Case Report

Isolated non-obstructive accessory mitral valve tissue in an adult mimicking ruptured chordae

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A B S T R A C T

Accessory mitral valve tissue is commonly associated with other congenital heart diseases and is usually detected in children causing left ventricular outflow tract obstruction. We present an adult patient with isolated non-obstructive accessory mitral valve tissue that was mimicking ruptured chordae of the mitral valve. Accessory mitral valve tissue in adults is very rare and can mimic various causes of left ventricular outflow tract obstruction. This patient represents the first case in literature wherein an unobstructive accessory mitral valve tissue simulated a ruptured chordae. This case illustrates that in patients with suspected mitral valve chordae rupture without any mitral regurgitation, this diagnosis should be considered, which can have therapeutic implications.

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1. Introduction

Accessory mitral valve tissue (AMVT) is a rare congenital cardiac anomaly, commonly associated with other congenital heart diseases, such as ventricular septal defects and transposition of the great arteries.1–3 It usually presents in childhood causing left ventricular outflow tract (LVOT) obstruction; presenting as exercise intolerance, chest pain, effort syncope or recurrent transient ischemic attack/stroke.1,2 It’s incidence in adults is one per 26,000 echocardiograms.2 We present an adult patient with isolated non-obstructive accessory mitral valve tissue mimicking ruptured chordae of the mitral valve.

2. Case report

A 35-year-old man was referred to our hospital for transesophageal echocardiography (TEE). He presented to a regional hospital with atypical chest pain. Physical examination revealed normal heart sounds with a faint, grade 2/6, mid-systolic murmur heard at the left sternal border. His ECG was normal. Transthoracic echocardiography (TTE) performed in the regional hospital reported ruptured chordae tendineae of the anterior mitral valve leaflet with trivial mitral regurgitation. TTE done in our institute demonstrated normal size cardiac chambers with normal thickness of mitral valve leaflets. There was a highly mobile sail-like structure attached to the ventricular part of the anterior mitral valve leaflet prolapsing in and out of LVOT, but without causing LVOT obstruction (Fig.1 A and B, arrowheads). The structure suggested AMVT. Chordae to anterior mitral leaflet were intact. There was no LVOT turbulence or aortic regurgitation and the peak pressure gradient across the LVOT was 7 mmHg at rest and on exercise (Fig.1 C). The left ventricular ejection fraction was 0.65. TEE revealed a mobile, echogenic, membrane-like structure attached to the ventricular side of the proximal...
part of the anterior leaflet that moved into the LVOT during systole and occupied the sub aortic area like a sail as well as sac-like structure suggesting AMVT (Fig. 2A and B, arrowheads, Video 1). It was attached to left ventricular wall apical area with a long well formed chordae. In view of no LVOT obstruction, patient was advised echocardiographic and clinical follow up. He was advised prophylactic aspirin therapy to prevent thromboembolism events.

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3. Discussion

Majority of patients with AMVT are diagnosed during the 1st decade of life with varying degree of LVOT obstruction with a median gradient of 50 mmHg. In an analysis of 90 patients with AMVT, 3.3% of patients presented without LVOT obstruction, as seen in this patient. AMVT has been variably described as sail-shaped, sac-like, balloon-like, parachute-like, and leaflet-like, or as a sheet, membrane, or pedunculated mass. Prifti et al have classified AMVT as Type I-Fixed type (A – nodular, B – Membranous), Type II-Mobile type (A – Pedunculated, B – leaflet-like). Type IIB is divided into 1) rudimentary chordae and 2) developed chordae. This patient had type II-mobile type of AMVT with well developed chordae attached to left ventricular wall. AMVT inserts into six locations in the left ventricle through its chordae tendinae: the left ventricular wall, interventricular septum, accessory papillary muscle, anterolateral papillary muscle, anterior mitral valve leaflet, and the anterior mitral valve chordae. AMVT causes LVOT obstruction by systolic ballooning into the LVOT resulting in a mass effect and the continuing turbulence produced this effect results in a fibrous tissue reaction with permanent deposits of fibrous tissue and progressive LVOT obstruction. Embryologically, AMVT results from an incomplete separation of the mitral valve from the endocardial cushion during cardiac development.

A comprehensive TTE and TEE when indicated can diagnose most of the cases of AMVT. Other LVOT masses like myxoma, papillary fibroelastoma, thrombus or vegetations can produce similar echocardiographic appearances, but a detailed TTE and TEE can define the morphology and attachments of these lesions and thus differentiate them from AMVT. In addition, AMVT may be misdiagnosed as a ruptured chordae as in this case. In a study, the diagnostic accuracy for ruptured mitral valve chordae was 96.7% for TTE and 100% for TEE compared with surgical findings. Anatomically, mitral valve anterior and posterior leaflets are attached through primary, secondary and tertiary chordae to both the anterolateral and posteriomedial papillary muscles with resultant

![Fig. 1](image1.png)

**Fig. 1** – Transthoracic echocardiogram of the left ventricle (A) and (B) showing the accessory mitral valve (arrowheads) attached to the ventricular side of the proximal part of the anterior leaflet of the mitral valve, moving into the left ventricular outflow tract (LVOT) during systole. On Doppler interrogation of LVOT (C) there was no LVOT turbulence or aortic regurgitation and the peak pressure gradient across the LVOT was 7 mmHg at rest and on exercise.

![Fig. 2](image2.png)

**Fig. 2** – Multi-plane transesophageal echocardiogram of the left ventricle showing a sail-like accessory mitral valve (A, arrowheads) attached to the ventricular side of the proximal part of the anterior leaflet of the mitral valve and (B) producing a sac-like appearance.
stretching during ventricular contraction. Rupture chordae is related to the strain forces exceeding the stretching threshold of the chordae.\(^8,9\) Chordal rupture is defined as the presence of the free and highly mobile, linear echoes associated with or without flail mitral leaflet fluttering coarsely in the left atrium during each systole.\(^8\) Rupture of a major chordae or rupture of multiple chords simultaneously may cause flail leaflet and severe acute or chronic mitral regurgitation.\(^9\) However, rupture of secondary or tertiary chordae or separation of a single chorda may lead to lesser degrees of mitral regurgitation or no MR.\(^9\)

In the case presented, the mobile structure was significantly larger than a chordae, was clearly seen to be attached to anterior mitral valve and left ventricular apical area by an intact chordae with absence of any flail leaflet. In addition, there was no significant MR, thus ruling out ruptured major chordae and suggesting AMVT. In conclusion, we present an adult patient with isolated non-obstructive accessory mitral valve tissue mimicking ruptured chordae of the mitral valve. Accessory mitral valve tissue in adults is very rare and can mimic various causes of left ventricular tract obstruction. This patient represents the first case in literature wherein an unobstructive accessory mitral valve tissue simulated a ruptured chordae. This case illustrates that in patients with suspected mitral valve chordae rupture without any mitral regurgitation this diagnosis should be considered, which can have therapeutic implications.

**Conflicts of interest**

All authors have none to declare.

**REFERENCES**