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MINI-FOCUS ISSUE: IMAGING

INTERMEDIATE

CASE REPORT: CLINICAL CASE

Parachute Mitral Valve

A Case of Isolated Accessory Mitral Valve Tissue

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ABSTRACT

A 69-year-old male presented to the emergency room with dyspnea on exertion lasting more than 2 weeks. Echocardiography showed an ill-defined subaortic structure. Subsequent transesophageal echocardiography revealed a parachute-like structure prolapsing into the left ventricular outflow tract causing subvalvular aortic obstruction. Surgical excision confirmed this structure as an accessory anterior mitral leaflet. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:1578–81) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 69-year-old male presented to the emergency room with progressive dyspnea on exertion associated with an intermittent cough lasting more than 2 weeks. He had no prior symptoms and denied weight gain, orthopnea, or paroxysmal nocturnal dyspnea. Although he had no history of a murmur, physical examination revealed a 3/6 systolic ejection murmur best heard at the right upper sternal border with no carotid radiation. He had bibasilar crackles with no extremity edema. His blood pressure was 130/72 mm Hg, heart rate was 58 beats/min, and his oxygen saturation was 91% on room air. The findings from the rest of his physical examination were normal. Electrocardiogram showed an old left bundle branch block and first-degree atrioventricular block with frequent premature ventricular complexes. For further evaluation of his murmur, transthoracic echocardiography (TTE) was ordered. TTE showed a depressed left ventricular ejection fraction of 35% to 40%, mild aortic regurgitation, and a mobile ill-defined subaortic structure prolapsing into the left ventricular outflow tract (LVOT) causing LVOT obstruction. LVOT peak velocity was 2.85 m/s with a gradient of 32.5 mm Hg, a post-premature ventricular complex LVOT gradient increased to 37.5 mm Hg with a peak velocity of 4.4 m/s (**Figure 1**). Parenthetically there was no change in

LEARNING OBJECTIVES

- AMTV is a rare congenital anomaly consisting of an endocardial cushion that typically arises from the anterior mitral leaflet and often a cause of LVOT obstruction.
- Patients with AMVT may have an asymptomatic murmur or present with chest pain, palpitations related to arrhythmia, congestive heart failure, cerebrovascular events, fatigue, or syncope.
- Echocardiography is pivotal for diagnosis and follow-up. Surgical resection is recommended in symptomatic patients and patients with significant LVOT obstruction.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* author instructions page.

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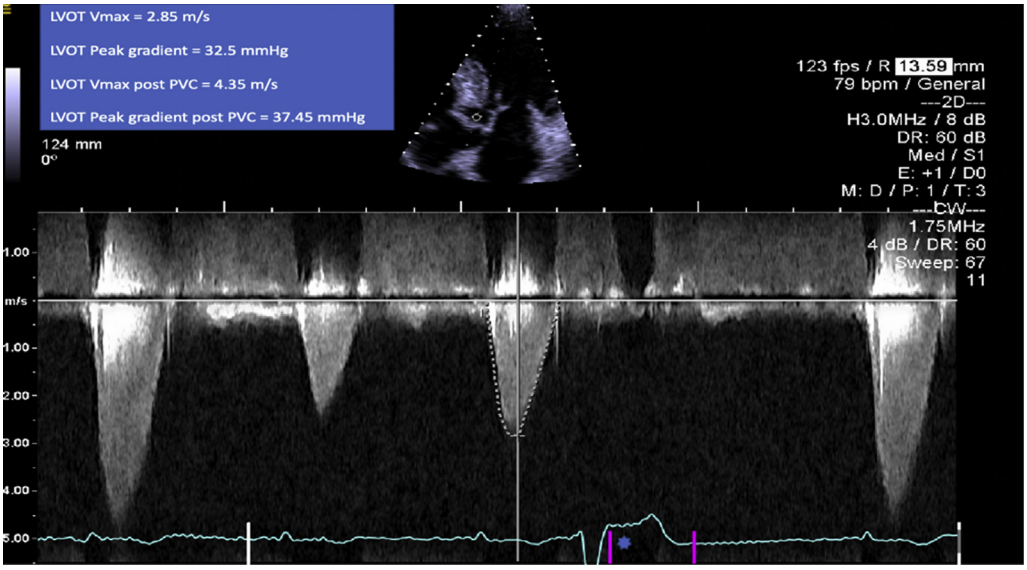
FIGURE 1 Continuous Wave Doppler Across the LVOT Showing High Velocity and Gradient Across the LVOT That Increased Post-Premature Ventricular Complex



LVOT = left ventricular outflow tract; PVC = premature ventricular complex; Vmax = maximum velocity.

the gradient with a Valsalva maneuver. There was no prior TTE for comparison.

