CASE REPORT

Yamaguchi syndrome presenting as atrioventricular nodal re-entrant tachycardia in an African-American patient

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SUMMARY

Apical hypertrophic cardiomyopathy (Yamaguchi syndrome) is a rare subtype of hypertrophic cardiomyopathy. The syndrome is more common in Japan where it was first described. Outside Asia, it is a very rare cause of hypertrophic cardiomyopathy. Apical hypertrophic cardiomyopathy is usually detected incidentally and has a good long-term outcome. We present a case of apical hypertrophic cardiomyopathy in an African-American patient manifesting as atrioventricular nodal re-entrant tachycardia.

BACKGROUND

To the best of our knowledge, after an extensive review of the literature, this is the first case report of apical hypertrophic cardiomyopathy (AHCM) presenting as atrioventricular nodal re-entrant tachycardia (AVNRT).^{1–3} It is important to recognise this type of arrhythmia given that catheter ablation of the arrhythmia has a good long-term outcome. AHCM is a rare type of hypertrophic cardiomyopathy (HCM) in western countries.

CASE PRESENTATION

A 63-year-old African-American man with no known medical history presented with persistent palpitations while having sexual intercourse. Emergency medical service was called and documented a supraventricular rhythm (figure 1). He was given three doses of adenosine which did not halt the arrhythmia necessitating synchronised cardioversion. Postcardioversion his ECG showed deep T waves in aVL, I and V4 to V6 suggestive of AHCM (figure 2). An echocardiogram confirmed the presence of apical type of HCM (figure 3). The patient denies any history of syncope or history of palpitations. He does not have any family history of sudden cardiac death or heart disease. The patient had a coronary angiogram performed which did not show any coronary artery disease. An electrophysiological (EP) study was carried out to identify and isolate the arrhythmia. During the EP study, arrhythmia induction showed that the VA time (which reflects the RP interval in a surface ECG) was 50 ms. A VA time <70 has a 99% positive predictive value for a diagnosis of AVNRT. Ventricular entrainment showed a VAVA response which is typical of an AVNRT. AVNRT ablation was subsequently mapped and ablated. There was no induced tachyarrhythmia induced following ablation suggesting successful ablation. The patient

improved and was discharged with referral to genetics to evaluate the familial nature of the disease.

INVESTIGATIONS

- ▶ ECG
- ▶ Echocardiography
- ▶ EP study

DIFFERENTIAL DIAGNOSIS

- ▶ Supraventricular tachycardia with aberrancy
- ▶ Idiopathic fascicular ventricular tachycardia

TREATMENT

AVNRT ablation.

OUTCOME AND FOLLOW-UP

The patient had successful ablation of AVNRT. He has had no recurrence of the arrhythmia. He was referred to genetics for further evaluation and management.

DISCUSSION

AHCM was described by Sakamoto and Yamaguchi *et al*⁴ as a form of non-obstructive HCM characterised in an ECG by giant negative T waves (>10 mm) associated with a high QRS voltage (R wave >26 mm in lead V5 or the sum of the S wave in lead V1 and R in lead V5 35 mm or more) in the absence of hypertension or significant coronary artery disease. This is a relatively rare form of HCM, more commonly seen in Japan where it constitutes 13–15% of all cases.⁵ A study by Kitaoka *et al*⁶ comparing the prevalence of AHCM in Japan and the USA showed that Japanese patients were more predisposed to develop AHCM (15% in Japan vs 3% in the USA).

A two-dimensional echocardiogram is generally used to diagnose structural heart disease like HCM and its variants. Cardiac MRI (CMRI) is generally used as an adjunct imaging test especially if there are repolarisation changes on a surface ECG but a normal echocardiogram. CMRI is able to identify regions of hypertrophy not seen in an echocardiogram. CMRI provides enhanced assessment of left ventricular hypertrophy in AHCM. Our patient had a diagnostic ECG and echocardiograph for AHCM, hence a CMRI was not performed.^{7–9}

Racial differences in ECG findings have been well documented. ECG findings that seem to be more prevalent among the African-American population include early repolarisation pattern, high amplitude R wave, ST elevation, T wave findings

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Figure 1 ECG showing a regular wide complex tachycardia with a right bundle branch block pattern. The heart rate is approximately 160 bpm. Encircled in this figure are the retrograde P waves which are very typical atrioventricular nodal tachycardia. The interval between the between the R wave and the retrograde P wave is <100 ms which is typical of short R-P tachycardia.

Figure 2 EKG shows presence of giant (over 10 mm) inverted T waves in leads V4 to V6 V; sum of R wave in V5 and S wave in V1 is >35 mm. To be noted in this resting ECG is the absence of the right bundle branch block which was seen previously from the EKG with a fast rhythm (AVNRT). AVNRT, atrioventricular nodal re-entrant tachycardia; EKG, echocardiogram.

