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CASE REPORT

Surgical Repair of an Isolated Parachute-like Asymmetric Mitral Valve Defect in an Adult

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ABSTRACT

Parachute mitral valve (PMV) is a congenital anomaly of the left ventricular papillary muscles. True PMV is characterized by the unifocal attachment of all chordae tendineae into a single papillary muscle group, while parachute-like asymmetric mitral valve (PLAMV) involves two papillary muscle groups with marked asymmetry in the distribution of chordae. Both defects often occur in association with other cardiac anomalies and, therefore, are typically diagnosed in early infancy or childhood. The diagnosis is rarely made in adults as only 1% of PMV is isolated. We report a case of isolated PLAMV in an adult in which more accurate preoperative diagnosis by echocardiography may have assisted with surgical planning.

Keywords: Parachute-like asymmetric mitral valve, Parachute mitral valve, Severe mitral regurgitation.

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INTRODUCTION

Parachute mitral valve (PMV) is a congenital anomaly of the left ventricular papillary (LUP) muscles affecting the attachment of the mitral valve chordae tendineae. True PMV is characterized by the unifocal attachment of all chordae into a single papillary muscle group. A subset of this anomaly, the PLAMV, involves two papillary muscle groups, one of which is dominant and receives attachments from the majority of chordae.[1-3] These defects are most commonly diagnosed in early childhood, as they often occur in association with other cardiac anomalies.[4] The anomaly is well-described as a component of the Shone's complex: PMV, supravalvular mitral membrane, subaortic stenosis, coarctation of the aorta, and, in many cases, a bicuspid aortic valve.[5,6] Because of its association with other significant cardiac anomalies, patients often present early in life with notable symptoms, including congestive heart failure, cyanosis, and/or murmurs with accompanying arm-to-leg bp gradients.[1] In one study of 84 patients diagnosed with PMV, the median age at the time of diagnosis was 3 days. In this study cohort, 99% of patients were found to have associated cardiac lesions, with only one patient having an isolated PMV defect.[1] The overall incidence of isolated PMV is estimated to be < 1%, but the true incidence is plausibly more substantial as these patients may live well into adulthood with minimal or no symptoms from their lesion.[7,8] As a result, adult patients may be diagnosed as having the PMV anomaly by echocardiographic studies obtained as a part of routine evaluation for nonspecific cardiac symptoms.[7,9,10] Echocardiography is an important diagnostic tool for PMV, although few studies describe echocardiographic findings of isolated PMV likely due to its rare diagnosis in adulthood. Clinicians should nevertheless be familiar with these findings on echocardiography. We present a case of isolated PLAMV in an asymptomatic adult in which the correct preoperative diagnosis of PMV on echocardiographic evaluation may have aided in proper surgical planning.

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CASE REPORT

A 51-year-old male presented 1 year prior to surgery for treatment of a nonhealing plantar ulcer, at which time a systolic murmur was noted on physical examination. His past medical history was notable for well-controlled hypertension, peripheral neuropathy, tobacco and alcohol abuse, and methadone use in the setting of prior opioid abuse. The patient had previously undergone multiple podiatric procedures under general anesthesia without problems, and he had no known prior cardiopulmonary history. A diagnostic transthoracic echocardiogram (TTE) was ordered for further evaluation of the murmur; this demonstrated a thickened mitral valve with moderate mitral regurgitation (MR). On repeat surveillance, TTE obtained 16 months later, however, interval progression to severe MR and prolapse of the posterior mitral valve leaflet were discovered. A transesophageal echocardiogram (TEE) was obtained for more precise evaluation of

the lesion and surgical planning. Transesophageal echocardiogram results noted normal left ventricular size and function, moderate left atrial enlargement, and normal right ventricular parameters. The mitral valve appeared to be thickened, demonstrating severe regurgitation and bi-leaflet prolapse, with what appeared to be a torn P2 chordae with flail segment. No significant mitral stenosis was present (transmitral mean pressure gradient 3 mm Hg). The patient was subsequently scheduled for robotic-assisted mitral valve repair. On the day of his surgery, induction of general endotracheal anesthesia was uneventful and followed by lung isolation and peripheral cannulation for cardiopulmonary bypass. An intraoperative TEE examination obtained prior to the surgical exposure demonstrated a thickened, myxomatous mitral valve with what appeared to be A1/A2 prolapse and P1/P2 prolapse (Fig. 1). Concern existed for possible chordae rupture at A2 resulting in a posteriorly directed eccentric regurgitant jet consistent with several MR. Three-dimensional TEE views confirmed a highly abnormal appearing valve with a tri-leaflet appearance (Figs 2 and 3). Intraoperative exposure of the mitral valve ultimately revealed a configuration consistent with PMV, in which all but one chordae were attached to a single posterior papillary muscle and the remaining chorda connecting the base of the P1 segment to a thickened portion of the left ventricular wall. Redundant P1 tissue resulted in the tri-leaflet appearance noted on echocardiography. Retrospective TEE review confirmed this single posterior papillary muscle (Fig. 4). The valve was repaired with resection of the redundant P1 leaflet tissue, placement of an annuloplasty ring, and implantation of artificial neochords connecting the P1 leaflet to the lateral left ventricle and P2-3 to the base of the existing papillary muscle. This more complex repair extended the operative time by an estimated 60 minutes, but was ultimately successful. Postoperative TEE showed only trace MR post-repair with no mitral stenosis. The patient recovered well from his surgery and was discharged home on postoperative day four.

