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Accessory Mitral Valve With Cordal Attachments to Mitral and Aortic Valves

An Unusual Cause of Left Ventricular Outflow Tract Obstruction and Both Mitral and Aortic Insufficiencies

António Gaspar, MD; Jorge Almeida, MD; Benjamim Marinho, MD; Vítor Monteiro, MD; Armando Abreu, MD; Paulo Pinho, MD

We introduce the case of a 72-year-old woman referred with exertional dyspnea and chest pain. On clinical examination, a grade III/VI harsh systolic murmur radiating to the neck was audible. Transthoracic echocardiography showed a structure attached to the proximal left ventricular outflow tract (LVOT) causing significant obstruction (maximum and median gradients of 55 and 33 mm Hg, respectively), which led to the initial diagnosis of subaortic membrane (Figure 1A and 1B; Movie I in the online-only Data Supplement). Transesophageal echocardiography allowed the visualization of a mobile structure in the proximal left ventricular outflow tract, with cordal attachments to the subvalvular mitral apparatus and apparently to an aortic cuspid, conditioning severe mitral and moderate aortic insufficiencies (Figure 1C and 1D; Movie II in the online-only Data Supplement). These findings were consistent with the diagnosis of accessory mitral valve tissue (AMVT). Coronary angiography revealed normal coronary arteries. Intraoperative three-dimensional echocardiography confirmed the presence of AMVT in the proximal left ventricular outflow tract and better delineated the cordal attachment to the aortic valve (Figure 1E and 1F; Movies III and IV in the online-only Data Supplement).

The patient underwent cardiopulmonary bypass. The ascending aorta was opened and the aortic valve inspected. The aortic insufficiency was due to cordal tissue coming from the AMVT and attached to the left coronary leaflet (Figure 2A). All the leaflets were thickened with a myxoid appearance. Left atriotomy was performed to directly inspect the mitral valve. The accessory tissue was attached to the anterior leaflet of the mitral valve, and through cords, to both the ventricle and aortic valve (Figure 2B). The tissue was difficult to individualize from the original valve, and it was not possible to enucleate all the tissue in security to realize aortic and mitral valve reconstruction. Both the valves and the excess

tissue were resected, and 2 bioprosthetic valves were implanted.

AMVT is a rare congenital cardiac anomaly, in particular in adults, that was first described by Mclean et al.¹ AMVT presents mostly with left ventricular outflow tract obstruction, usually being diagnosed in the first or second decade of life, with exercise intolerance, dyspnea, chest pain, and syncope as the main symptoms.^{1,2} Echocardiography, both transthoracic and transesophageal, has been widely recognized as the most valuable imaging technique for identification and characterization of this anomaly.³ It has been estimated to be present in 1 per 26 000 echocardiograms.⁴ Although little is known about the embryological mechanism of AMVT, it is thought to result from the abnormal development of endocardial cushion tissue.⁵ Direct and simultaneous involvement of both mitral and aortic valves, as in the present case, has been very rarely described.⁶

Disclosures

None.

