

Case Report

Apical Hypertrophic Cardiomyopathy and Persistent Thebesian Circulation

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INTRODUCTION

We report a unique case of apical hypertrophic cardiomyopathy in conjunction with persistent Thebesian circulation (TC) of both coronary arteries. The pathophysiology and clinical implications are highlighted with a literature review. Apical hypertrophic cardiomyopathy (AHC) is an unusual form of hypertrophic cardiomyopathy (HCM). TC refers to an anomalous vascular network that allows coronary arteries to drain directly into the heart chambers.

CASE REPORT

A 52-year-old Latin female was referred for coronary angiography because of new onset chest pain on exertion, associated with shortness of breath and diaphoresis. The patient had no significant prior cardiac risk factors other than being postmenopausal and a heavy smoker for many years. Physical examination was negative. The electrocardiogram showed voltage criteria for left ventricular hypertrophy, ST segment depression suggestive of subendocardial injury and pronounced T-wave inversion (Fig. 1). Subsequently, three sets of cardiac enzymes were drawn with no evidence of myocardial damage.

Two-dimensional echocardiography showed apical hypertrophy (15 mm) in all standard views. Coronary angiography revealed normal sized arteries with no obstructive lesions, however, the coronary arteries drained directly into the left ventricle from distal segments of the left anterior descending artery, left circumflex artery, and right coronary artery (Fig. 2 and 3). Left ventriculography in the right anterior oblique projection during end diastole revealed a spade-like configuration (Fig. 4), with apical end systolic obliteration. Nuclear imaging scanning revealed minimal perfusion defect at the infero-apical region.

The patient was subsequently discharged with the diagnosis of apical hypertrophic cardiomyopathy (AHC) and persistent Thebesian circulation (TC).

The initial treatment included B-adrenergic blocking agents and risk factor modification.

DISCUSSION

AHC was described initially in Japan in 1976^[1]. Further investigations have subdivided this entity into the Japanese form, in which the hypertrophy is located mainly at the apex, and the non-Japanese form, in which the hypertrophy extends under the level of the chordae tendineae^[1]. The first reports described a typical angiographic finding at the end of diastole, in which the left ventricular cavity appeared as an "ace of spades", along with characteristic ECG changes including giant negative T-waves^[2]. This form of hypertrophic cardiomyopathy (HCM) accounts for 25% of Japanese cases vs. 2% in Western populations^[3]. The morphological and clinical differences observed between the two forms are possibly related to genetic, racial, and environmental factors^[4].

Kereiakes et al., described the first reported case of apical hypertrophy in the US in 1982^[5]. This was only the third female described, consistent with prior data demonstrating a 93% male preponderance suggestive of a sex-linked recessive mode of transmission^[5]. Barbosa et al,^[3] studied 14 patients and found a higher incidence of females in their study population and less prominent T-wave inversions than the classic Japanese form.

Various studies indicate a mean age of 44-47 years. Mass electrocardiographic screening in the Japanese population revealed an increasing incidence with age since there were no cases reported in persons < 15 years of age^[6].

The other entity described involves the presence of TC. Over the years since Thebesius initially described it in the 18th century, much of the discussion has focused on the etiologic factor that predisposes to such abnormality. In the original paper, Thebesius described small branches of veins that empty their blood directly into the heart cavities. Later on, Wearn et al.^[7] described vessels

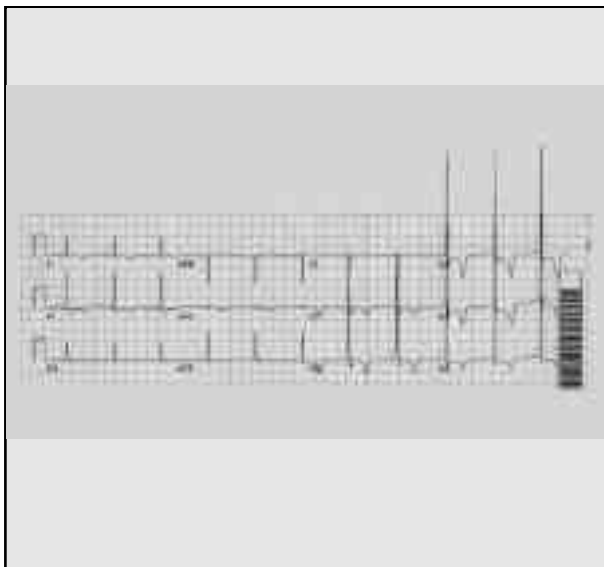


Fig. 1: Typical electrocardiographic features of apical hypertrophic cardiomyopathy including voltage criteria for left ventricular hypertrophy and deep T wave inversion.



