Apical Hypertrophic Cardiomyopathy: Calling a Spade a Spade
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Clinical History
A 53-year-old asymptomatic man presenting for a routine physical examination was found to have an abnormal EKG, with deep T-wave inversions across the anterior precordial leads. The patient subsequently had a cardiac ultrasound (ECHO) at an outside institution, which demonstrated apical hypertrophic cardiomyopathy (ApHCM). A cardiac magnetic resonance imaging (CMR) study was requested for further evaluation.

Findings
CMR confirmed the ECHO diagnosis of ApHCM. A focal area of fibrosis/scar is suggested by the apical defect on cine perfusion sequencing and myocardial late gadolinium enhancement involving segment 17 (based on the AHA 17-segment model). Additionally, high T2 signal at the apex suggests focal edema at the site of hypertrophy.

Discussion
The case presented is an excellent demonstration of ApHCM or Yamaguchi syndrome, an uncommon but well recognized variant of HCM originally described in Japan. ApHCM is characterized as concentric, apical left ventricular thickening, resulting in a spade-like morphology of the left ventricular cavity during end diastole. Diagnostic criteria for ApHCM includes an LV wall thickness >15mm or an apical-to-basal LV wall thickness ratio of 1.3-1.5.1

ECHO is the primary noninvasive tool for diagnosis and hemodynamic assessment of patients suspected of having HCM. However, ECHO is operator-dependent, and thus may occasionally result in non-diagnostic studies. In such cases, CMR can helpful in establishing a definitive diagnosis.2

In our case, CMR was vital in demonstrating focal areas of fibrosis in the apical wall of the left ventricle, which is thought to be a reflection of chronic myocardial ischemia stemming from the inadequate perfusion of hypertrophied myocardium.3 The presence of fibrosis is a known risk factor for adverse sequelae such as arrhythmias, and aneurysm formation.

REFERENCES