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A Case of Bicuspid Aorta with Stanford Type a Aortic Dissection

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ABSTRACT

Bicuspid aortic valve disease (BAV) is a congenital disease that frequently produces complications during adulthood, and is considered not only a problem of valvulogenesis, but also a more complex genetic alteration that involves the development of the heart and the aorta, which entails a high morbidity and mortality in these patients. In 50% of adults with this pathology, non-valvular anomalies are observed, with dilation of the ascending aorta being the most common. Genes such as NOTCH1, UFDL1, ACTA2, eNOS, among others, have been identified as responsible for the appearance of aneurysms, which increases the risk in these patients of complications such as aorticdissection and its subsequent rupture. Painless aortic dissection may be seen in less than 5% of patients and may have an atypical clinical presentation. A case and review of the literature is presented in which we can observe the great complexity of this pathology, in the case of a patient with BAV, annulo-ectatic dilation of the aortic root and ascending aorta who developed heart failure,syncope and presented as a complication Stanford type A aortic dissection, requiring highly specialized surgical treatment.

KEYWORDS: bicuspid aortic valve disease, aortic dissection, acute aortic syndrome

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INTRODUCTION

Bicuspid aortic valve (BAV) is the most common congenital heart disease, found in 1% of the generalpopulation, with a clear male preponderance, accounting for up to 80% of cases. In recent years, a family association has been described, with a prevalence of BAV of 24% in relatives of affected people. Various mutations have also been identified, such as NOTCH1 (which acts in cardiac development and years later in the regulation of tissue calcium deposition), ACTA 2 (which codes for muscle alpha actin, intervening in the development of aneurysms) and FBN1 or TGF-β pathway β mutations. For this reason, the guidelines recommend screening for this pathology in first-degree relatives of affected people[1,2]. In this case there was no family history. Typically, the bicuspid aorticvalve comprises two leaflets of unequal size, due to the fusion of the right and left coronary leaflets in up to 85%, the fusion of the right and non-coronary leaflets with a right-left orientation are of an atypical pattern present in 12% and fusion of the left and non-coronary leaflets (3%)[3,4].

Accelerated cystic degeneration of the tunica media begins in childhood and is progressive, the dilation rate ranges from 0.2-2.3 mm/year depending on morphology, characteristics of the individual,pathologies, and associated complications. The presence and type of BAV fusion was associated withchanges in regional distribution, shear stress, systolic flow eccentricity, and expression of BAV aortopathy [5,6]

CLINICAL CASE

33- Year-old male with a history of systemic arterial hypertension and hypercholesterolemia. He beganhis current clinical condition 5 days prior to his hospital admission when he presented a sensation ofdyspnea with medium exertion which progressed to rest, orthopnea, profuse diaphoresis,

palpitations and nausea. On physical examination he was conscious, restless and diaphoretic, precordium with the presence of a GII/IV systolic murmur in the aortic focus and a long holodiastolic murmur in the accessory focus. Lung fields with presence of crackles at both bases. Extremities with presence of Celer pulse. During the first hours of his stay in the emergency department, he suddenly experienced syncope, with recovery ad integrum. The chest x-ray (fig.1A) and transthoracic echocardiogram showed the presence of hypertrophy and severe dilation of the left ventricle, severe global hypokinesia with LVEF of 26%, both atria severely dilated. Bileaflet aortic valve with failure in coaptation due to severe dilation of the aortic root generating severe aortic insufficiency. Aortic dissection flap from the root of the aorta extending to the arch and descending aorta, PSAP 49 mmHg. Simple and contrast-enhanced CT and angiotomography (fig. 1B) show dilation of the ascending aorta with a transverse diameter of 10.8 cm, which reduces to 3.6 cm in the aortic arch and 3.5 cm inthe descending aorta.

Given the data of heart failure and type A aortic dissection, the patient was referred to the cardiosurgery service, performing a debranching procedure and Bentall and Bono surgery (fig. 1C), with tubular valve replacement. The intraoperative findings were asymmetric keel thoracic deformitywith protrusion of the right hemithorax, grade III cardiomegaly, pulmonary aorta ratio 4:1, generalized hypokinesia, aneurysmal dilation of the ascending aorta with dissection flap 4mm from the ostium of the right coronary artery that continues through the aortic arch affecting supra-aortic trunks, bileaflet aortic valve with dilated annulus, non-coapting valves.

