Case Report
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Pseudoaneurysm of the mitral aortic intervalvular fibrosa secondary to treatment of tunnel aortic subvalvular stenosis

Pseudoaneurisma de la Fibrosa Intervalvular Mitro-Aórtica Secundaria al Tratamiento de Estenosis Subvalvular Aórtica en Túnel

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SUMMARY

Twelve year-old child referred for echocardiographic evaluation after surgical treatment of subaortic stenosis. Transthoracic and transesofageal (2D and real time 3D) showed a large pseudoaneurysm of the mitral-aortic intervalvular fibrosa associated with a small perforation at the base of the anterior leaflet of the mitral valve. She had neonatal coarctation of the aorta associated with ventricular septal defect and required multiple surgical procedures in infancy due to an inappropriate treatment of the predisposing anatomical structures of subvalvular aortic stenosis. In this report, the mechanisms of these iatrogenic lesions and the abnormal anatomical features that predispose to left sided obstructive lesions are discussed

Descriptors: Aortic Coarctation; Discrete Subaortic Stenosis; Heart Septal Defects, Ventricular; Echocardiography

RESUMEN

Niño con 12 años de edad, encaminado para ecocardiografía transtorácica y transesofágica 2D y 3D para control postoperatorio de corrección de estenosis subaórtica, que evidenció un gran pseudoaneurisma de fibrosa intervalvular mitro-aórtica, asociada a la perforación de la base del folleto anterior de la válvula mitral. Se trababa de un caso de coartación de la aorta (CoAo) neonatal asociada a la comunicación interventricular (CIV), que necesitó de múltiples intervenciones quirúrgicas en la infancia, debido al no tratamiento adecuado de las estructuras que predisponen la estenosis subvalvular aórtica. En este relato, son discutidas las particularidades anatómicas que predisponen a lesiones obstructivas izquierdas y a probable causa de desarrollo de esas lesiones iatrogénicas.

Descriptores: Coartación Aórtica, Estenosis Subaórtica, Comunicación Interventricular, Ecocardiografía

INTRODUCTION

Obstructions of the outflow tract of the left ventricle (LV) usually present at various levels, and can affect, concomitantly, the inflow route, the subaortic region, the aortic valve, and the aortic arch. Although anatomical substrates are already present from fetal life, the lesions tend to evolve and manifest clinically over time. Aortic subvalvular stenosis is rarely present at birth having a progressive and recurrent characteristic.[1,2]

The identification of the mechanisms of subaortic stenosis during the echocardiographic study is essential for planning the treatment of patients with left obstructive lesions. These,

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if not properly addressed during surgery, lead to repeated cardiac operations throughout lifetime.

Herein, we report a case of neonatal aortic coarctation, which evolved with tunnel aortic subvalvular stenosis, requiring multiple surgical interventions in infancy due to improper treatment of the structures predisposing to left obstructive lesions. In the late postoperative period of the last operation, an aneurysm emerged in the mitral-aortic intervalvular fibrosa, possibly due to the inadvertent handling of the left ventriculo-infundibular fold (LVIF).

CASE REPORT

SPSC, female, 12 years-old, asymptomatic with respect to cardiovascular system, in postoperative follow-up after correction of aortic coarctation, ventriculoseptoplasty, expansion of the LV outflow tract and the ascending aorta, associated with aortic valve correction. The control echocardiography revealed the presence of a large aneurysm of the mitral-aortic intervalvular fibrosa associated with perforation of the base of the anterior leaflet of the mitral valve.

Background

Patient was referred to our institution at 15 days of life due to congestive heart failure by severe aortic coarctation and hypoplastic aortic isthmus associated with large and persistent perimembranous VSD of the ductus arteriosus. Soon after the diagnosis, the patient underwent termino-terminal isthmus plasty, ligation of the ductus arteriosus, and banding of the pulmonary trunk with good outcome. At 40 days of life, it was decided to proceed with the correction of all defects, being performed the closure of the VSD with flap and removal of pulmonary banding.

At the age of nine months, the patient already showed signs of aortic coarctation, being also observed obstruction in the LV outflow tract. Cardiac catheterization was performed, which identified aortic subvalvular stenosis by posterior deviation of the infundibular septum associated with persistent LVIF and aortic coarctation with hypoplastic isthmus. There was a poor response to the attempt of dilatation with balloon catheter. The patient remained in clinical evolution until the fourth year when a significant increase in the subaortic gradient was noted (104 mmHg measured by echocardiography). A new surgical correction was indicated, performing resection of the fibrotic membrane occluding 70% of the LV outflow tract. This gradient was reduced to 38 mmHg in the early postoperative period.

