Complete Heart Block in Hypertrophic Cardiomyopathy: A Rare Association

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ABSTRACT

Ventricular tachyarrhythmias are common in hypertrophic cardiomyopathy that may lead to syncope and sudden death. Bradyarrhythmia such as atrioventricular conduction disturbance, a relatively rare complication associated with hypertrophic cardiomyopathy, may also cause syncope and sudden death in hypertrophic cardiomyopathy. We report a 28-year old man who was diagnosed as a case of hypertrophic cardiomyopathy presented with syncope and complete heart block. Subsequently, a permanent pacemaker was implanted to the patient.

KEY WORDS

Complete heart block, hypertrophic cardiomyopathy, permanent pacemaker, syncope

INTRODUCTION

Hypertrophic cardiomyopathy (HCM), an autosomal dominant inherited genetic disease, is defined by the unexplained myocardial hypertrophy and is present in 1 in 500 adults in the general population.[1] Arrhythmias are common in HCM which may lead to syncope and sudden death. Although in most patients with HCM syncope and sudden death is caused by ventricular arrhythmias (sustained ventricular tachycardia or ventricular fibrillation), it can also be caused by bradyarrhythmia such as atrioventricular (AV) conduction disturbance, a relatively rare complication associated with HCM.[2] In this paper we describe a 28-year old man who was a diagnosed case of HCM presented with syncope and complete heart block. He underwent a permanent pacemaker implantation. To the best of our knowledge, this is the first reported case of complete heart block in HCM from Nepal.

CASE REPORTS

A 28-year old man presented to our emergency room with a history of syncope and palpitation. The first syncope occurred in 2012. After having recurrent syncope he visited another hospital for evaluation of loss of consciousness. Cardiac evaluation, including conventional echocardiography showed left ventricular hypertrophy and electrocardiogram (ECG) revealed complete right bundle branch block with left anterior fascicular block. The neurologic exams for differential diagnosis showed no evidence of seizure disorder or any other diseases inducing loss of consciousness and he was diagnosed as HCM and was discharged with metoprolol. After taking metoprolol, he felt uncomfortable and frequent episodes of dizziness and stopped taking medicine.

In 2014 he again developed syncopal attack and visited our hospital. On arrival to our hospital, he was conscious, afebrile with pulse rate of 30 beats/minute and blood pressure of 100/70 mmHg. Initial ECG revealed complete heart block with ventricular rate of 30 beats/min (Fig. 1). He reported that his father and mother died suddenly at the age of 44 years and 40 years respectively. The baseline two-dimensional echocardiography showed HCM with asymmetric septal hypertrophy (septal wall thickness during diastole 25 mm and posterior wall thickness of 12 mm) (Fig. 2) without Doppler evidence of significant

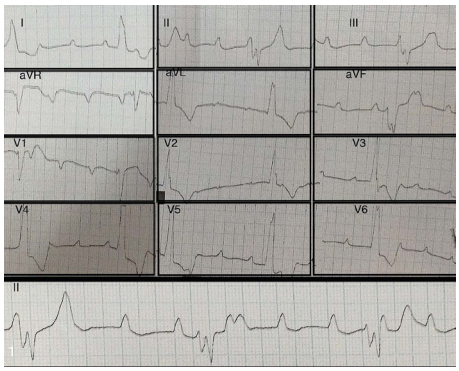

Figure 1. Initial electrocardiogram on presentation showed complete heart block with ventricular rate of 30 beats/min

obstruction of the left ventricular outflow tract. Left atrium was mildly dilated (41 mm) but cavity dimensions and systolic function of the left ventricle were within normal limits.

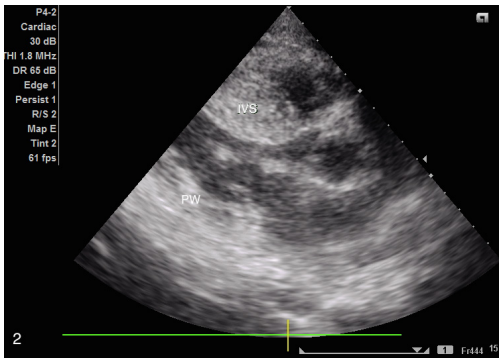

Figure 2. Baseline echocardiography showed asymmetric septal hypertrophy of left ventricle in parasternal long axis view. Thickness of interventricular septum (IVS) was 25 mm and posterior left ventricular wall (PW) was 12 mm.

After seeing complete heart block in ECG we thought this conduction disturbance could play a role in repeated loss of consciousness and he was taken to the catheterization laboratory and a temporary pacemaker was inserted via right femoral vein and kept in the right ventricular apex. The patient refused dual chamber permanent pacemaker implantation and four days later a single chamber permanent pacemaker (VVIR) was implanted. His subsequent stay in the hospital was uneventful and discharged on oral bisoprolol 2.5 mg a day. His elder brother (45-year old only available family member) was screened for HCM and he also had echocardiographic evidence of HCM.