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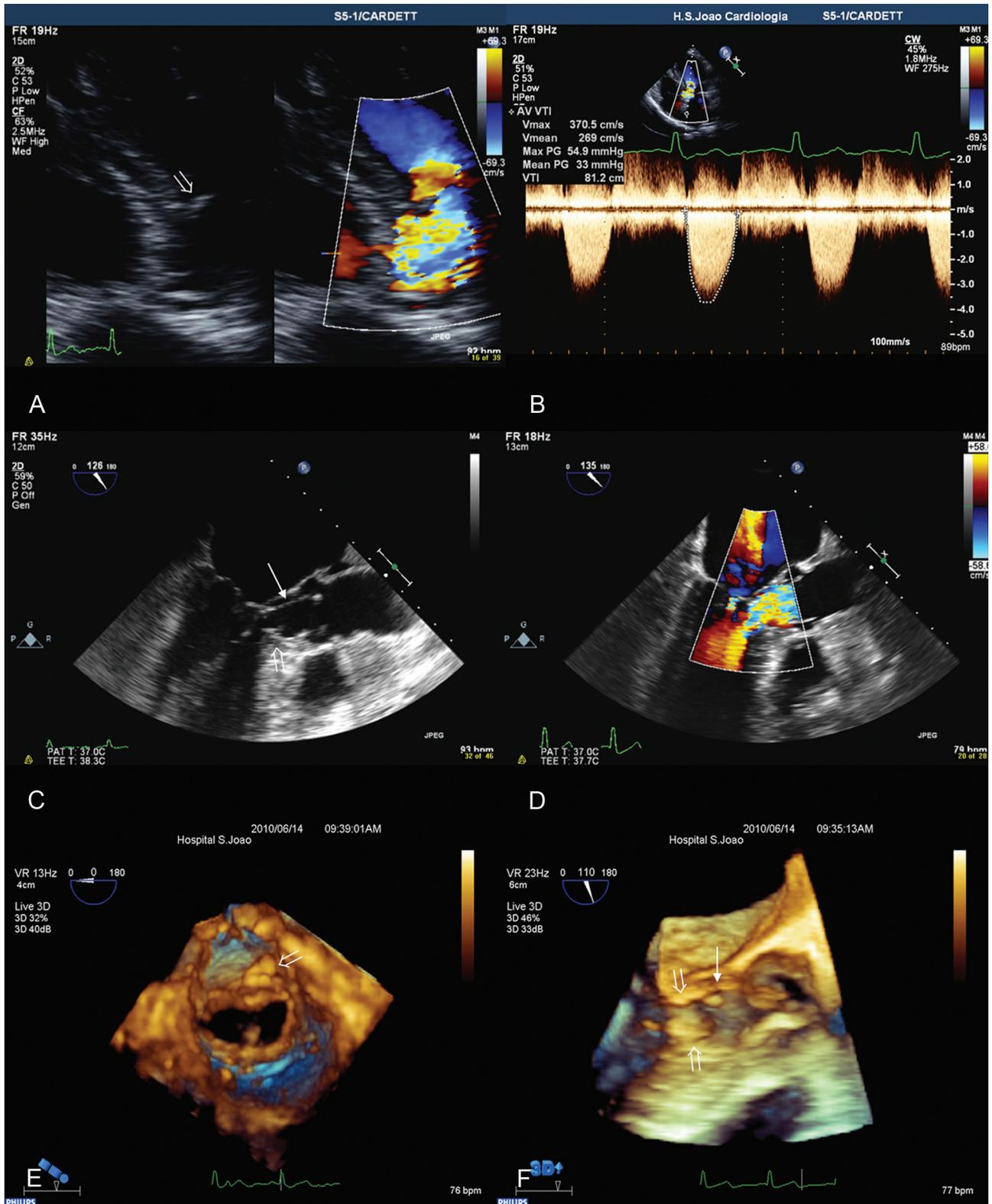


Figure 1. **A**, Preoperative transthoracic echocardiography (apical 5-chamber view) showing an obstruction of the proximal LVOT (double arrow) associated with flow acceleration. **B**, Continuous Doppler measurement at the proximal LVOT showing instantaneous and mean gradients of 55 and 33 mm Hg, respectively. **C**, Preoperative transesophageal echocardiography (midesophageal long-axis view) showing AMVT (double arrow) in the proximal LVOT, with cordal attachments to the subvalvular mitral apparatus and apparently to an aortic cuspid (single arrow). **D**, Preoperative transesophageal echocardiography (midesophageal long-axis view) with color Doppler showing flow acceleration at the proximal LVOT. **E**, Three-dimensional echocardiography showing AMVT (double arrow) in the proximal LVOT with the mitral valve just below (viewed from the left ventricle side). **F**, Three-dimensional echocardiography showing AMVT (double arrow) in the proximal LVOT (cross-sectioned in its long axis) with a cordal attachment to an aortic cuspid (single arrow). LVOT indicates left ventricular outflow tract; AMVT, accessory mitral valve tissue.

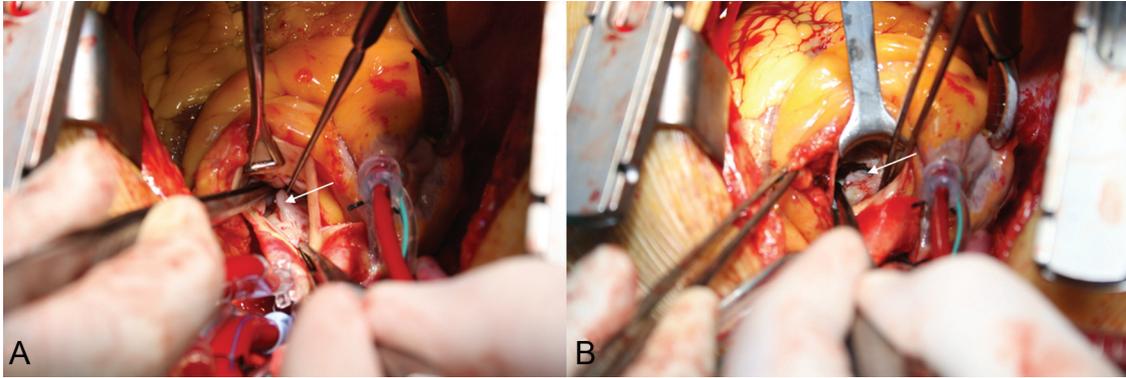


Figure 2. **A**, After opening the ascending aorta, the inspection of the aortic valve showed cordal tissue (single arrow) coming from the AMVT and attached to the left coronary leaflet. **B**, Inspection through left atriotomy showed AMVT attached to the anterior leaflet of the mitral valve (single arrow). AMVT indicates accessory mitral valve tissue.