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YAMAGUCHI APICAL HYPERTROPHY SYNDROME-A CLINICIAN'S DILEMMA

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ABSTRACT
Apical hypertrophic cardiomyopathy (AHC) or Yamaguchi syndrome is a variant of hypertrophic cardiomyopathy which primarily involves left ventricular apex. In 1976 Sakamoto described a condition in Japanese patients with giant T waves as asymmetric apical hypertrophy (AAH).[1] Later it was elaborated on by Yamaguchi in 1979.[2] In this case we report a 51 year old male patient with AHC who was referred to the authors erroneously diagnosed as a case of acute coronary syndrome. AHC is a rare form of non-obstructive cardiomyopathy which has characteristic findings on electrocardiography, echocardiography and cardiac magnetic resonant imaging. Treating physicians should recognize the typical electrocardiographic changes and differentiate it from the more common and serious diagnosis of acute coronary syndrome.

KEYWORDS: Apical hypertrophy, Yamaguchi syndrome.

INTRODUCTION
AHC is a rare form of non-obstructive cardiomyopathy which has characteristic findings on electrocardiography, echocardiography and cardiac magnetic resonant imaging. It has been commonly described in Japanese patients with an incidence of 13-25% of all the cases of hypertrophic cardiomyopathy.[3] Although rare in rest of the world but now with the understanding of this disorder it is being increasingly recognized in other parts of the world.[4] It is characterized by presence of giant inverted T-waves and characteristic 'spade-like' shape of the left ventricular cavity on ventriculography in the end-diastole.[2] Early changes may not show the classical 'spade-like' pattern but may be recognized by free lateral wall hypertrophy on echocardiography in a patient who has giant T-wave inversion on electrocardiography.[1,3] This disorder has a relatively benign course with an overall survival of 95% at 15 years.[5] Less than one third patients experience cardiovascular morbidity in the form of atrial fibrillation and myocardial infarction.[5,6,7] Medical specialists should recognize the typical electrocardiographic changes and differentiate it from the more common and serious diagnosis of acute coronary syndrome, so that patient may not be subjected unnecessary invasive diagnostic procedures and mental agony.

CASE REPORT
A 52 year old man was referred by a general physician to our institute with a diagnosis of acute coronary syndrome and an advice to get an urgent coronary angiography. He had presented with palpitation and heaviness in the chest. On interviewing him again it was found that his palpitation was recurrent and present for the past 6 years. Heaviness of the chest was also recurrent and present for the last one year. He had got himself evaluated 6 months earlier and he was put on anxiolytics and proton pump inhibitors. There was little benefit and he continued to have symptoms till he got his electrocardiography done. There-after he was referred to us. There was no significant past history. The patient was a non-smoker and rarely took alcohol. There was no history of substance abuse. There was no significant family history and he was not currently taking any medications. On examination patient was well oriented and he was not distressed. His vital parameters were within normal limits with a blood pressure of 154/96 mm of Hg, pulse rate of 88 beats/minute (with no pulse deficit), respiratory rate of 14 per minute and he was clinically afebrile. General physical examination was unremarkable so was abdominal and neurological examination. Cardiovascular and respiratory system examination did not reveal any abnormality.

Electrocardiogram (ECG) revealed giant T-wave inversions in V_2, V_3, V_4, V_5 and V_6 (>10 mm) and tall QRS waves in the lateral precordial leads. QRS duration was within normal range (89 msec). Worth noting was diffuse absence of septal Q-waves. There was an ST-segment depression of >1mm seen in V_4 to V_6.

According to Sokolow Lyon criterion the ECG was suggestive of left ventricular hypertrophy ($RV_5 + SV_1$ = 49 mm).[Figure 1] These changes represent the typical pattern of apical hypertrophic cardiomyopathy (AHC). Routine laboratory investigations were normal including serum electrolytes. Cardiac specific creatinine phosphokinase (CPK-MB) and troponin-T were within normal limits.

