



HYPERTROPHIC CARDIOMYOPATHY (HCM) PRESENTED AS UNSTABLE ANGINA COMPLICATED BY SERIOUS VENTRICULAR ARRHYTHMIAS CASE REPORT AND REVIEW LITERATURE

Lusyun Kumar Yadav* and Jin li Jun

Department of Cardiology, Clinical Medical College, Yangtze University, Jingzhou, Hubei, 434023, P.R. China

ABSTRACT

Most patients with hypertrophic cardiomyopathy (HCM) have asymmetric septal hypertrophy and among them, 25% present dynamic subaortic obstruction. Apical HCM is unusual and mid-ventricular HCM is the most infrequent presentation, but both variants may be associated to an apical aneurysm. An even more rare presentation is the coexistence mid-ventricular and apical HCM. This case is a combination of obstructive HCM with mid-ventricular HCM. Till date there is not any literature review.

Case presentation: The patient is a 66 year-old female who presents a combination of septal asymmetric hypertrophic cardiomyopathy (HCM) and biventricular HCM, a sub aortic gradient of 60mm Hg and a midventricular gradient of 20 mm Hg. Her clinical presentation was an unstable angina in March 2014. A month after hospital discharge, the electrocardiogram (ECG) showed a right bundle branch block (RBBB) with no Q waves or ST segment elevation. Coronary angiography revealed normal coronary arteries, left ventricular hypertrophy and moderate apical aneurysm.

Conclusion: The case is a rare example of an asymptomatic patient with sub aortic and mid-ventricular hypertrophic cardiomyopathy, who presents as normal coronary arteries with moderate apical aneurysm.

Key features: Hypertrophic cardiomyopathy, Unstable Angina, Apical aneurysm

INTRODUCTION

HCM has a prevalence of 0.2 % (1 in 500) in the general population and presents with marked clinical polymorphism, given by the extent and degree of hypertrophy of the myocardium involved and by the location and magnitude of the intra-ventricular gradient. Doppler-echocardiography plays an important role in the diagnosis, identifying the distribution of hypertrophy and the site of intraventricular obstruction.

Most patients with HCM (95%) have asymmetric septal hypertrophy and among them, 25% present with dynamic sub aortic obstruction [2]. Apical HCM is unusual and mid-ventricular HCM is the most infrequent presentation, but both variants may be associated to an apical aneurysm. An even more rare presentation is that of coexistent mid-ventricular and apical HCM. This case represents a combination of asymmetric septal and mid-ventricular HCM.

Case presentation:

The patient is a 67 year-old female, with a family history of HCM and no coronary risk factors, who in march 2014 was admitted to the coronary care unit at clinical hospital of Yangtze University due to an episode of oppressive and prolonged chest pain. Her ECG at admission showed sinus rhythm, left ventricular hypertrophy and ST segment elevation in leads V4 to V6, which later normalized, without developing Q waves.

A fourth sound and a 4/6 systolic murmur along the mesocardium were noted in the physical exam. Twelve hours after the beginning of chest pain.

Laboratory data are as follow:

Myocardium Enzymogram

Time	CPK	LDH	CK-MB
At presentation	115U/L	325U/L	52U/L
After 12hrs	3.090U/l	305U/L	3.43U/l

Blood routine examination

Date	WBC	Neutrophil	L	RBC	HB	PLT
On admission day	12.0m g/dl	74%	23 %	5.4 T/L	17. 0 gm /dl	145 G/L

Renal function test:

Date	BUN	Cr	UA
On admission day	9.35mmol/ L	128umol/ L	350umol/ L

The ECG at hospital discharge showed sinus rhythm, right bundle branch block (RBBB) with no Q waves, right ventricular hypertrophy and/or posterior necrosis (Figure 1B). The vector cardiogram was consistent with apical necrosis (Figure 1C). The chest X-ray showed a mild increase in cardiothoracic index and the lower left arch. The 24-hour Holter showed sinus rhythm, permanent RBBB and 234 monomorphic premature ventricular contractions (PVC's), without complex arrhythmias.

