Complete Atrioventricular Block and Infective Endocarditis in a Patient with Hypertrophic Obstructive Cardiomyopathy

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Abstract

A 62-year-old man with hypertrophic obstructive cardiomyopathy (HOCM) had complete atrioventricular (AV) block and subsequent cardiac standstill. A previous electrocardiogram revealed a bifascicular block pattern. Because he also suffered from infective endocarditis of the native aortic valve, surgical therapy (dual-chamber permanent pacing, myectomy of the left ventricular outflow tract, and valve replacement) was performed. Complete AV block unrelated to a procedure is a rare complication in patients with HOCM, but it may be life-threatening. Therefore, a pre-existing cardiac conduction disturbance should be specifically recognized as the aura of a higher degree of AV block.

Key words: hypertrophic obstructive cardiomyopathy, complete atrioventricular block, dual-chamber pacemaker implantation, infective endocarditis

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Introduction

Hypertrophic cardiomyopathy (HCM) is commonly complicated by arrhythmias; in particular ventricular arrhythmias have been shown to be associated with sudden cardiac death (1). On the other hand, complete atrioventricular (AV) block unrelated to a procedure is a rare complication in patients with HCM (2-7), and its association with sudden cardiac death remains unclear. Here, we report the case of a patient with hypertrophic obstructive cardiomyopathy (HOCM) who experienced complete AV block and subsequent cardiac standstill. In addition, the patient suffered from infective endocarditis of the native aortic valve.

Case Report

A 62-year-old man was admitted to the division of neurology in our hospital because of hemiparesis in the right upper limb. He had no family history of cardiac disease, and his past medical history was unremarkable. On admission, physical findings revealed height of 159 cm, weight of 64 kg, blood pressure of 175/82 mmHg, regular pulse of 96 beats per minute and temperature of 38.0°C. There were no rales in the chest and a grade 3 systolic murmur was audible in the lower left sternal border. Light motor paresis and hypesthesia were detected in the right upper limb. Oxygen saturation was 97% under room air, and laboratory tests showed white blood cell (WBC) count of 12,900/μL with neutrophil of 83%, hemoglobin of 11.4 g/dL and C-reactive protein (CRP) of 4.2 mg/dL (Fig. 1). Transaminase, blood urea nitrogen, serum creatinine, and urinalysis were normal. An electrocardiogram revealed sinus rhythm, right axis deviation and complete right bundle branch block (Fig. 2a). A chest radiograph showed a cardiothoracic ratio of 59% and no remarkable findings in the lung fields. A diffusion-weighted magnetic resonance imaging scan showed a new infarct lesion extending from the left frontal lobe to the parietal lobe. Aspirin was administered for treatment of cerebral infarction, and rehabilitation was begun. He became afebrile spontaneously at 3 days of hospitalization, but had shortness of breath during level walking. An echocardiogram

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in parasternal long axis view showed left atrial diameter of 58 mm, left ventricular end diastolic diameter of 52 mm, systolic diameter of 31 mm, fractional shortening of 40%, septal and posterior wall thickness of 18 mm. Reduced wall motion of the posterior wall in the left ventricle, a pressure gradient of 58 mmHg in the left ventricular outflow tract (LVOT), mild aortic regurgitation, moderate mitral regurgitation, systolic anterior movement of the mitral valve and no intracardiac thrombus were revealed. A cardiologist had planned on examinations including cardiac catheterization for HOCM and left ventricular asynergy.

