CASE REPORT
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Bentall and de Bono procedure in the context of double aortic valve lesion with right sinus of Valsalva aneurysm and ischemic cardiomyopathy: case report

Procedimiento de Bentall y de Bono en el contexto de doble lesión aórtica con aneurisma de seno de Valsalva derecho y miocardiopatía isquémica: a propósito de un caso

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ABSTRACT

The sinuses of Valsalva are subtle dilatations of the ascending aorta that correspond to the portion of the left ventricular outflow tract located between the aortic valve annulus and the sinotubular junction. We present a case of a right sinus of Valsalva aneurysm associated with ischemic cardiomyopathy, characterized by a 15-year history of cardiovascular symptoms, which prompted referral to our center in January 2024 following the incidental discovery of a cardiac murmur during a preoperative evaluation. The etiology, anatomy, histology, and diagnostic and surgical treatment guidelines for this condition are discussed.

Keywords: aorta, aortic sinus, aortic valve, Bentall procedure, coronary artery bypass grafting, sinus of Valsalva.

RESUMEN

Los senos de Valsalva son ligeras dilataciones de la aorta ascendente y corresponden a la porción anatómica del tracto de salida ventricular izquierdo que se localiza entre el anillo de la válvula aórtica y la unión sinotubular. Se presenta un caso de un aneurisma de seno de Valsalva derecho, asociado a miocardiopatía isquémica, con presencia de aproximadamente 15 años de síntomas cardiovasculares que es referido a nuestro centro asistencial en enero de 2024 debido al hallazgo de un soplo cardíaco en una evaluación preoperatoria. Se discute su etiología, anatomía, histología y pautas para el diagnóstico y tratamiento quirúrgico.

Palabras clave: *aorta, senos aórticos, válvula aórtica, procedimiento de Bentall, revascularización coronaria, seno de Valsalva.*

INTRODUCTION

The sinuses of Valsalva are slight dilatations of the ascending aorta that correspond to the anatomical portion of the left ventricular outflow tract located

Abbreviations:
CABG = coronary artery bypass grafting
ESC = European Society of Cardiology
LVEF = left ventricle ejection fraction

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between the aortic valve annulus and the sinotubular junction. There are three Valsalva sinuses: the right coronary sinus, the left coronary sinus, and the non-coronary sinus, each one connected to its respective coronary ostium. The anomalies of these sinuses can be explained by their histological characteristics. Notably, the superior margin of each sinus, at the peak of the semilunar insertion line, is known as the supravalvular ridge, which is mainly composed of elastic and collagen fibers interspersed with smooth muscle cells and fibroblasts.[1,2]

The left coronary sinus ridge contains a greater number of smooth muscle cells embedded within a dense extracellular matrix rich in type I collagen fibers. In contrast, the right coronary sinus contains fewer smooth muscle fibers immersed in type III collagen. It is known that the walls of the Valsalva sinuses are primarily composed of type I collagen fibers in their lower portions, closest to the insertion of the aortic leaflets, where there is also a slight presence of left ventricular muscle fibers. It is important to note that the amount of type I collagen fibers gradually decreases in an ascending direction while the amount of elastic fibers increases progressively within each sinus.[1,2]

Although aortic valve disease is relatively common, its association with sinus of Valsalva aneurysm is rare and only a limited case series have been reported in the past years. Nevertheless, the coexistence of these entities, particularly when accompanied by ischemic cardiomyopathy poses a unique diagnosis and therapeutic assessment. All of this underscores the relevance of the present case, in which a double aortic valve lesion, a right sinus of Valsalva aneurysm and ischemich cardiomyopathy were successfully managed with the Bentall and de Bono procedure.

CASE DESCRIPTION

A 78-year-old male patient with a past medical history of a transient loss of consciousness 15 years ago and an episode of chest pain 10 years ago was referred in January 2024 after a preoperative evaluation for inguinal hernioplasty, during which a cardiac murmur was detected. At a follow-up appointment in the outpatient clinic, a transthoracic echocardiogram was performed exhibiting significant leaflet thickening and calcification, severely restricting both opening and closure. These findings were associated with an eccentric systolic jet traversing an aneurysmal segment of the right sinus of Valsalva. The lesion resulted in mixed aortic valve disease, predominantly stenotic, with a peak transvalvular velocity of 3.7 m/s, a peak gradient of 55 mmHg, a mean gradient of 28 mmHg, and a left ventricular outflow tract velocity of 0.6 m/s. Mild aortic regurgitation was also present. Aortic measurements included an aortic annulus diameter of 2.0 cm (1.3 cm/m^2), sinus of Valsalva diameter of 3.9 cm (2.6 cm/m^2), sinotubular junction of 2.9 cm (1.9 cm/m^2), and ascending aorta diameter of 3.4 cm (2.2 cm/m^2). All of the following findings confirmed a tricuspid aortic valve with double aortic lesion and preserved left ventricle ejection fraction (LVEF), as well as the incidental finding of a right Valsalva sinus aneurysm *(Figure 1)*.

