Left ventricular aneurysm complicating apical hypertrophic cardiomyopathy

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DESCRIPTION

A 44-year-old previously well man presented to our emergency department with exertional chest pain. An ECG showed widespread deep T-wave inversion and left ventricular hypertrophy by voltage criteria, [figure 1](#). High-sensitivity troponin I was persistently elevated at 48–58 ng/L (normal <20 ng/L) and he was managed as a non-ST elevation acute coronary syndrome.

Coronary angiography demonstrated patent coronary arteries with mild luminal irregularities. Left ventriculography demonstrated a ‘spade-like’ left ventricular cavity in diastole and systole and a large apical aneurysm, [figure 2](#) and [video 1](#). Invasive pressure measurements demonstrated a peak-to-peak midventricular systolic gradient of 141 mm Hg (284/15 mm Hg in the aneurysmal apex and 143/18 mm Hg in the left ventricular base).

A transthoracic contrast echocardiogram demonstrated a large apical aneurysm, with left ventricular hypertrophy extending from mid to distal left ventricular segments, with midventricular cavity obliteration in systole. There was reduced contrast enhancement in the thinned apical myocardium, indicating reduced perfusion, [figure 3](#) and [video 2](#).

A diagnosis of apical hypertrophic cardiomyopathy (HCM, Yamaguchi syndrome) complicated by a large left ventricular aneurysm was made. The

Figure 3 A transthoracic contrast echocardiogram demonstrating a large apical aneurysm (white arrow), with left ventricular hypertrophy extending from mid to distal left ventricular segments (red arrows), with midventricular cavity obliteration in systole. There was reduced contrast enhancement in the thinned apical myocardium (blue arrow). See [video 2](#).

patient was commenced on a beta-blocker. The patient’s clinical presentation was attributed to demand ischaemia and small vessel angina. Given the incidental mild irregularities demonstrated on coronary angiography the patient was also commenced on a statin and aspirin.

HCM is a spectrum of disease encompassing primary left ventricular hypertrophy of several morphologic subtypes. The genetic basis of the disorder involves sarcomeric protein mutations.¹ The clinical features include heart failure,

Figure 1 ECG showing widespread T-wave inversion and left ventricular hypertrophy by voltage criteria.

Figure 2 Left ventriculography demonstrating a ‘spade-like’ left ventricular cavity (black arrow) and a large apical aneurysm (white arrow) in diastole ([figure 2A](#)) and systole ([figure 2B](#)). See [video 1](#).

Video 1. Left ventriculography demonstrating a ‘spade-like’ left ventricular cavity and a large apical aneurysm

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Video 2 A transthoracic contrast echocardiogram demonstrating a large apical aneurysm with left ventricular hypertrophy extending from mid to distal left ventricular segments, with mid ventricular cavity obliteration in systole

ischaemic chest pain, arrhythmia and pulmonary hypertension.¹ The apical subset is often defined as left ventricular predominating at the apex with a wall thickness ≥ 15 mm and a ratio of maximal apical to posterior wall thickness of ≥ 1.5 detected on echocardiography or cardiac MRI.² The apical left ventricular hypertrophy may cause large midventricular pressure gradients, confers an elevated risk of atrial fibrillation and is less associated with left ventricular outflow tract obstruction.² Apical HCM may be further complicated by aneurysm formation resulting in increased risk of thromboembolism, ventricular arrhythmia and sudden cardiac death.³ Patients suspected of HCM may be further investigated with cardiac MRI to assess the distribution and severity of left ventricular hypertrophy or aneurysm in a

more sensitive manner compared with echocardiography. In addition, cardiac MRI may demonstrate the presence and distribution myocardial fibrosis which is of diagnostic and prognostic importance.²

Learning points

- ▶ Patients with hypertrophic cardiomyopathy may present with exertional chest pain.
- ▶ Apical hypertrophic cardiomyopathy may be complicated by left ventricular aneurysm formation.
- ▶ Such aneurysms confer an increased risk of thromboembolism, ventricular arrhythmia and sudden cardiac death.

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