At 23 days of hospitalization, orthopnea suddenly developed. Physical findings revealed a systolic blood pressure of 180 mmHg, regular pulse of 50 beats per minute, respiratory rate of 30 breaths per minute, temperature of 37.0°C and wheezing in the chest. Oxygen saturation was 86% under room air. A chest radiograph revealed butterfly shadow, and an electrocardiogram showed complete AV block with a heart rate of 50 beats per minute (Fig. 2b). Laboratory test revealed WBC count of 21,500/μL, CRP of 6.5 mg/dL and brain natriuretic peptide (BNP) of 1,820 pg/mL (Fig. 1). Tracheal intubation and mechanical ventilation were necessary for acute respiratory failure. Since cardiac standstill subsequently developed, temporary intravenous ventricular pacing was immediately performed. Coronary angiogram revealed no organic stenotic lesions. Norepinephrine and a diuretic were administered for the treatment of cardiogenic shock and pulmonary congestion, respectively. An echocardiogram showed no remarkable change of left ventricular systolic function and mitral regurgitation, but moderate aortic regurgitation. Severe liver and renal injury developed, and an antibiotic (ceftriaxone 1 g/day) was empirically administered for a week to prevent central venous catheter-related bacteremia and ventilator associated pneumonia. At 33 days of hospitalization, high fever of 39.0°C developed, and another antibiotic (piperacillin/tazobactam 4.5 g every 12 hours) was continued for 10 days as central venous catheter-related bacteremia was suspected (Fig. 1). No organism was isolated from 2 blood culture sets prior to administration of piperacillin/tazobactam. He became afebrile 7 days after administration of the antibiotic, and pulmonary edema, and liver and renal dysfunction also improved. A temporary pacing lead and central venous catheter were repeatedly exchanged, and permanent pacemaker implantation had been planned.

At 52 days of hospitalization, leg edema and bilateral pleural effusion on a chest radiograph were observed, and diagnosis of congestive heart failure was made. An echocardiogram revealed a thin structure (size, 13 mm) attached to the aortic valve with severe aortic regurgitation (Fig. 3). He did not have a fever or skin lesions. Laboratory data revealed WBC of 4,400/μL, CRP of 1.0 mg/dL and BNP of 1,271 pg/mL (Fig. 1). In addition to a diuretic, dual antibiotics (cefazolin 2 g every 6 hours and gentamicin 60 mg every 12 hours) were initiated because of suspicion of infective endocarditis. No organisms were isolated from 3 blood culture sets prior to administration of the antibiotics. Based on Duke’s criteria in which a major and two minor criteria were fulfilled, diagnosis of possible infective endocarditis was made (8).

At 59 days of hospitalization, the patient was transferred to the surgical department, where he underwent myectomy of the LVOT, mitral and aortic valve replacement, and dual-chamber pacemaker implantation using epicardial pacing.

Figure 1. Clinical course. BT: body temperature, C-AVB: complete atrioventricular block, CEZ: cefazolin, CRP: C-reactive protein, CTRX: ceftriaxone, GM: gentamicin, PIPC: piperacillin, TAZ: tazobactam, WBC: white blood cell.
leads. Mitral valve replacement was performed because of infective thickening in the anterior leaflet of mitral valve and anatomical variant of papillary muscle and chordate tendineae. A vegetation and valvular destruction in left coronary cusp of aortic valve were found, but no ring abscess was seen. Pathological study of the left ventricular myocardium revealed mild hypertrophy and variant form of myocardial cells’ nuclei and interstitial fibrosis. Although the diagnosis of infective endocarditis was histologically defined, no organism was isolated from tissue culture. He was discharged after additional antibiotic therapy for 4 weeks, and has been clinically well 34 months after surgical therapy.

Discussion

Little has been reported on HCM with both complete AV block and infective endocarditis; to our knowledge the present case is the first report. Complete AV block unrelated to a procedure in patients with HCM is also a rare complication, and previous cases have been observed in a relatively high proportion of young adult individuals or individuals with familial HCM (4, 5). Previous reports showed that pathological studies of conduction system revealed marked

Figure 2. a: Electrocardiogram on admission showing sinus tachycardia, right axis deviation and complete right bundle branch block. b: Electrocardiogram at 23 days of hospitalization showing a heart rate of 50 beats per minute and complete atrioventricular block with wide QRS wave of left bundle branch block.

