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LIVE THREE-DIMENSIONAL ECHOCARDIOGRAPHY OF APICAL HYPERTROPHIC CARDIOMYOPATHY

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INTRODUCTION

Apical hypertrophic cardiomyopathy (AHC) is a subtype of hypertrophic cardiomyopathy that was first described in 1976. [1] It predominantly occurs in the Japanese population, where it accounts for up to 25% of patients found to have hypertrophic cardiomyopathy, but can also occur in non-asian patients. [2] The diagnostic features of AHC include electrocardiographic findings of giant inverted T waves and a characteristic “ace of spades” morphology on left ventriculography. [3] It is clinically important to distinguish AHC from other variants of hypertrophic cardiomyopathy, as AHC carries a more benign long term prognosis. [4] Conventional two-dimensional echocardiography is typically non-diagnostic for AHC secondary to foreshortening of the left ventricular apical segment. [5] We report a case of a patient with known AHC and non-diagnostic two-dimensional echocardiography in whom diagnostic imaging of AHC was achieved with the use of live three-dimensional trans-thoracic echocardiography.

CASE REPORT

A 63 year old white male was diagnosed with apical hypertrophic cardiomyopathy during a hospital admission eight years ago. He had been admitted with symptoms of prolonged chest discomfort located in the mid chest with radiation to the left neck and left upper extremity. He had previously had symptoms of non-exertional chest discomfort during the three months prior to admission. He had also experienced symptoms of dyspnea with exertion, without symptoms of dyspnea at rest, orthopnea or paroxysmal nocturnal dyspnea. He reported occasional symptoms of palpitations and lightheadedness but no episodes of syncope or near syncope. The patient’s cardiac risk factors included a history of hypertension and hyperlipidemia, but no family history of cardiomyopathy or premature coronary heart disease.

On physical examination, the blood pressure was 124/72, the heart rate 62 and the respiratory rate 18. There was no carotid bruit. The lungs were clear to auscultation. On cardiac examination, there was a regular rhythm with a soft systolic murmur at the left sternal border. On extremity exam, there were intact peripheral pulses and no pedal edema.

