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Accessory Mitral Valve Tissue with Mitral and Aortic Regurgitation in an Adult

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Abstract

Accessory mitral valve tissue (AMVT) as a rare cause of congenital subaortic stenosis is commonly associated with other congenital malformations. Echocardiography is the preferred imaging modality for the diagnosis and evaluation of AMVT. Unless in patients with a significant LVOT obstruction and in those undergoing cardiac surgery for other reason surgery is not indicated. We report here an asymptomatic 19 year old male referred to our hospital for the evaluation of cardiac murmur. Transthoracic and then transesophageal echocardiography revealed a cystic mobile mass in LVOT which was an AMVT with mitral and aortic regurgitation and mild LVOT obstruction.

Keywords

accessory mitral valve tissue**;** congenital anomaly**;** echocardiography

Abbreviations

AMVT: Accessory Mitral Valve Tissue; AR: Aortic Regurgitation; CW: Continuous wave; LA: Left Atrium; LV: Left Ventricle; LVOT: Left Ventricular Outflow Tract; MV: Mitral Valve; MPG: Mean Pressure Gradient; PPG: Peak Pressure Gradient; 2D: Two-dimensional; 3D: Three-dimensional; TTE: Transthoracic Echocardiography; TEE: Transesophageal Echocardiography

Introduction

AMVT is a rare congenital anomaly [1-3]. Embryologically, it develops as a result of an incomplete separation of the mitral valve from the endocardial cushion tissue [1]. The first case of this peculiar lesion and first surgical treatment were described in the literature in 1842 and 1963, respectively [4,5]. Two-dimensional (2D) echocardiography, both transthoracic (TTE) and transesophageal (TEE), and the recent introduction of three-dimensional (3D) echocardiography play an important role in the diagnosis, management, and follow-up of patients with this abnormality [6-11].

Case Presentation

A 19 year old male with pilonidal l cyst planned for surgery referred to our hospital for the evaluation of heart murmur. The patient had no history of palpitation, chest pain, dyspnea or syncope. Physical examination revealed a grade 3/6, mid-systolic murmur best heard at the left sternal border, not radiating to the neck. The 1st, 2nd heart sounds and electrocardiogram were normal. Two-dimensional TTE performed with a Vivid S6 ultrasound scanner that showed a small mobile cyst-like structure attached to the anterior mitral valve leaflet oscillating in LVOT suggestive of AMVT. (Figure-1). Color Doppler study revealed moderate to severe mitral regurgitation (MR) and moderate aortic regurgitation.

Continuous wave Doppler (CW) study of LVOT revealed mild obstruction (PPG/MPG= 43/23 mm Hg). No obstruction in mitral inflow or other associated congenital abnormalities were observed. Transesophageal echocardiography confirmed the diagnosis (Figure-2).

Discussion

AMVT is rare and may be an unusual cause of subvalvular LVOT obstruction [12]. This entity is usually seen in symptomatic children with other congenital abnormalities of the heart and great vessels [13-15]. Herein, we describe the rare presentation of AMVT in an asymptomatic adult with mitral and aortic regurgitation and with mild degree of LVOT obstruction.

AMVT may affect one or both of the atrioventricular valves simultaneously; however, the MV is most often involved [2]. It is commonly associated with other congenital intracardiac and vascular malformations, [12,16] with a higher incidence of ventricular septal defects, subaortic membrane, and transposition of great arteries [21].

From a pathophysiological point of view, AMVT is recognized as a cause of LVOT obstruction [6,7,16-19]. Two mechanisms underlying obstruction in patients with AMVT were recognized: (i) mass effect of the accessory tissue and (ii) progressive deposition of fibrous tissue due to the turbulent flow created by AMVT [3,20]. In the first case, obstruction can be immediately significant while, in the other case, outflow gradient increases progressively, probably due to AMVT enlargement and LVOT narrowing [12]. However, cases with no obstruction also were reported [21].

AMVT has been variably described as sac-like, balloon-like, parachute-like, sail-shaped, and leaflet-like, or as a sheet, membrane, or pedunculated mass. In addition, AMVT has been classified on the basis of intraoperative descriptions and anatomic presentation. Type I AMVT, defined as a fixed mass, can have a nodular (type IA) or membranous (type IB) presentation. Type II AMVT occurs as a mobile mass and is classified as pedunculated (type IIA) or leaflet-type (type IIB). Type IIB AMVTs are further divided into those with rudimentary chordae tendineae (type IIB1) or well-developed chordae tendineae (typeIIB2) [12,22]. Type IIB has been the most frequent presentation reported (33/58 patients, 56.8%) [12]. Preoperative findings have shown 6 different locations of the insertion of the chordae tendineae of the AMVT: the left ventricular wall, interventricular septum, accessory papillary muscle, anterolateral papillary muscle, anterior mitral valve leaflet, and the anterior mitral valve chordae. The most common location has been the anterolateral papillary muscle (14/32, 44%) [12, 15].

Echocardiography has been considered to be the optimal imaging technique for the diagnosis of AMVT since its use for this purpose was first reported in 1985 by Alboliras and colleagues [18,23-25].

