

A rare finding of giant accessory mitral valve tissue: a case report

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Background

Accessory mitral valve tissue (AMVT) is a rare anomaly that can be detected in the first decade. It is associated with other congenital cardiac abnormalities, such as ventricular septal defect. When detected in adulthood, it is usually an incidental finding on echocardiography. Symptomatic individuals can present with breathlessness, syncope, and features of distal tissue embolization. Cardiac surgery is indicated in those with significant left ventricular outflow tract obstruction.

Case summary

A 45-year-old man without any significant medical history was referred due to an abnormal electrocardiogram. He was asymptomatic from a cardiac perspective. Echocardiography revealed the presence of a giant mobile mass attached to the anterior mitral valve leaflet and prolapsing into the left ventricular outflow tract (LVOT). This was classified as Type IIB2 AMVT. As there was no dynamic outflow tract obstruction on subsequent treadmill stress echocardiography, and in the absence of other coexistent congenital abnormality, surgical excision was not performed.

Discussion

It is important to exclude significant obstruction when a large AMVT is seen to be prolapsing into the LVOT. Three-dimensional echocardiography is the tool of choice for anatomical classification and to assess for concomitant congenital cardiac abnormalities.

Keywords

Accessory mitral valve tissue • Congenital • Echocardiography • Case report

Learning points

- Accessory mitral valve tissue (AMVT) is a rare congenital cardiac abnormality with an estimated incidence of 1:26 000 in adulthood.
- It is important to exclude significant left ventricular outflow tract obstruction when a large AMVT is detected.
- Three-dimensional echocardiography is the tool of choice for anatomical classification and to assess for concomitant congenital cardiac abnormalities.

Introduction

Accessory mitral valve tissue (AMVT) is a rare congenital cardiac abnormality that was first described in 1842.¹ Patients can present with a variety of symptoms ranging from phenomena of distal tissue embolization to being completely asymptomatic.² Cardiac surgery is indicated in those with significant left ventricular outflow tract obstruction (LVOTO).³ Here, we present the case of a gentleman with an incidental finding of giant AMVT, who after thorough investigation remained asymptomatic.

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Timeline

The patient was followed-up between November 2017 and November 2018

November 2017	Referred by general practitioner to Cardiology Department after routine health screening which reveals right bundle branch block pattern electrocardiogram.
January 2018	Reviewed in the cardiology clinic. Electrocardiogram pattern felt to be normal variant. Patient is asymptomatic.
October 2018	Transthoracic echocardiogram shows abnormal mobile mass attached to anterior mitral valve leaflet
November–December 2018	Transoesophageal echocardiogram reveals structure to be 8 cm Type IIB2 accessory mitral valve tissue prolapsing into left ventricular outflow tract (LVOT) Treadmill exercise stress echocardiogram shows only mild increase in LVOT gradient from 7.1 to 24 mmHg without dynamic LVOT obstruction Patient remains asymptomatic and discharged to future follow-up