On September 19, 2024, a diagnostic coronary angiography was performed; finding a left-dominant coronary artery pattern was identified. The left anterior descending artery exhibited diffuse disease in the proximal and mid segments, with a focal lesion causing 70% stenosis in the distal segment. The circumflex artery also demonstrated diffuse disease, with a maximal stenosis of 70% in the distal segment. The right coronary artery showed diffuse proximal disease with

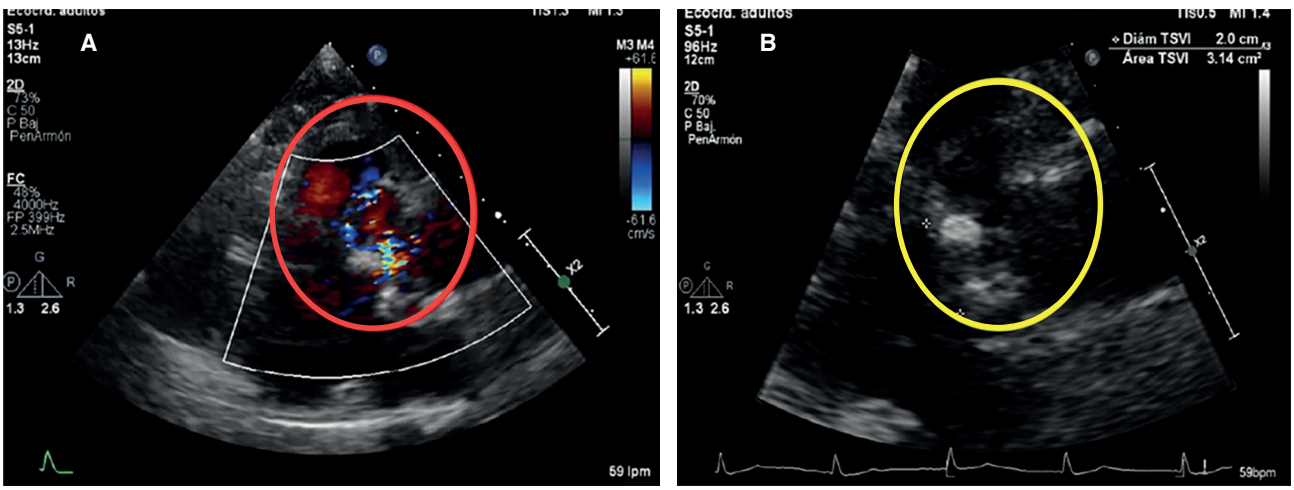

Figure 1: A) Dilatation of the sinus of Valsalva observed via color Doppler. **B)** Transthoracic echocardiogram image.

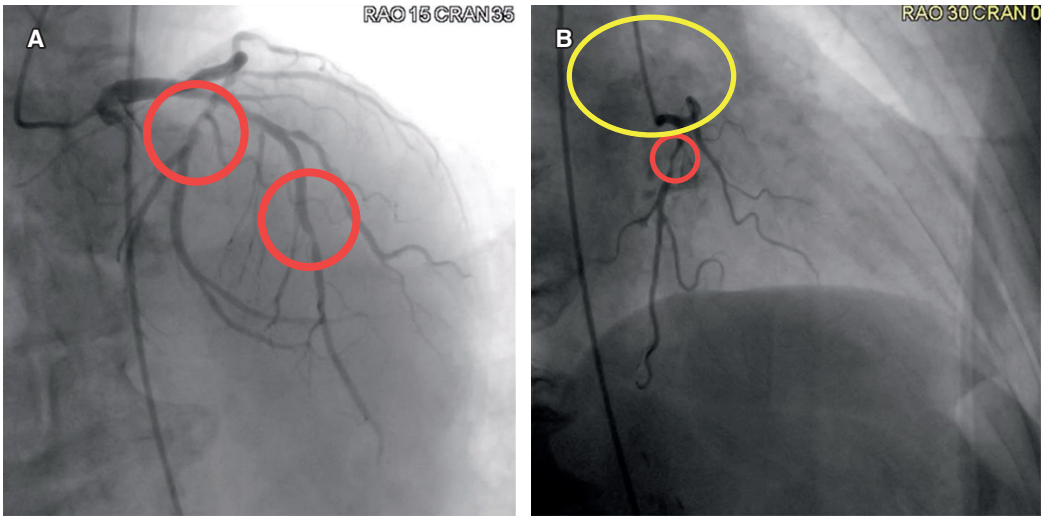

Figure 2:

