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【 CASE REPORT 】

Cardiac Magnetic Resonance Identified the Fibrotic Lesion Associated with Syncope Attack Due to Complete Atrioventricular Block in a Patient with Hypertrophic Cardiomyopathy and Aortic Stenosis

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Abstract:

An 84-year-old man presented with syncope. Prior to admission, ambulatory electrocardiogram had demonstrated non-sustained ventricular tachycardia. Echocardiography showed severe aortic stenosis. He was also diagnosed with hypertrophic cardiomyopathy (HCM) by cardiac magnetic resonance (CMR) showing remarkable inhomogeneous left ventricular hypertrophy and extensive late gadolinium enhancement (LGE) in the lesions at the upper border and right-ventricular side of the basal-mid septal wall. Finally, he showed complete atrioventricular (AV) block followed by a long pause and syncope several times after admission. In this case with several possible causes of syncope, the CMR findings suggested a clue concerning the etiology of his syncope: complete AV block in HCM.

Key words: cardiac magnetic resonance, complete atrioventricular block, hypertrophic cardiomyopathy, late gadolinium enhancement, syncope

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Introduction

Ventricular arrhythmia is a major cause of syncope and sudden death in hypertrophic cardiomyopathy (HCM). Myocardial fibrosis is a pathological hallmark of HCM and is considered a substrate for ventricular arrhythmia. Recently, cardiac magnetic resonance (CMR) has helped visualize myocardial fibrosis and scarring *in vivo* through late gadolinium enhancement (LGE), and the prognostic role of the presence of LGE has been demonstrated. However, complete atrioventricular (AV) block in HCM is a relatively rare complication, and the details concerning the etiology are unclear (1).

Case Report

An 84-year-old man was referred to our hospital for the evaluation of new-onset recurrent syncope attacks. He had no history of coronary artery disease. His family history showed a sister with pacemaker implantation, a brother with heart failure, and a daughter with cardiac hypertrophy.

An examination revealed a blood pressure of 135/80 mmHg and a heart rate of 84 bpm. A grade 3/6 systolic murmur was best heard in the third intercostal space, right of the sternum. An electrocardiogram (ECG) showed first-degree AV block with left bundle branch block. Echocardiography showed severe aortic stenosis (AS) with a peak velocity of 4.5 m/s and an aortic valve area of 0.72 and 0.67 cm^2 by the continuity equation and planimetry, respectively (Fig. 1A and B). It also showed inhomogeneous remarkable LV hypertrophy with a septal thickness of 26 mm and no left ventricle outflow tract (LVOT) pressure-gradient. The LV cavity dimensions and systolic function were within normal limits (Fig. 1C and D). The ambulatory ECG prior to admission showed the non-sustained ventricular tachycardia (VT). CMR showed remarkable inhomogeneous LV hypertrophy with extensive LGE. Namely, LGE lesions were

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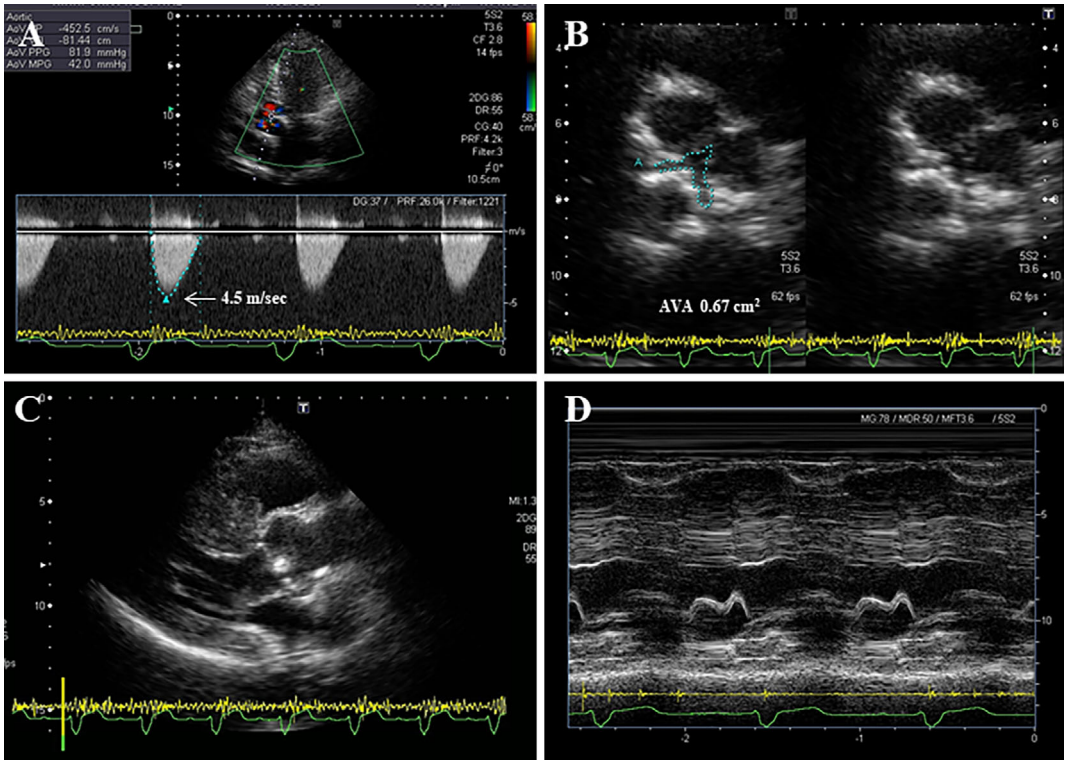