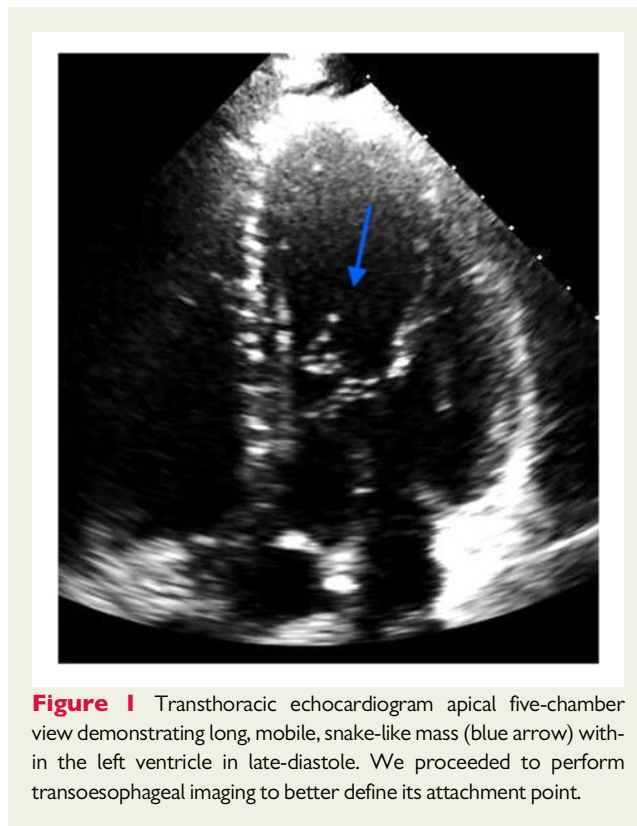


Figure 1 Transthoracic echocardiogram apical five-chamber view demonstrating long, mobile, snake-like mass (blue arrow) within the left ventricle in late-diastole. We proceeded to perform transoesophageal imaging to better define its attachment point.

Case presentation

A 45-year-old man presented to the cardiology outpatients clinic, following a routine health screening. He was referred by his general practitioner as his electrocardiogram (ECG) revealed a right bundle branch block (RBBB) pattern. He was asymptomatic from a cardiac perspective and was training for a long-distance marathon. There was no chest pain, exertional breathlessness, palpitations, or syncope. Aside from a simple Maxillofacial operation, there was no significant past medical history. He was not taking any regular medication and there was no family history of congenital cardiac disease, sudden cardiac death, or cardiomyopathy.

He undertook between 30 and 40 miles of long-distance running on a weekly basis.

On examination, his blood pressure was 135/80 mmHg and his pulse was regular at 64 b.p.m. Heart sounds were normal on auscultation with no audible murmur. The rest of the clinical examination was unremarkable.

Routine blood tests performed including a full blood count, renal and liver function tests, were all within normal range. His 12-lead ECG showed sinus rhythm with partial RBBB and left axis deviation, but a normal PR interval. As there was no history of pre-syncope or syncope, Holter monitoring was not performed.

We proceeded to a transthoracic echocardiogram (TTE), which revealed normal biventricular cavity size and systolic function. Left ventricular wall thickness was normal with an estimated ejection fraction of 65%. There was Grade 1 diastolic dysfunction with a mildly dilated left atrium.

Most strikingly, however, there was a long, mobile, echogenic mass seen attached to the anterior leaflet of the mitral valve (*Figure 1*). This did not appear to cause left ventricular outflow tract (LVOT) obstruction at rest (peak velocity through the LVOT was 1.1 m/s with a peak gradient of 5.6 mmHg and no significant turbulent flow on colour Doppler imaging) or interfere with mitral valve closure significantly (*Supplementary material online, Videos S1 and S2*).

We went on to characterize the anomaly further with a transoesophageal echocardiogram (TOE) (*Figures 2 and 3*). The highly mobile redundant tissue measured 8 cm in length and was seen to prolapse freely into the LVOT during mid-to-late systole (*Figure 4*) but not extend beyond the aortic valve (*Supplementary material online, Video S3*).

The mitral valve itself was thin and mobile with no stenosis and only trivial mitral regurgitation. All other valves were structurally normal.

We also performed a treadmill stress echocardiogram to investigate dynamic LVOTO. He undertook 8 min of exercise under Bruce protocol and achieved 100% of his age-predicted heart rate without chest pain or breathlessness. Left ventricular outflow tract peak velocity and peak gradient increased from 1.3 m/s and 7.1 mmHg to 2.4 m/s and 24 mmHg, respectively. Thus, there was no haemodynamic compromise from this giant AMVT.

In the case described this asymptomatic gentleman was referred due to an RBBB pattern on his ECG. We used TOE to confirm that the patient had a Type IIB2 leaflet-like AMVT, with no dynamic LVOT gradients during exercise. There were no other congenital heart disease malformations detected on imaging.

