APICAL HYPERTROPHIC CARDIOMYOPATHY IN AN INDIAN MALE

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SYNOPSIS

A case of apical hypertrophic cardiomyopathy occurring in a 53 year-old Indian man is described. So far only 39 cases have been reported in the english literature to the best of our knowledge. This case is the first reported in a non-Japanese and outside of Japan, suggesting that this condition may be encountered in future in other parts of the world.

INTRODUCTION

The subvariety of non obstructive hypertrophic cardiomyopathy called "apical hypertrophic cardiomyopathy" (AHC) was first described in Japan in 1976 (1, 2). So far only 39 cases have been reported in the english literature and all are from Japan. In this paper, we describe another case occurring in a middle-aged Indian man.

CASE REPORT

The patient was a 53 year-old Indian man who was born in Peninsular Malaysia. His parents were of Indian ethnic origin and had migrated to Malaysia from Sri Lanka 40 years ago. He complained of typical angina pectoris each time following severe physical exertion for 6 months prior to being referred to us for clinical evaluation. There was no previous history of hypertension or diabetes mellitus and he led an essentially sedentary life.

Clinical examination revealed an obese middle-aged Indian male. Examination of the heart was essentially normal except for an apical fourth heart sound. The blood pressure was 130/84 mmHg. The chest X-ray showed an enlarged heart (cardio-thoracic ratio 55%). The resting 12-lead electro-cardiogram showed a normal frontal plane axis, bifid P waves, tall R wave in lead V5 (36 mm) and giant T wave inversion in multiple leads, but especially in leads V2 (16 mm), V3 (15 mm) and V4 (13 mm). (Fig. 1) A clinical diagnosis of hypertrophic cardiomyopathy was made but M-Mode echocardiography surprisingly showed none of the hallmark features of this condition. The interventricular septum just below the mitral valve measured 11 mm and the posterior wall of the left ventricle measured 10 mm. The mitral valve was normal. An attempt was made to examine the apex of the left ventricle. However, this aspect of the examination was technically difficult and the tracings obtained were suboptimal. A treadmill exercise stress test was done using Bruce's protocol and a modified bipolar lead at V5 position. The patient completed 5 stages before complaining of chest pain. At that time, the ST segment was noticed to be 8 mm depressed (Fig. 2).

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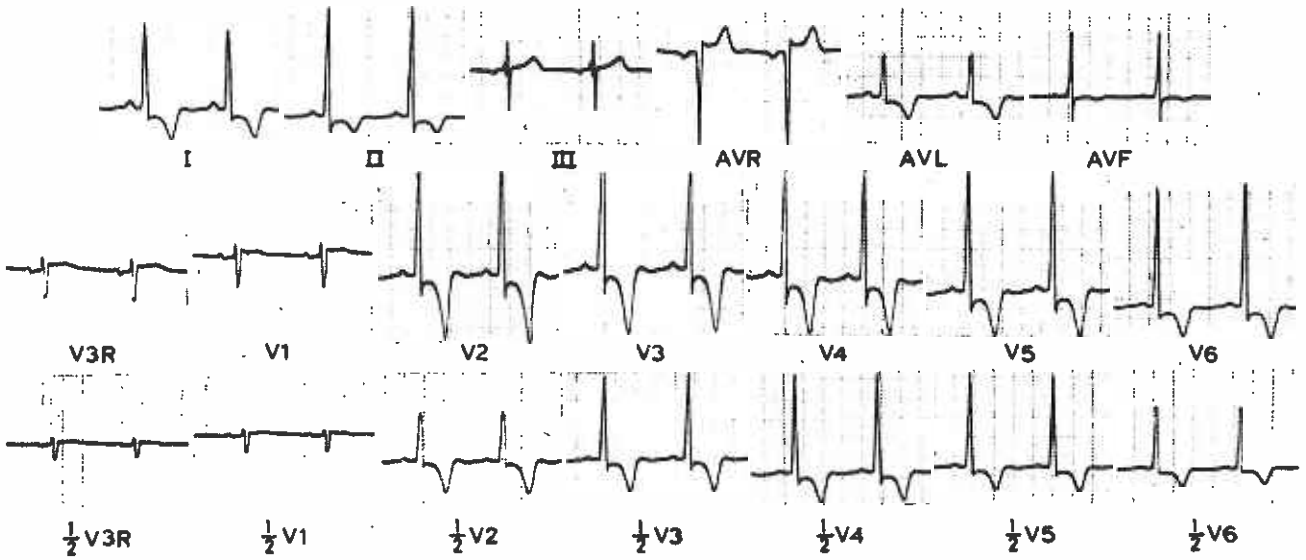


Figure 1 Resting electrocardiogram showing widespread giant T wave inversion and high QRS voltage.

