

European Society https://doi.org/10.1093/ehjcr/ytac169

Aberrant mitral valve chord in the left atrium causing moderate regurgitation

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Received 18 February 2022; first decision 16 March 2022; accepted 13 April 2022; online publish-ahead-of-print 16 April 2022



A 54-year-old woman presented with moderate mitral regurgitation (MR) and was referred to our hospital for further diagnosis and treatment. She had a history of left breast cancer resection, with radiotherapy and chemotherapy, 17 years ago. Mild MR was found at that time. Five years ago, regional wall motion abnormality with moderate MR was noted upon transthoracic echocardiography (TTE), but the coronary arteries were unremarkable on the coronary angiogram. Thus, regular follow-up was recommended.

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Handling Editor: Christoph Jensen

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The patient had no obvious clinical symptoms, and a systolic murmur could be heard during routine physical examination. Blood pressure, pulse, and heart rate were unremarkable.

Transthoracic echocardiography showed enlargement of the left atrium (LA) and ventricle (LV) with moderate MR and a left ventricular ejection fraction of 40%. Estimated pulmonary artery pressure by tricuspid regurgitation was 47 mmHg. Transthoracic echocardiography revealed a cord-like structure in the LA. It originated from the A2 scallop of the mitral valve (MV) leaflet, with its other end connected to the aortic sinotubular junction. The anterior MV leaflet was restricted in diastole due to the tethering of the cord-like structure, and a prolapse-like appearance of the A2 scallop was noted in the systole. Colour Doppler echocardiography showed moderate MR with vena contracta of 5 mm (Panels A-C, Supplementary material online, S1). Two- and three-dimensional transoesophageal echocardiographic examinations confirmed that the cord-like structure extended from the anterior wall of the LA to the left atrial aspect of the A2 scallop with moderate MR (Panels D and E, Supplementary material online, S2). Because the patient's symptoms were not obvious, a close follow-up was carried out.

The aberrant MV chord is Type I of accessory mitral valve tissue (AMVT), which is a rare congenital anomaly.^{1–2} It is also the least common type of AMVT and has the most devastating clinical presentation.¹ Aberrant MV chord in the LA usually results in MR, which may deteriorate over time. Moderate to severe MR may lead to LA enlargement and left ventricular dysfunction.^{3,4} The aberrant MV chord in the LA is still challenging by TTE. The differential diagnosis includes MV prolapse resulting from the chordal rupture or infective endocarditis. Transoesophageal echocardiography is useful for differentiating these entities.

(Panel A) (Supplementary material online, Video S1) The left ventricle long-axis view on the transthoracic echocardiography showed a cord-like structure (white arrow) in the left atrium. It originated from the A2 scallop of the mitral valve leaflet, with its other end connected to the aortic sinotubular junction. (Panel B) Colour Doppler echocardiography showed moderate mitral regurgitation with vena contracta of 5 mm. LA, left atrium; LV, left ventricle. (*Panel C*) The short-axis view of the cord-like structure showed the hyperechogenicity foci (white arrow) in the left atrium. LV, left ventricle. (*Panel D*) Transoesophageal echocardiography showed that the cord-like structure (white arrow) extended from the anterior wall of the left atrium to the left atrial aspect of the A2 scallop. LV, left ventricle. (*Panel E*) (Supplementary material online, *Video S2*) Real-time three-dimensional transoesophageal echocardiography showed that the cord-like structure extended from the anterior wall of the left atrium to the left atrial aspect of the A2 scallop of the mitral valve. AO, aorta.

Supplementary material

Supplementary material is available at European Heart Journal – Case Reports online.

Consent: The authors confirm that written consent for submission and publication of this case report, including images and associated text, has been obtained from the patient, in line with COPE guidance, which has been added in the text.

Conflict of interest: None declared.

Funding: None declared.

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