Fig. 2: Right coronary angiogram in anterior-posterior projection showing contrast material filling the left ventricular cavity.

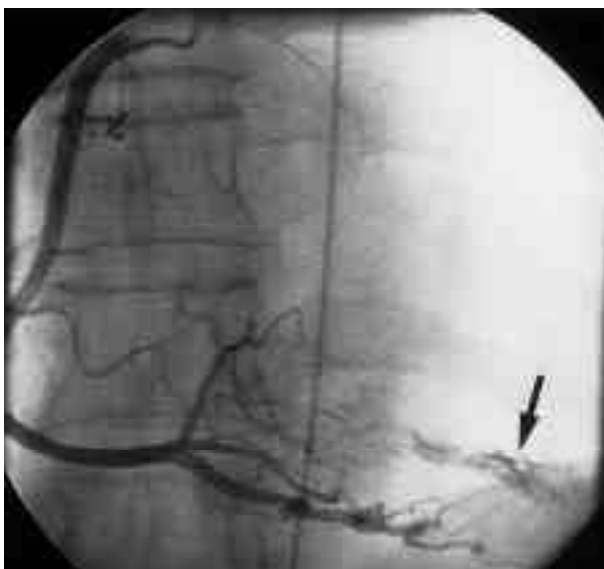


Fig. 3: Left coronary angiogram, in the right anterior oblique projection, showing left ventricular cavity opacification by contrast.



Fig. 4: End diastolic-left ventriculogram, in the right anterior oblique projection, showing spadelike configuration.

that had the histological appearance of veins, which drain arterial blood to the heart cavities from branch arteries. These connections have also been referred to as arterioluminal vessels^[7-9]. Finally, these are vessels that collect blood from the capillaries and drain directly into the heart cavities. In the myocardium, there is also a richly developed network of irregular fissures regarded as myocardial sinusoids, or small intertrabecular spaces. The vessels emptying blood directly to the heart cavities may evacuate their contents to the myocardial sinusoids or to the intertrabecular spaces^[10]. The initial thought on the development or expression of these vessels was merely myocardial hypoxemia, since morphological and pathophysiological investigations have shown convincing evidence that

anastomotic vessels are developed and widened and new capillaries built up on a hypoxemic myocardium^[10]. However, these vessels have also been present in hearts of normal subjects, as well as infants, in whom no evident cardiac pathology was encountered, leaving the etiology in doubt^[11].

Taylor and Taylor^[12], in a study on embryonic, fetal, infant, and adult human hearts, suggested that the heart is composed of simple repeats, or triplets, of conducting, valvar, and vascular tissues. Nodal conducting tissue dominates right-heart structure while non-nodal conducting tissue fashions the shorter and plainer left ventricle. They hypothesized that radial expansion of the trabecular-sinusoidal component of each triplet away from nodal tissues located in the lesser curve,

gives the mature heart its greater curvature. In this model, the septal junction forms as nodal conducting tissue bulges into the bloodstream, dragging with it its own primary nutrient supplies in the form of intertrabecular spaces and sinusoids. These findings support the view that although the heart is indeed an external pump, it is primarily constructed for internal pumping^[12]. Moreover, Ardehali et al showed in 1995 that although as much as two-thirds of retrograde cardioplegia is shunted through thebesian and arterio-sinusoidal channels into the ventricular cavities, some has traversed capillary beds and, therefore has some nutritive properties. The total nutritive fraction in their study was approximately 55%^[13].

In our case, the reason for admission and further workup was chest pain. In both of these entities, the cause of chest pain was unclear. In apical hypertrophy, the postulated origin of chest pain is a substantial increase in muscular mass relative to standard coronary blood supply, resulting in a demand/supply mismatch. Moreover, diminished coronary vasodilatory reserve has been shown in this condition^[14,15]. Patients with persistent TC on the other hand, may experience chest pain secondary to a steal phenomenon from diversion of blood into a low resistance channel especially under conditions of stress or exercise. This mechanism has been supported by the demonstration of perfusion defects during nuclear imaging scanning in the absence of angiographic evidence of coronary artery disease^[16].

The prognosis of patients with AHC is benign relative to other forms of HCM^[6]. There are only three prior cases of both apical hypertrophy and TC combined described in the medical literature, however, none of these involved all major coronary arteries^[17-19].

In summary, our case is unique due to the coexistence of two rare cardiac conditions; apical hypertrophy and persistent TC of both coronary arteries. This poses a greater challenge in the management of symptoms, which may be ascribed to either anomaly, or even a combination of the two. Fortunately, despite sparse published observations of their natural history or progression due to rarity, both appear to be relatively benign. Understanding the pathophysiologic implications of both entities should enhance the medical management of patient symptoms.

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