

An Extremely Rare Cause of Mitral Regurgitation—Accessory Commissural Mitral Tissue with Anomalous Left Atrial Chordal Attachment



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INTRODUCTION

Accessory mitral valve tissue (AMVT) is an uncommon congenital cardiac anomaly often detected during the first decade of life, with an estimated incidence in the adult population of 1:26,000 echocardiograms.¹⁻³ This condition is commonly associated with other congenital cardiac abnormalities including ventricular septal defect, patent ductus arteriosus, and transposition of the great arteries. AMVT is most commonly found adjacent to the medial aspect of the mitral valve often resulting in left ventricular outflow tract (LVOT) obstruction, with mitral stenosis also having been described.^{3,4} We report a previously undescribed congenital mitral valve complex consisting of the P1 scallop with associated AMVT being suspended by an anomalous chord extending from the posterior medial papillary muscle through to the anterior left atrial wall. The complex was found incidentally in the operating room in a 61-year-old having coronary artery bypass surgery and was associated with moderate mitral regurgitation (MR) resulting in a change in the surgical plan.

CASE PRESENTATION

A 61-year-old male with past medical history significant for hypertension and type 2 diabetes presented to his general practitioner with chest pain and dyspnea on exertion. The patient was scheduled for elective triple vessel coronary artery bypass grafting after appropriate workup revealed severe multivessel coronary artery disease. Prolapse of the P1 scallop with associated mild regurgitation was seen on preoperative transthoracic echocardiogram (TTE), which was not deemed to necessitate surgical intervention (Videos 1 and 2). TTE evaluation of MR severity was made difficult by the eccentricity of the jet path. Estimated left ventricular ejection fraction was 40%, with no other significant structural abnormalities. Intraoperative transesophageal echocardiogram (TEE) unexpectedly

revealed moderate MR. Multiple jets were seen with quantitation of the dominant jet via the proximal isovelocity surface area method demonstrating a regurgitant volume of 34 mL and an estimated effective regurgitant orifice area of 0.14 cm² (Figure 1). On closer three-dimensional (3D) interrogation of the mitral valve, a coaptation defect was observed at the lateral commissure (Figure 2) caused by an anomalous chord-like structure (Figures 2 and 3), extending from the left ventricle through the mitral valve, attaching at the anterior wall of the left atrium and creating a hammock-like appearance (Figure 2; Videos 3 and 4). The chord-like structure was not appreciated on retrospective review of the preoperative TTE. The suspended P1 segment was associated with AMVT, which was noted on direct surgical inspection and became evident after bypass imaging (Video 5). Neither mitral valve nor LVOT obstruction was evident as a consequence of the AMVT, and no other valve anomalies were present.

Given the unexpected intraoperative findings, the decision was made to proceed with removal of the accessory chordae with the goal of restoring adequate lateral mitral valve coaptation. After atriotomy, visual inspection demonstrated an accessory chord originating from the posterior medial papillary muscle, passing through the mitral valve to the left atrial free wall with impingement of the accessory mitral leaflet (Figures 4 and 5). The anomalous chord was resected with what appeared to be acceptable mitral valve coaptation (Video 5); however, mild to moderate MR remained. Residual MR occurred as a consequence of the suspended previously suspended P1 scallop remaining in a somewhat fixed position even after removal of the impinging aberrant cord possibly secondary to fibrosis and loss of normal leaflet elastance and pliability (Video 5).

Three-dimensional imaging postresection demonstrated more clearly the AMVT on the ventricular side of a still somewhat fixed P1 leaflet (Figures 6 and 7; Video 5). The remainder of the intraoperative course was uneventful. The patient's postoperative course was unremarkable, and he was discharged home on postoperative day 7. Six-month outpatient follow-up TTE similarly showed mild-moderate MR.

