Asymptomatic giant ascending aortic aneurysm: a challenging surgical strategy for a silent bicuspid aortopathy

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Abstract

We report the case of an incidental finding of a giant aneurysm of the ascending aorta with a congenital bicuspid aortic valve type 0-lateral. This severe condition was totally unknown to the patient, who was asymptomatic for cardiovascular disease. The aneurysmal mass involved the entire mediastinum, altering the normal anatomical relations, so the operative strategy was modified intraoperatively, tailoring the surgical technique to the anatomical conditions found. Despite a delayed awakening, the patient had an uncomplicated postoperative course. Therefore, this case highlights the importance of not underestimating nonspecific, seemingly harmless symptoms and signs that may reveal potentially catastrophic pathologies while also focusing on the surgical technique used. The modified Cabrol procedure, while an underutilized technique, if present in the cardiac surgeon's "arsenal" can represent a life-saving strategy in complex cases requiring an aortic valve and ascending aorta replacement.

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Introduction

Giant ascending aortic aneurysm (GAAA) is defined as an aneurysm greater than 10 cm in diameter [1]. This is a life-threatening but very unusual entity, as in most cases complications, such as dissection or rupture, occur before the aneurysm reaches this size. Considering that the risk of complications is related to the aneurysm size, current guidelines recommend surgery for ascending aortic aneurysms (AAA) with a diameter greater than 55 mm; this parameter drops to 50 mm in cases of concomitant bicuspid aortic valve (BAV). BAV is the most common congenital heart defect, occurring in 0.5-2.0% of the general population. BAV-related histological changes include abnormal processing of fibrillin 1 by vascular smooth muscle cells (VSMCs), increased release of metalloproteinases, increased apoptosis of VSMCs, and abnormalities in the medial layer, probably due to collagen and elastin. Mutations in genes including *NOTCH1* and *GATA5*, *FBN1* and *GATATGFBR1*/2, and *ACTA2* are implicated in the etiology of BAV [2]. The prevalence of AAA is reported to be between 40% and 70% in patients with BAV [3]. This condition is therefore considered a predisposing factor for the occurrence of AAA, as hemodynamic changes resulting from atypical valve morphology predispose individuals to turbulent hemodynamics and the appearance of valve degeneration earlier [4]. Bicuspid aortopathy, which is clinically manifested by dilatation of the ascending aorta, is the most common complication of BAV. The increased risk of aortic dilatation highlights the importance of accurately assessing the size and shape of the aorta in individuals diagnosed with BAV [5]. Therefore, considering that for aneurysms greater than 60 mm in diameter, the annual rupture rate is 14%, the diagnosis of an unruptured or dissected GAAA is an extremely rare clinical finding [1].

Case Report

A 56-year-old male was referred to our division after an episode of dyspnea and chest pain arising for the first time following physical exertion. Past medical history revealed no previous or familiar pathologies, among cardiovascular disease; only hypertension was found, already under pharmacological treatment, and blood tests were normal. A physical examination revealed a 4/6 Levine holodiastolic murmur in the Erbs area, so transthoracic echocardiography (TTE) was performed, which to our surprise, showed a monstrous aneurysmal dilatation involving the aortic root and the entire ascending thoracic aorta. The aortic valve was bicuspid with a moderate eccentric regurgitation jet. The aortic arch and descending aorta had normal size.

Therefore, transesophageal echocardiography (TEE) confirmed the presence of a voluminous AAA, in the absence of dissection, with estimated dimensions of 118×116 mm, a BAV, type 0-lateral, according to Sievers classification, with a 30 mm dilated aortic ring (Figure 1). Biventricular systolic function was preserved (ejection fraction 60%, end-diastolic volume 130 mL). An urgent angiocomputed tomography (ACT) confirmed, also with three-dimensional rendering, the real expansion of the fusiform aneurysm. The mass involved the entire mediastinum, such as to displace the heart in a posterior position and crushing it on the diaphragm (Figure 2). Having assessed the life-threatening condition given by the enormous aneurysm size and the consequent increased risk of rupture, an emergency surgical approach became mandatory, thus a modified Cabrol operation was performed. At the time of surgery, the right axillary artery and femoral vein were cannulated and cardiopulmonary bypass was established prior to opening the sternum.