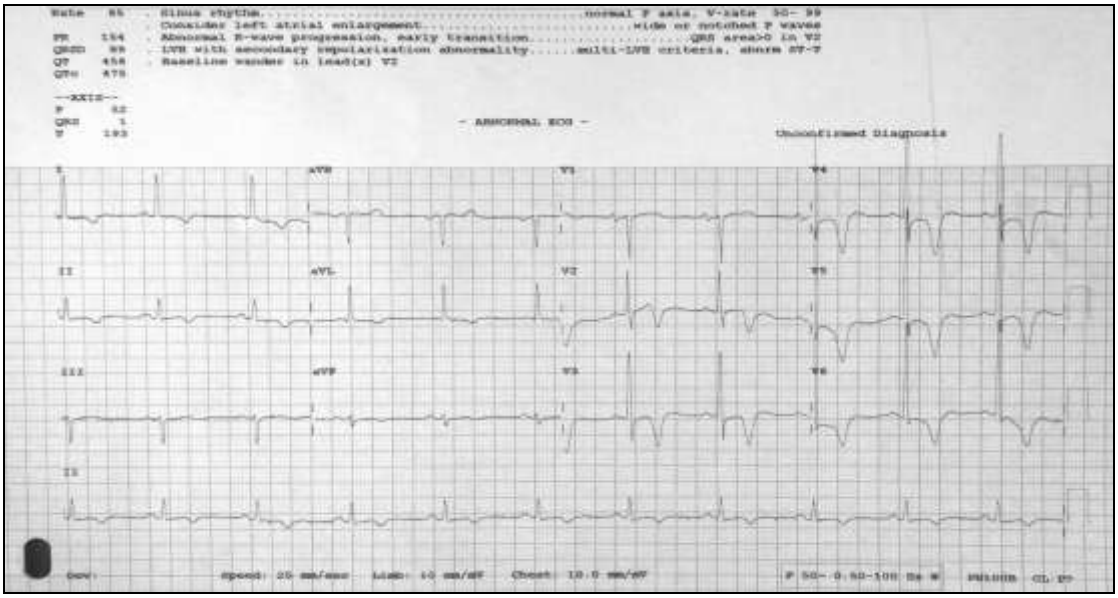

Figure 1. Characteristic ECG changes in the patient.

Chest radiography showed no variation from the normal. A two dimensional echocardiography was done immediately which revealed a situs solitus heart with no regional wall motion abnormality. There was no valvular abnormality. Ejection fraction was 73%. Slight left atrial enlargement was seen. Right ventricle and right atrium were not dilated. There was little hypertrophy of the inter-ventricular septum or of the posterior wall of the left ventricle. The hypertrophy spared lateral wall of the left ventricle except the portion near the apex which was thickened (27mm) and the ventricular cavity showed a classical spade pattern.[Figure 2]

The patient was diagnosed as a case of apical hypertrophic cardiomyopathy and was put on beta blockers. He reported a improvement in his symptoms on follow up.

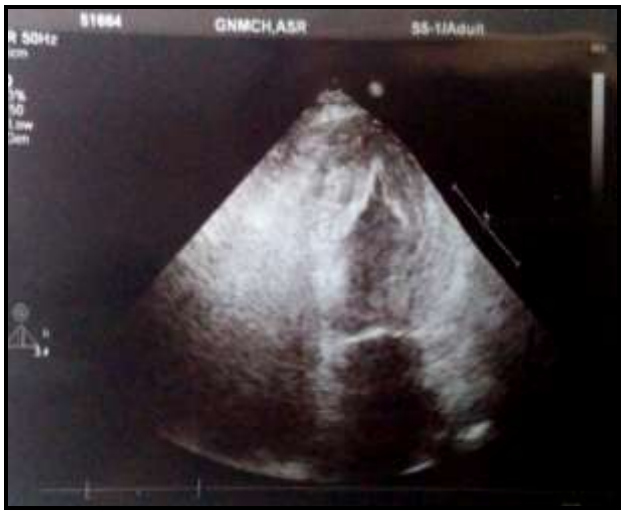

Figure 2. Echocardiograph of the patient.

DISCUSSION

Apical hypertrophic cardiomyopathy (AHC) is a relatively uncommon form of hypertrophic cardiomyopathy which was first described by Sakamoto and Yamaguchi in Japanese patients.[1,2] It is a common form of hereditary hypertrophic cardiomyopathy pattern seen in Japan accounting for upto one fourth of the patients diagnosed to be having hypertrophic cardiomyopathy.[3] Familial cases have been reported but phenotypic presentation may not be seen in all relatives. This form of hypertrophic cardiomyopathy has been rarely described in Western countries although with the characterization of the disorder it is being increasingly recognized.[4] In a relatively large retrospective study done at Toronto General Hospital 105 patients with