MEDICAL HISTORY

He had a history of hypertension and left bundle branch block that was seen on prior electrocardiograms. He had no prior workup for his left bundle branch block.

DIFFERENTIAL DIAGNOSIS

Identification of a mobile structure in the LVOT raises concerns for vegetation, subaortic membrane, redundant mitral valve chordae, primary or secondary cardiac tumors, and accessory mitral valve tissue (AcMVT).

INVESTIGATIONS

Because his TTE was nondiagnostic, transesophageal echocardiography (TEE) was pursued to better define the ill-defined structure in the LVOT. On the standard mid-esophageal 4-chamber TEE view there was a parachute-like subaortic structure prolapsing into the LVOT, consistent with an AcMVT (Videos 1 and 2). As the aortic valve opened, the parachute-like structure was noted to prolapse in a "peek-a-boo" manner toward the aortic valve (Video 3). Color comparison imaging showed flow acceleration around but not through this structure (Video 4). Associated mild aortic insufficiency was noted (Video 5). Given the high suspicion for AcVMT on TEE, no further diagnostic imaging was pursued.

MANAGEMENT

Because the patient was symptomatic with echocardiographic findings of significant LVOT obstruction, surgical intervention was recommended. Preoperative cardiac catheterization showed nonobstructive coronary artery disease. Intraoperative inspection of the LVOT showed a completely formed mitral valve reminiscent of an anterior mitral leaflet affixed to the basal interventricular septum with chordal attachments to the papillary muscle. The complete structure was excised, and histopathology confirmed the diagnosis of accessory mitral valve tissue with degenerative changes (**Figures 2 to 5**). His perioperative period was uncomplicated, and he was discharged to cardiac rehabilitation on goal-directed heart failure therapy for his depressed left ventricular ejection fraction.

DISCUSSION

AcMVT is a rare congenital anomaly of the endocardial cushion that typically arises from the anterior mitral

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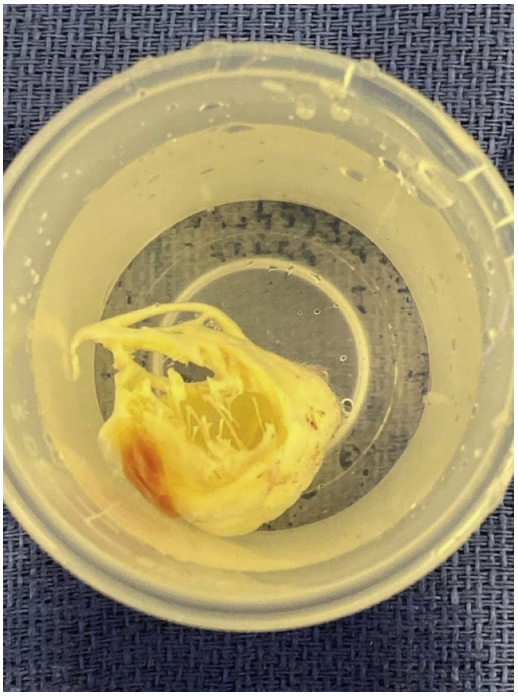
FIGURE 2 Excised Accessory Mitral Valve Leaflet With Chordae

FIGURE 3 Excised Parachute Like Structure Reminiscent of an Accessory Mitral Valve Leaflet With Chordae