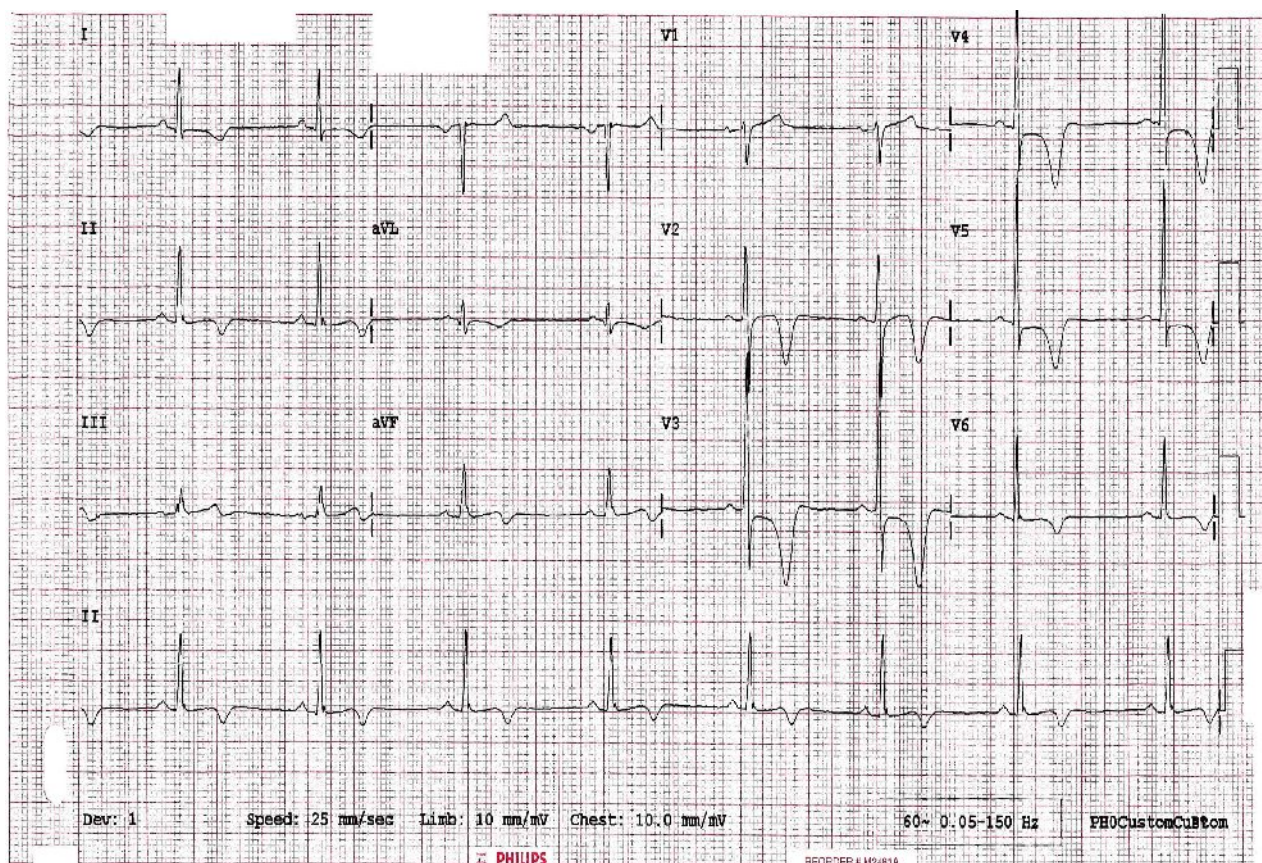


FIGURE 1

The electrocardiogram (Figure 1) demonstrated sinus rhythm, left ventricular hypertrophy and giant T wave inversions in the precordial and lateral leads. Chest radiography showed moderately enlarged cardiac size and clear lung fields. Laboratory studies included normal electrolytes, renal function and complete blood count. Serial creatine kinase (CK-MB) and troponin-I levels were within normal limits. A trans-thoracic echocardiogram demonstrated left ventricular wall thickness at upper limits of normal, normal left ventricular systolic contractility with a normal calculated ejection fraction of 65% but inability to fully visualize the left ventricular apical endocardium. Mild mitral and tricuspid insufficiency was detected on Doppler examination.

Diagnostic cardiac catheterization was subsequently performed. The left ventricular pressure was 128/9 with no intra-ventricular or trans-aortic systolic gradient.



FIGURE 2

Left ventriculography (Figure 2) showed normal left ventricular contractility and an end-systolic “ace of spades” appearance. Coronary arteriography demonstrated no evidence of significant coronary disease.

The patient was diagnosed with AHC and treated with a regimen which included a beta blocker agent (Propranolol), a calcium channel blocking agent (Verapamil) and Aspirin. He was subsequently able to resume full activities without symptoms of exertional chest pain or dyspnea, and without further

symptoms of palpitations or lightheadedness. A recent electrocardiogram was unchanged, with continued presence of giant inverted T waves.

Recently, trans-thoracic echocardiography was performed, including live three-dimensional assessment of the left ventricle. A Philips iE33 ultrasound system (Bothell WA, USA) with an xMATRIX x3-1 array probe for cine-loop acquisition and *Cardiac 3D Quantification Advanced* for processing three-dimensional images.

