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Case Report

Cardiac sarcoidosis, the complete atrioventricular block of which was completely recovered by intravenous steroid pulse therapy

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ABSTRACT

Atrioventricular block (AVB) in individuals with cardiac sarcoidosis (CS) is one of the major complications caused by inflammation of the conducting system of the heart, as a sign of worse prognosis. We report the case of a 53-year-old Japanese woman whose electrocardiogram showed complete AVB by the clinical diagnosis of CS. We administered intravenous methylpredonisolone (1 g/day) for 3 days. On the second day of steroid pulse therapy, the complete AVB improved to sinus rhythm of 1st degree AVB and complete right bundle branch block. Normal sinus rhythm was then observed after oral steroid therapy. These results suggest that in cases of complete AVB, steroid pulse therapy with a strong anti-inflammatory effect may be recommended first.

<**Learning objective:** This case illustrates a typical case of CS with complete AVB, but the cardiac contraction was normal. In this setting, steroid pulse therapy may be effective when (1) the active inflammation of the conduction system can be suppressed by steroid pulse therapy; (2) the time to start steroid therapy is short enough to recover.>

Introduction

Atrioventricular block (AVB), a common complication of cardiac sarcoidosis (CS), is caused by inflammation of the conducting system of the heart. AVB is a sign of worse prognosis, and thus its early diagnosis and treatment are important. There have been some reports of CS patients in which atrioventricular (AV) conduction disturbances were improved by steroid therapy [1]. However, there have been few reports of patients with CS who underwent steroid pulse therapy for AVB. We report herein the case of a patient with CS whose complete AVB was completely recovered by intravenous steroid pulse therapy followed by oral steroid treatment.

Case report

A 53-year-old Japanese woman was admitted to our hospital with exertional shortness of breath and dizziness that had begun one month earlier. She had no history of cardiovascular disease. On physical examination, the Cannon sound was audible on the chest. An electrocardiogram (ECG) showed complete AVB with an escaped rhythm of the left bundle branch block type (Fig. 1A). The left ventricular ejection fraction (LVEF) on echocardiography was 66% with no abnormality in the size of the heart. Interventricular septum was thickened to 13 mm without morphological changes. Chest X-ray showed no cardiomegaly or hilar lymph-adenopathy. The patient's plasma brain natriuretic peptide level was elevated (174.3 pg/mL), but other laboratory tests such as serum calcium and angiotensin-converting enzyme were within normal range. Cardiac magnetic resonance imaging (CMR) demonstrated late gadolinium enhancement (LGE) at the anteroseptal and lateral walls of the left ventricle (LV) (Fig. 2).

T2-weighted CMR showed high-intensity signal areas at the anteroseptal wall of the LV (Fig. 1D). ^{67}Gallium imaging and ^{18}F-fluorodeoxyglucose positron emission tomography (^{18}F-FDG PET), performed under long-fasting state >18 h [2], showed increased uptake in the lateral and anteroseptal wall of the LV and in the hilar and mediastinum lymph nodes (Fig. 2). Endomyocardial biopsy was performed from the right ventricle, but typical non-caseating granulomatous changes were not detected.

