

[ Case Report ]

## Focal myocarditis with complete atrioventricular block as an initial presentation of rheumatic fever

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### SUMMARY

We encountered a case of rheumatic fever with focal myocarditis and complete atrioventricular block. A cardiac MRI, which enhances inflammation, enhanced the focal dyskinetic lesion. Therefore, one should consider rheumatic fever when complete atrioventricular block occurs with a focal myocarditis, which can be detected and tracked by a cardiac MRI.

**Key words:** rheumatic fever, focal myocarditis, complete atrioventricular block, cardiac MRI

### I. Introduction

Focal myocarditis with complete atrioventricular block is an extremely rare complication of acute rheumatic fever. We encountered a case of focal myocarditis in a portion of the basal interventricular septum that was complicated by a complete atrioventricular block as a result of acute rheumatic fever. To the best of our knowledge, this is the first report of a focal myocarditis, in which a cardiac MRI was used to track its progress, with complete atrioventricular block that was a complication of acute rheumatic fever.

### II. Case

A 10-year-old girl was admitted after presenting with convulsive syncope. She had a fever prior to her convulsive syncope, but was afebrile at the time of syncope. Her electrocardiogram showed complete atrioventricular block, a P rate of 167 beats per minute, and a QRS rate of 30 beats per minutes without abnormal Q waves or ST changes (Fig. 1). An echocardiogram showed a normal left ventricular ejection fraction (55%), but focal dyskinetic wall motion of the interventricular septum at a region below the membranous septum. Laboratory tests revealed a plasma brain natriuretic peptide level of 253.5 pg/mL and plasma troponin I level of 2.90 ng/mL, suggesting myocardial injuries. Plasma anti-streptolysin O level was 296 IU/mL and a rapid antigen detection test for group A streptococcus was positive. These clinical findings fulfilled Jones criteria; therefore, she was diagnosed with rheumatic carditis with complete atrioventricular block.

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Fig. 1 The electrocardiogram performed on admission showed complete atrioventricular block with an incomplete right bundle branch block and left posterior hemiblock. The P rate was 167 beats per minute, while the QRS rate was 30 beats per minute. There was neither an abnormal Q wave nor ST changes.

We emergently inserted a temporary pacemaker via the right internal jugular vein. Prednisolone, aspirin, and immunoglobulin were administered to control the cardiac inflammation. We concurrently administered aminobenzyl penicillin for group A *streptococcus* elimination. On the 2nd hospital day, the complete atrioventricular block resolved and the pacing rate was gradually decreased. The pacemaker was removed on the 5th hospital day. She was started on enalapril on the 6th hospital day and prednisolone was decreased every 7 days without atrioventricular block recurrence.

A cardiac MRI, obtained 10 days after admission, revealed a high intensity signal in a region of the basal interventricular septum on black blood short-tau inversion recovery image, just inferior to the membranous septum (Fig. 2A), suggesting focal myocardial edema caused by acute myocarditis. This lesion was also dyskinetic, while echocardiogram showed similar findings as described previously.

Twenty-nine days after admission, she was discharged with amoxicillin (10 mg/kg/day) as a secondary prophylaxis.

Eight months after discharge, the high intensity signal region on black blood short-tau inversion recovery images or late gadolinium enhancement of left ventricular myocardium including interventricular septum on cardiac MRI were not observed (Fig. 2B, C), suggesting that there was no myocardial inflammation or myocardial fibrosis.

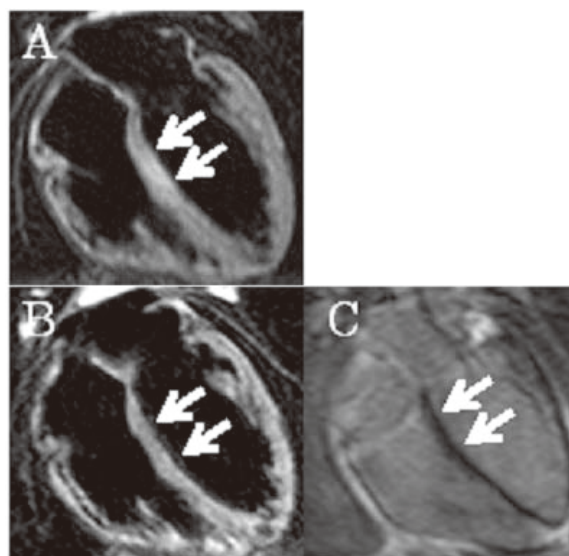


Fig. 2 Cardiac MRI demonstrates a high intensity signal on black blood short-tau inversion recovery image (A, arrows) in a region of the base of the interventricular septum, 8 days after admission. The high signal intensity on black blood short-tau inversion recovery image disappeared (B, arrows) and late gadolinium enhancement of left ventricular myocardium including interventricular septum was not detected (C, arrows) at 8 months (7 months after discharge).

