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Systemic Juvenile Idiopathic Arthritis Accompanied by Immune Myocarditis

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We report here the case of a 13-year-old boy with refractory systemic juvenile idiopathic arthritis, characterized by acute myocarditis. The patient had various symptoms, including fever, rash, and polyarthritis, and subsequently, he developed chest pain, nausea, and vomiting. From the biochemical tests, it proved the existence of NT-proBNP (N-terminal fragment of B-type natriuretic peptide) 3450 pg/mL, cTnI (cardiac troponin I) 0.635 μg/L, interleukin 6 (IL-6) 167.20 pg/mL, and ferritin 16,500 ng/mL. Electrocardiogram showed ST segment down and a decrease in T-wave amplitude. Echocardiography revealed thickened ventricular septum and ventricular wall and a moderate pericardial effusion (Figs. 1A, B).

Cardiac magnetic resonance illustrated subendocardial patchy perfusion defect of the left ventricular inferior wall, inferior septum, and inferior lateral wall (Figs. 2A, B), indicating the clinical signs of myocardial ischemia. The delayed myocardial enhancement exhibited that the left and right ventricles were scattered and strengthened (Figs. 3A, B), representing myocardial fibrosis.

The patient did not respond to high-dose methylprednisolone and γ-globulin and was subsequently adjusted to tocilizumab (IL-6 monoclonal antibody). After 48 hours of tocilizumab administration, his symptoms were relieved. The cardiac dysfunctions recovered during the 18 months of follow-up.

The systemic juvenile idiopathic arthritis–related cardiac complications are critically affecting pericardium, myocardium, endocardium, coronary artery, heart valves, and cardiac conduction.[1] However, they are obviously underestimated for the absence of specific means to detect the subclinical forms of cardiovascular system damage.[2,3] Cardiac magnetic resonance is a powerful technology to detect cardiovascular system damage at an early stage, thereby stratifying the risk of patients and formulating individualized treatment.

Interleukin 6 is involved in myocardial inflammation and ventricular remodeling.[4] Up to now, tocilizumab has achieved significant clinical affects with promising benefits.[5] Most importantly, more researches are still needed to identify IL-6–sensitive individuals at an early stage and determine the best time to initiate the therapy.

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Patient has provided informed consent for publication of the case.

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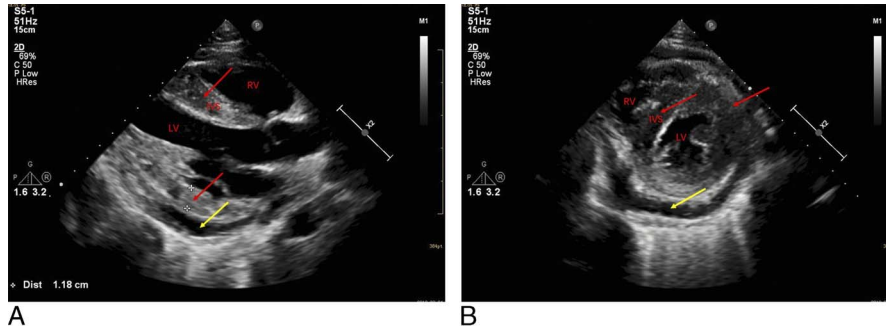

FIGURE 1. Echocardiography. Parasternal long-axis view (A) and short-axis view (B) of papillary muscle of left ventricle. Thickened interventricular septum and posterior wall of left ventricle (red arrows) and moderate to large amount of pericardial effusion (yellow arrows).

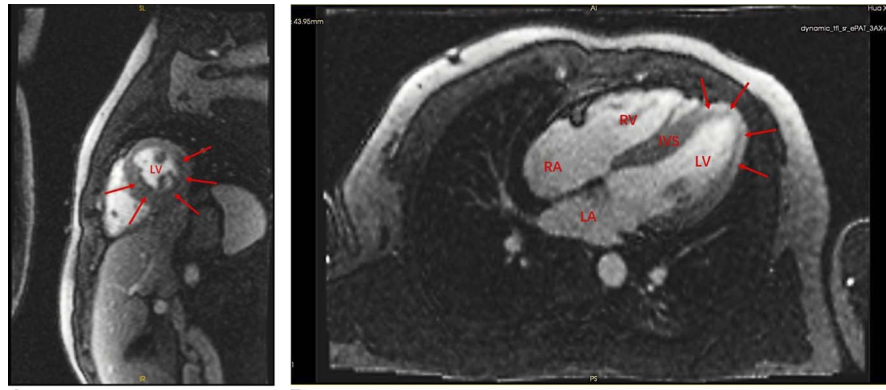

FIGURE 2. Cardiac magnetic resonance. A and B, First-pass perfusion imaging suggests that the left ventricular inferior wall, inferior septum, and inferior lateral wall have subendocardial patchy perfusion defects (red arrows).

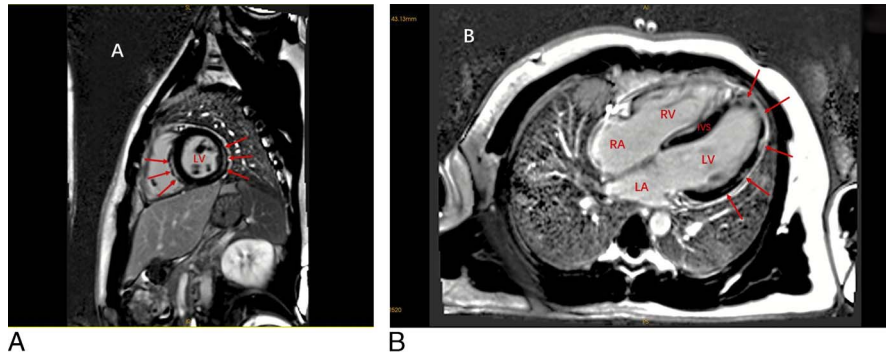

FIGURE 3. Cardiac magnetic resonance. A and B, Delayed myocardial enhancement indicates that the left and right ventricles are scattered and have delayed enhancement, which is obvious under the epicardium and middle myocardium (red arrows).