Yamaguchi syndrome is a non-obstructive and relatively rare subtype of hypertrophic cardiomyopathy. It has a wide range of clinical manifestations from being asymptomatic to sudden cardiac death. Moreover, they commonly presented as chest pain, dyspnoea, palpitation, syncope, and heart failure symptoms.

The most frequent symptom is chest pain and thus apical hypertrophic cardiomyopathy can mimic the symptoms and electrocardiographic changes indicative of acute coronary syndrome. Given the time-sensitive nature and possible iatrogenic risks of reperfusion therapy, it is important to recognize apical hypertrophic cardiomyopathy as one of the differential diagnoses in a young patient presented with chest pain.

**Keywords:** Acute coronary syndrome, apical hypertrophic cardiomyopathy, Yamaguchi syndrome

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**Introduction**

Yamaguchi syndrome (apical hypertrophic cardiomyopathy) is a non-obstructive and relatively rare subtype of hypertrophic cardiomyopathy. It has a wide range of clinical manifestations from being asymptomatic to sudden cardiac death. Moreover, they commonly presented as chest pain, dyspnoea, palpitation, syncope, and heart failure symptoms.

The most frequent symptom is chest pain and thus apical hypertrophic cardiomyopathy can mimic the symptoms and electrocardiographic changes indicative of acute coronary syndrome. Given the time-sensitive nature and possible iatrogenic risks of reperfusion therapy, it is important to recognize apical hypertrophic cardiomyopathy as a potential imitator of an acute myocardial infarction.

**Case Report**

A 35-year-old man who is a non-smoker and works as a teacher in school presented with a sudden onset of chest pain, while he was sitting down in staff room. He described the pain as stabbing in nature and radiating to the left arm. The pain persisted for more than 30 minutes and hence brought him to the emergency department. His vital signs were stable, and physical examination reveals no abnormality. No significant family history. An electrocardiogram at presentation showed evidence of left ventricular hypertrophy and deep T wave inversion over the inferior and anterolateral leads. His initial blood investigations were normal. The acute coronary syndrome was the provisional diagnosis made by the attending physician and the patient was referred to our cardiac centre for coronary angiography. Transthoracic echocardiography was done at our centre and it showed asymmetric left ventricular hypertrophy localized to LV apex and no left ventricular outflow obstruction. He then underwent coronary angiography which reveals normal coronary arteries, and also a left ventriculogram, which shows a “spade-like shape” left ventricular filling. The patient was diagnosed with...
Apical hypertrophic cardiomyopathy (ApHCM) and was started on oral bisoprolol 2.5 mg once a day to control the heart rate and also oral Ramipril 2.5 mg once a day to reduce left ventricular afterload.

**Discussion**

Apical hypertrophic cardiomyopathy (ApHCM) is a form of non-obstructive hypertrophic cardiomyopathy localized to the left ventricular apex that was first described in Japanese patients with precordial deep T wave inversions referred to as giant T wave inversion in 1976.\[1\] It was first described in detail by Yamaguchi in the year 1979, and thus the other given name for ApHCM is Yamaguchi syndrome.\[1\] It has historically been considered a disease of the Asian population, where it accounts for 13% to 41% of all hypertrophic cardiomyopathies.\[2-4\]

As many as half of patients with ApHCM are mildly symptomatic or asymptomatic, and many patients have been diagnosed only when giant T wave inversions in the precordial leads are noted incidentally during electrocardiography.\[5-7\] Patients with ApHCM who are symptomatic may complain of chest pain, palpitation, dyspnoea, fatigue or syncope. Although severe clinical manifestations including sudden cardiac death, malignant arrhythmias, and apical infarction with apical aneurysm have been described, cardiovascular mortality is lower than in other variants of hypertrophic cardiomyopathies.\[8\]

Patients with ApHCM may present with symptoms mimicking of the acute coronary syndrome, as was the case with the patient presented above. In this context, the presence of angina chest pain and deep T wave inversions on ECG often prompts physicians to follow an early invasive route.

The diagnosis of ApHCM is made by the presence of typical findings over the ECG, echocardiography, and ventriculogram. The typical ECG changes in ApHCM are called “giant T wave inversions” which is an inverted T wave greater than 10 mm and a high R wave voltage in precordial leads and often mistaken for the acute coronary syndrome. The characteristic findings in transthoracic echocardiography is asymmetrical hypertrophy localized to left ventricular apex causing a “spade-like” configuration at end-diastole over the left ventricular cavity during left ventriculogram.\[9\]

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 ACE inhibitors to reduce left ventricular after-load.\[10\] For a patient who is persistently symptomatic or has a 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 echocardiography also.

Fortunately, ApHCM generally runs on a benign course. There are some predictors of poor prognosis that have been identified such as young age at diagnosis, positive family history of sudden cardiac death, presence of heart failure symptoms NYHA class 2 and above.\[5\]

**Conclusion**

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. For that reason, most of the patients will be subjected to multiple investigations repeatedly before they underwent a ventriculogram to arrive to diagnosis.

**Take home message**

Whenever a patient who is young and has low risk of coronary artery disease presented with symptoms of acute coronary syndrome along with giant T wave inversions in precordial leads, it may be important to keep ApHCM as one of the possible causes, in order to avoid diagnostic delay.
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Conflicts of interest
There are no conflicts of interest.

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