

Accessory mitral leaflet: an uncommon form of subaortic stenosis

Folheto mitral acessório: uma causa incomum de estenose subaórtica

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Abstract

Three-years-old boy presenting with a subvalvar aortic stenosis without a precise definition by echocardiography, where the surgical approach revealed an accessory mitral leaflet.

Descritores: Mitral valve/surgery. Aortic stenosis, subvalvular. Heart valve diseases.

Resumo

Criança de três anos, com estenose subvalvar aórtica sem definição precisa ao ecocardiograma, apresentando como diagnóstico cirúrgico um folheto mitral acessório.

Descritores: Valva mitral/cirurgia. Estenose aórtica subvalvar. Doenças das valvas cardíacas.

INTRODUCTION

Subvalvar aortic stenosis (SAS) corresponds to about 1% of congenital heart disease and usually presents in the form of membrane (narrow segment) or tunnel (longest segment) [1]. It is usually acquired and progressive. A rare cause of congenital SAS is the obstruction by accessory mitral valve tissue (AMT), approximately half of these corresponding to an accessory mitral leaflet (AML) [2]. There are few reports in the literature of patients with AMT, so that its incidence in children is not well established. It is known that about 70% of cases the diagnosis is performed in childhood (including neonatal) and often by signs or symptoms related to obstruction [3]. Performing echocardiograms in the same american institution for six years, Rovner A. et al. found an incidence of one case per 26,000 examinations [4]. They may present as isolated defect in 30% of cases, but usually is associated with other heart

diseases, the most common being VSD, ventricular aneurysm and complex congenital heart disease [2,5].

We treated in our institution a 3-year-old brown boy, weighing 13 kilograms. He was the third child of healthy mother, aged 25. The story did not include notifiable gestational maternal complications, denying use of medications during pregnancy, except for vitamins prescribed by his physicians. There was no family history of congenital heart disease. The child's medical history included epilepsy (tonic-clonic seizures), initially for one year, six months and controlled by the use of phenobarbital.

At eight months in a routine pediatric visit, a systolic murmur was heard, and the patient was referred to a cardiologist. Due to structural limitations of the public health system, only three years after he came to our clinic for correction. The patient never presented symptoms suggestive of heart failure or cyanosis.

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Abbreviations, acronyms & symbols

AML	accessory mitral leaflet
AMT	accessory mitral valve tissue
CPB	cardiopulmonary bypass
LVOT	left ventricular outflow tract
SAS	subvalvar aortic stenosis

In the evaluation, the child was asymptomatic, stained, and cyanosis. Atypical facies. Cardiac auscultation revealed an ejective systolic murmur 3 + / 6 + best heard in the medium right sternal border, without irradiation, with absence of other sounds and a normal second sound. No reports of change in characteristics of sound during the Valsalva maneuver. Blood pressure was 110 x 70 mmHg, with adequate perfusion. He presents eupneic and without visceromegalies. No significant changes in other systems. Chest radiography was normal and electrocardiogram showed sinus rhythm with no signs of ventricular hypertrophy. An echocardiogram during hospitalization showing situs solitus, atrioventricular and ventriculoarterial concordance, interatrial and interventricular intact septa. But, calling attention to the presence of a parachute-like structure that bulged to the left ventricular outflow tract (LVOT) during systole (Figure 1), generating a whirling flow, with significant subvalvar stenosis (peak gradient of 77

mmHg) with slight acceleration of blood passing through the aortic valve (2.3 m / s). The mitral valve was morphologically and functionally normal. The aortic valve presented mild thickening without valve insufficiency under color flow. Was asked if the mitral valve chordae could generate redundant obstruction, and he was referred for surgical correction.