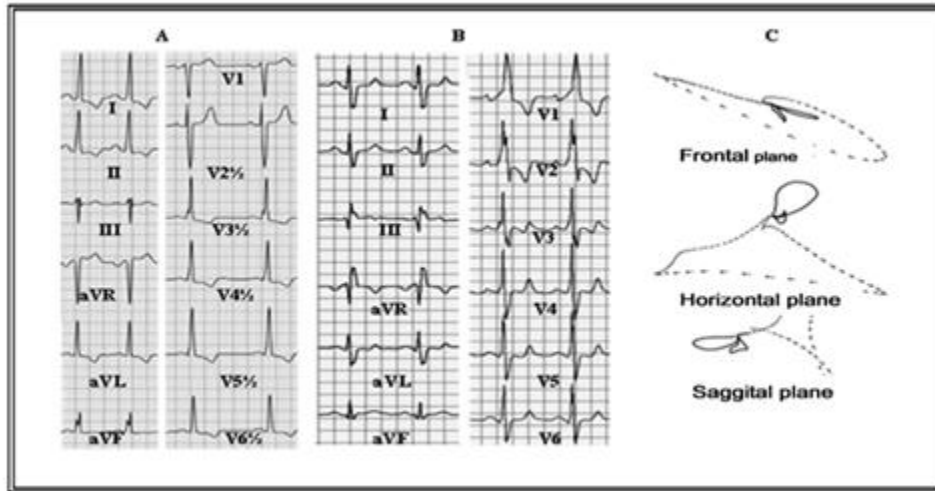


Figure 1 A: The electrocardiogram (ECG) shows sinus rhythm, biatrial enlargement, and left ventricular hypertrophy. **A:** The electrocardiogram (ECG) shows sinus rhythm, biatrial enlargement, and left ventricular hypertrophy. **B and C:** ECG and vectorcardiogram (VCG) one month after acute myocardial infarction. The ECG (B) shows right bundle branch block, right ventricular hypertrophy and/or posterior necrosis. The VCG (C) shows biatrial enlargement with prevalence of the left atrium left ventricular hypertrophic and apical necrosis.

The Doppler-echocardiogram showed a combination of asymmetric septal and mid-ventricular HCM (Figure 2), and the anterobasal septum measured 21 mm. There was systolic anterior motion of the mitral valve with a sub aortic gradient of 65 mm Hg (Figure 3), anteromedial septal thickness was 19 mm, there was a mid-ventricular systolic gradient of 20 mm Hg. Ejection fraction was 55%. The presence of a restrictive mitral flow indicated an increase in left ventricle end-diastolic pressure due to diastolic dysfunction.

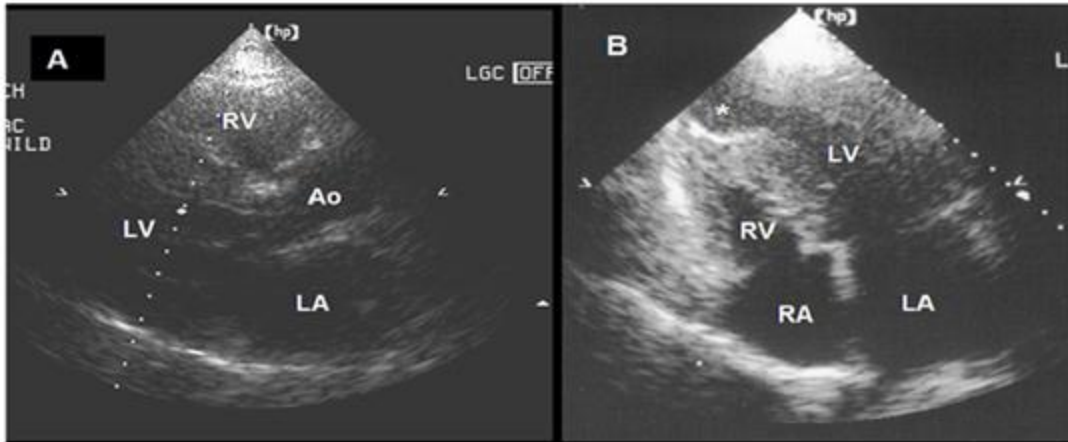


Figure 2: Transthoracic echocardiogram A: Parasternal long axis view showing asymmetric septal hypertrophic cardiomyopathy in end diastole. B: A four-chamber view showing a combination of basal and mid-ventricular hypertrophic cardiomyopathy and an apical aneurysm (*). (LA = left atrium, RA = right atrium, RV = right ventricle, LV = left ventricle, Ao = aorta).