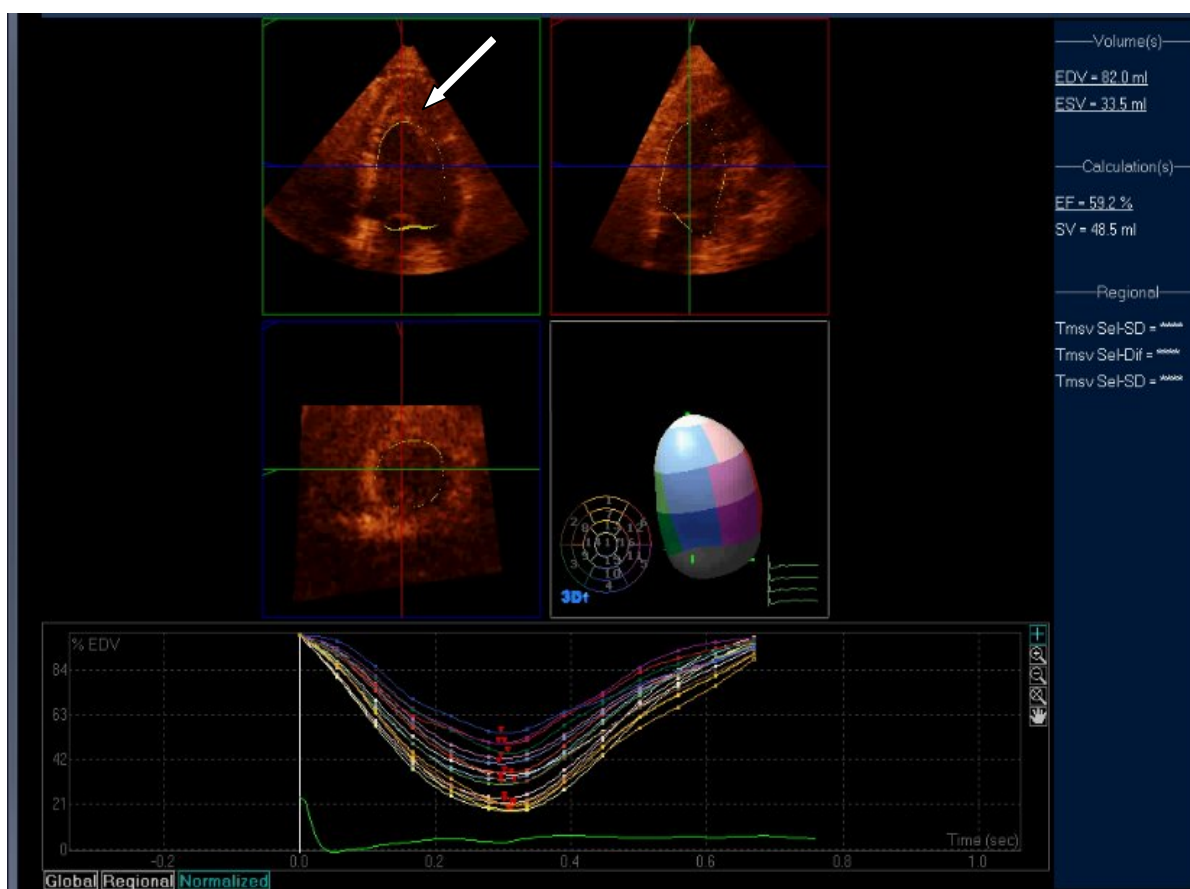


FIGURE 3

The acquired image demonstrated significant isolated apical hypertrophy consistent with AHC (Figure 3, arrow).

DISCUSSION

Conventional two-dimensional trans-thoracic echocardiography is often an initial examination performed for evaluation of patients with electrocardiographic abnormalities suggestive of AHC. However, the left ventricular apex is typically difficult to visualize with two-dimensional echocardiography secondary to foreshortening. [5] Cardiac magnetic resonance imaging has been found to be an accurate alternative diagnostic modality, [6] but is more costly and not widely available. The use of an intravenous microbubble contrast agent in conjunction with two-dimensional echocardiography may improve the accuracy of AHC diagnosis, [7] but the safety of intravenous contrast agents has recently been questioned. [8]

Live three-dimensional echocardiography has been shown to accurately assess left ventricular volumes and function. [9] A previous report of live three-dimensional echocardiography in a patient with AHC utilized an earlier generation of transducer and required intravenous contrast to achieve diagnostic images. In our patient, live three-dimensional non-contrast echocardiographic images utilizing a more advanced transducer was diagnostic for AHC.

The capability of performing live three-dimensional echocardiography is becoming increasingly available as newer transducer technology and three-dimensional software are incorporated into echocardiography systems. Our case suggests that routine live three-dimensional imaging of the left ventricular apex in conjunction with standard two-dimensional trans-thoracic echocardiography may improve the diagnostic accuracy for the AHC variant of hypertrophic cardiomyopathy.

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