The aortic arch was not addressed in this surgery, because the recoarctation was believed to be little significant clinically. The follow-up continued at the institution, with reappearance of subaortic stenosis (subaortic gradient again above 100 mmHg) and concomitant worsening of the aortic coarctation (subaortic gradient of 57 mmHg), thus being referred for surgical treatment. In this last operation, at 11 years old, the coarcted region was expanded with bovine pericardium flap, a new subaortic membrane was resected, and the LV outflow tract was expanded. The operation was completed by aortic commissurotomy and valvuloplasty in the ascending aorta.

The patient evolved clinically well without significant narrowing in the regions approached, however, the control echocardiography performed late after operation (twelve years-old) showed a pseudoaneurysm in the region of the mitral-aortic intervalvular fibrosa associated with a perforation of the anterior leaflet of the mitral valve, specifically, in the base of the component A3. The two-dimensional study was complemented by three-dimensional transesophageal echocardiography which elucidated the mechanism of lesion development and the location of the iatrogenic hole in mitral valve (Figure 1).

DISCUSSION

This case is quite illustrative to show the classical evolution and stories of natural and modified left heart obstructive lesions. What appeared only neonatal aortic coarctation associated with VSD was a compendium of abnormalities of the structures on the left side of the heart, resulting in progressive and recurrent obstructive lesions.

The neonatal aortic coarctation generally occurs due to the reduction of effective anterograde aortic flow, which prevents the optimal development of the transverse arch and aortic isthmus during fetal life. The aortic isthmus is particularly susceptible to obstructions, since with circulations occurring in parallel only 10% of the cardiac output pass through this region [3]. Any injury that further restricts the anterograde aortic flow, either the presence of large VSDs that steal flow from left to right or interposed structures in the outflow tract, the chances of neonatal coarctation are very high.

In this case, three important factors could justify arch hypoplasia and neonatal aortic coarctation: a large VSD, the posterior deviation of the infundibular septum in subaortic region, and persistent LVIF. These two latter change the geometry of the left outflow tract, stretching it and forming a true subaortic tunnel, which restricts the effective flow to the ascending aorta [4].

There are several mechanisms of aortic subvalvular stenosis: isolated fibromuscular membrane; tissue interposition of the mitral valve in the LV outflow tract; hypertrophic obstructive cardiomyopathy; tricuspid valve tissue herniation by VSD to the subaortic region; posterior deviation of the infundibular septum, and VSDs associated with overriding aorta in which the crest of the trabecular septum deviates to subaortic region [5]. Abnormal anatomical configurations, such as increased LVIF (fibrous discontinuity

