may also occur, due to pressure on retroperitoneal structures. US scanning is the first level examination for follow up and allows an easy and timely identification of this late complication although other imaging methods may be needed to demonstrate an aortic PA. CT or magnetic resonance angiographies is necessary for planning conventional or endovascular correction [17].

Aortic PA requires prompt intervention to prevent subsequent complications. Progressive enlargement with rupture, thrombosis, distal embolism or aortoenteric fistula insurgence can be catastrophic. Mortality rate for elective surgery is 8–28% with a morbidity of 36–73% [19–21]. When performed emergently, surgery can be extremely difficult with mortality rates ranging from 24% to 67% [18–20].

Endovascular surgery should be considered the treatment of choice for every kind of aortic PA considering morbidity and mortality of open surgical repair [4]. Benefits of a mini-invasive approach are especially notable in high-risk patients with prohibitive comorbidities who may not tolerate general anesthesia or in those with hostile abdomen where open surgery carries higher risk of complications. Despite the favorable morphology of the proximal neck, open surgical option treatment was preferred in this case because of the small diameters of the iliac accesses, making them unsuitable for endovascular approach with devices available at the time. Recent evolution toward lower-profile EVAR delivery systems should allow endovascular interventions for nearly all patients although SAS's challenging anatomy could be, in our opinion, a place for conventional surgery even today.

4. Conclusion

Surgical technical problems in SAS cases, related to the unfavourable sizes of the arteries, are always challenging for the

vascular surgeon despite of therapeutic strategy. AE has been widely used for correction of SAS and PA can develop when patch graft is performed for arteriotomy closure. Anyway, coexistence of both conditions makes difficult the correction of this unusual late post-operative complication.

Conflicts of interest

All authors declare that they have no conflicts of interest to disclose.

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Ethical approval

This is a report of a case; no research was conducted on patients that needed ethical approval.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Authors contribution

Maurizio Domanin: Literature Review, Case presentation, main Surgeon, submitting, corresponding and main author.

Daniele Bissacco: Literature review, surgical first assistant.

Silvia Romagnoli: Review of final manuscript.

Adelaide Buora: Literature review.

Guarantor

MD.

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