The choice of this surgical technique was modified intraoperatively when the aneurysmal sac was transversally opened and approximately 1400 cc of blood was aspirated because we observed that the distance between coronary artery ostia (CAO) and aortic annulus was less than 1.5 cm, the aortic root was heavily calcified, and CAO was also very dislocated, fixed, and difficult to mobilize to perform a direct anastomosis on the aortic valved conduit. Assessed that anatomy of aortic annulus and CAO, distorted by the presence of the aneurysm, was unsuitable for the surgical technique of replacement with valved conduit and direct reimplantation of the coronary buttons on the graft and considering the risk of laceration and bleeding of conventional coronary artery anastomosis, a modified-Cabrol technique was adopted to interpose two separate short 8 mm Dacron tubular grafts between CAO and aortic valved conduit. The surgical procedure was performed without complications. Subsequently, genetic tests were performed which did not reveal any mutations.

Despite a delayed awakening in the intensive care unit, the patient had an uncomplicated postoperative course. On the 16th postoperative day, he was discharged home following an ACT examination which confirmed good functioning and positioning of the prosthesis and excellent graft patency, scheduling follow-up with ACT and TTE 1, 6, and 12 months after surgery.

Discussion

An unruptured, asymptomatic GAAA is extremely rare in clinical practice. Initially they can develop subtly being asymptomatic, however, when excessive dimensions are reached, a variety of clinical manifestations can occur, including compressive symptoms such as dyspnea, dry cough, dysphagia, dysphonia and chest pain. Only two cases of unruptured and asymptomatic GAAAs are reported in literature [1-6].

Because the risk of catastrophic rupture or dissection is proportional to the diameter of the aneurysm, aneurysmal size is the criterion for elective surgical repair. Nevertheless, operative mortality for GAAA remains high, especially when surgery is performed on an emergency basis and the aneurysmal size exceeds those recommended for surgery by guidelines, because GAAA presents with extreme clinical variability, it is difficult to establish standardized principles for their treatment, therefore it needs to be tailored on an individual basis, adapting surgical strategy to the variability of this pathology. Hence, their surgical management is a technical challenge [7].