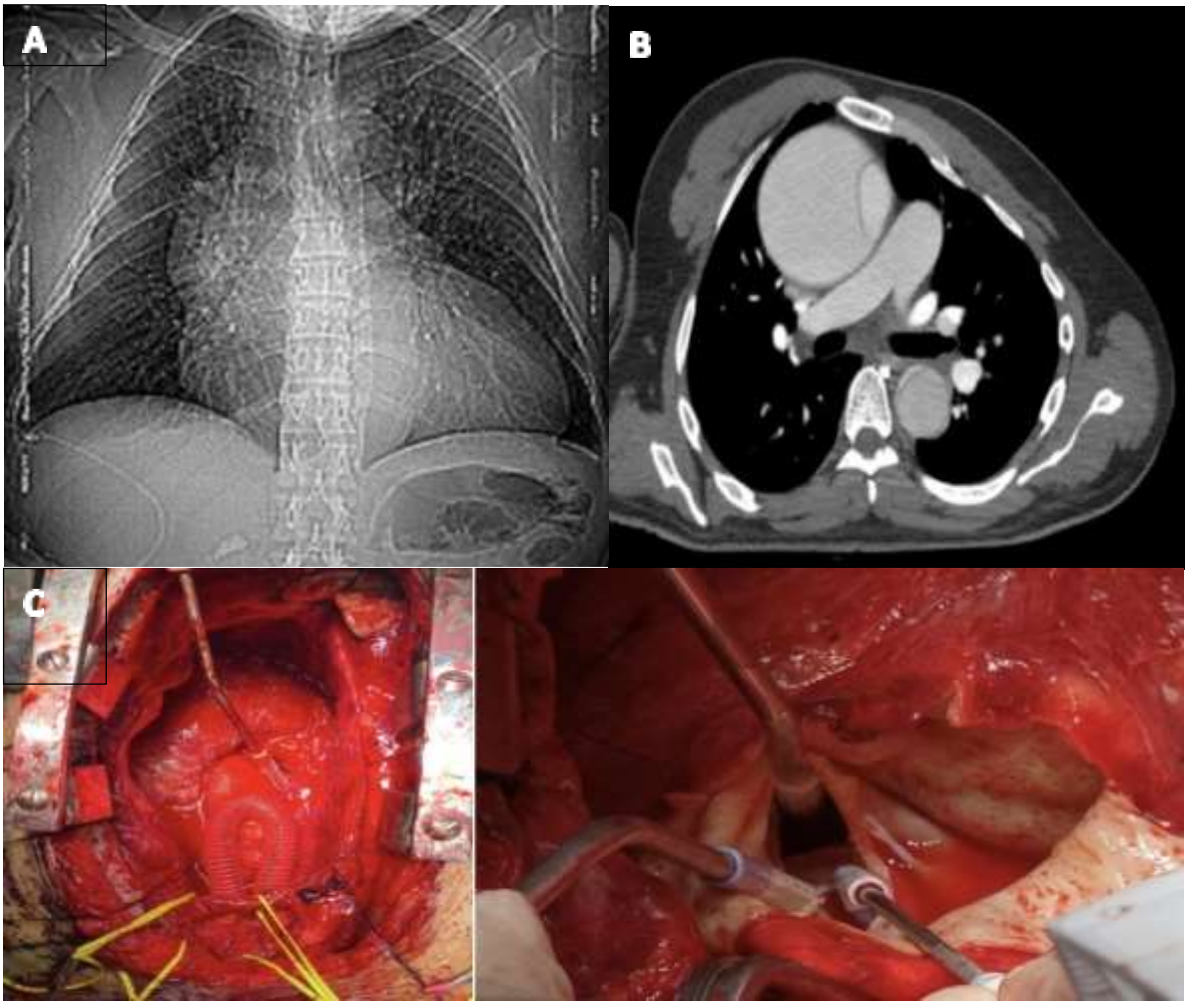

Figure 1. A) Chest x-ray with aortic aneurysm and cardiomegaly. B) Contrast-enhanced tomography with aneurysmal dissection in the aortic root. C) Bentall and Bono technique in truelumen and aortic dissection flap.

DISCUSSION

The aneurysm is defined as the diameter greater than two standard deviations above the mean,according to adjusted normograms, based on specific segment; Whether it is the aortic root, the sinus of Valsalva or the ascending aorta, it is the main factor associated with aortic dissection and also leads to premature valve dysfunction, with stenosis being more common than severe aortic insufficiency (associated with young people, like the patient). Rupture of the media culminates in aortic dissection, creating a new channel, the false lumen. Once the dissection begins, the pulsatile flow of blood withinthe aortic wall causes it to extend[6]. Underlying hypertension, found in the patient and in the general population is 75%, is considered an independent predictor of

mortality[7]. The phenotype that occurs is the most frequent, type I; with dilation of the tubular ascending aorta along its convexity with mild to moderate root dilation. Type II: isolated tubular dilation of the ascending aorta and Type III refersto isolated dilation of the aortic root[8,9]. The risk of dissection is described as 3.8% with an ascendingdiameter of 5.3 cm (10.8 cm in this case)[10].