The surgery was performed without complications, with cardiopulmonary bypass (CPB) as current protocol at the hospital. The approach was by median sternotomy. The LVOT was approached by an oblique aortotomy (J-shape), and surgical findings presented as a subaortic stenosis by accessory mitral leaflet fixed on the ventricular face of the anterior leaflet of the mitral valve and spanned to the anterior septal region which was fixed by means of thickened chordae and a well-formed accessory papillary muscle (Figure 2). Finally, an atriotomy was also performed for better assessment of mitral valve and its leaflets. As the AML does not contribute to the maintenance of the mitral valve function, it was dried collectively, together with the wire ropes and the papillary muscle (Figure 3). The CPB time was 60 minutes, with aortic clamping for 45 minutes. The postoperative course was favorable, leaving room extubated without complications in the postoperative period and remained in intensive care unit for 48 hours and was discharged on the fifth day. The echocardiogram performed on the fourth postoperative day showed no residual stenosis or valvular LVOT.

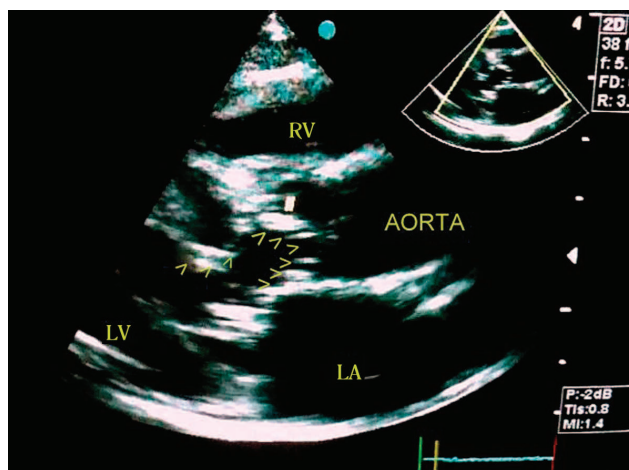


Fig. 1 - Two-dimensional echocardiography in parasternal longitudinal cutting, showing the systolic bulging of the accessory mitral leaflet (arrows), obstructing the left ventricular outflow tract. LA = left atrium, LV = left ventricle, RV = right ventricle

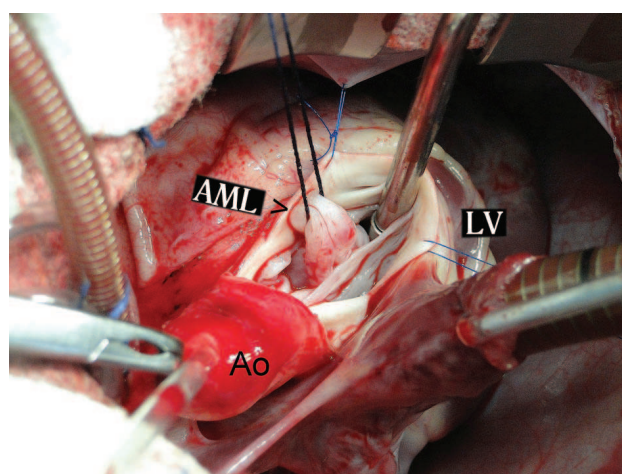


Fig. 2 - Oblique aortotomy showing the accessory mitral leaflet (AML), being suspended by wires. Ao = aorta, LV = left ventricle