Figure 3. Parasternal long axis view of transthoracic echocardiogram at 52 days of hospitalization showing a thin vegetation (13 mm in size) attached to the aortic valve (arrow). Ao: aorta, LA: left atrium, LV: left ventricle, RV: right ventricle.
fibrotic change (9, 10), however, it was not performed in the present case. Pathological findings of the left ventricular myocardium did not contradict those of HCM, and there was no other finding suggesting secondary cardiomyopathy.

Complete AV block caused acute pulmonary congestion and subsequent cardiac standstill as clinical manifestations, thus it may be a life-threatening complication associated with HCM. Fundamentally, loss of AV sequence and bradycardia in addition to left ventricular diastolic dysfunction may cause reduction of cardiac output followed by congestive heart failure in HOCM with complete AV block. A previous report showed that decreased afterload caused by prolongation of RR interval was a more important variable for LVOT gradient than decreased preload caused by loss of AV sequence (3). In the present case, LVOT gradient was not measured at the onset of complete AV block, however, the increase of aortic regurgitation might have contributed to the deterioration of pulmonary congestion. A few reports have also described HCM patients who had complete AV block followed by cardiopulmonary arrest (3, 7). Fananapazir and Epstein showed that among 30 survivors of sudden cardiac arrest with HCM, abnormal AV nodal and His-Purkinje conduction was detected in 1 (3%) and 7 (23%) patients, respectively (11), however, the relation between AV block and sudden cardiac death in the HCM patients remains unclear. It is important that a pre-existing AV conduction disturbance such as a bifascicular block in the present case should be recognized as the aura of a higher degree of AV block. In fact, of 6 previous cases for whom electrocardiogram prior to complete AV block was performed, left bundle branch block in 3, left axis deviation plus right bundle branch block in 1, and left axis deviation plus intraventricular conduction delay in 1 were revealed (2, 4-7). In HOCM patients with conduction disturbance, the use of negatively inotropic agents such as beta-blockers, Ca channel blockers, and anti-arrhythmic agents to reduce LVOT stenosis worsens conduction disturbance. Hence, dual-chamber pacemaker implantation may be selected as a treatment for not only conduction disturbance but also reduction of LVOT stenosis (12). In the present case, myectomy was added to pacemaker implantation, because it can be performed simultaneously with valve replacement for endocarditis and certainly reduce LVOT stenosis.

Endocarditis complicating HOCM may be related to damage of the mitral and aortic valve endocardium caused by turbulence of blood flow during systole accompanying LVOT stenosis and mitral valve regurgitation. Although endocarditis complicating HOCM has been shown to occur predominantly on the left ventricular aspect of the anterior mitral valve leaflet, some cases with endocarditis in aortic valve alone have been reported (13). Accordingly, we think that aortic valve endocarditis was associated with HOCM in the present case. Complete AV block may rarely develop in patients with infective endocarditis if infection extends from the valve leaflets into the surrounding myocardium, and that is more common in the aortic valve than in mitral valve (14). On the other hand, extension of infection was not a cause of complete AV block in the present case because there was no ring abscess of the aortic valve. Regarding the onset time of infective endocarditis, we had supposed that catheter-related bacteremia occurring after the onset of complete AV block caused infective endocarditis, because the patient was almost afebrile for 20 days after 3 days of hospitalization. However, an elevation of inflammatory markers on admission and an increase of aortic regurgitation at the onset of complete AV block suggests that cerebral infarction on admission resulted from infective embolization. It is possible that no organism was isolated from the blood cultures because of prior administration of antibiotic. We recognized that infective endocarditis should be suspected in the HOCM patients with fever of origin unknown.

**Conclusion**

We encountered a rare case of the HOCM patient with both complete AV block and infective endocarditis. Complete AV block is a rare but life-threatening complication in patients with HOCM. Therefore, a pre-existing AV conduction disturbance should be specifically recognized as the aura of a higher degree of AV block.

The authors state that they have no Conflict of Interest (COI).

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