A) Occlusion areas identified in the left coronary artery, specifically in its circumflex branch. **B)** The red circle delineates an occlusion area in the right coronary artery. The yellow circle identifies a slight deviation in the flow of contrast medium, caused by the dilation of the right sinus of Valsalva.

a maximal stenosis of 90%, all of this revealing triple-vessel coronary artery disease with a SYNTAX score of 22 *(Figure 2)*.

The patient was hospitalized for monitoring and was considered a candidate for Bentall and De Bono surgery with coronary artery bypass grafting (CABG), due to the presence mixed aortic valve disease. He underwent successful CABG and Bentall-De Bono operation.

A simultaneous dual approach was performed. The first consisted of a median sternotomy for the harvesting of the left internal thoracic artery in a caudo-cephalic direction, from its bifurcation up to the first costal arch, ensuring meticulous hemostasis of its branches through ligation. The second approach involved the dissection and procurement of the left great saphenous vein in a caudal direction. Systemic heparinization was administered, and purse-string sutures with 3-0 Prolene were placed: double sutures on the ascending aorta, and single sutures on the right atrium, right superior pulmonary vein, and the aortic root. Once an optimal activated clotting time was achieved, the aforementioned structures were cannulated, and cardiopulmonary bypass was initiated under normothermia. The ascending aorta was cross-clamped, and antegrade crystalloid cardioplegia was administered. CABG was first performed. Subsequently, a transverse aortotomy was performed, exposing both the left and right coronary ostia. The native aortic valve and ascending aorta were excised, preserving both ostia using a "button" technique. A modified Bentall procedure was performed using a 30 mm woven Dacron graft with a 27 mm aortic valve prosthesis. U-stitches with 2-0 Ethibond were placed and passed through the valved conduit. The graft was deployed using the parachute technique and secured. Coronary reimplantation was performed using 5-0 Prolene sutures for both the left and right coronary arteries. Adequate valve function and hemostasis were confirmed. The distal anastomosis of the Dacron graft to the native aorta was completed using 4-0 prolene.

Following this, the aortic cross-clamp was removed, and the patient returned to nodal rhythm, with temporary epicardial pacing leads placed. Weaning from cardiopulmonary bypass was successfully achieved on the first attempt, with stable hemodynamic parameters and adequate left heart decompression. Decannulation was performed, followed by protamine administration. Mediastinal packing was carried out for hemostasis. Chest drains were placed. Due to bleeding from the posterior surface of the pulmonary root, additional packing with two surgical rolls was required. The procedure was concluded, and the patient was transferred to the intensive care unit.

COMMENTARY

Aneurysms of the Valsalva sinuses are abnormalities of the aortic root that can be identified between the aortic valve annulus and the sinotubular junction. They may be of either congenital or acquired origin. Congenital aneurysms are associated with connective tissue disorders such as Marfan syndrome (characterized by fibrillin-1 deficiency, resulting in medial layer involvement) and Loeys-Dietz syndrome, caused by mutations in transforming growth factor beta (TGF-β) receptors. Both conditions contribute to the weakening of the tunica media. Alterations in elastin and collagen are the primary congenital causes affecting the normal histology of the Valsalva sinuses.[3,4]

Acquired forms arise from factors that lead to weakening of the tunica media and the extracellular matrix, most commonly due to ischemic processes affecting the vascular wall or conditions causing wall thickening. Infectious etiologies are

associated with propagation of infection, septic emboli, or direct invasion of the vascular wall. Likewise, non-infectious inflammatory processes, such as various forms of arteritis, are strongly associated. Valsalva sinus aneurysms frequently coexist with other pathologies such as ventricular septal defects, aortic valve disease, or infections like syphilis, endocarditis, or even trauma.[5,6]