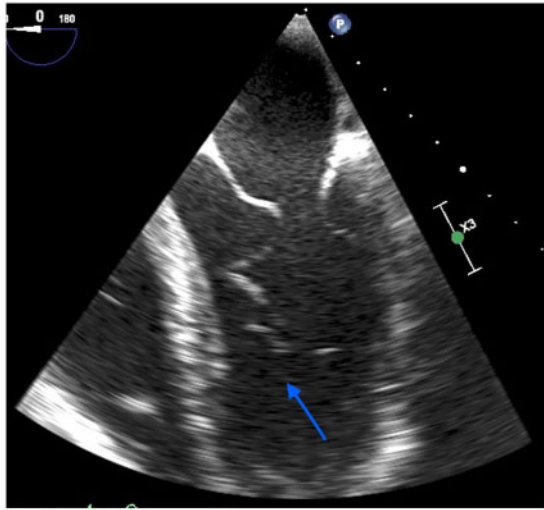


Figure 2 Two-dimensional transoesophageal echocardiogram: mid-oesophageal four-chamber view showing mobile leaflet-like redundant tissue (blue arrow) attached to anterior mitral valve leaflet.

Thus in a patient who has remained well despite a regular high level of activity, we were happy that he did not require surgery. The rationale for this was his lack of significant LVOTO. Surgery is indicated at an LVOT gradient above 25 mmHg, or if there is co-existent congenital cardiac abnormality also requiring surgery.

He was not treated with anticoagulation. The data to support this approach in the asymptomatic individual is lacking. The decision would likely have been different if the presentation was one of distal tissue embolization; this may also be an indication for surgery.

Follow-up will involve annual surveillance with a TTE with assessment of LVOT gradient. The literature on adequate follow-up time periods is scarce and mostly limited to case series but most advise a surveillance programme for asymptomatic patients. The gentleman has been advised to report symptoms of pre-syncope, syncope, chest pain, or undue breathlessness which would prompt earlier assessment.

Discussion

Accessory mitral valve tissue is an anomaly that can be detected in the first decade of life. It can be associated with other congenital cardiac abnormalities, such as ventricular septal defect, subaortic stenosis, transposition of the great arteries, atrial septal defect, coarctation of the aorta, and coronary artery anomalies.⁴

Estimated incidence in adulthood is 1:26 000 echocardiograms with a preference for males (male-to-female ratio = 1.75:1).⁴

The embryological mechanism behind AMVT formation is uncertain but is thought due to incomplete separation of the mitral valve from the endocardial cushion; hence, the association with other congenital tissue abnormalities.⁵



Figure 3 Three-dimensional transoesophageal echocardiogram: in this long-axis view, we appreciate the degree of prolapse into the left ventricular outflow tract during mid-systole but not through the aortic valve. Three-dimensional transoesophageal echocardiogram is important in demonstrating the point of attachment of the structure to the ventricular surface of the anterior mitral valve leaflet.

Accessory mitral valve tissue has been classified anatomically.⁶ *Type IA* is a fixed, nodular mass normally attached to the ventricular side of the anterior mitral valve leaflet (AMVL) or the mitroaortic continuity. *Type IB* is usually attached to the latter and is more membranous. The more mobile AMVT comes in the 'pedunculated' form as in *Type IIA* and is normally attached to the mitroaortic continuity or the ventricular side of the AMVL. The mobile, 'leaflet-like' form is more common (*Type IIB*) and is sub-classified further depending on whether the chordae is rudimentary (*Type IIB1*) or well developed (*Type IIB2*).

In a case series, the mobile AMVT variant which tended to prolapse into the LVOT appeared dysplastic and displayed thickening of the sponge layer. Histology of the fixed variant (attached to the ventricular septum) in the same series, revealed normal morphology of the valve tissue.⁶

Most adult cases are detected in asymptomatic adults undergoing echocardiography for a different indication. However, patients can present with chest pain, exertional breathlessness, and syncope when LVOT gradients exceed 50 mmHg.² Less common presentations include manifestations of distal tissue embolization and endocarditis.

