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CASE REPORT

Yamaguchi Syndrome – A Pseudoacute Coronary Syndrome of the Young: A Case Report on Apical Hypertrophic Cardiomyopathy

Alvin Oliver Payus[1], Farisha Mohd Sholeh[2], Norlaila Mustafa[2]

[1]*Faculty of Medicine and Health Science, Universiti Malaysia Sabah (UMS), Jalan UMS, Kota Kinabalu, Sabah,*
[2]*Department of Internal Medicine, Universiti Kebangsaan Malaysia Medical Centre (UKMMC), Jalan Yaacob Latif, Cheras, Kuala Lumpur, Malaysia*

Yamaguchi syndrome, or apical hypertrophic cardiomyopathy (ApHCM), is a nonobstructive subtype of hypertrophic cardiomyopathy which predominantly affects the left ventricular apex. Due to the nature of its presentation that mimics acute coronary syndrome and also to the unfamiliarity of the condition by some physicians, the diagnosis of ApHCM is frequently missed or delayed. Here, we report a case of a young male who presented with chest pain and breathlessness. His cardiac enzyme was normal and electrocardiogram showed giant negative T-waves. He was treated as unstable angina and was then subjected to a line of diagnostic procedures including coronary angiogram before he subsequently underwent ventriculogram which reveal a characteristic "spade-like" configuration over the left ventricle, in keeping with the diagnosis of ApHCM. The purpose of this case report is to emphasize the importance of keeping ApHCM as one of the differential diagnoses in a young patient presented with chest pain.

Key words: Yamaguchi syndrome, hypertrophic cardiomyopathy, acute coronary syndrome, unstable angina, giant-negative T-wave

INTRODUCTION

Apical hypertrophic cardiomyopathy (ApHCM) is a nonobstructive and relatively rare subtype of hypertrophic cardiomyopathy (HCM). It has a wide range of clinical manifestation, but commonly presented as chest pain, dyspnea, palpitation, syncope, and heart failure symptoms.[1] Rarely, it can also present as a sudden cardiac death. The management of ApHCM is mainly symptomatic using medications such as beta-blockers or nondihydropyridine calcium-channel blockers to control the heart rate, and also angiotensin-converting enzyme inhibitors (ACEIs) to reduce the left ventricle afterload and adverse cardiac remodeling. In a patient with high risk of sudden cardiac death, implantable cardioverted-defibrillator may be warranted. ApHCM is usually benign as compared to other types of HCM, and this subtype of HCM will be the center of discussion in this case report.

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Corresponding Author: Dr. Alvin Oliver Payus, Department of Medicine Based, Faculty of Medicine and Health Science, Universiti Malaysia Sabah, Jalan UMS, 88400 Kota Kinabalu, Sabah, Malaysia. Tel: +6018-8703503; Fax: +6088-320 000. E-mail: dralvinpayus@ums.edu.my

CASE REPORT

A 29-year-old male who is a noncigarette smoker and works as an office clerk presented with a sudden onset of chest pain, while he was sitting down in the office. He described the pain was constant, stabbing in nature, and radiates to the left arm. He was also slightly breathless and sweating profusely at that time. The pain persisted for more than 30 min and hence brought him the emergency department. Upon arrival to the emergency department, the pain resolved after a single sublingual glyceryl trinitrate. His vital signs were stable, and physical examination reveals no abnormal finding. His preliminary blood investigation reveals no elevated cardiac enzymes even in the repeated sample. Serial electrocardiogram (ECG) showed evidence of left ventricular hypertrophy and deep T-wave inversion over the

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inferior and anterolateral leads, but otherwise, there was no Q-wave or ST-segment changes [Figure 1]. Further history taking noted that the patient's father has passed away at the age of 40 due to heart attack a few years back. The patient was treated as unstable angina at that time and was discharged with single antiplatelet medication. He was then subjected to exercise stress test a few months later, and the result was inconclusive. He also had echocardiography (ECHO) done, and it shows left ventricular hypertrophy with mild dilatation of the left atrium, but otherwise, no left ventricular outflow obstruction [Figure 2]. He then underwent coronary angiogram which reveals patent coronary artery [Figure 3], and also ventriculogram, which shows a "spade-like shape" left ventricular filling, in keeping with HCM (ApHCM) [Figure 4]. The patient was diagnosed as ApHCM and was started on oral bisoprolol 2.5 mg daily to control the heart rate and also oral perindopril 2 mg daily to reduce the left ventricle afterload.

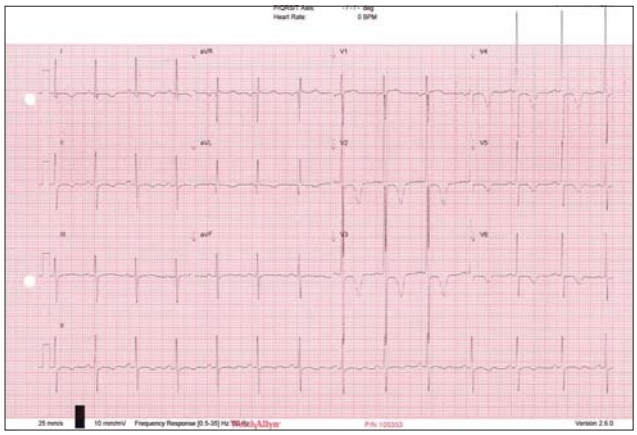

Figure 1: Electrocardiogram showing sinus rhythm with voltage criteria for left ventricular hypertrophy as well as diffuse T-wave inversions over the inferior and anterolateral leads