DISCUSSION

About 25-30% of patients with HCM experience symptoms of syncope or near syncope.[1] A history of recurrent syncope episodes is one of the predictable risk factors for sudden cardiac death in younger patients.[3,4] It is important to know the mechanism causing loss of consciousness in patients with HCM because of the different management strategy, which can be explained by two underlying mechanisms: hemodynamic mechanism and arrhythmic complications. Among the arrhythmic causes for syncope, sustained ventricular tachycardia can be the cause of syncope and even sudden cardiac death in HCM.[5] However, besides these tachyarrhythmia, bradyarrhythmia such as atrioventricular (AV) block also cause syncope or presyncope in HCM.[2,6]

In HCM, bradyarrhythmias has been considered an uncommon and atrioventricular (AV) block an infrequent complication and there are only a very few case reports of HCM combined with atrioventricular (AV) block causing syncope both in pediatric and adult patients.[2] First report of atrioventricular (AV) block associated with HCM is from Luisada in 1965.[7] They reported atrioventricular (AV) block in a 10-year old boy who presented with headache during a clinic follow-up. In 1977, Spilkin and colleagues described the case of a 20-year old boy with HCM who subsequently developed AV block.[8]

The cause of atrioventricular (AV) block in HCM is not clear. Histopathologic reports can describe possible causes. Maron et al. in their study described histopathologic examination of the atrioventricular (AV) nodal tissue was normal,[9] however, continuity of the conduction system was interrupted in the bundle of his. Others reported interstitial fibrosis or myocardial necrosis in the conduction system and abnormally small intramural coronary arteries with thickened walls, luminal narrowing in HCM and advanced conduction system disorders.[10]

It has been reported that if atrioventricular (AV) block occurs and progresses rapidly to high-grade block and then to severe syncope death will be inevitable. It is possible that the sudden appearance of high grade AV block is a more frequent cause of sudden death in adults with HCM than previously suspected. Neuromuscular disease, muscular dystrophy also has the similar presentation (HCM and AV block) but our patient has no symptoms and signs of neuropathy/myopathy. It should be noted that acute and subacute complete heart block are also a sequelae of alcohol septal ablation for HCM.

CONCLUSION

This case describes syncopal episodes caused by complete heart block in a patient with HCM who was managed successfully with a permanent pacemaker implantation. Although a rare complication, we should keep in mind the probability of atrioventricular (AV) block as a cause of syncope in a patient with HCM.

REFERENCES

1. Gersh BJ, Maron BJ, Bonow RO, Dearani JA, Fifer MA, Link MS, et al. American College of Cardiology Foundation/American Heart Association Task Force on Practice; American Association for Thoracic Surgery; American Society of Echocardiography; American Society of Nuclear Cardiology; Heart Failure Society of America; Heart Rhythm Society; Society for Cardiovascular Angiography and Interventions; Society of Thoracic Surgeons, 2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guide lines. *J Thorac Cardiovasc Surg* 2011;142:e153-203.
2. Kim KH, Yang DH, Kim CY, Kim NK, Choi WS, Bae MH, et al. Recurrent syncope episodes and exercise intolerance in hypertrophic cardiomyopathy combined with atrioventricular conduction disturbance. *J Cardiovasc Ultrasound* 2013;21:148-51
3. Maron BJ, Bonow RO, Cannon RO 3rd, Leon MB, Epstein SE. Hypertrophic cardiomyopathy. Interrelations of clinical manifestations, pathophysiology, and therapy (2). *N Engl J Med 1987;316:844-52.*
4. McKenna WJ, Franklin RC, Nihoyannopoulos P, Robinson KC, Deanfield JE. Arrhythmia and prognosis in infants, children and adolescents with hypertrophic cardiomyopathy. *J Am Coll Cardiol* 1988;11:147-53.
5. Spirito P, Bellone P, Harris KM, Bernabo P, Bruzzi P, Maron BJ. Magnitude of left ventricular hypertrophy and risk of sudden death in hypertrophic cardiomyopathy. *N Engl J Med* 2000;342:1778-85.
6. Williams L, Frenneaux M. Syncope in hypertrophic cardiomyopathy:mechanisms and consequences for treatment. *Europace* 2007;9:817-22.
7. Luisada AA. Sub-aortic muscular stenosis and complete heart block in an adolescent. Pediatric-surgical-cardiac conference case presentation. *Chic Med Sch Q* 1965;25:169-75.
8. Spilkin S, Mitha AS, Matisonn RE, Chesler E. Complete heart block in a case of idiopathic hypertrophic subaortic stenosis: noninvasive correlates with the timing of atrial systole. *Circulation* 1977;55:418-22.
9. Maron BJ, Connor TM, Roberts WC. Hypertrophic cardiomyopathy and complete heart block in infancy. *Am Heart J* 1981;101:857-60.
10. Yesil M, Bayata S, Susam I, Dinckal H, Postaci N. Rare association of hypertrophic cardiomyopathy and complete atrioventricular block with prompt disappearance of outflow gradient after DDD pacing. *Europace* 1999;1:280-2.