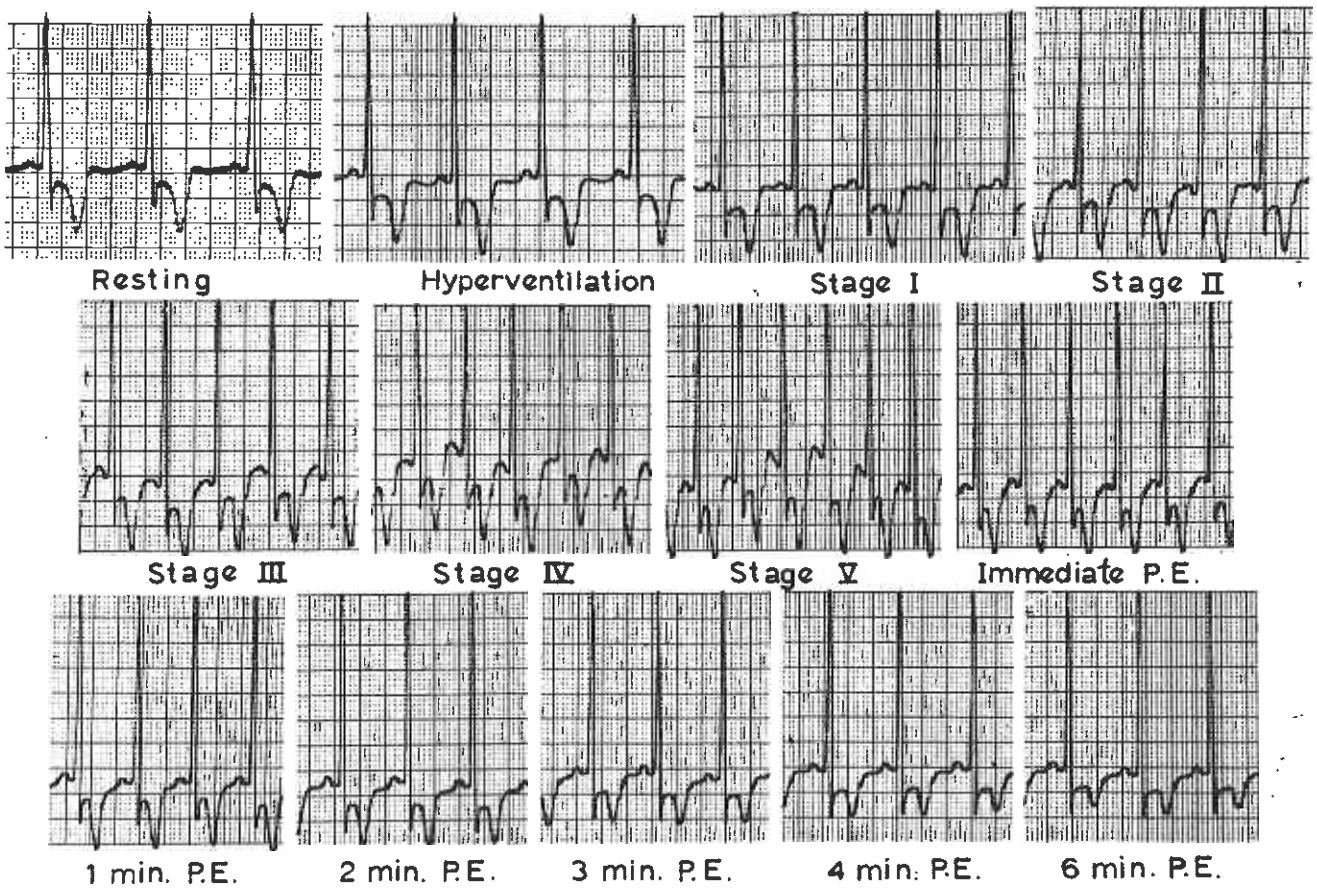


Figure 2 Treadmill exercise stress test showing 8 mm ST segment depression at Stage V of the Bruce's protocol. P.E. = post exercise.

Right and left heart catheterization was then performed using the percutaneous technique. The haemodynamic data is shown in Table 1. The left ventricular end-diastolic pressure was elevated at 20 mmHg, but no gradient was obtained (even after a post-ectopic ventricular beat) in the outflow tract of the left ventricle or across the aortic valve. Biplane left ventricular angiography was done in the 30 degrees right anterior oblique and 60 degrees left anterior oblique position. No mitral regurgitation was seen. The shape of the left ventricle in end-diastole in the right anterior oblique position exhibited a peculiar configuration resembling the spade on a playing card, due to apical obliteration (Fig. 3). In end-systole, the ventricle showed vigorous symmetrical contraction, maximal at the apex. (Fig. 4). These angiographic findings were identical to those described for the sub-variety of non-obstructive hypertrophic cardiomyopathy which Yamaguchi and co-workers termed "apical hypertrophic cardiomyopathy" in 1979 (3). Selective coronary angiography using the Judkin's technique showed widely patent right and left coronary arteries. (Figs. 5 and 6).