Transthoracic echocardiogram and TOE are important in visualizing the abnormal structure. In particular, three-dimensional TOE

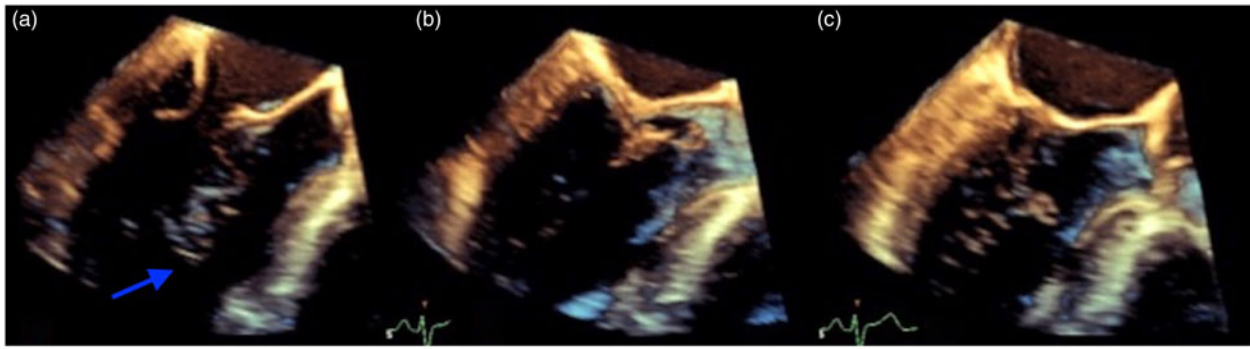


Figure 4 Three-dimensional transoesophageal echocardiogram: large spiral anterior mitral valve leaflet (blue arrow) attached to anterior mitral valve leaflet (A) in mid-diastole before prolapsing into the left ventricular outflow tract in mid-systole (B) and returning to fill the left ventricular cavity (C) by the end of the systolic phase.

allows assessment of AMVT attachment location and the presence of concomitant mitral valve pathology. Tissue echogenicity is similar to that of native endocardial structures. Cardiac magnetic resonance imaging can play an important role in better delineating obstructive AMVT, in particular, prior to surgery. Asymptomatic patients without LVOTO at rest may undergo exercise echocardiography to exclude dynamic gradients, as was the case in this gentleman.

Cardiac excision surgery is indicated in patients with significant LVOTO gradients (>25 mmHg) or those undergoing cardiac surgery for other congenital lesions. Overall mortality after surgery in reported cases is 8.9%, due primarily to persistence of LVOTO after excision, or the consequence of concomitant congenital anomalies.³

Conclusion

Accessory mitral valve tissue is a rare but important congenital cardiac lesion. Most commonly, its detection will be incidental, but thorough work-up is required to determine if surgical intervention is warranted. Due to its rare nature, the evidence to guide surveillance of the patient managed conservatively is limited, but new and adverse symptoms should prompt repeat imaging, with a focus on outflow tract gradients both at rest and on exertion.

Patient perspective

Due to being referred to as a consequence of what was a fairly routine health screening, our patient was relatively surprised to have undergone so many subsequent tests. He remarked upon feeling fit and not having any adverse effects from the discovered AMVT to date. His most pressing query was whether he would be medically fit to continue exercising to the level he currently did and, therefore, was reassured when the treadmill stress echocardiogram helped confirm this.

Lead author biography



Dr Joseph Okafor is a Cardiology Registrar in North West London. He graduated from the University of Sheffield Medical School in 2013. He has a keen interest in Cardiac Imaging and is currently involved in research within Cardiac CT, Stress Echocardiography, and Virtual Reality.

Supplementary data

[Supplementary material](#) is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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