CASE REPORT

Rare association of hypertrophic cardiomyopathy and complete atrioventricular block with prompt disappearance of outflow gradient after DDD pacing

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Although arrhythmias are common in hypertrophic cardiomyopathy (HCM), complete atrioventricular (AV) block is very unusual. A 27-year-old female presented with a recent history of syncope and exercise intolerance. ECG demonstrated complete AV block. Two-dimensional Doppler echocardiography revealed HCM with a 60 mmHg left ventricular outflow tract (LVOT) gradient. A temporary transvenous ventricular pacemaker was inserted urgently, and subsequently replaced by a permanent DDD pacemaker. All symptoms were eliminated. This symptomatic improvement was associated with complete disappearance of LVOT gradient at the time of implantation. No gradient was observed during early follow-up and at 6 months after DDD pacemaker implantation.

Key Words: Hypertrophic Obstructive cardiomyopathy, atrioventricular block, DDD pacemakers.

Case history

A 27-year-old woman was referred to the cardiology clinic because of new-onset recurrent syncope and exercise intolerance. She had had three episodes of loss of consciousness during the week before hospitalization. There was no family history of cardiac disease and her past medical history was unremarkable. Definable causes of AV block in a young person were excluded. She was not taking any medication. Clinical examination was normal except for a regular pulse rate of 37 beats.min⁻¹ and a soft ejection systolic murmur in the left sternal border. On admission the ECG demonstrated complete AV block with a QRS duration of 145 ms and a left bundle branch block pattern ventricular escape rhythm. A temporary transvenous ventricular pacemaker was inserted as an emergency. Doppler echocardiographic examination revealed hypertrophic cardiomyopathy (HCM) with a septal thickness of 16 mm and a 60 mmHg left ventricular outflow tract (LVOT) gradient. Cavity dimensions and systolic function of the left ventricle were within normal limits. Cardiac catheterization disclosed a 52 mmHg LVOT gradient as the left heart catheter was pulled back (Fig. 1). For the purpose of aetiological evaluation, coronary angiography was performed and found to be normal. Right heart catheterization parameters were normal with a pulmonary artery wedge pressure of 12 mmHg. A permanent DDD pacemaker was implanted. Following the implantation, exercise intolerance recovered completely. Elimination of symptoms was associated with sudden and complete abolition of outflow obstruction. Haemodynamic (Fig. 2), echocardiographic and angiographic examination revealed complete disappearance of the LVOT gradient and systolic anterior motion of the mitral anterior leaflet after pacemaker implantation. She has been symptom free since then. No gradient was observed during early follow-up or at 6 months post-pacing. Complete AV block has persisted.

Discussion

Patients with HCM frequently have arrhythmias and haemodynamic abnormalities and are prone to syncope.
and sudden death. Arrhythmias, are most usually complex ventricular tachyarrhythmias and may be associated with syncope in this disorder, but there is a possibility of other causes of syncope. This report describes a patient with HCM, who developed recurrent syncopal attacks associated with AV block. In this patient, symptoms and LVOT gradient simultaneously and completely disappeared at the time of DDD pacemaker implantation. The occurrence of this combination is rare\cite{1–4}. According to our knowledge, 11 cases of HCM in adults and six cases in childhood, which were complicated by complete AV block, have been reported in the literature. Familial occurrence of this combination is common in these reported cases\cite{3}.

The treatment of obstructive HCM is based on the major pathophysiological problem presented by each patient. Therapy is directed at lessening the severity of the obstruction by negative inotropic agents, AV sequential pacemaker therapy, surgery (ventriculotomy–mitral valve replacement) or percutaneous transluminal septal myocardial ablation. Fananapazir et al.\cite{5} implanted dual chamber pacemakers in 84 patients with symptomatic obstructive HCM. During a follow-up period of up to 2-3 years, symptoms, especially syncope and presyncope, were improved and the gradient was reduced by an average of about 70%.

This patient with HCM demonstrated complete AV block almost certainly involving the His-Purkinje system. A causal relation between conduction disease and HCM will be difficult to establish and the cause of AV block in HCM is unknown. Some histopathological reports describe possible causes for the development of conduction abnormalities. Maron et al.\cite{6} describe normal AV nodal tissue but interruption in the bundle of His as it coursed through the central fibrous body. Gavrilescu et al.\cite{7,8} and Bharati et al.\cite{7,8} demonstrated interstitial fibrosis or myocardial fibrosis in the conduction system and abnormally small intramural coronary arteries with thickened walls, luminal narrowing in HCM and advanced conduction system disorders. In addition to these reports, James et al.\cite{9} demonstrated cystic and vascular abnormalities in the AV node and bundle of His as well as persistent fetal dispersion of these structures in 22 HCM patients, who died suddenly.

The literature, therefore, suggests that patients with HCM may rarely develop AV block. This combination must be considered in the differential diagnosis of syncope.

Atrioventricular block may be the first manifestation of HCM and DDD pacing may be life-saving by treating both the outflow obstruction and AV block. Therefore, during follow-up of patients, with HCM, it is prudent to check for development of abnormal AV conduction. In this patient with an uncommon association of HCM and complete AV block there was prompt and total resolution of LVOT gradient after DDD pacemaker implantation.

Figure 1 Simultaneous ECG and left heart pressure tracings showing LVOT gradient during complete heart block.
Figure 2  Simultaneous ECG and left heart pressure tracings showing complete disappearance of LVOT gradient during DDD pacing with an appropriate AV delay.

References