TABLE 1

Haemodynamic Findings

	Pressures in mm Hg
Right atrium	8 (mean)
Right ventricle	40/4
Pulmonary artery	40/15, 22 (mean)
Pulmonary capillary wedge	15 (mean)
Aorta (root)	120/70, 80 (mean)
Left ventricle	124/20

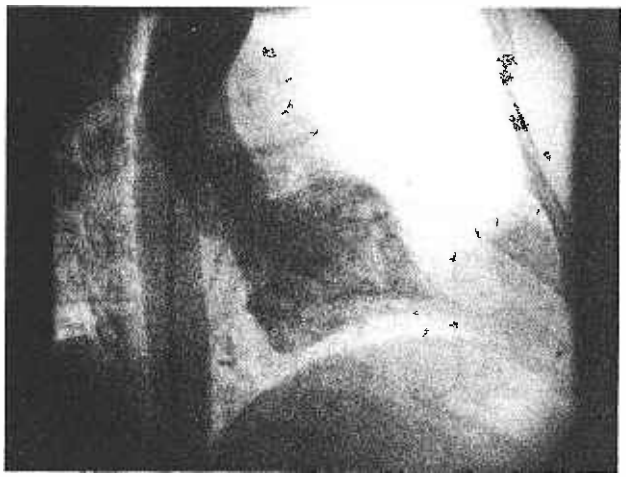


Figure 3 Left ventricular angiogram in the 30 degrees right anterior oblique position showing a unique spade shaped configuration of the left ventricular cavity in end-diastole.

The patient was started on oral propranolol because of his angina pectoris. A review of his family history revealed that both his parents died in their old age of unknown causes. He has 4 younger brothers (ages ranging from 32 years to 46 years old) all of whom are outside Singapore. None has any cardiovascular problems as far as the patient is aware. He is married to a Chinese woman and all his 3 sons, ages ranging from 11 to 20 years, are normal with respect to clinical examination, chest X-ray, resting electrocardiogram and echocardiography.

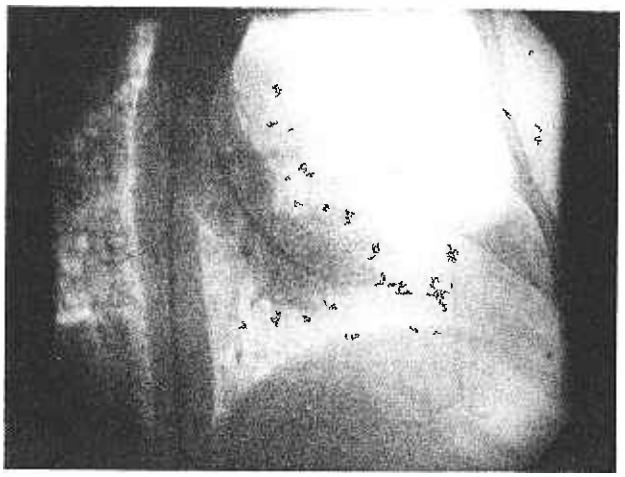
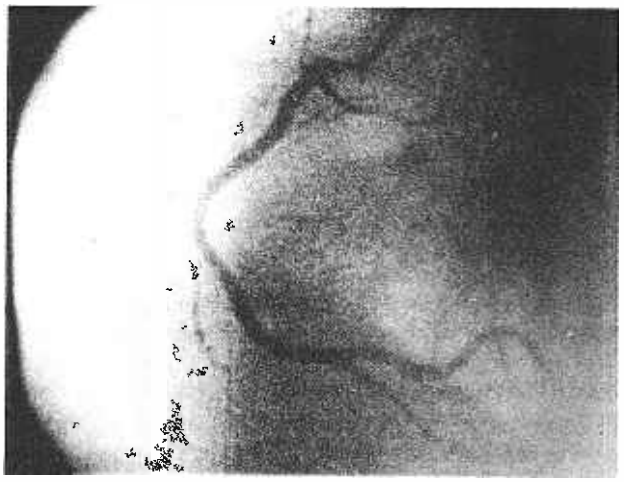
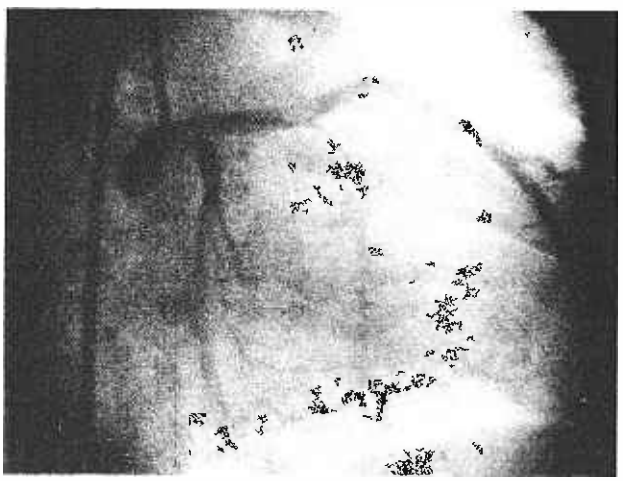