Figure 3 Echocardiogram images. (A) Long parasternal view during diastole, evidence of left ventricle apical hypertrophy (truncated given the ultrasound depth used during the evaluation) pointed by the red-shaded area. (B) Long parasternal view during systole identifying with red shade the hypertrophic left ventricular apex (truncated due to the ultrasound depth used during the evaluation). (C) Four-chamber view depicting the left ventricular apical hypertrophy (red shade).

and prolonged PR interval.¹⁰ A peculiar finding in the patient's ECG showing the atrioventricular tachycardia is the presence of a bundle branch block, specifically a pattern of a right bundle branch block. The bundle branch block is not seen in resting surface ECG. This could be explained by tachycardia-dependent bundle branch block. This is due to increased bundle branch refractoriness in the setting of a fast heart rate. In the setting of tachycardia, cardiac impulse reaches the bundle branch at the absolute refractory period thus causing a delay in conduction of impulse. This phenomenon is generally seen in the setting of ischaemia or stretching of the conducting tissue due to increased myocardial mass as our case with HCM.^{11 12}

HCM is a complex primary and genetically transmitted cardiac disease with a diverse clinical course varying from a benign or stable clinical course over many years or progressively worsening to congestive symptoms requiring therapeutic intervention and the possibility of sudden and unexpected death. Cardiovascular death could be due to sudden cardiac death from a ventricular arrhythmia, heart failure or stroke.^{13 14} AHCM has generally a benign course based on studies done in Japan.^{4 5}

In a study carried out by Eriksson and colleagues in North American population, patients with AHCM presented with angina, atypical chest pain, palpitations, dyspnoea and presyncope/syncope. The overall cardiovascular mortality among these patients was 1.9%. Overall survival was 95% at 15 years.¹⁵ The most common sustained supraventricular tachycardia for HCM and AHCM is atrial fibrillation.^{14 15} There have been several

case reports of supraventricular tachycardia in HCM resulting in sudden cardiac death. This is particularly more common among patients with left ventricular outflow tract LVOT obstruction.^{16 17}

AVNRT is the most common form of regular narrow complex tachycardia. It is due to dual atrioventricular nodal conduction over two pathways with different EP properties. It has a 'fast' pathway that conducts faster but has a longer refractory period than a second 'slow' pathway. A typical AVNRT is initiated when a premature atrial beat arrives at the AV node when the fast pathway is in its refractory period. In this case, the antegrade flow of the impulse is blocked. A premature cardiac impulse can be conducted through the AV node when the slow pathway is excitable due to its shorter refractory period. When the fast pathway recovers and is excitable by the time the slow pathway impulse reaches the junction where both pathways converge, there is a retrograde conduction up the fast pathway. This process can become repetitive to induce a tachyarrhythmia.¹⁸ The incidence in patients with HCM has not been established. A study carried out by Schiavone *et al*¹⁹ looking at the EP studies among patients with HCM, 65% had supraventricular tachycardia. This included either atrial fibrillation or atrial flutter.

To the best of our knowledge, this is the first case report of AHCM presenting with AVNRT. Given this scenario, our case is likely an incidental presentation rather than a real variant presentation of Yamaguchi syndrome. A study of the long-term outcome of patients with AHCM showed the occurrence of

atrial fibrillation as the most common supraventricular tachycardia.¹⁵ The presentation in this case is a very rare manifestation of the disease. It is important to recognise this type of arrhythmia because this type of supraventricular tachycardia has a very good outcome with catheter ablation. Catheter ablation is performed with either radiofrequency ablation or cryoablation.^{20 21} Catheter ablation targets the slow pathway along the posteroseptal region of the tricuspid annulus. Slow pathway ablation has a lower incidence of complete atrioventricular block and PR interval prolongation. Success rate with ablation of an AVNRT is 96–97% with a recurrence rate of 5%.^{20 21} Our patient underwent successful radiofrequency ablation of the abnormal pathway causing the AVNRT.

Given the rarity of AHCM in the USA, there is no clinical practice guidelines tailored for this subset of HCM. Risk stratification among patients with AHCM is based on the American College of Cardiology Foundation (ACCF)/American Heart Association (AHA) guideline for the diagnosis and treatment of HCM. Although there is no recognised relationship between the pattern or distribution of left ventricular hypertrophy and clinical outcome, it is important to know the extent of hypertrophy. Sudden cardiac death risk is highest with wall thickness more than or equal to 30 mm.⁹ Patients with AHCM may develop the typical symptoms and complications of HCM.^{22 23} Patients with AHCM are at risk of apical myocardial infarction and apical aneurysm development.^{23 24} It is recommended to follow patients with an annual echocardiography.²⁴

Learning points

- ▶ Apical hypertrophic cardiomyopathy is a rare type of hypertrophic cardiomyopathy that generally has a good long-term outcome.
- ▶ Close follow-up among patients with hypertrophic cardiomyopathy is important given that a small number of patients may have complications of the classic hypertrophic cardiomyopathy.
- ▶ Atrioventricular nodal re-entrant tachycardia is a rare manifestation of hypertrophic cardiomyopathy especially in the case of apical hypertrophic cardiomyopathy. This type of supraventricular tachycardia is very responsive to catheter ablation.

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