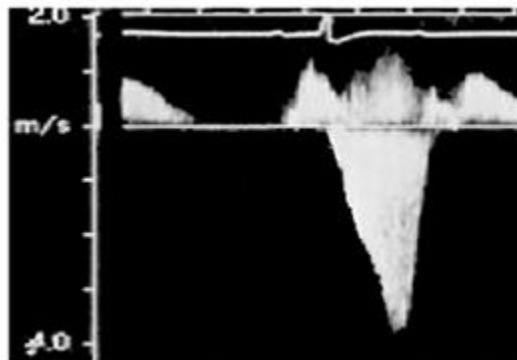


Figure 3: systolic anterior motion of the mitral valve with a subaortic gradient of 65 mm Hg, anteromedial septal thickness was 19 mm, there was a mid-ventricular systolic gradient of 20 mm Hg

Hemodynamic study revealed septal and mid-ventricular thickening (Figure3) with normal epicardial coronary arteries, and apical aneurysm. Coronary angiogram ruled out systolic myocardial bridging of the left anterior descending artery and septal perforation branches.

Laboratory tests and serology for Chagas disease were normal.

During follow-up, the patient was treated with beta-blockers and remained clinically stable.

DISCUSSION

Echocardiogram and left ventriculography of our patient showed an asymmetric septal and mid-ventricular hypertrophy (although the epicardial coronary arteries were angiographically normal), a moderate subaortic gradient and a mild mid-ventricular gradient. Apical infarction is not a rare finding in patients with mid-ventricular HCM. Several reports have shown an association of mid-ventricular and apical HCM [3,4,5]. The mechanisms responsible for an apical infarction are not completely understood, but such patients are known to have normal epicardial coronary arteries, usually present with microvascular dysfunction and a decrease in coronary reserve, attributed to narrowing of the small intramyocardial coronary arteries. The combination of an increase in oxygen demand due to increased ventricular thickness and the decrease in oxygen supply due to a decrease in the capillary network predispose to ischemia. Additionally, the apical chamber is subject to greater and sustained systolic stress due to the high intraventricular gradient. In our patient, in the presence of normal epicardial coronary arteries and in the absence of coronary spasm, could be due to microvascular dysfunction and increased stress in the apical chamber, consequent to the combination of sub aortic and mid-ventricular obstruction [6]

To the best of our knowledge, no cases of combined sub-aortic and mid-ventricular obstructive HCM, associated with apical aneurysm have been described.

Treatment of symptomatic HCM is aimed at reducing the intraventricular gradient which, in most cases, is achieved with beta-blockers and calcium antagonists (diltiazem or verapamil) [12]

Cases of HCM with an apical aneurysm may be complicated by serious ventricular arrhythmias, which usually respond to antiarrhythmic drugs (amiodarone); however, when they are refractory to medical treatment, the insertion of an implantable defibrillator may be warranted [7], to prevent sudden death. In some patients with an apical aneurysm and very thin walls, the size of the aneurysm may increase rapidly, which entails a high risk of spontaneous rupture and, hence the surgical resection of the aneurysm may be required [3]. On rare occasions, a thrombus may form inside the aneurysm, which may cause embolization [8] and thus, in such cases, anticoagulation is indicated.

CONCLUSION

Several reports have shown the association between mid-ventricular and apical HCM and apical aneurysms, but this is the first case presenting a special phenotypic expression of HCM, characterized by a combination of asymmetric septal and mid-ventricular HCM that presents clinically as an unstable angina with normal coronary arteries and develops an apical aneurysm[8,12].

ACKNOWLEDGMENTS

This work has been supported by Professor Jin Li Jun. I'm very thankful for his support and help.

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