Figure 1. **Standard transthoracic echocardiography. Continuous-wave Doppler measurement of peak aortic transvalvular velocity (A), aortic valve area (AVA) by planimetry (B), parasternal long-axis view (C), and M-mode recording at the mitral valve level (D).**

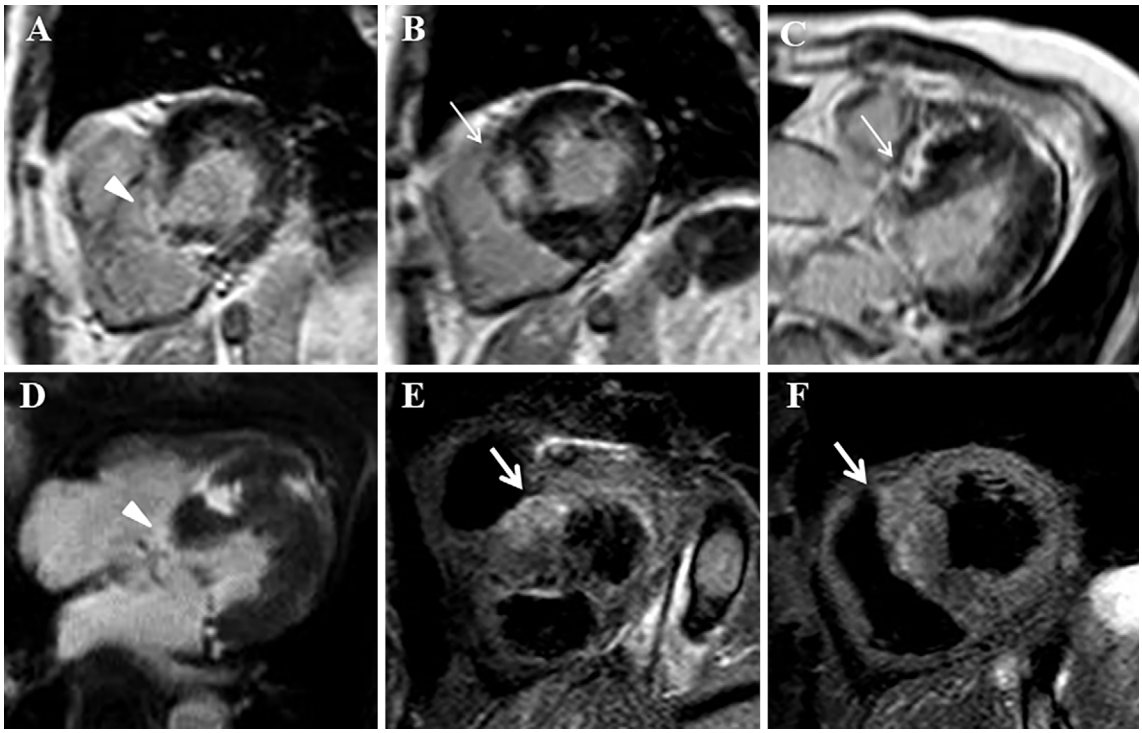

Figure 2. **Cardiac MR images with late gadolinium enhancement (LGE) (A-D) and T2-weighted images (E, F). Basal (A, E) and basal-mid short-axis images (B, F), and long-axis images (C, D). The arrowheads indicate LGE at the upper border of the ventricular septum, the thin arrows indicate LGE at the right-ventricular side of the basal-mid septal wall, and the thick arrows indicate the high-signal-intensity area of T2 images.**