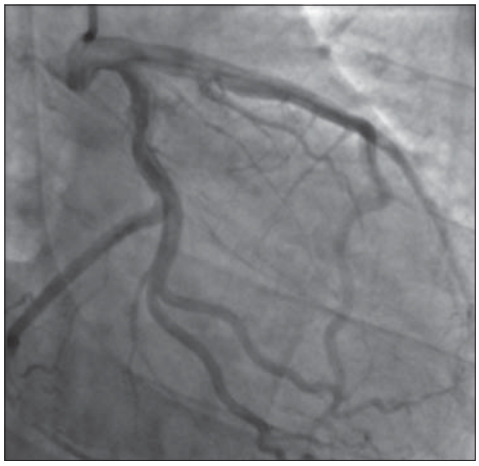

Figure 3: Coronary angiography showing patent coronary arteries

He was also planned for elective insertion of dual-chamber implantable cardioverter-defibrillator (ICD) in 1-month time. The patient has three siblings in which all will be screen for the possibility of having the same condition.

DISCUSSION

ApHCM represents a relatively small proportion of all cases of HCM where it is nonobstructive and characterized by the localized hypertrophy over the left ventricular apex. It was first described in detail by Yamaguchi in the year 1979, and thus the other given name for ApHCM is Yamaguchi syndrome. It has a variable clinical presentation which ranges from being asymptomatic to sudden cardiac death. Moreover, the commonly presenting complaints are chest pain, breathlessness, palpitations, syncope, and also heart failure symptoms.[1]

The diagnosis of ApHCM is made by the presence of typical findings over the ECG, ECHO, and ventriculogram. The typical ECG changes in ApHCM are called "giant T-wave negativity" which is an inverted T-wave over the

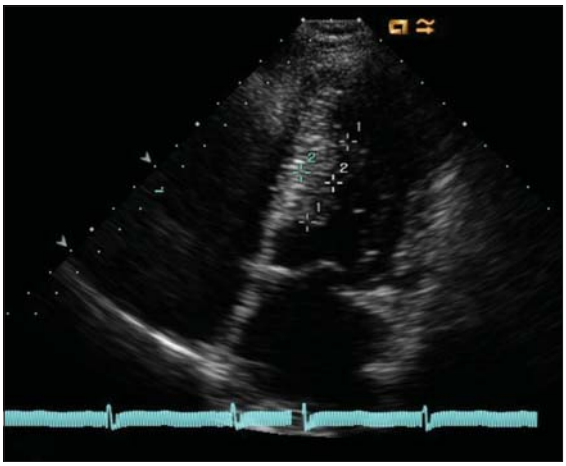

Figure 2: Two-dimensional echocardiography showing thickening of the left ventricular wall measuring 20 mm

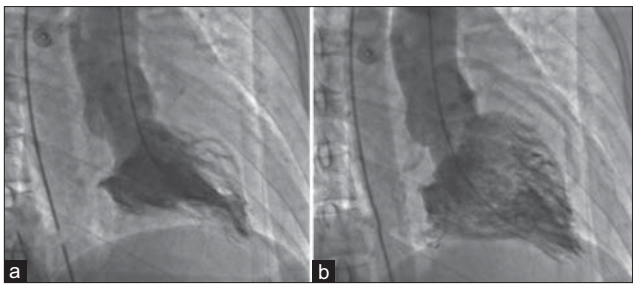

Figure 4: Ventriculogram showing left ventricle apical hypertrophy with spade-like configuration of the ventricular cavity during systole (a) and diastole (b)

precordial lead and often mistaken for acute coronary syndrome. The ECHO will reveal asymmetrical thickening of the left ventricular apex. The ventriculogram will show a pathognomonic "spade-like" configuration at end-diastole over the left ventricular cavity.[2]

The aim of treatment for ApHCM is mainly to control symptoms and prevent complications, which are done by means of medications such as beta-blockers or calcium-channel blockers to control the heart rate, and also ACEI to reduce the left ventricular afterload. Recent discovery also showed that ACEI will reduce the adverse cardiac remodeling and fibrosis in ApHCM.[3] For a patient who is persistently symptomatic or has high risk of sudden cardiac death, an ICD will be inserted. The first-degree family member of the patient will be screened for having a similar condition with genetic testing and also ECHO. In our patient, the decision for ICD implantation was made on the basis of having strong family history of sudden cardiac death where his father died due to cardiac event at the age of 40. Apart from that, his ECHO showed dilated left atrium and thickened left ventricular wall. Given all these findings and his young age, this makes him at high risk of sudden cardiac death and warrants an ICD implantation.

ApHCM leads to a few complications, namely, diastolic function impairment, left atrium dilatation that causing atrial fibrillation, apical clots formation with subsequent embolic events, ventricular wall aneurysm, arrhythmias, and myocardial infarction. Fortunately, ApHCM generally runs on benign course. There are some predictors of poor prognosis that has been identified such as young age at diagnosis, positive family history of sudden cardiac death, present of heart failure symptoms at New York Heart Association Class II and above, and others.[4]

In view of the nature of its presentation that mimics acute coronary syndrome, along with the low index of suspicion among some physicians due to unfamiliarity with the condition, the diagnosis of ApHCM is frequently missed or delayed.[5] For that reason, most of the patient will be subjected to multiple investigations repeatedly before they do a ventriculogram to arrive to the diagnosis. And although myocardial infarction is one of the morbid sequalae of ApHCM, the pathology is usually outside the coronary artery.[4] Therefore, coronary angiogram usually will reveal a normal or only mild stenosis over the coronary artery which fails to explain the symptoms and ECG changes.

CONCLUSION

The objective of this case report is to remind the reader that whenever a patient who is young and has low risk of cardiac disease presented with symptoms of acute coronary syndrome, it is very important to keep ApHCM as one of the possible causes, in order to avoid diagnostic delay.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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