They are described as typical clinical data in 90% of acute dissections; Sudden, severe pain, referred to as tearing or stabbing in the anterior or posterior chest. The pain may migrate as the dissection progresses along the aorta or the branches of the vessels and is accompanied by anxiety[10]. On physicalexamination, pulse deficits can be found in 10-30%, with a considerable variation > 20 mmHg in SBP. When comparing blood pressure in the arms, it may be intermittent, because the dynamic movement of the flap interferes with the perfusion of the branches of the glass. The murmur is diastolic in aortic insufficiency related to aortic dissection, heard along the right sternal border, compared to the left border for aortic insufficiency due to primary aortic valve disease. The durationmay be short due to rapid ventricular filling and early equilibration of aortic and left ventricular diastolic pressures[10].

The patient had an atypical clinical presentation, consisting of painless dissection, syncope (relatedto hypotension due to cardiac tamponade or aortic rupture, obstruction of cerebral vessels, activation of cerebral baroreceptors), heart failure (associated with dissection of the ascending aorta), so predominant, which makes diagnosis more difficult and delays medical care[11].

As an initial study, an ECG should be requested, finding findings consistent with associated manifestations. Complications related to the dissection can also be observed, such as coronary involvement. The chest x-ray revealed widening of the aortic silhouette, the most common anomaly in 56-63%. The diagnosis of aortic dissection, in addition to clinical data, is confirmed by imaging; the choice of study depends on availability. In the CT angiography and echocardiogram performed on the patient, the false lumen was observed[12,13]. Contrast-enhanced computed tomography, magnetic resonance imaging, and transesophageal echocardiography have high sensitivity and specificity and are useful when additional evaluation is required[14].

According to the DeBakey classification, based on the site of origin, we found type I in the case; aortic dissection that begins in the ascending aorta and extends at least to the aortic arch or beyond. Type II involves only the ascending aorta, type III dissections begin in the descending aorta, most often only distal to the left subclavian artery. The Stanford classification divides it into type A, present in the patient, described as; The intimal flap originating from the ascending aorta remains a highly lethal condition: more than half of all patients die within 30 days of the index event, management is emergency open surgical intervention. Without surgery, it can cause infarction, tamponade or acute coronary ischemia. Type B; In the descending aorta, treatment is usually conservative with control of systemic hypertension; beta-adrenergic blockade is the mainstay of therapy[14,15]. Long-term survival rates after acute type B dissection have been reported of 56% to 92% at 1 year and 48% to 82% at 5 years[16].

The decision for surgical treatment is multifactorial and is established by the anatomical conditions of the aorta, the underlying disease, the risk of anticoagulation, age and the presence of active infection, among others. Replacement of the root and ascending aorta with a valved tubular graft, called the Bentall procedure, is considered the option for the treatment of aneurysmal disease of the ascending aorta that is accompanied by annuloectasia. This technique has shown low morbidity and mortality, from 1.7 to 17%, and a five-year survival of 73 to 92% and a 10-year survival of 60 to 73%. However, surgical mortality can vary significantly depending on the hospital center, due to theexperience of the medical teams, available resources, and patient heterogeneity[17,18,19]. As used in thecase of our patient when considering the gold standard treatment, the Bentall and Bono Technique, consists of replacing the root and the aortic valve with a composite graft formed by a Dacron vasculargraft (straight or with a morphology that imitation of the sinuses of Valsalva) and a valve prosthesis;The coronary arteries must be reimplanted to the Dacron vascular graft. twenty.

Pseudoaneurysm of the ascending aorta, as a complication in cardiac surgery, has an incidence of less than 0.5%, with mortality ranging from 6.7-60% 21. Graft infection and mediastinitis are the main predisposing factors. 1–3. The symptoms of these complications are subject to their size and location, but they can be asymptomatic and be a casual finding in follow-up studies. 22. Moderate tosevere aortic stenosis and dissection involving the ostium of the coronary artery could be associated with adverse clinical outcomes and is recommended as part of basic follow-up after surgery, with echocardiography [23,24].

CONCLUSION

The timely detection of the bicuspid aortic valve and the risk factors for its progression to aortopathy are of great importance for the prevention of outcomes with a high impact on the patient's quality oflife and mortality.

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