

Accessory Mitral Valve Tissue: An Unusual Echocardiographic Finding



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INTRODUCTION

Accessory mitral valve tissue (AMVT) is a rare congenital cardiac malformation, which is diagnosed most frequently during the first decade of life and rarely detected in adulthood.¹ This condition is reported in the literature commonly associated with other congenital cardiac anomalies, such as ventricular septal defects, patent ductus arteriosus, and transposition of the great arteries.¹ AMVT may be detected as an incidental finding or, in some cases, may cause left ventricular outflow tract (LVOT) obstruction.

We report the case of a 52-year-old man referred for dyspnea, without any history of cardiac malformations.

CASE PRESENTATION

A 52-year-old man, obese, with no significant comorbidities, sought medical assistance because of exertion dyspnea. Physical examination revealed irregular cardiac rhythm and mitral systolic murmur 2+/6+. Resting electrocardiography showed ventricular ectopic beats.

Transthoracic echocardiography identified mild left atrial volume enlargement (37 mL/m²), but the other cavity diameters were normal. Echocardiography also revealed preserved biventricular systolic function with normal segmental contractility. The mitral valve appeared as a serpiginous structure (leaflet-like), with great mobility and only mild to moderate mitral regurgitation (Figures 1 and 2, Videos 1–4). There was mild aortic regurgitation, without LVOT obstruction (Video 5).

Transthoracic three-dimensional (3D) echocardiography was performed as well to evaluate mitral valve morphology (Figure 3, Video 6). There was no evidence of any other cardiac malformation, and the morphology was suggestive of AMVT. Two-dimensional and 3D echocardiographic images were excellent, so it was not necessary to perform transesophageal echocardiography. In addition, 3D echocardiography provided anatomic information about other cardiac structures through full-volume acquisition. In this case, AMVT did not cause obstruction, so the physicians decided to follow the patient over time.

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Keywords: Accessory mitral valve tissue, Echocardiography, Mitral valve, Congenital heart disease

Conflicts of interest: The authors reported no actual or potential conflicts of interest relative to this document.

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2468-6441

<https://doi.org/10.1016/j.case.2017.09.004>

DISCUSSION

AMVT is a rare anomaly in adults and may be associated with other congenital intracardiac and vascular malformations, such as atrial septal defects, septal aneurysms, coronary artery anomalies, persistent left superior vena cava, aortic coarctation, bicuspid aortic valve, mitral valve leaflet cleft, and dextrocardia.¹⁻³ The estimated incidence of AMVT in adults is 1:26,000 echocardiograms.⁴ More frequently identified in men, this entity has a male-to-female ratio of 1.75:1.¹ The embryologic mechanism of AMVT formation is not clear and may be related to abnormal or incomplete separation of the mitral valve from the endocardial cushions.^{3,5}

AMVT may affect one or both atrioventricular valves simultaneously; however, the mitral valve is more often involved.⁶ Published data show a high prevalence of the mobile type of AMVT, which projects itself into the LVOT, in most cases originating from the anterior mitral valve leaflet, with a higher incidence of parachute-like or balloon-like structures.^{3,6,7} Morphologically, AMVT may be classified as type I (fixed) or type II (mobile). Type I presents in two forms: IA (nodular) and IB (membranous). Type II is divided into two subtypes: IIA (pedunculated) and IIB (leaflet-like). The latter corresponds to 46% of cases and may be further subdivided into leaflets with rudimentary chordae and chordae with well-developed tissue.³

Patients with AMVT may be asymptomatic, and diagnosis may be an incidental finding during routine echocardiography. However, patients may become symptomatic, usually when the mean gradient across the LVOT reaches 50 mm Hg.^{4,7,8} In these cases, physical examination often shows aortic systolic murmur, radiating toward the neck and causing symptoms such as dyspnea, chest pain, palpitations, and syncope. Other complications described are endocarditis and cardioembolic events due to the excessive mobility of AMVT.^{4,9,10}

Both transthoracic and transesophageal echocardiography may help in the diagnosis, revealing possible associated lesions and complications. Both 3D transthoracic echocardiography and 3D transesophageal echocardiography enable more precise anatomic characterization, allowing visualization of the accessory tissue attachment to the interventricular septum or the myocardium, which can help in its classification.^{3,11-13} Three-dimensional echocardiography enables en face visualization of the mitral valve leaflets from both left ventricular and left atrial perspectives.