2D echocardiography can be considered the gold standard modality for the evaluation of AMVT [21]. It plays a fundamental role as it enables anatomical characterization of AMVT and, also, shows possible associated lesions and complications such as LV hypertrophy, dilatation, and systolic dysfunction [25]. Furthermore, Doppler evaluation of the LVOT allows a non-invasive assessment of the degree of obstruction [26]. With TTE, AMVT may look like a mobile parachute-like leaflet floating in the LVOT and usually undergoes a progressive thickening or a fixed structure attached to the interventricular septum by a short chordal apparatus [2, 20]. Large redundant AMVT may appear as a globular or even cystic mass. The AMVT typically prolapses into the LVOT or aortic valve during systole and retracts during

diastole owing to its attachment to various mitral valve and subvalvular apparatus [21]. In summary, echocardiography is the preferred tool for the morphological and functional study of AMVT, with no need for other imaging modalities in the majority of cases [7].

The current approach is to submit for intervention only patients with a significant LVOT gradient (mean gradient of ≥25 mmHg) and those undergoing correction of other congenital malformations or exploration of an intracardiac mass [6,20,27,28]. For patients without significant LVOT obstruction, a follow-up with serial echocardiography to assess progression of the gradient is indicated [6].

In our asymptomatic patient there was neither significant LVOT obstruction, nor other congenital malformation requiring surgery; therefore he was planned to be followed up by repeated echocardiographic examinations for progressive LVOT obstruction and valvular regurgitation.

Figures

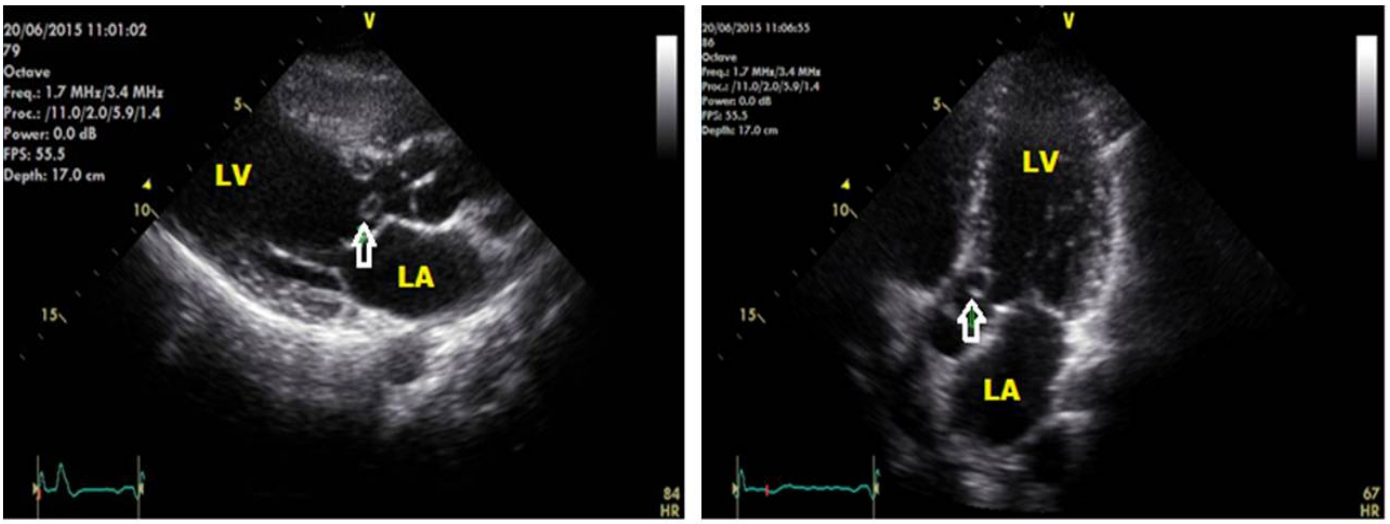

Figure 1: Two-dimensional transthoracic echocardiogram (parasternal long-axis view left and apical 5 chamber view right) shows spherical cyst-like mass suggestive of AMVT (white arrow) in the left ventricular outflow tract.

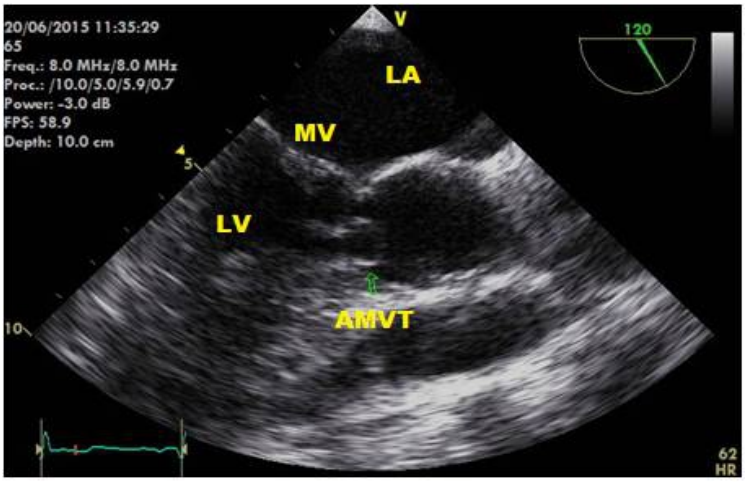

Figure 2: Two-dimensional transesophageal echocardiogram at 120°shows AMVT (green arrow) in the left ventricular outflow tract.

Conclusion

AMVT is rare and may cause LVOT obstruction or valvular regurgitation. Most reported cases are children and echocardiography is the diagnostic modality of choice. Unless significant LVOT obstruction or other congenital anomaly requiring surgery intervention is not required. Although similar cases

previously had been reported but AMVT as a cause of LVOT obstruction and both mitral and aortic insufficiencies is extremely rare that emphasizes the importance of acquaintance of this rare congenital anomaly for the proper diagnosis and management.

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