apical hypertrophy syndrome were recognized and there long term outcome was studied.[5] 54% patients having this form of cardiomyopathy were symptomatic at presentation. The main presenting features were chest pain (30%), palpitations (10%), dyspnoea (6%) and syncope (6%).[5,6,7] Diagnostic modalities include electrocardiography (ECG), echocardiography, ventriculography, nuclear myocardial perfusion studies and MRI. In such situations, understanding of the characteristic ECG changes of AHC can be of great value in diagnosing this uncommon condition. The electrocardiographic findings include presence of giant T-wave inversions in precordial leads (V_2 to V_6) and loss of septal Q-wave and these changes are pathognomonic for AHC. The absence of septal Q-waves is due to intra-ventricular conduction defects and be dysfunctional activation of the ventricle. The giant T-wave inversions are due to the reversal of repolarization order within the hypertrophied ventricular wall near the apex.[8] Although these ECG changes are considered pathognomnic for AHC, it is pertinent to rule out other more common causes of ST-T changes which may closely mimic this condition especially myocardial ischemia, cerebral T waves (in sub-arachnoid haemorrhage) and certain drugs like digoxin.[9] Considering these differentials cardiac echocardiography has an important role in confirmation of the diagnosis. Echocardiography, although operator dependent, is the most accepted form of evaluation of a suspected case based on ECG findings. It is the most cost effective imaging modality available to delineate the ventricular anatomy.[10,11] Ventriculography is the best in demonstrating the spade shaped ventricular cavity.[3] Cardiac MRI is now emerging as the first line diagnostic procedure for AHC.[12] Main drawbacks of these new diagnostic techniques are the lack of availability and high costs.

Ease of performing this simple study, availability, ease of interpretation and cost make ECG an indispensible instrument in diagnosing this condition which can be confirmed on echocardiography. In this case report we want to emphasize the fact that the diagnosis was clinched upon ECG findings alone who had been erroneously labeled as a case of acute coronary syndrome. A misdiagnosis is fully avoidable with an apt ECG interpretation. It is evident from this case that there is need of developing appropriate and adequate ECG interpretation skills in family physicians so that AHC may not be misconstrued as acute coronary syndrome.

REFERENCES

1. Sakamoto T, Tei C, Murayama M, et al. Giant T wave inversion as a manifestation of asymmetrical apical hypertrophy (AAH) of the left ventricle: echocardiographic and ultrasono-cardiotomographic study. Jpn Heart J 1976;17:611-29.
2. Yamaguchi H, Ishimura T, Nishiyama S, et al. Hypertrophic nonobstructive cardiomyopathy with giant negative T waves (apical hypertrophy): ventriculographic and echocardiographic features in 30 patients. Am J Cardiol 1979;44:401-12.
3. Yamaguchi H, Nishiyama S, Nakanishi S, Nishimura S. Electrocardiographic, echocardiographic and ventriculographic characterization of hypertrophic nonobstructive cardiomyopathy. Eur Heart J 1983;4 Suppl F:105-19.
4. Kitaoka H, Doi Y, Casey SA, Hitomi N, Furuno T, Maron BJ. Comparison of prevalence of apical hypertrophic cardiomyopathy in Japan and the United States. Am J Cardiol 2003;92:1183-6.
5. Eriksson MJ, Sonnenberg B, Woo A, Rakowski P, Parker TG, Wigle ED and Rakowski H. Long-term outcome in patients with apical hypertrophic cardiomyopathy. J Am Coll Cardiol 2002;39:638–45.
6. Sakamoto T, Amano K, Hada Y, et al. Asymmetric apical hypertrophy, ten years experience. Postgrad Med J 1986;62:567–70.
7. Suganuma Y, Shinmura K, Hasegawa H, et al. Clinical characteristics and cardiac events in elderly patients with apical hypertrophic cardiomyopathy. Nippon Ronen Igakkai Zasshi 1997;34:474 –81.
8. Chen CH, Nobuyoshi M and Kawai C. ECG pattern of left ventricular hypertrophy in nonobstructive hypertrophic cardiomyopathy: the significance of the mid-precordial changes. Am Heart J 1979;97:687-95.
9. Kasirye Y, Manne JR, Epperla N, Bapani S, Garcia-Montilla R. Apical Hypertrophic Cardiomyopathy Presenting as Recurrent Unexplained Syncope. Clinical Medicine & Research. 2012;10(1):26-31.
10. Thanigaraj S, Pérez JE. Apical hypertrophic cardiomyopathy: echocardiographic diagnosis with the use of intravenous contrast image enhancement. J Am Soc Echocardiogr. 2000;13:146-9.
11. Moukarbel GV, Alam SE, Abchee AB. Contrast-enhanced echocardiography for the diagnosis of apical hypertrophic cardiomyopathy. Echocardiogr. 2005;22:831-3.
12. Pons-Lladó G, Carreras F, Borrás X, Palmer J, Llauger J, Bayés de Luna A. Comparison of morphologic assessment of hypertrophic cardiomyopathy by magnetic resonance versus echocardiographic imaging. Am J Cardiol. 1997; 79:1651-6.