The progression of this condition typically begins with an aneurysm that is initially asymptomatic. As the disease advances, valvular prolapse may occur, most commonly into the left ventricular outflow or inflow tract. Subsequent rupture results in a left-to-right shunt, the severity of which is proportional to the size of the rupture area. The classification proposed by Sakakibara and Konno remains the only formal system used to differentiate the various types of sinus of Valsalva aneurysms based on the affected coronary sinus and the anatomical region into which the aneurysm protrudes or ruptures *(Table 1)*.[4]

Diagnosis of this condition is primarily made using echocardiography, which is considered the gold standard. However, other diagnostic modalities, such as aortography, are also clinically valuable. Transthoracic echocardiography enables visualization of the aortic root, allowing identification of aneurysmal dilatation. It also facilitates the estimation of aneurysm size and assessment of associated abnormalities, such as aortic valve insufficiency or ventricular septal defects. On the other hand, transesophageal echocardiography provides superior imaging resolution, allowing better assessment of aneurysm extension and its effects on adjacent structures. These echocardiographic modalities have demonstrated a diagnostic accuracy of approximately 90% for ruptured sinus of Valsalva aneurysms and 75% for unruptured ones. They are also instrumental in determining the originating sinus, the severity of the condition, and the associated mechanisms contributing to the pathophysiology.[6,7]

Table 1: Classification of sinus of Valsalva aneurysms proposed by Sakakibara and Konno.

Type I	Connects the right sinus of Valsalva and the right ventricular outflow tract below the pulmonary valve
Type II	Connects the right sinus of Valsalva and the right ventricle at the crista supraventricularis
Type III	
IIIa	Connects the right sinus of Valsalva and the right atrium
IIIv	Connects the posterior area of the right sinus of Valsalva and the right ventricle
IIIa+v	Connects the right sinus of Valsalva to both the right atrium and the right ventricle
Type IV	Connects the non-coronary sinus of Valsalva and the right atrium

Classification for aneurysms of the sinus of Valsalva proposed by Sakakibara and Kono according to Arenaza.[1]

Color Doppler imaging may also be utilized to visualize systolic and diastolic flow patterns, considering the physiological principle that the aorta functions as a high-pressure system. This allows for the observation of flow diversion from the aortic outflow into the aneurysmal sac. Other imaging techniques, such as magnetic resonance imaging, contrast-enhanced aortography, and computed tomography, may also be employed. Aortography, in particular, offers the advantage of evaluating other aortic regions and facilitates the inclusion or exclusion of differential diagnoses.[7,8]

The treatment of choice in these cases is always surgical intervention. According to the 2024 European Society of Cardiology (ESC) Guidelines on the diagnosis and treatment of aortic diseases, published by ESC, various surgical techniques are available to appropriately address this condition. It is crucial to emphasize that conservative measures are both ineffective and insufficient for achieving proper management of this pathology.

Based on the underlying pathophysiology of sinus of Valsalva aneurysms, ruptured cases are associated with more rapid clinical deterioration, often leading to congestive heart failure as a result of the aforementioned left-to-right shunt. Consequently, the European guidelines recommend surgical repair even for unruptured aneurysms when they are associated with malignant arrhythmias, infection, coronary artery obstruction, or obstruction of the ventricular outflow tract. Therefore, the aneurysm's rate of progression must be considered a key factor when determining the timing of surgical intervention, along with any coexisting pathologies.

In the present case, CABG was performed due to the patient's significant coronary artery disease. A Bentall and De Bono procedure was selected to address the associated aortic valve involvement and right sinus of Valsalva dilatation. In accordance with the ESC guidelines, the selected approach allowed for complete management of the coronary artery disease and correction of the aneurysmal dilation and valvular dysfunction through replacement with a prosthetic valve. Therefore, it's important to emphasize in the Need of an individualize approach to every case that allows the attendings to get a more realistic scenario while applying the knowledge and algorithms established by different type of guideline as the ones of ESC allowing to have a more punctual approach to the patient case type, and how that can be a determining factor in identifying the differences in mortality rates, and the type of surgical management that is required. It's essential to understand that identifying and classifying sinus of Valsalva aneurysms is key to achieving the right approach and the best surgical choice.

CONCLUSIONS

The coexistence of a double aortic valve lesion, right sinus of Valsalva aneurysm and ischemic cardiomyopathy is unusual. This case highlights the need for a comprehensive evaluation of the hemodynamic impact of each condition and their surgical approach. The successful outcome after a Bentall and de Bono procedure supports its role as an effective option in comparable cases.

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