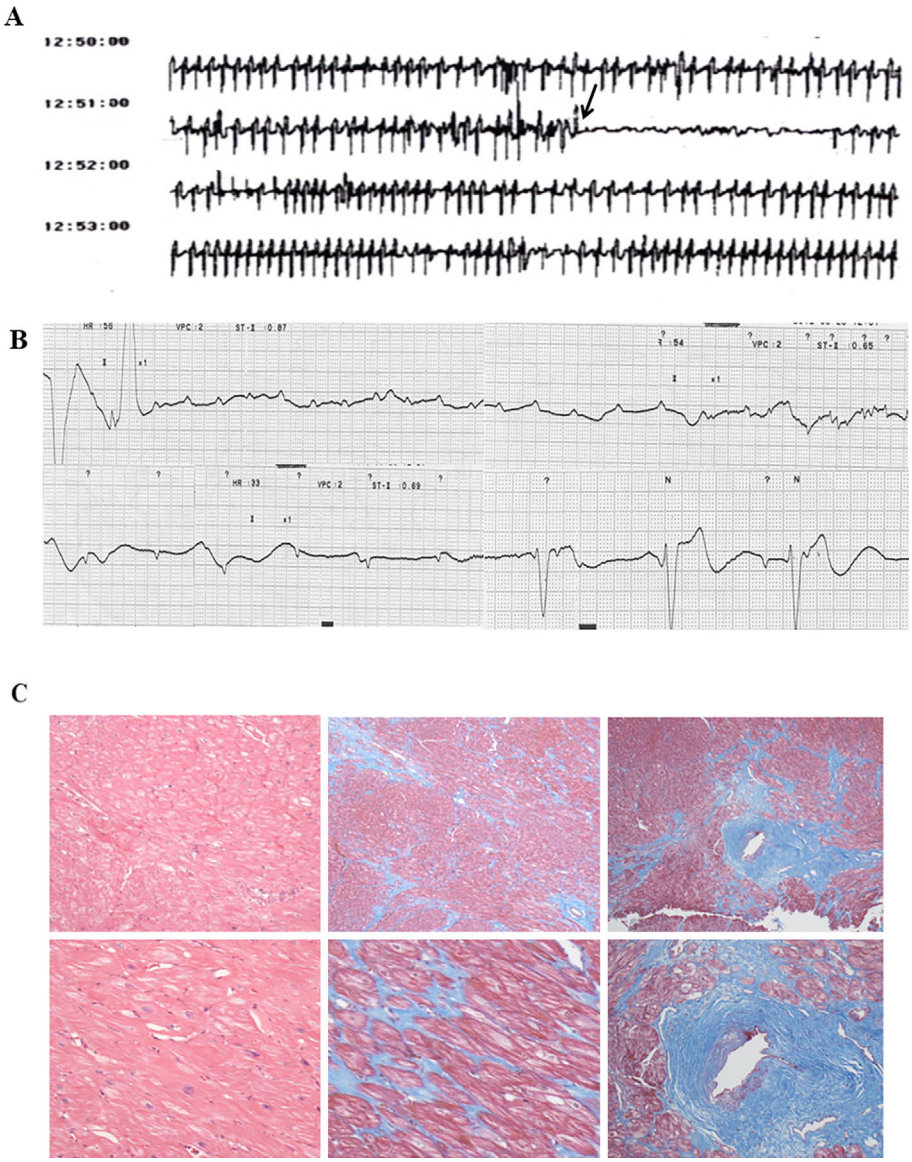

Figure 3. Continuous ECG-monitoring records when a syncope attack occurred (A, arrow). Complete AV block with a ventricular pause for 19 seconds was noted (B). Representative images stained with Hematoxylin and Eosin staining or Masson's trichrome of myocardial tissues removed at surgery (C). ECG: electrocardiogram, AV: atrioventricular

apparent at the upper border of the ventricular septum just below the membranous septum and right-ventricular side of the basal-mid septal wall, and subsequent T2-weighted imaging showing high signal intensity (Fig. 2). Cardiac catheterization showed no LVOT pressure gradient, and coronary angiography showed no significant coronary stenosis. However, after admission, he presented with complete AV block followed by long pause and syncope several times (Fig. 3A and B).