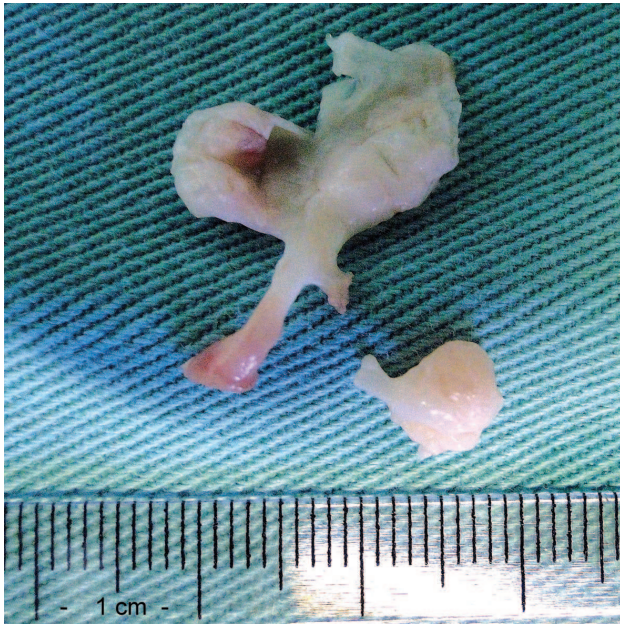


Fig. 3 - Resected accessory mitral leaflet demonstrating the accessory papillary muscle (separately) and thickened rudimentary chordae

DISCUSSION

The first report of AMT was performed in 1842 and the first description of surgical correction performed in 1963 [4]. The embryological origin of the defect comes from the incomplete separation of the mitral valve of the endocardial cushions [6-7]. AMT can be found on the ventricular face of the anterior leaflet of the mitral valve, the chordae or papillary muscle attachments.

A classification of types of AMT was suggested by E. Prifti et al, based on review of 90 cases described, depending on the deployment and the morphology of the defect. Type I (fixed), presents itself in two forms: AI (nodular) and IB (membranous). Type II (Mobile), is divided into two subtypes: IIA (pedunculated) and IIB (leaflet-like). The latter corresponds to 46% of cases and is called the accessory mitral leaflet and may be further subdivided into leaflets with rudimentary chordae and chordae with well development [2].

In the case reported by us, the patient is classified as type II, subtype B, with rudimentary chordae.

An AMF may or may not produce LVOT obstruction. In cases where there is obstruction or it is mild, patients usually present asymptomatic and diagnosed accidentally while performing an echocardiogram. When the

obstruction is more important, it can be heard the typical murmur of subvalvular aortic stenosis of ejection and audible character in the second or third right intercostal space, and may radiate to the neck vessels. Besides the obstructive effect of the mass, the turbulence generated at the site of stenosis leads to gradual fibrosis, increase of the obstruction and secondary valve insufficiencies. Symptomatic cases usually manifest themselves in the first decade of life and symptoms are related to obstruction presenting as dyspnea, syncope, or angina on effort [2]. Stroke is also a symptom related in some cases. Symptoms tend to appear when the obstruction is more important, identified by echocardiography in a high pressure gradient in the LVOT (peak gradient > 50 mmHg) [4].

The echocardiogram is considered the method of choice for the diagnosis of obstruction by AMT, allowing the quantification of stenosis and its hemodynamic effect, identifying the site of obstruction and also the classification of AMT in their different types [4,8]. The transthoracic technique is usually sufficient for this setting, but the TEE can facilitate analysis of the braids. In the specific case of accessory mitral leaflet, it can get a typical image on which parachute bulges LVOT during systole [9]. Catheterization adds very little to the findings, being suitable only for investigation of associated defects [2].

The management depends on the degree of obstruction and the clinical patient. In asymptomatic patients and usually without obstruction or mild obstruction, clinical and imaging approach can only be performed.

The surgery is oriented in symptomatic cases, also in those with moderate to severe cases of obstruction and those associated with other heart diseases requiring approach. Aortic failure is another indication regardless of the degree of obstruction [3].

The identification of a AMT during surgery can be difficult using only the aortotomy, and the left atriotomy can be a great help [10]. In a review covering 68 surgical patients found a mortality rate of 9%. Such mortality was associated with poor surgical outcome: the presence of other complex congenital heart disease, fail to complete removal of the tissue and surgeries performed in the neonatal period. The most common complications were mild residual stenosis in 13% of cases, and the reoperation for severe residual stenosis was also necessary in 13% of cases. Mitral regurgitation (10%) or aortic (7%) [2] were the less common complications.

This case alerts us to the importance of prior knowledge of this pathology and clinical-surgical implications for the echocardiographic diagnosis can be performed more objectively, facilitating the classification of the type found in MRI, to a prior suitable handling and consistent surgical indication.

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