Mitral annulus disjunction, a rare form of focal arrhythmogenic cardiomyopathy

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The rate of sudden cardiac death in mitral valve prolapse (MVP) patients is roughly twice that observed in the general population. Arrhythmogenic bileaflet MVP syndrome has been recently described and mitral annulus disjunction (MAD) has been found associated with papillary muscle fibrosis and ventricular arrhythmias even without severe mitral regurgitation. An underlying primary structural myocardial disease might be expected and the non-invasive imaging techniques are playing an increasingly important role in the detection of myocardial fibrosis as a focal arrhythmogenic substrate. Standard 2D-, 3D- and speckle tracking echocardiography and cardiac MRI can help to differentiate between the benign and malignant forms of MVP.

A 52-year-old man with a history of hypertension was referred for evaluation of frequent palpitations and presyncope. Ambulatory Holter monitoring showed paroxysmal atrial fibrillation and non-sustained ventricular tachycardias. Dilated left ventricle (LVEDD: 65mm, LVESD: 45 mm) and left atrium (LAVi: 52mL/m2), moderately reduced left ventricular (LV) systolic function (LVEF: 42%), and reduced LV global longitudinal strain (GLS: -13.5%) with severely decreased segmental longitudinal strain in the basal inferolateral segment were measured by transthoracic echocardiography. The diastolic function was preserved, however Pickelhaube sign was noticed in the lateral mitral annulus TDI curves. MAD (17 mm) and curling motion of the inferolateral basal segment of the left ventricle was observed. Transoesophageal echocardiography showed bileaflet mitral valve prolapse with the involvement of all the six scallops (mitral valve prolapse volume: 10.2 mL). Severe mitral regurgitation (3D vena contracta area: 1cm2) and extremely dilated and dyssynchronous mitral annulus (diameters: 55x72x12 mm, area: 34 cm2) was measured by 3D echocardiography. MRI showed transmural late gadolinium enhancement in the underlying myocardium of both papillary muscles and midmyocardial enhancement in the basal inferior, inferolateral and anteroseptal LV segments. During electrophysiology study ventricular fibrillation was easily induced and ICD was placed for primary prevention of sudden cardiac death. The patient was referred to the heart surgeon for mitral valve replacement.

This case highlights the importance of early recognition of MAD and arrhythmogenic MVP syndrome using novel non-invasive imaging techniques. The presence of MAD, the curling motion of the inferolateral basal LV segment, the Pickelhaube sign, the decreased LVEF and GLS, the characteristic segmental longitudinal strain pattern and the presence of late enhancement in the papillary muscles and the surrounding myocardium could be warning signs of the malignant form of MVP.
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