DISCUSSION

AMVT and anomalous left atrial chordal attachments are rare congenital anomalies with AMVT usually detected in childhood.³ The embryology of AMVT is not completely understood but seems to be related to the abnormal and incomplete separation of the mitral valve from the endocardial cushion during cardiac development.^{3,5,6} AMVT has been classified as fixed (type I) and mobile (type II), and morphologically it has been described as sail-like, sac-like, balloon-like,

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Keywords: Mitral regurgitation, Accessory mitral valve tissue, Anomalous chordae, Congenital

Conflicts of interest: The authors reported no actual or potential conflicts of interest relative to this document.

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2468-6441

<https://doi.org/10.1016/j.case.2019.05.007>

VIDEO HIGHLIGHTS

Video 1: Preoperative transthoracic apical two-chamber view of the mitral valve showing movement of the lateral posterior leaflet (P1 scallop) above the annular plane. Caution should be taken in the diagnosis of prolapse in this view.

Video 2: Preoperative transthoracic apical two-chamber view of the mitral valve with color showing a medially directed eccentric regurgitant jet originating from the P1 scallop.

Video 3: A 3D en face left atrial view of the mitral valve with P1 suspended by an anomalous cord extending from the left ventricular posterior medial papillary muscle to the left atrium.

Video 4: A 3D left atrial mitral valve view with color tilted slightly from the en face view showing the regurgitant jet originating at the usual P1/P2 position and being directed medially.

Video 5: 3D en face left atrial view of the mitral valve post-operation showing the inferior AMVT layer of the bilayered mitral valve appreciated at the lateral commissure. On mitral valve closure, the P1 scallop can still be seen partially retracted (black arrow) with the AMVT seen on the ventricular side of the valve in diastole (red arrow).

View the video content online at www.cvcasejournal.com.

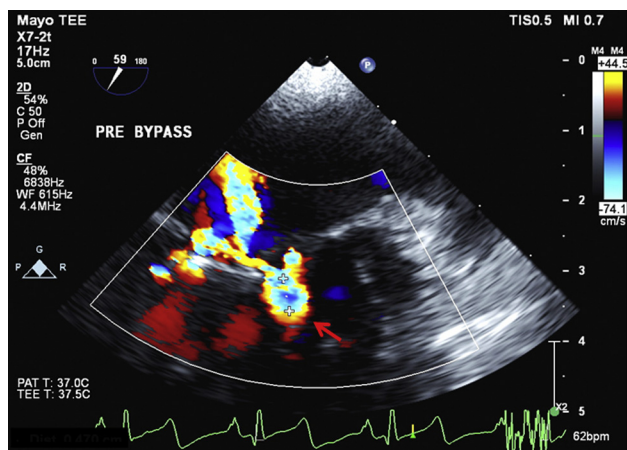


Figure 1 Midesophageal commissural view zoomed on the mitral valve to show the proximal isovelocity surface area head (red arrow) of the regurgitant jet pre-bypass. The regurgitant jet originates from the point where P1 was suspended above the annular plane resulting in malcoaptation where P1/P2 meets A2.

parachute-like, and leaflet-like in shape.^{2,6} In cases with LVOT obstruction, exercise intolerance, exertional dyspnea, chest pain, syncope, and heart failure can occur along with other secondary complications including endocarditis and cardioembolic events, which appear to be related to the excessive mobility of the redundant accessory tissue.^{3,6,7} Although our patient presented with chest pain and exertional dyspnea, his incidentally found mitral valve lesion was unlikely to be the primary cause of his symptoms given that there were no hemodynamically significant features on echo. Echocardiography has been