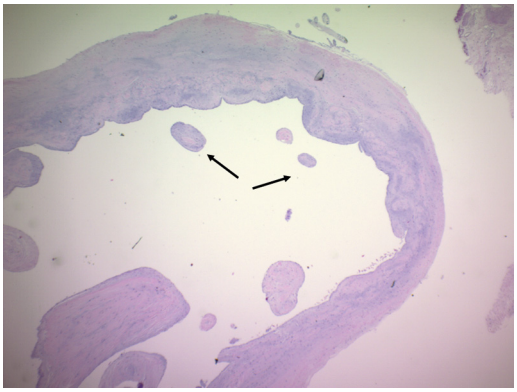


leaflet or less commonly both leaflets simultaneously. It is usually diagnosed in childhood because it is often associated with other congenital intracardiac or vascular anomalies. AcMVT as an isolated finding, as described in the present patient, is rare. The incidence of AcVMT in adulthood is 1 in 26,000 persons (1). AcMVT can be obstructive or nonobstructive, with the patient having an asymptomatic murmur (2). Symptoms of LVOT obstruction include exertional dyspnea, chest pain, syncope, stroke, thromboembolism, low cardiac output due to subaortic obstruction, and decompensated heart failure, as in this patient. The pathophysiology of LVOT obstruction is thought to be related to the mass effect of the AcMVT in the LVOT. As the AcMVT expands, there is progressive narrowing of the LVOT. Also, continuous turbulent flow in the LVOT leads to deposition of fibrous tissue and scarring, which contributes to LVOT obstruction (3,4). As with a fixed subaortic membrane, flow acceleration across the valve may lead to aortic insufficiency as seen in the present patient. Echocardiography is pivotal in the diagnosis of AcMVT, evaluation of associated congenital anomalies, LVOT obstruction, and other valvular abnormalities. In 2003, Prifti et al. (5) reviewed 90 published cases of patients with AcMVT. Severe LVOT obstruction with a trans-LVOT gradient more than 50 mm Hg was present in most cases, and mild LVOT obstruction was found in 15 patients, 11 of whom had a LVOT mean gradient <31 mm Hg and 3 other patients who presented with no LVOT obstruction (5). Although echocardiography remains the gold standard for the diagnosis of AcMVT, cardiac computer tomography and magnetic resonance imaging are useful modalities in its assessment and may better evaluate cardiac tumors and other cardiac anomalies. Surgical resection is recommended for patients with a significant LVOT obstruction mean gradient of >25 mm Hg and for those undergoing cardiac surgery for other cardiac pathology (6). There are no clear guidelines for management of asymptomatic patients (7).

FOLLOW-UP

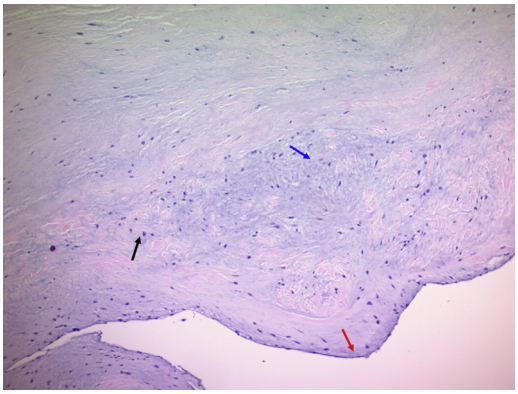
At his 2 weeks follow-up, this patient was completely asymptomatic. Repeated TTE 1 month after surgery showed resolution of LVOT obstruction but

FIGURE 4 **Low-Power View Showing Parachute-Like Structure With Chordae Tendinea Consistent With Valvular Tissue**



Low power view showing parachute like structure with chordae tendinea consistent with valvular tissue **(black arrows)**.

FIGURE 5 **Valvular Tissue With Normal Endothelium, Nuclei, and Bluish Myxoid Intracellular Material Consistent With Loosely Organized Connective Tissue**



Valvular tissue with normal endothelium **(red arrow)**, nuclei **(black arrow)** and bluish myxoid intracellular material consistent with loosely organized connective tissue **(blue arrow).**

unfortunately did not show any improvement in his left ventricular systolic function. It is hoped that further from his surgery his left ventricular systolic function may improve.

CONCLUSIONS

AcMVT is a rare congenital cardiac anomaly that can cause LVOT obstruction. The present case describes an adult male who presented with heart failure symptoms and was found to have LVOT obstruction in the setting of an isolated AcMVT. Echocardiography played an important role in this diagnosis, surgical resection was required as he was symptomatic and had significant LVOT obstruction.

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KEY WORDS acute heart failure, chordae, mitral valve, murmur

APPENDIX For supplemental videos, please see the online version of this paper.