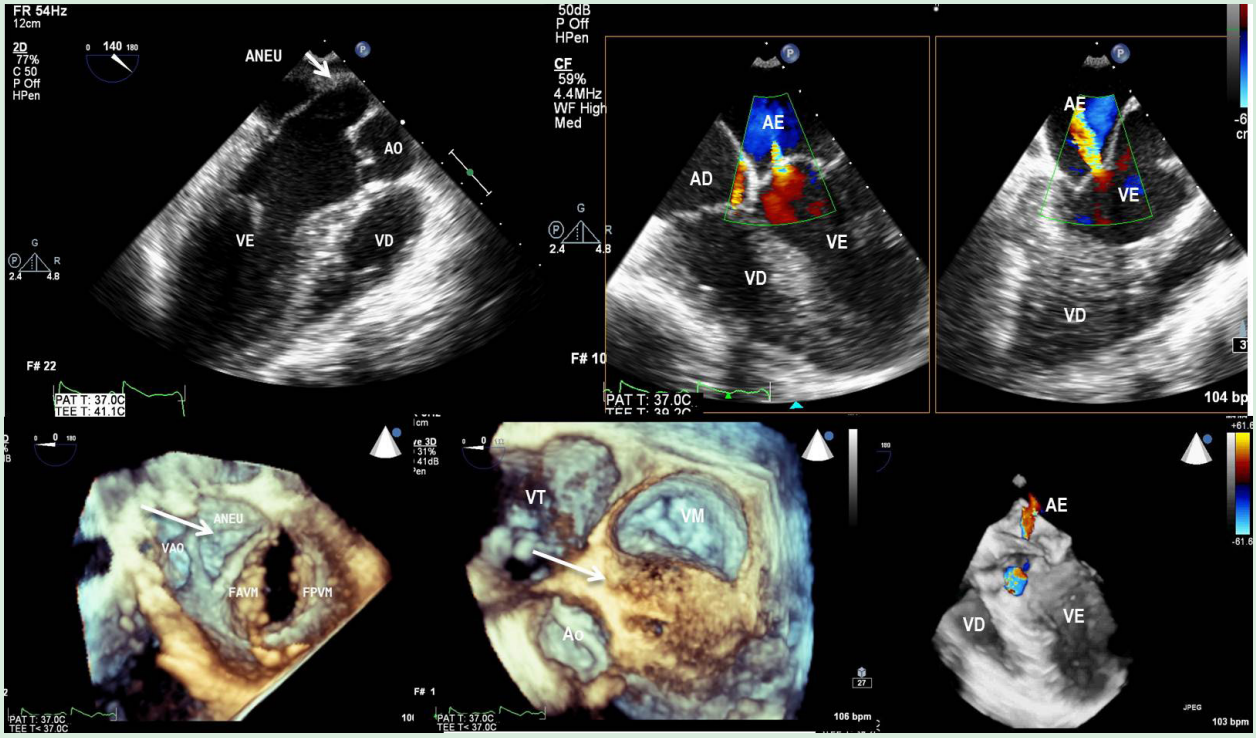

Figure 1: *Ao: aorta, VD: right ventricle, AD: right atrium, AE: left atrium, VT: tricuspid valve.*
Transesophageal echocardiography performed at 12 years-old. a) Two-dimensional study of the left ventricle long axis (LV), showing great dilatation of mitral-aortic continuity (arrow). b) In color flow mapping, there is mild mitral regurgitation due to a perforation in the anterior leaflet of the mitral valve (MV). c) Real-time three-dimensional echocardiography showing the base of the left heart seen from the left ventricular side. Note the anterior and posterior leaflets of the mitral valve (MVAL and MVPL) open, the aortic valve closed, and a large dilated area separating the two valvular rings, wherein the pseudoaneurysm appeared (ANEU). d) Atrial view of the heart base showing the pseudoaneurysm from the top. e) Color 3D demonstrating the perforation site in the base of the MVAL.

between the aortic and mitral valves), decreased angle between the interventricular septum and the aorta (which should be around 180 degrees under normal conditions) are responsible for forming the subaortic tunnel and change the direction and flow rate in this region [6]. This unfavorable geometry predisposes the development of fibrous tissue, forming the aortic subvalvular membrane or subaortic fibromuscular ring [7,8]. It is believed that there is also a genetic substrate contributing for abnormal proliferation of the endothelium in this region when subjected to flow changes [9].

Although the anatomical substrate is already present from birth, most subaortic stenoses manifest only after the neonatal period and the diagnosis is often made during the late follow-up of patients who had been treated for other injuries such as aortic coarctation and VSD. In the reported case, although these components have not been identified or described in the neonatal period, they were observed in subsequent examinations. This would explain the appearance of subaortic stenosis not reported in previous examinations and its reappearance after the first surgical treatment of subaortic stenosis, in which the aortic subvalvular membrane was resected, but the anatomical substrates predisposing for such condition were not addressed.

It is known that in this type of anatomical configuration, the definitive treatment of subaortic obstruction must be performed by applying the modified Konno procedure, wherein the subvalvular region is increased by resecting the left portion of the interventricular septum (that one protruding to the subaortic region) and placing a flap on the right to close the VSD. Sometimes it is necessary to expand the outflow tract of the right ventricle due to the positioning of the flap in VSD [10].

The inadvertent handling of the LVIF region causes serious injury. This fold is derived from the internal curvature of the primitive heart tube and the resection of this region causes the surgeon to get out of the heart [9]. Another complication described is the formation of a false aneurysm. Possibly, what

was reported by the surgeon as resection of the subaortic region combined with the inadvertent handling of the LVIF region and triggered the formation of the pseudoaneurysm observed in the last examination performed at twelve years--old. This means that this patient will need a fifth surgery to correct this potentially serious lesion due to risk for rupture.

CONCLUSION

This case illustrates the classic presentation of a complex aortic coarctation which has been formed during intrauterine life due to the presence of anatomical substrates that may trigger obstructive anomalies of the left heart and had late evolution complicated by recurrent aortic subvalvular stenosis.

Although undergoing various surgical procedures, this patient followed the natural history of the anomaly due to a not initial approach of the mechanisms predisposing to obstruction of the LV outflow tract. Belatedly, the evolution was complicated by pseudoaneurysm of the mitral-aortic intervalvular fibrosa, probably due to inadvertent handling in the LVIF region.

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