DISCUSSION

Isolated PMV in the adult population is a rare finding, and accurate diagnosis of this defect requires not only high clinical suspicion but also a detailed examination and a thorough understanding of how the lesion appears on echocardiography. In the pediatric population, PMV has traditionally been highly associated with mitral stenosis[11] and is cited as one of four predominant causes of congenital mitral stenosis.[12] In a study of 12 pediatric patients diagnosed with PMV, all 12 patients presented with moderate to severe mitral stenosis, whereas less than half of these also demonstrated MR on echocardiogram.[13] Though MR is less commonly associated with pediatric PMV, this may not be true when the diagnosis is made in adults.[14]

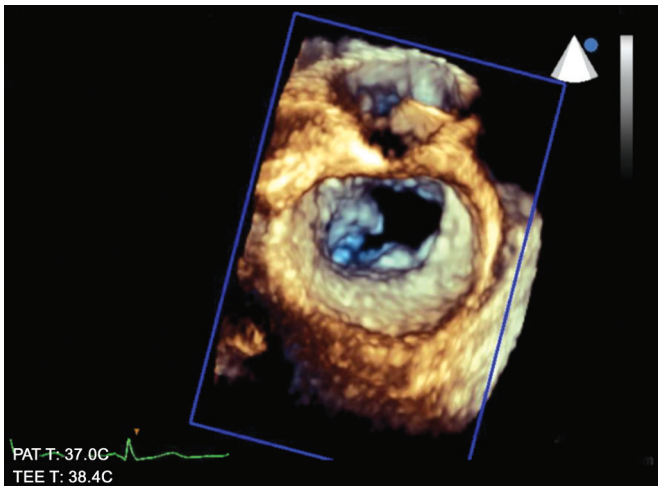


Fig. 2: Surgeon's view three-dimensional acquisition of mitral valve during diastole showing tri-leaflet appearance

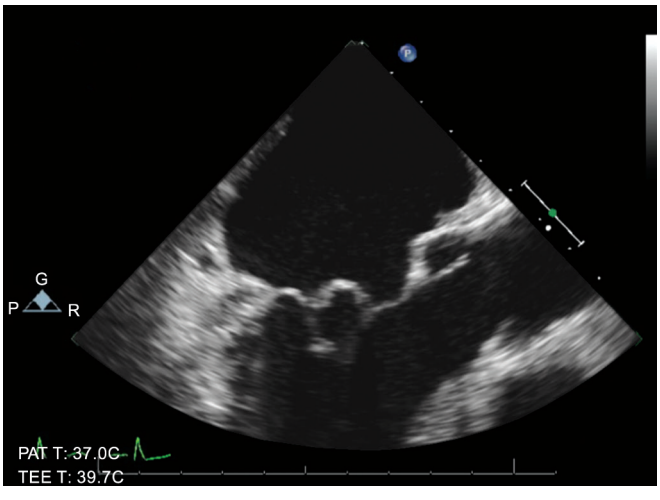


Fig. 1: Mid-esophageal long-axis showing prolapse of the anterior and posterior leaflets