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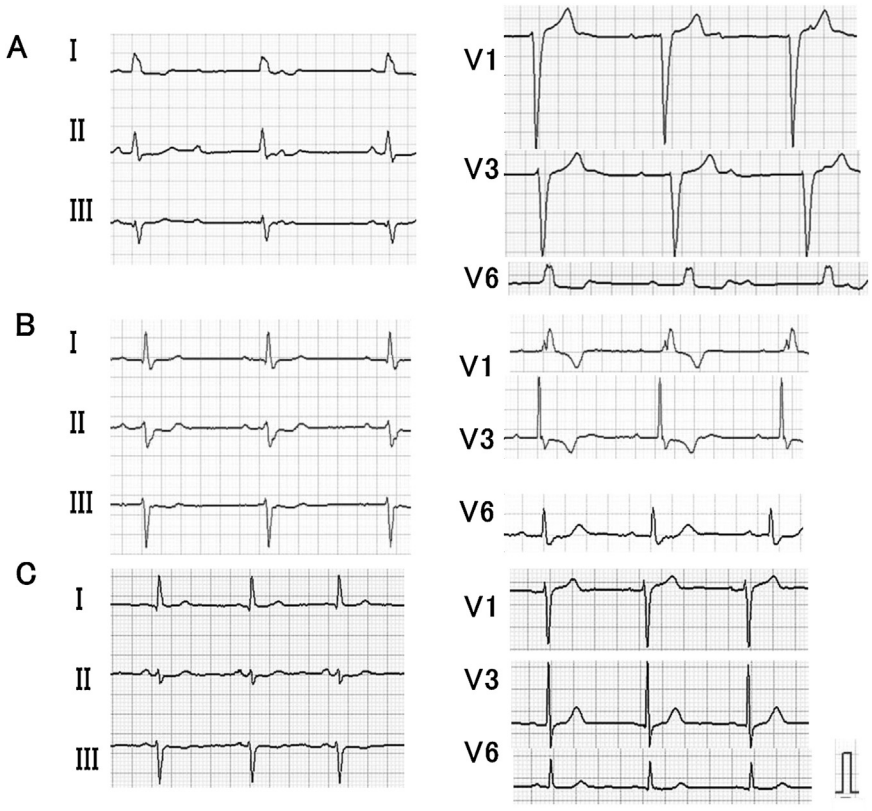

Fig. 1. The electrocardiogram (ECG) of the patient, a 53-year-old woman, showed complete atrioventricular block (AVB) with escape beats of the left bundle branch block type (A). ECG showed 1°AVB with complete right bundle branch block on the second day of steroid pulse therapy (B). ECG, after 2 months of oral steroid therapy, showed normal sinus rhythm (C).

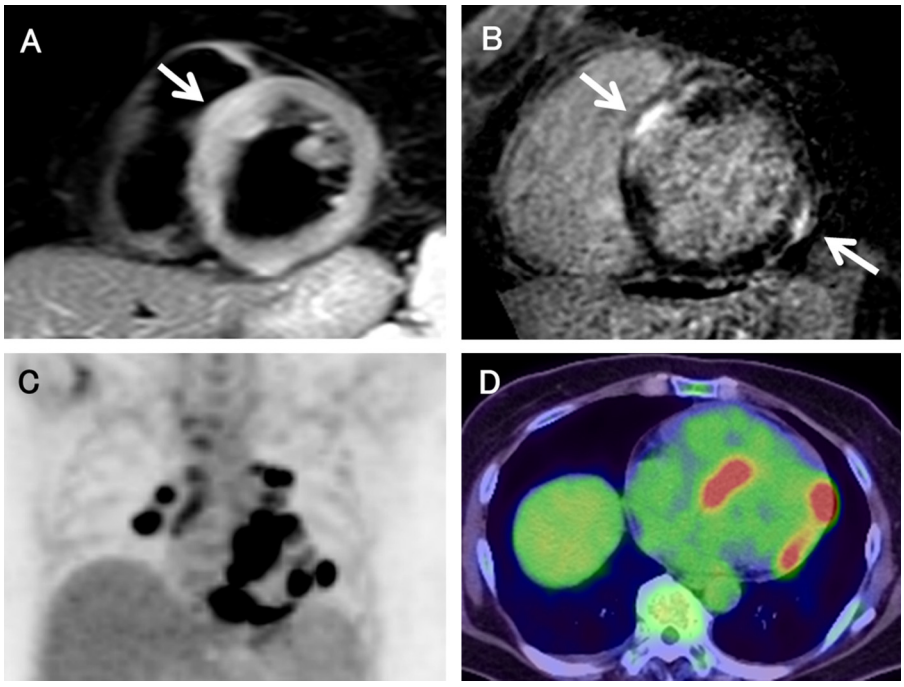

Fig. 2. T2-weighted cardiac magnetic resonance (CMR) showed high-intensity signal areas and wall thickening at anteroseptal wall of the left ventricle (arrow) (A). CMR imaging demonstrated late gadolinium enhancement at the anteroseptal and lateral walls of the left ventricle (arrow) (B). ^{18}F-fluorodeoxyglucose positron emission tomography images demonstrating abnormal uptake in the hilar lymph nodes and the anterior and inferior wall of the left ventricle (C), horizontal plane, the interventricular septum and the lateral wall of the left ventricle (D).