He successfully underwent permanent dual-chamber (DDD) pacemaker implantation to prevent syncope due to complete AV block, followed by aortic valve replacement with septal myocardial resection for the AS and HCM. Histology showed remarkable myocardial hypertrophy and disarray with extensive interstitial and perivascular fibrosis (Fig. 3C). After the surgery, echocardiography showed no aortic-valvular malfunction or LV dysfunction, including LVOT obstruction. His clinical course has been uneventful without syncope, VT events, or HF symptoms for six and a half years following the DDD pacemaker implantation and surgery.

Discussion

Patients with HCM frequently have arrhythmia and hemodynamic abnormalities and are prone to syncope and sudden death. Complex ventricular tachyarrhythmias are usually associated with syncope, but LVOT obstruction must be considered as another potential cause of syncope. While apparent LVOT obstruction was not shown by echocardiography or catheterization in the present case, the patient was complicated by severe AS and was difficult to diagnose. Eventu-

ally, the development of complete AV block was revealed to be the cause of syncope. We discussed whether DDD pacemaker or implantable cardioverter defibrillator (ICD) implantation would be more appropriate in this case and ultimately chose a DDD pacemaker because he developed syncope with complete AV block repeatedly. We therefore diagnosed his syncope as being due to complete AV block. We also concluded that his non-sustained VT on ambulatory ECG did not suggest a high risk for sudden death because he lacked any other major factors indicating a high risk, such as a family history, LVOT obstruction, wall thickness ≥ 30 mm, and abnormal blood response to exercise (2).

The incidence of arrhythmia in HCM is well documented (3). Life-threatening VT is associated with syncope and sudden cardiac death in HCM. In contrast, reports about AV-conduction disease are rare. However, Fananapazir et al. reported an abnormal His-Purkinje conduction in 23% of HCM patients who survived sudden cardiac death (4). It suggests that some patients with HCM may have syncope or sudden cardiac death related to complete AV block. The cause and etiology of AV block in HCM are unclear. Specific mutations concerning AV block have not been identified in studies on genetics of HCM. Several histopathologic reports in patients with HCM accompanied by advanced AV conduction disorders have shown interstitial fibrosis or myocardial necrosis in the conduction system and abnormalities in the small intramural coronary arteries with thickened walls and luminal narrowing (5). Myocardial ischemia, autonomic dysfunction, and an abnormal vascular response may also be underlying mechanisms of complete AV block. CMR facilitates the visualization of myocardial fibrosis and scarring *in vivo* by virtue of LGE.

In addition, T2-weighted imaging presents the visualization of edematous myocardium or inflammation *in vivo*, which may be indicative of recently sustained myocyte injury in HCM (6). In the present case, CMR showed inhomogeneous remarkable LV hypertrophy with extensive LGE, which was apparent in the lesions at the upper border of the ventricular septum and right-ventricular side of the basal-mid septal wall; the former may correspond to the branching portion of the His bundle and upper portion of the left bundle branch, and the latter may correspond to the proximal portion of the right bundle branch. High signal intensity on T2-weighted imaging suggested ongoing inflammation or myocardial injury of the AV conduction system, leading to development of complete AV block. Interestingly, since feline cardiomyopathies, including HCM, are occasionally associated with complete AV block, Kaneshige et al. examined the conduction systems histologically in 13 feline cases of HCM and complete AV block (7). They reported the anatomical basis of complete AV block, where marked degeneration and fibrous replacement of the AV conduction system were consistently observed in the combined regions of the branching portion of the His bundle and the upper portion of the left bundle branch. CMR findings suggest that our case may correspond to these feline cases with regard to histology. In addition, the findings in our case may be similar to those in cases of complete AV block occurring after septal myectomy or septal alcohol ablation in HCM patients. However, the pathological process may instead be related to the aging phenomenon seen in elderly patients with idiopathic or primary heart block.

Although the literature suggests that patients with HCM may rarely develop AV block, this combination must be considered in the differential diagnosis of syncope, as it affects the approach to the treatment. During the long-term follow-up of patients with HCM, it is prudent to be alert for the development of abnormal AV conduction. When patients with HCM show bundle block, it may suggest the possible development of AV block in the future. In addition, CMR with LGE and T2-weighted imaging may be helpful for assessing the possibility of AV block development. While the presence of LGE by CMR is generally associated with ventricular arrhythmia and its prognosis, it may also be useful for detecting conduction abnormalities.

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

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