Left atrial mitral valve chordae; an important congenital abnormality mimicking endocarditis on transthoracic echocardiography

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Abstract

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Data statement

The data that support the findings of this study are available from the corresponding author, Dr Alan Appelbe, upon reasonable request.

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Conflict of interest

There is no conflict of interest to disclose.

Ethics approval

Local ethics committee approval at Barwon Health, Victoria was obtained prior to data collection.

Patient consent

Patient information has been deidentified so consent was not obtained.

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Abstract

A 59-year-old male was incidentally diagnosed with a left atrial mitral valve chordae involving the junction of the A1 and A2 mitral valve leaflets and resulting in moderate mitral regurgitation. The recognition of this extremely rare congenital malformation prevented over diagnosis and overtreatment.

Case report

An otherwise well 59-year-old male was admitted to hospital with undifferentiated migratory polyarthralgia, fevers, raised inflammatory markers and a new pansystolic murmur. Transthoracic echocardiogram was performed to investigate for infective endocarditis and showed localised thickening of the anterior mitral valve leaflet with mild prolapse with moderate mitral regurgitation (see Figure 1). The findings were suspicious for endocarditis. Left ventricular cavity size and systolic function were normal. A transoesophageal echocardiogram revealed a left atrial mitral valve chordae. This was attached from the left atrial wall, near the ostium of the left atrial appendage, to the anterior leaflet near the junction of A1 and A2 (see Figures 2 and 3). Mild-moderate mitral regurgitation was present with multiple jets. The patient was discharged home with a plan for repeat echocardiography in 2 years to reassess the severity of his mitral regurgitation and left ventricular function.

Left atrial mitral valve chordae is a rare congenital abnormality of which is the prevalence is largely unknown. There have been few case reports worldwide and the clinical significance remains uncertain, ranging from incidental findings with mild-moderate mitral valve pathology managed conservatively(1-4) to severe mitral regurgitation requiring surgical intervention(5-13). There has been an isolated report of left atrial mitral valve chordae causing complex endocarditis(14). In our patient, recognition of this congenital abnormality was imperative to avoid over diagnosis and unnecessary surgical intervention, particularly given the differential diagnosis of infective endocarditis.

The origin of left atrial mitral valve chordae is proposed to be a result of a developmental defect during embryogenesis between the 14^{th} and 17^{th} weeks of gestation as the papillary muscles and chordae develop(4). Left atrial mitral valve chordae have been identified in children as young as 8-years-old(12), while one case was only diagnosed at the age of 85-years-old(6). At 59-years-old, our patient was asymptomatic from his valvular pathology and his finding was incidental. It has been hypothesised that symptoms may be delayed by progressive growth and dilatation of the left atrium, resulting in gradual traction of the aberrant chord eventually resulting in leaflet prolapse(6). Other suggested mechanisms include the chord holding the free edge of the leaflet in a scallop during diastole with the atrial kicking motion resulting in the development of mitral regurgitation(7) and traumatic injury to the free edge of the leaflet resulting in tethering and restricted mobility(10). Most cases in the literature describe involvement of the A2 leaflet of the mitral valve(3, 4, 8, 10-13), with one case involving the A3 leaflet(7) and one case involving P2 leaflet(6). We describe the first case which involves the junction of A1 and A2.

Conclusion

Left atrial mitral valve chordae is a rare congenital abnormality which has a broad range of clinical presentations. In our patient it mimicked a vegetation on transthoracic echocardiography. Recognition of this pathology is essential to avoid over diagnosis and treatment. In patients with mild-moderate mitral regurgitation, close follow-up with serial echocardiography is recommended given the risk of progression to severe mitral regurgitation.

Figure 1 – Transthoracic echocardiogram showing thickening on anterior mitral valve leaflet, suspicious for infective endocarditis

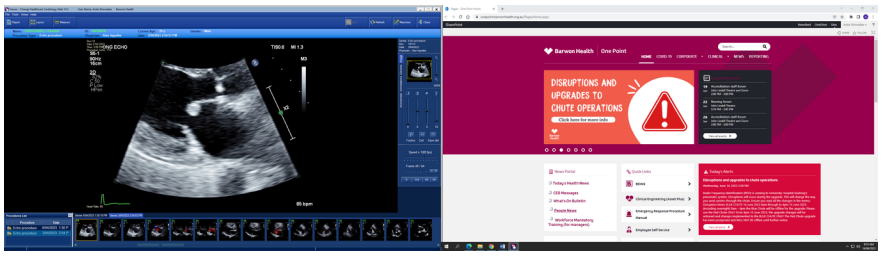


Figure 2 – Transoesophageal echocardiogram showing left atrial mitral valve chordae attached from the left atrial wall, near the ostium of the left atrial appendage, to the anterior leaflet of the mitral valve.

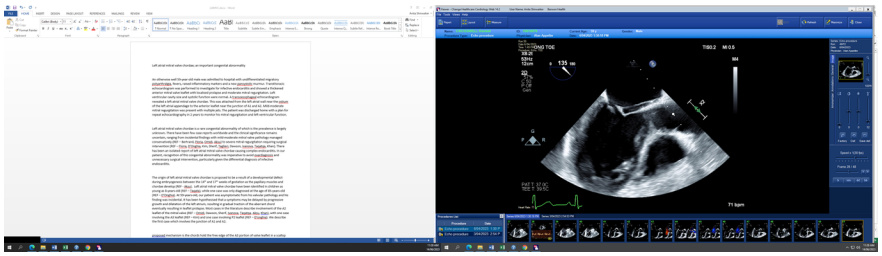
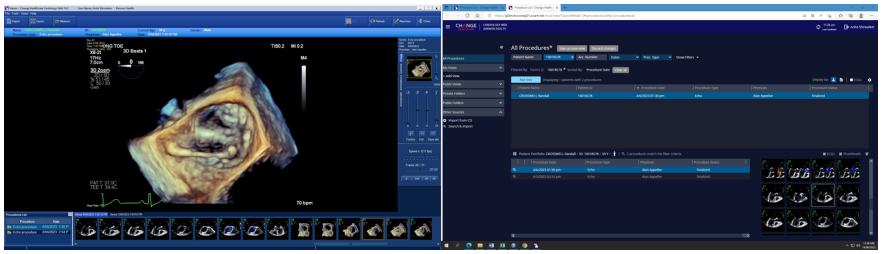


Figure 3 – 3D transoesophageal echocardiogram showing left atrial mitral valve chordae attached to the anterior leaflet of the mitral valve at the junction of A1 and A2



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