A rare case of isolated accessory mitral tissue in an asymptomatic adult female

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Abstract
Accessory mitral valve tissue (AMVT) is a rare congenital cardiac anomaly, which is associated with other congenital heart diseases. It is diagnosed in neonates or childhood and rarely in adulthood. Nevertheless, AMVT is an incidental finding or described as isolated. Echocardiography, especially three-dimensional echocardiography is considered as an optimal imaging technique for AMVT diagnosis. We herein presented an asymptomatic adult AMVT case with significant left ventricular outflow tract obstruction and surgical excision was recommended.

Keywords: Mitral valve disease • Accessory mitral valve tissue • Congenital heart disease • Left ventricular outflow tract obstruction

INTRODUCTION

Accessory mitral valve tissue (AMVT) was first reported as early as 1842 by Chevers et al., as a congenital cardiac lesion. Although often associated with other cardiac anomalies, it might be seen as isolated and rarely detected in adulthood [1]. Symptomatology of the patient commonly manifests asymptomatic heart murmur or symptoms of left ventricular outflow tract (LVOT) obstruction, such as chest pain, syncope or palpitations.

CASE REPORT

A 46-year-old woman was transferred from the Cardiovascular Surgery Department to our clinic for the reassessment of patient before surgery. She denied any history of chest pain, dyspnoea or syncope. The physical examination was unremarkable except for the presence of 3/6 systolic ejection murmur with faint radiation to the neck. Transthoracic echocardiography showed normal-sized cardiac chambers and the left ventricular ejection fraction was 79%. Concurrently, ascending aorta dilatation was found with diameter up to 37 mm with mild to moderate aortic regurgitation. An abnormal membranous structure, mobile, attached to the ventricular side of the anterior mitral leaflet was seen and caused LVOT obstruction during systole. The maximal pressure gradient measured was 63 mmHg and velocity was 4.0 m/s (Fig. 1A). Mitral valve insufficiency was not found. No other congenital heart anomalies were presented. A diagnosis of AMVT with severe obstruction of the left ventricular outflow tract based on echocardiographic characteristics was made.

The patient underwent surgery under standard cardiopulmonary bypass. Intraoperative transoesophageal echocardiography revealed a movable membranous structure that prolapses into the LVOT during diastole with sac shape and occupied the LVOT in systole as an expanded parachute structure (Fig. 1B). Aortotomy in combination with transeptal approach through right atriotomy was undertaken. Trans-aortic excision of a mobile mass that attached to the anterior mitral leaflet without well-developed chordae tendineae was applied. After the excision, the saline test showed mild mitral valve regurgitation, which prompted the mitral valve repaired with a 30-mm Physio ring (Fig. 2A). A saline test demonstrated good coaptation with no leakage. Histological examination showed myxomatous degeneration analogous to dysplastic valvular tissue (Fig. 2B). No residual tissue and mitral regurgitation were demonstrated in the postoperative transthoracic echocardiography with only mild aortic regurgitation. The patient was discharged on the 12th day after the operation and had no symptoms after 6 months of follow-up, further follow-up at 1 year showed no progression of aortic pathology.

DISCUSSION

AMVT is a rare congenital malformation, which may be caused by abnormal or incomplete separation of the mitral valve from the endocardial cushions. It may be isolated or combined with other congenital heart anomalies, such as ventricular septal defect.
and transposition of the great arteries. Symptoms of patients with AMVT depend on the degree of LVOT obstruction, knock-on effects on the aortic and mitral valves and on concomitant cardiovascular malformations. The usual age range for diagnosis of AMVT was from newborn to 77 years (average 8.6 years), while the incidence of AMVT in adults was 1/26 000 based on echocardiography [2]. As to AMVT morphology, saclike, balloon-like, parachute-like, sail, leaflet-like, sheet, membrane or pedunculated mass were demonstrated. Prifti et al. [3] provided a classification of this anomaly based on intraoperative description and anatomic presentation: Type I (fixed) and Type II (mobile). Mobile AMVT includes a pedunculated form (Type IIA) and a leaflet-like form (Type IIIB). The leaflet type was further subdivided into the rudimentary chordae (free-edge chordae) (Type IIB1) and the well-developed chordae (Type IIB2). The patients in our case had a mobile leaflet-like structure and no well-developed chordae tendineae. Therefore, our case was classified as Type IIB1. Echocardiography can clarify the morphology and attachment points of the AMVT, particularly in patients scheduled for surgery [4]. Typically, we need to differentiate AMVT from redundant mitral valve chordae and other structures in LVOT. For example, redundant mitral valve chordae may be involved in chordal systolic anterior motion with dynamic LVOT.

Figure 1: (A) Preoperative transthoracic echocardiography, continuous Doppler measurement traced the instantaneous gradient of 64 mmHg at the proximal left ventricular outflow tract. (B) Intraoperative transoesophageal echocardiography, 162° view, demonstrated the movement of accessory mitral valve tissue being folded into left ventricular outflow tract. Aliasing was clearly seen in left ventricular outflow tract with colour Doppler flow frame. (C) Three-dimensional rendering of the accessory mitral valve tissue viewed from the base of the heart to the apex in the long-axis view.

Figure 2: (A) Intraoperative photograph showing accessory mitral valve tissue attached to the anterior mitral annulus and part of its chordae attached to the membranous part of interventricular septum. (B) Histological examination (haematoxylin-eosin stain) showed myxomatous degeneration. The resected tissue was almost identical with the normal valve tissue.

Video 1: 3D rendering of the AMVT viewed from the base of the heart to the apex in the longaxis view.

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obstruction and similar to AMVT in case of chordal rupture. Identifying AMVT in an arrested heart can be challenging. The patient we reported was asymptomatic and eventually underwent surgery to prevent thrombosis and aggravation of the LVOT obstruction. The presence of AMVT places the patients at risk of embolic stroke, which in itself can be considered an indication for surgical intervention [5]. For patients without severe LVOT obstruction, a serial echocardiographic follow-up was recommended to assess the progression of the gradient.

CONCLUSIONS

AMVT is a rare congenital cardiac abnormality with various clinical manifestations, which results in one of the rare causes of LVOT obstruction, mitral valve or aortic valve regurgitation. It is very rare and not easily found because of the lack of the relevant symptoms and associated cardiac anomalies. We present this clinical case in which AMVT was incidentally diagnosed during standard echocardiography and highlighting the usefulness of echocardiography in diagnosis, indication for surgery and follow-up.

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