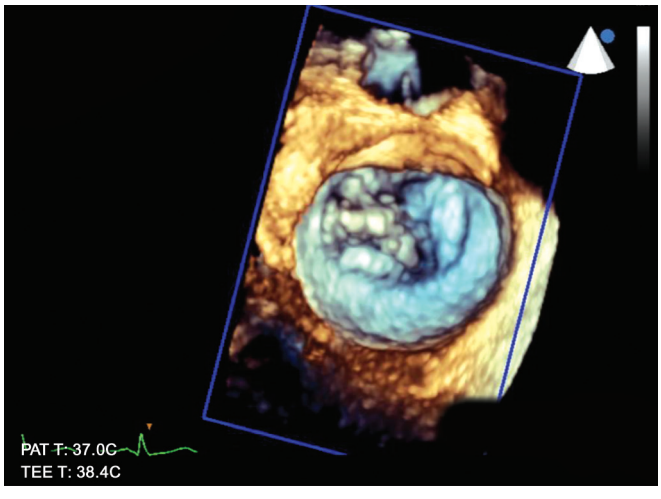
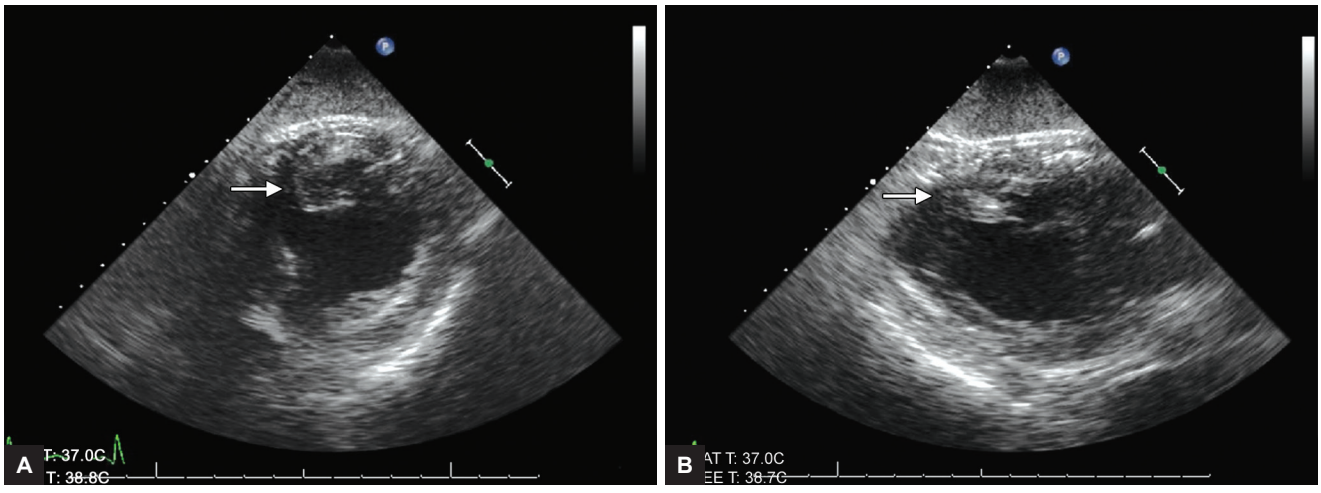


Fig. 3: Mitral valve in systole demonstrating A1/A2 and P1 prolapse. Anterior leaflet cleft is apparent



Figs 4A and B: Transgastric midpapillary view of the left ventricle in (A) short axis, and (B) long axis demonstrating only one well-defined papillary muscle (arrows)

Marino et al., noted that among patients with congenital PMV undergoing biventricular palliation who had progressive MR on follow-up echocardiogram, most did not demonstrate MR on initial studies. Only 6 of 49 patients (13%) demonstrated even trivial or mild MR on initial echocardiogram, and the authors found a significant increase in the incidence of MR from initial to most recent echocardiogram.[4] Only a small number progressed to hemodynamically significant MR, but the trend toward worsening regurgitation may explain differences in examination findings among adult patients with PMV. In the case presented here, significant MR was present without mitral stenosis, and in a study of 9 adult patients with PMV, 56% had regurgitant lesions on initial diagnosis.[7] This suggests that the hemodynamic consequences of PMV may also differ between pediatric and adult populations.

Echocardiographic findings may also differ from that reported in the literature. Classically, the PMV takes on a "pear-shaped" configuration in the four-chamber view, with the left atrium forming the base of the pear, the mitral leaflets the converging sides, and redundant chordae forming the apex.[9] However, this may be difficult to appreciate using TEE, given the inability to visualize the entire left atrium. Previous case reports also describe a "domed" valve in diastole, best seen in the apical four- or two-chamber views.[9] This case demonstrates, however, that anatomic variations of this defect may not produce classically appearing PMV on echocardiography. Failure to identify two independent papillary muscles remains the absolute diagnostic criteria for PMV, although this may be difficult to reliably ascertain. Redundancy of the chordae, and in this case of the leaflet tissue, may be the defining feature on echocardiographic examination. In our case, tissue redundancy was interpreted as torn chordae on multiple examinations. Individual anatomic variants of the congenital defect, therefore, may appear considerably different on echocardiography, and clinical suspicion remains vital to accurately diagnosing the defect, particularly when it occurs as an isolated finding. Certainly, the presence of two papillary muscles should be verified when any other component of the Shone's complex is present, but clinicians should remain suspicious of the defect in adults with mitral valve disease in whom tissue or chordal redundancy is present or suspected.

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