Figure 4 Left ventricular angiogram in the 30 degrees right anterior oblique position in end-systole showing vigorous symmetric contraction of the left ventricle, maximally at the apex.





Figures 5 & 6 Right and left coronary angiogram showing normal coronary arteries.

DISCUSSION

Hypertrophic cardiomyopathy, with or without left ventricular outflow tract obstruction, is characterised by uniform asymmetrical septal hypertrophy and has

been the subject of numerous publications and reviews in the last two decades (4-8). In 1967, Falicov and co-workers described two patients presenting with a subvariety of hypertrophic cardiomyopathy where the obstruction was in the mid-ventricular area rather than at the junctions of the inflow and outflow tracts of the left ventricle (9).

In 1976, Yamaguchi and co-workers delineated another variety of hypertrophic cardiomyopathy (1). In this condition which they termed apical hypertrophic cardiomyopathy, the hypertrophy primarily involves the apex of the left ventricle. Apical hypertrophic cardiomyopathy occurs predominantly in middle-aged males and is characterised by giant T wave inversion and high QRS voltage in the resting electrocardiogram and a unique spade-like configuration of the left ventricle seen in the right anterior oblique angiogram in end-diastole, due to apical cavity obliteration. In a subsequent publication, Yamaguchi and co-workers reported on a consecutive series of 30 patients with AHC and concluded that this sub-variety of non-obstructive hypertrophic cardiomyopathy appeared to be fairly common in Japan (3).

The presentation of our patient is identical to the description of the disease as reported by Yamaguchi and co-workers in 1979 (3). Twenty nine out of thirty of their patients were males and their ages ranged from 3 to 60 years old. Our patient fits into this profile with respect to sex and age. He complained of classical angina pectoris which was present in twenty percent of Yamaguchi's patients. A characteristic electrocardiographic finding in AHC is giant T wave inversion and high QRS voltage which is also demonstrated in our patient. None of Yamaguchi's thirty patients or Sakamoto's nine patients had positive results on a double Master's two step test or treadmill exercise stress test, respectively (2, 9). However, in our patient, marked ST segment depression was seen during treadmill exercise stress testing.

M mode echocardiography has been shown to be extremely useful for the diagnosis of the usual type of hypertrophic cardiomyopathy (10). Because the whole septum is markedly and uniformly thickened in this condition, asymmetric septal hypertrophy is easily demonstrated. In patients with AHC however, asymmetric septal hypertrophy and a left ventricular outflow tract gradient are both absent because the hypertrophy is confined only to the apex of the left ventricle. In the study by Yamaguchi and co-workers (3) and Sakamoto and co-workers (2), examination of the left ventricular apex, using both M-mode and two dimensional echocardiography showed marked increased in both septal and posterior wall thickness in this area of the heart. Attempts at echocardiographic examination of the left ventricular apex in our patient were unsuccessful.

The angiographic findings of AHC are highly characteristic. Due to the marked apical hypertrophy, the right anterior oblique ventricular angiogram seen in end-diastole looks like a spade on a playing card (3) and this is clearly illustrated in our patient. (Fig. 3).

The actual aetiology of AHC is presently unknown. The large preponderance of men seen with this condition suggests a sex-linked recessive transmission. In one of the thirty patients reported by Yamaguchi and co-workers, biopsy of the hypertrophic apical wall obtained during coronary artery bypass surgery revealed findings of myofibrillar disarray and the presence of degeneration of muscle cells on light microscopic examination (3). These histological findings are compatible with hypertrophic cardiomyopathy and support the contention that AHC is a sub-variety of non-obstructive hypertrophic cardiomyopathy.

So far, only 39 cases of AHC have been reported in the english literature as far as we are aware and all are from Japan (2, 3). Our patient is the first case of AHC described in a non-Japanese and suggests that this condition may be encountered in future in other parts of the world.

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