Regarding treatment of this anomaly, cardiac surgery is indicated only in patients with significant LVOT gradients and those undergoing correction of other congenital cardiac defects.^{8,14} Surgery includes accessory tissue removal and, sometimes, artificial chordae implantation and annuloplasty, depending on the severity of the disease.¹⁵ For patients without significant LVOT obstruction, follow-up with serial echocardiography to assess progression of the gradient is indicated.⁵ Possible explanations for how the gradient may appear after

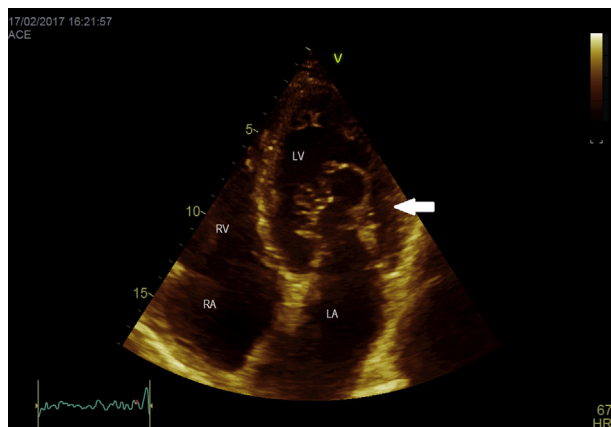


Figure 1 Transthoracic apical four-chamber view showing accessory mitral valve tissue (*white arrow*) and mild left atrial enlargement. LA, Left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

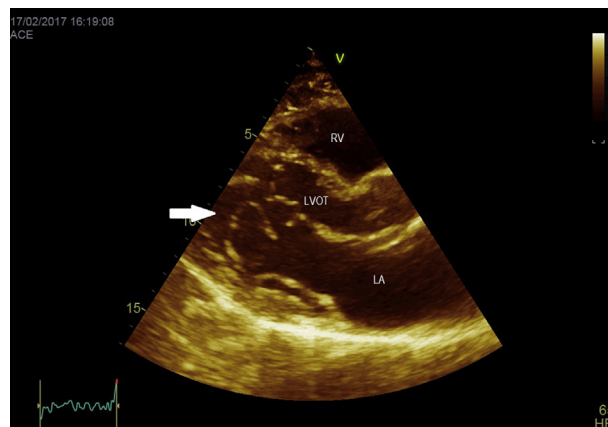


Figure 2 Transthoracic parasternal long-axis view. Visualization of accessory mitral valve tissue (*white arrow*) without LVOT obstruction. LA, Left atrium; RV, right ventricle.

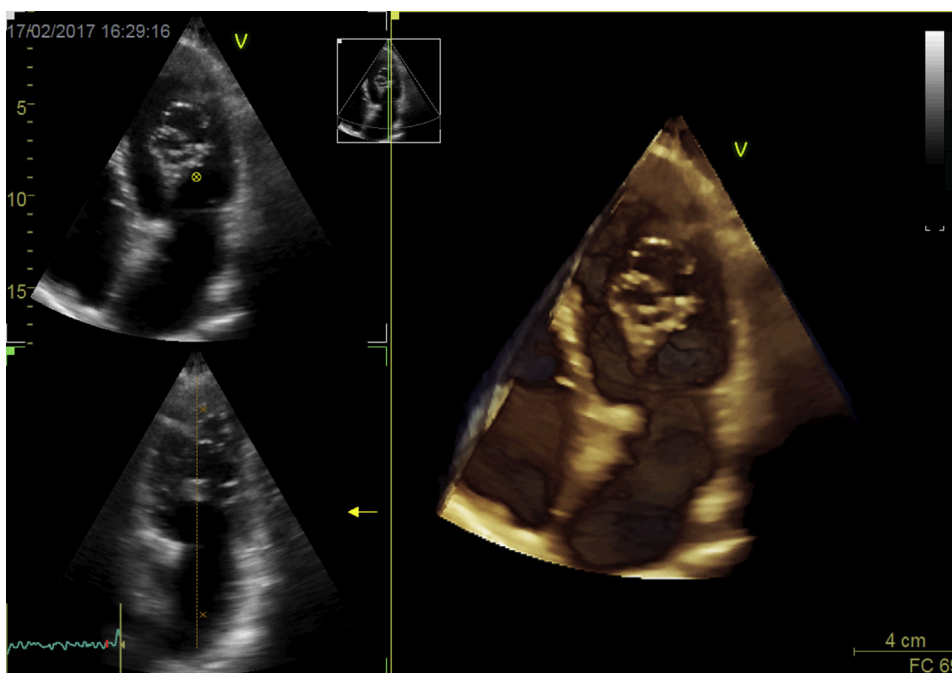


Figure 3 Three-dimensional transthoracic echocardiographic imaging displaying accessory mitral valve tissue in apical four-chamber view.

childhood would be elongation of chordae tendineae over time or its abrupt stretch, causing obstruction.

CONCLUSION

AMVT is a congenital cardiac malformation still poorly known, and it is recognized as a differential diagnosis of LVOT obstruction. Both transthoracic and transesophageal echocardiography and, more recently, 3D echocardiography, play a fundamental role in diagnosis, management, and follow-up of patients with this anomaly, enabling accurate anatomic characterization and revealing possible associated lesions and complications.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2017.09.004>.

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