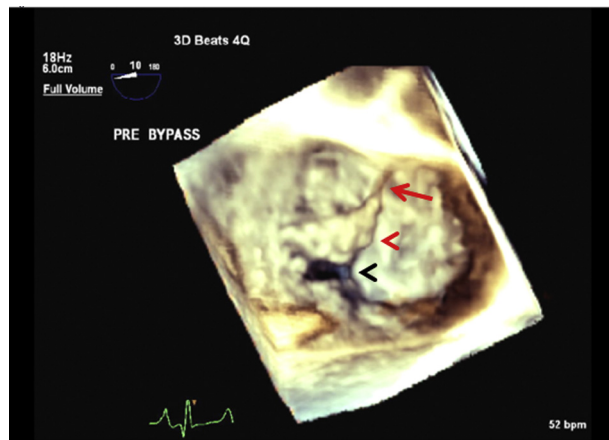


Figure 2 En face 3D LA view of the mitral valve showing the hammock-like appearance created by an anomalous cord (red arrow) suspending the P1 scallop of the mitral valve (red arrow-head) with a resulting coaptation defect (black arrowhead).

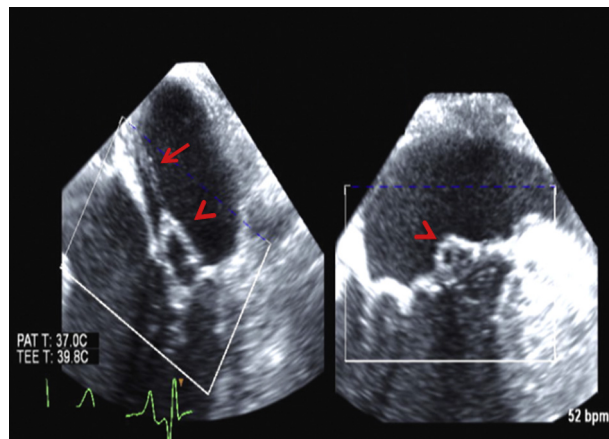


Figure 3 Three-dimensional generated biplane 2D images of the mitral valve. Orthogonal planes show the equivalent of a five-chamber view on the left with the anomalous chord (red arrow) and suspended scallop (red arrowhead).

considered the gold standard in the diagnosis of AMVT since it was first described for this purpose in 1986.⁸ In the last few decades an increased number of AMVT cases have been reported, likely secondary to the widespread use of echocardiography.⁵ Although both TTE and TEE may help with the diagnosis of AMVT, TEE better defines the nature, morphology, and attachment of the AMVT.^{5,6,9} Furthermore, as highlighted by our case, TEE may identify AMVT not seen on TTE examination.

TEE may also help differentiate AMVT from other entities including myxomas, papillary fibroelastomas, and vegetations, which can have a similar echocardiographic appearance and produce similar LVOT mass effect on TTE images.^{9,10} Baek *et al.*¹⁰ for example, reported a case of LVOT obstruction secondary to cardiac myxoma that was misdiagnosed as AMVT on preoperative TTE.

Cases of left atrial anomalous chordae are also a rare occurrence, with only several case reports in the literature describing associated MR and only one in which the anomalous chord extended from the ventricular side of the mitral valve through to the left atrium.^{11,12}

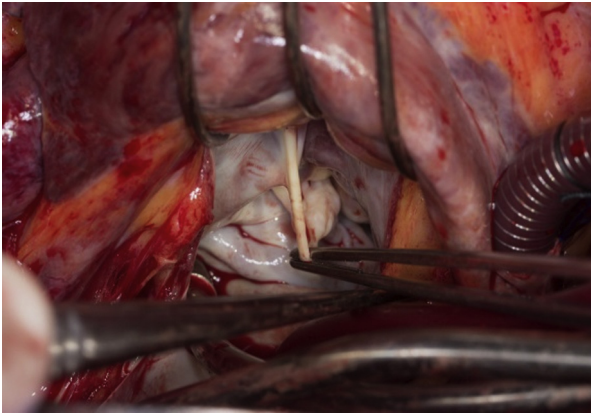


Figure 4 Intraoperative photo showing the anomalous chord extending from the posterior medial papillary muscle through the mitral valve, attaching to the left atrium.

