

Image of the month: 'Hourglass left ventricle' in mid-ventricular hypertrophic cardiomyopathy

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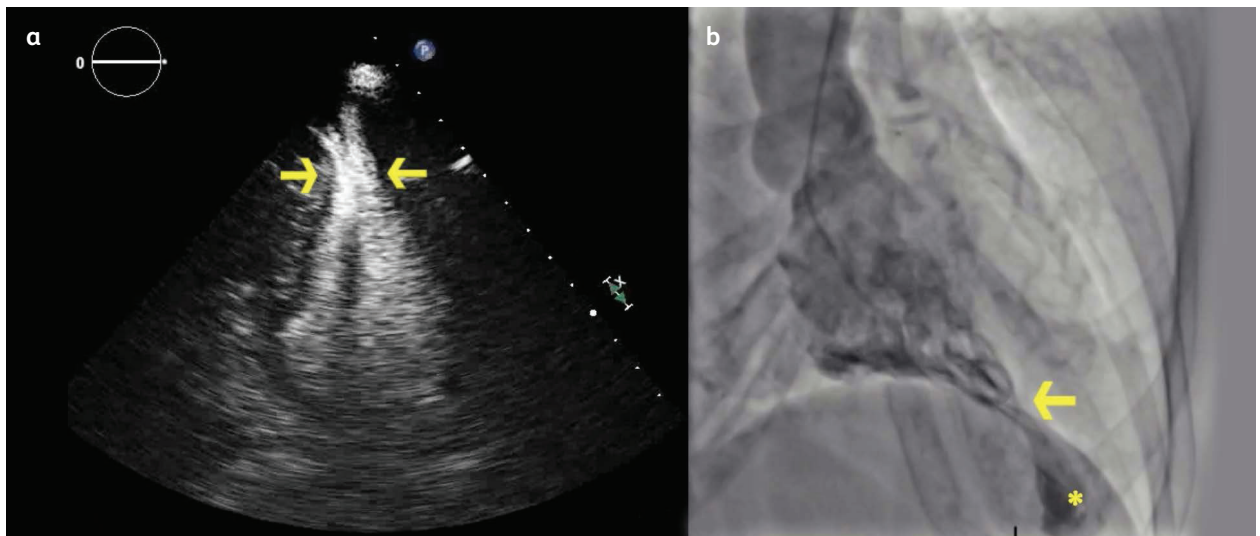


Fig 1. a) Transthoracic echocardiography with contrast showing midventricular narrowing of the left ventricle. b) Left ventriculography showing midventricular narrowing and 'hourglass' appearance of the left ventricle. Asterisk points to the apical aneurysm.

KEYWORDS: hypertrophic cardiomyopathy, echocardiography

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Case presentation

A 69-year-old woman with a past medical history of hypertension presented with dyspnoea. Electrocardiography showed left ventricular (LV) hypertrophy and diffuse T wave abnormality. Echocardiography showed systolic mid-cavity narrowing of LV with turbulent flow (Fig 1a). Coronary angiography showed normal coronaries and left ventriculography showed mid-cavity obliteration with 80 mmHg gradient and characteristic 'hourglass' appearance (Fig 1b). Cardiac magnetic resonance imaging showed complete obliteration of the midventricular cavity during systole

with aneurysm of the true apex consistent with a diagnosis of mid-ventricular hypertrophic obstructive cardiomyopathy (MVHOCM).

Discussion

Hypertrophic cardiomyopathy is a heterogenous disease with several described phenotypes. MVHOCM is a rare variant characterised by hypertrophy mainly confined to the midmyocardial segments. This results in characteristic hourglass appearance of LV with distinct proximal and distal chambers. LV apical aneurysms occur in 20% of MVHOCM. It is suggested that chronic subendocardial ischaemia due to the persistently elevated pressures in the distal cavity leads to scarring and thinning and eventually aneurysm formation. Presence of apical aneurysm in these patients is associated with increased risk of thrombosis and ventricular tachycardia. ■

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