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 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,
ischaemic chest pain, arrhythmia and pulmonary hypertension.\(^1\) The apical subset is often defined as left ventricular predomi-
nating at the apex with a wall thickness \(\geq 15\) mm and a ratio of
maximal apical to posterior wall thickness of \(\geq 1.5\) 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 distri-
bution 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.

**Contributors**

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