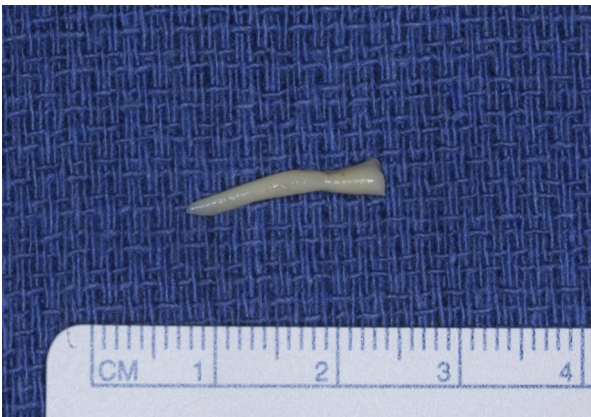


Figure 5 Surgical picture of the anomalous chord.

In a report by Ivanova *et al.*¹³ the aberrant chord resulted in severe MR and attempted open resection resulted in anterior mitral valve injury necessitating mitral valve repair. In our case, although the resection of the anomalous chordae was uncomplicated, the superior layer of the underlying tethered bilayered commissural mitral valve leaflet remained in a somewhat retracted position. Removal of the impinging anomalous chordae therefore improved coaptation, but a degree of incompetence remained, resulting in residual mild-moderate MR. This along with the reported case by Ivanova *et al.*¹³ suggests MR associated with anomalous chordal impingement of the mitral valve may lead to a more complicated repair than simple resection of the anomalous chordae.

CONCLUSION

AMVT is a rare congenital cardiac anomaly usually diagnosed during childhood and rarely reported in the adult population. To our knowledge this complex congenital mitral valve pathology involving AMVT of the lateral mitral commissure combined with an associated anomalous left atrial chordae has hitherto gone unreported. Echocardiography plays an important role in diagnosing this condition, with TEE often providing more information than TTE. AMVT remains a complex congenital cardiac anomaly, and this case highlights a previously unreported manifestation.

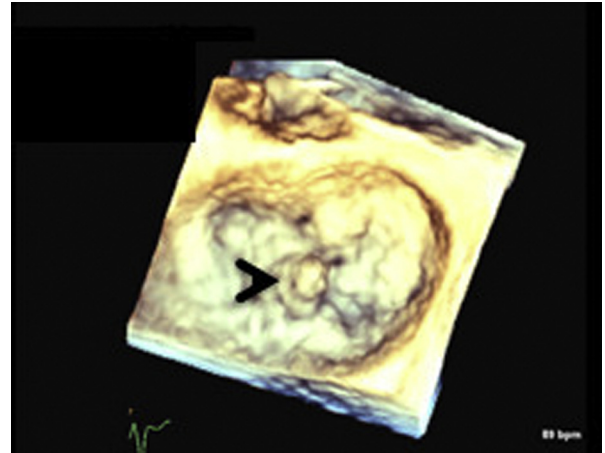


Figure 6 A 3D en face left atrial view of the mitral valve post-operation showing P1 partially stuck in a retracted position (black arrowhead).

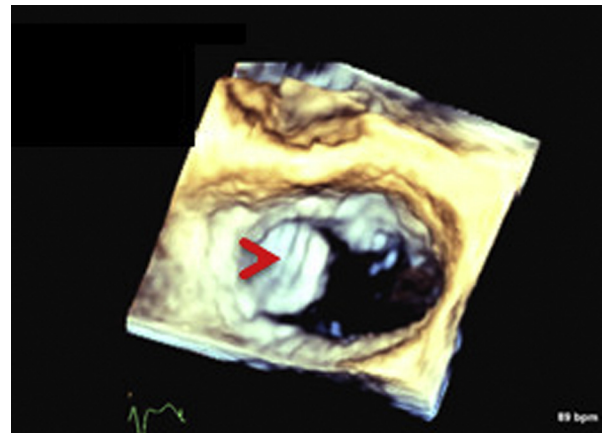


Figure 7 A 3D en face left atrial view of the mitral valve post-operation showing the AMVT layer on the ventricular side of P1 (red arrowhead).

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2019.05.007>.

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