Pseudoaneurysm of the aortic isthmus involving a right aberrant subclavian artery long after multiple coarctation repairs

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Received 21 January 2015; received in revised form 10 February 2015; accepted 24 February 2015

Keywords: Aortic coarctation • Pseudoaneurysm • Aberrant subclavian artery

CLINICAL PROBLEM

Coarctation of the aorta (CoA) is a common congenital cardiac malformation, usually treated by open surgical procedures. Open surgery via left thoracotomy (LT) is still considered the standard procedure although balloon angioplasty and stent placement are nowadays considered a suitable option after 3 months of age and in the adult population [1].

Even though it is classically considered a disease of infancy, and usually repaired before the person reaches school-age, CoA is also seen in the adult population as either delayed diagnosis, restenosis of cases treated in childhood or failure of previous surgical treatments [1, 2]. Lifelong surveillance of repaired infantile CoA is mandatory because of recurrence or pseudo-aneurysmal growth with an inherent risk of rupture and lethal outcome [3]. Both complications can occur during long-term follow-up, exposing the patient to multiple operations [2–4]. The presence of a coexisting aberrant right subclavian artery (ARSA), a rare entity with a reported prevalence, as high as 2%, might further complicate the decision-making process [5]. A new corrective procedure on the distal arch and proximal descending aorta, with its related morbidity and mortality rates, may be warranted in these cases.

CASE DESCRIPTION

A 48-year old female was referred to our hospital in 2014 for a huge pseudoaneurysm of the aortic isthmus involving an ARSA long after multiple coarctation repairs. Medical history consisted of a bicuspid normo-functioning aortic valve and aortic coarctation. In 1976, she underwent an end-to-end anastomosis through a LT. Due to recurrence of the CoA in 1988, a redo operation (patch enlargement) was performed through the same access. The operation was complicated by permanent paraplegia. Two years later, an extra-anatomical (ascending to descending aorta) bypass was carried out through a right thoracotomy. All these procedures were performed in another centre (the last one, 24 years ago). The patient came to our hospital with a very limited medical documentation, without data about pressures in the higher and lower compartments or residual gradients concerning previous operations. Preoperative angio-CT scan showed a pseudoaneurysm of the aortic isthmus (67 mm) with aortic arch hypoplasia and patency of the retro-pharyngeal extra-anatomical bypass (white arrow in A). In B, the white arrow shows the origin of the left and of the right aberrant subclavian artery from the pseudoaneurysm (posterior view).

Conflict of interest: none declared.
REFERENCES


Treatment solution by Botta et al.

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doi: 10.1093/icvts/ivv056
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Lifelong surveillance of repaired infantile CoA is mandatory because of possible recurrence, failure of surgical treatments and pseudoaneurysmal growth with an inherent risk of rupture and lethal outcome. Multiple technical options are nowadays available to treat a pseudoaneurysm of the aortic isthmus involving an ARSA after previous repairs, including endovascular, conventional surgical and hybrid procedures. Despite a variety of options, several doubts can arise: would we go for an open or an endovascular approach? Is a single-stage correction or are multiple steps preferable? What is better: a first-time sternotomy or third redo left thoracotomy? Is partial or total arch repair required? What about cerebral protection? Should we close the extra-anatomical bypass or not? Many clinical features can influence the final decision: age, permanent paraplegia, previous operations and surgical accesses, the ARSA, a patent ascending-to-descending bypass, a gothic hypoplastic arch, uncertain landing zones, small but healthy femoral vessels, also keeping in mind the potential cost of the chosen procedure. We decided to treat this patient by conventional open surgery through a left thoracotomy at fifth intercostal space without cardiopulmonary bypass. Patient was positioned in the right semi-lateral decubitus. Intubation was performed by using a selective bronchial tube. Cerebral monitoring was obtained by means of continuous near-infrared spectroscopy (NIRS) and electro-encephalogram (EEG) registration. The pseudoaneurysm was isolated as well as the transverse arch and the descending thoracic aorta after careful lysis of dense pulmonary adhesions (Fig. 1A). The left carotid artery was clamped. After 2 min without NIRS and EEG changes, the left subclavian artery, the middle arch (between the carotid arteries) and the descending aorta were clamped. Perfusion of the lower body depended on the patent extra-anatomical bypass that was neither isolated nor ligated at the end of the procedure. The pseudoaneurysm was opened and a Foley catheter was inserted into the ARSA (Fig. 1B). A residual coarctation was evident close to the previous patch (Fig. 1C). Both the subclavian arteries were transected at the origin. The distal arch was enlarged by a transverse incision with exclusion of the residual coarctation. The proximal part (18 mm) of a bifurcated vascular graft (18/ 9 mm) was used to replace the pseudoaneurysm. The aortic clamp was moved from the arch to the prosthesis after proximal anastomosis, allowing bilateral cerebral perfusion. The remaining portion of the bifurcated graft (18/9 mm) was anastomized to the descending prosthesis using a partial clamp. The two legs were then connected in a termino-terminal fashion to the subclavian arteries (Fig. 1D). The patient was extubated after 8 h and discharged by ICU after 22 h. The postoperative course was uneventful despite a mild left pleural effusion. Discharge occurred after 8 days. Good clinical conditions were observed at 1-year follow-up.

Figure 1: Intraoperative view. In (A), the white arrow shows the isthmic pseudoaneurysm while the green and yellow arrows show the left subclavian artery and the descending thoracic aorta, respectively (also in B). In (B), a Foley catheter is inserted into the right aberrant subclavian artery (blue arrow) while the orange arrow shows a residual coarctation. (C) The patch previously used for aortoplasty. The straight part (18 mm) of a bifurcated graft (18/9 mm) was used to replace the pseudoaneurysm. Proximal and distal anastomoses were reinforced with external Teflon felt. The bifurcated segment was used to perform the reimplantation of both subclavian arteries as shown by the final result in (D).