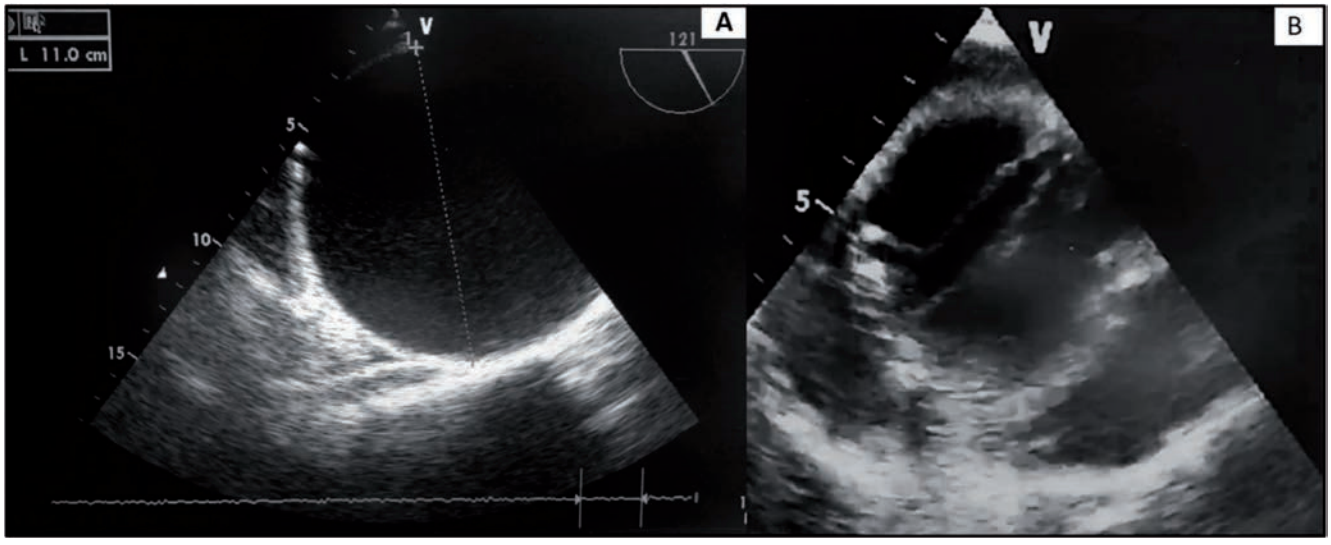

Figure 1. A) Transesophageal echocardiography in long axis aortic view shows giant ascending aortic aneurysm (11 cm) parcelar calcifications of the aortic wall, in the absence of dissection and pericardial effusion; B) zoomed view of the aortic valve at 45° in short axis, showing "true" aortic valve bicuspid type 0-lateral, without raphe with moderate regurgitation jet. Presence of calcifications of the aortic ring.