### III. Discussion

To the best of our knowledge, this represents the first report of a focal myocarditis with complete atrioventricular block, which was followed using a cardiac MRI, associated with rheumatic fever. This case demonstrated two important clinical issues.

First, rheumatic fever can cause a focal myocarditis with complete atrioventricular block. We diagnosed rheumatic fever in our patient based on the fulfillment of one major (carditis) and three minor (fever, elevated acute-phase reactants, and prolonged PR interval) Jones criteria[1] with evidence of a preceding group A *streptococcus* infection.

Rheumatic carditis occurs in approximately 65% of patients with rheumatic fever[2] and can cause any degree of atrioventricular block. A recent study described first-degree block in approximately 72.3% of rheumatic fever patients, second-degree block in 1.5%, and complete heart block in 4.6%. [3] Generally, rheumatic carditis has been considered as a pancarditis involving

the heart valves, myocardium, and pericardium.[4] Carano et al. reviewed 25 complete atrioventricular block cases and reported rheumatic carditis with complete atrioventricular block and normal contractility. However, the presence of a focal myocarditis was not mentioned.[5]

In the present case, using a cardiac MRI, we detected the inflammatory lesion just below membranous septum, which is consistent with a diagnosis of focal myocarditis. The inflammatory lesion involved a route of the cardiac conduction system, originating from the Koch triangle and the bundle of His on the interventricular septum.[6] Therefore, we should not overlook the possibility of focal myocarditis induced by rheumatic fever in a patient with an acquired sudden onset of a complete atrioventricular block.

Second, cardiac MRI is useful for detecting the inflammatory lesion of this disorder. The cardiac MRI demonstrated a high intensity signal on black blood short-tau inversion recovery images at the base of the interventricular septum, in a region that was just below the membranous septum. We diagnosed a focal myocarditis using the cardiac MRI findings in conjunction with elevated levels of troponin I and brain natriuretic peptide. As shown by Abdel-Aty et al, T2-weighted images offer high single-sequence accuracy in the identification of myocarditis with areas of high intensity signal. This was the only cardiac MRI parameter that correlated with serum markers of acute myocardial injury.[7,8]Moriwaki et al. have also reported a case of isolated myocarditis in adult-onset acute rheumatic fever, in which the clinical course was followed by cardiac MRI.[9]High intensity signal on T2-weighted images represent interstitial edema. This lesion was recognized during the acute phase of myocarditis, but subsequently, it completely disappeared. Late gadolinium enhancement was not observed in the convalescent phase, suggesting that cardiac MRI is a useful tool for tracking the clinical course of myocarditis caused by rheumatic fever.

In the present case, we concluded that the inflammation of a region of the basal interventricular septum led to complete atrioventricular block. Similar

phenomena have already been reported. Some tumors, such as in lung cancer, occasionally metastasize to the myocardium. When a metastasized cardiac tumor infiltrates into the right atrium, it can cause complete atrioventricular block.[10]Since the cardiac conduction system originates from the Koch triangle and the bundle of His is on the interventricular septum,[6]a neoplastic change protruding into the atrium can inhibit the cardiac conduction system, in the same manner as inflammation.

Almost all reports of complete atrioventricular block caused by a cardiac metastatic tumor have been diagnosed by autopsy; however, a cardiac MRI may have detected these lesions while these patients were still alive.

In conclusion, rheumatic fever can cause a focal myocarditis with complete atrioventricular block. We should not overlook focal myocarditis with complete atrioventricular block and normal contractility as a potential complication of rheumatic fever. A cardiac MRI is a useful tool for detecting the inflammatory lesion, tracking the clinical course, and clarifying the pathogenesis of myocarditis associated with rheumatic fever.

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