The patient was a 43-year-old asymptomatic Indian male referred for evaluation of an abnormal ECG obtained at a routine check-up. His cardiovascular risk factors included hyperlipidemia and a family history of premature coronary artery disease. His physical examination was unremarkable. The ECG (Figure 1) showed sinus rhythm with evidence of left ventricular hypertrophy, ST-segment elevation in the inferior leads III and aVF, and giant negative T waves in the precordial leads. Two-dimensional (Movie I) and contrast echocardiography (Movie II) with DEFINITY (Perflutren lipid microspheres, Bristol-Myers Squibb Medical Imaging, Inc, N. Billerica, Mass) were performed, revealing severe apical hypertrophy with the absence of a dynamic left ventricular outflow tract gradient. Radiocontrast left ventriculography (Figure 2) demonstrated the characteristic “ace-of-spades” left ventricular cavity, confirming the diagnosis of apical hypertrophic cardiomyopathy. The coronary arteries were free of obstructive disease.

This case demonstrates the classical electrocardiographic, echocardiographic, and ventriculographic features of apical hypertrophic cardiomyopathy (frequently referred to as “Yamagushi Syndrome”). This unusual form of hypertrophic cardiomyopathy localized to the left ventricular apex was first described in Japan1 in 1976 and is particularly uncommon in the United States. The concave upward ST-segment elevation is likely secondary to repolarization changes caused by extensive myocardial hypertrophy. The patient was reassured and discharged on lipid-lowering therapy with instructions to keep a copy of his “signature” ECG in his wallet. He denied experiencing any palpitations or syncope and his family history was negative for premature sudden death. His family members were advised to seek medical attention for screening purposes.

Disclosures
None.

Reference
Figure 2. Characteristic “ace-of-spades” left ventriculogram demonstrating marked apical hypertrophy.
Contrast Echocardiography in Apical Hypertrophic Cardiomyopathy
Harun Otieno, Yoel Vivas, Darren Traub, Asha Raman and Chandra Polam

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