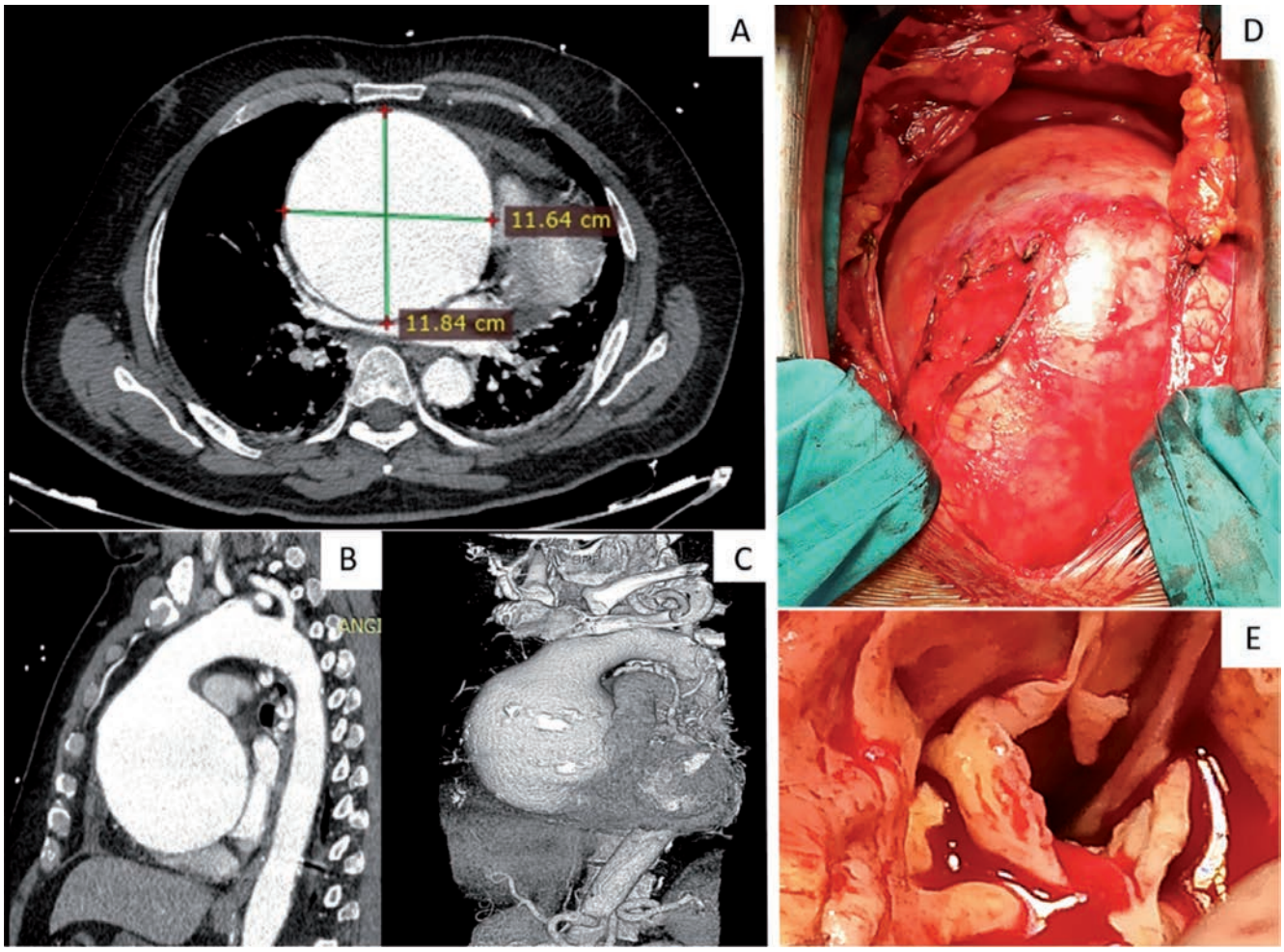

Figure 2. A) Angiocomputed tomography sagittal scan shows expansion of the aneurysm into the mediastinum; B) longitudinal scan; C) three-dimensional rendering shows that physiological anatomical relation is altered by the aneurysm, which has displaced the heart into a more posterior-lateral position; D) surgical view after sternal opening, the aneurysm occupies the entire mediastinum, preventing the vision of the heart, which is crushed on the diaphragm; E) intraoperative view of the aortic valve, which is confirmed as bicuspid without raphe, with sclerotic and calcified cusps. The subtype 0-lateral is confirmed by viewing the right coronary ostium as this phenotype has one coronary artery arising from each sinus.

The modified Cabrol procedure has a smaller indication for aortic surgery, but in some particularly indicated cases, it may be the most appropriate choice. This technique still remains an important surgical procedure used as an option to treat widely spaced coronary buttons in aortic root surgery; however, it is not usually applied as a first choice technique as its use is related to complications such as kinking or thrombosis of the graft, resulting in reduced graft patency.

The strengths of the modified Cabrol technique lay in its ability to provide a safe and tension-free anastomosis of the CAO to the aortic conduit when conventional reimplantation techniques fail to do so. The conditions that make use of the modified Cabrol anastomosis necessary are: poor mobilization of the CAO which can occur when ostia are less than 1.5 cm above the annulus, extreme aortic dilatation, friable or torn CAO that cannot be implanted with the conventional button approach, and heavily calcified aortic root.

When CAO is very dislocated, fixed, and difficult to mobilize to perform a direct anastomosis on the aortic valved conduit, the modified Cabrol procedure can be used to obtain tension-free anastomosis [8]. In our case all the conditions described were present; furthermore, the aortic annulus was strongly dilated, and the aortic valve was bicuspid, classifiable as type 0-lateral with no raphe. Although BAV is related to the development of AAA, evidence of type 0-lateral BAV with insufficiency occurs in only 2% of cases, making it a very rare subcategory of BAV [9].

Conclusions

Despite the modified Cabrol technique may be considered obsolete, when the conventional button reimplantation technique is not feasible, it can be used as a bailout procedure that can be pulled by cardiac surgeons as a "trump card", for complex aortic root replacements, in order to achieve tension-free anastomoses, limit bleeding and reduce development of suture-line pseudoaneurysms.

Considering that GAAAs can develop insidiously with extreme clinical variability, even in the absence of symptoms, and

that their finding is usually incidental, this case also highlights the importance of not underestimating nonspecific symptoms such as dyspnea on exertion and always deepening the evaluation of diastolic murmurs that may underlie BAV aortopathy, also investigating family medical history, even in young and healthy subjects without a clinical history of cardiovascular disease.

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