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Case Report

Active cardiac sarcoidosis in a patient with adult-onset Kawasaki disease

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ABSTRACT

Adult-onset Kawasaki disease is a rare condition. Cardiac sarcoidosis is an uncommon cardiomyopathy which is characterized by progressive cardiac dysfunction, and abnormality on electrocardiography and morphological aberration of the heart. We report a first case of a combination of these rare conditions. The patient was initially diagnosed with Kawasaki disease based on the coronary artery aneurysms and a past medical history at the age of 20 years which was typical of Kawasaki disease. Decades later, he developed progressive cardiac dysfunction and a sudden-onset atrioventricular block. Laboratory and imaging results revealed severe myocardial damage and inflammation which were unexplainable by coronary artery ischemia. We diagnosed him with cardiac sarcoidosis based on a Japanese guideline to diagnose cardiac sarcoidosis. A cardiac resynchronization therapy defibrillator was implanted and he received oral steroid therapy. This rare combination of adult-onset Kawasaki disease and cardiac sarcoidosis may suggest the causative association of these conditions.

<**Learning objective:** This is the first report of a rare combination of adult-onset Kawasaki disease and cardiac sarcoidosis. Kawasaki disease is not just a disease of children. Physicians should include Kawasaki disease in the list of differentials for unknown fever or eruptions. In patients with progressive heart failure and atrioventricular block, the possibility of cardiac sarcoidosis should be examined using various imaging modalities even if they had a known cause of cardiac dysfunction.>

Introduction

Kawasaki disease is a type of vasculitis which preferentially affects children. Kawasaki disease can occur rarely in an adult. A previous report showed that coronary aneurysm occurs in 5% of patients with adult-onset Kawasaki disease, and the patients often suffered from ischemic complications, leading to severe ischemic cardiomyopathy [1].

Cardiac sarcoidosis, which is a granulomatous disease of unknown etiology characterized by non-caseating granulomas in involved organs, is also rare and difficult to diagnose because of its various clinical courses. In fact, only 40–50% of patients with cardiac sarcoidosis diagnosed at autopsy have the correct diagnosis made during their lifetime [2]. The most common presented findings of cardiac sarcoidosis are progressive ventricular dysfunction and arrhythmia, including mainly atrioventricular block.

In the current report, we present an extremely rare combination of adult-onset Kawasaki disease and cardiac sarcoidosis. This case tells us one should suspect cardiac sarcoidosis in case of progressive cardiac dysfunction and atrioventricular block even in the patients with a known cause of cardiac dysfunction.

Case report

A 58-year-old man with a following past medical history came to the emergency room with dyspnea worsening over the previous 4 days.

At the age of 20 years, he suffered from a high fever sustained for 18 days, polymorphous rash, cervical lymphadenopathy, strawberry tongue, and desquamation of the hands. He was treated with some intravenous antibiotics as for a bacterial infectious disease, but antibiotics were not effective. About half a

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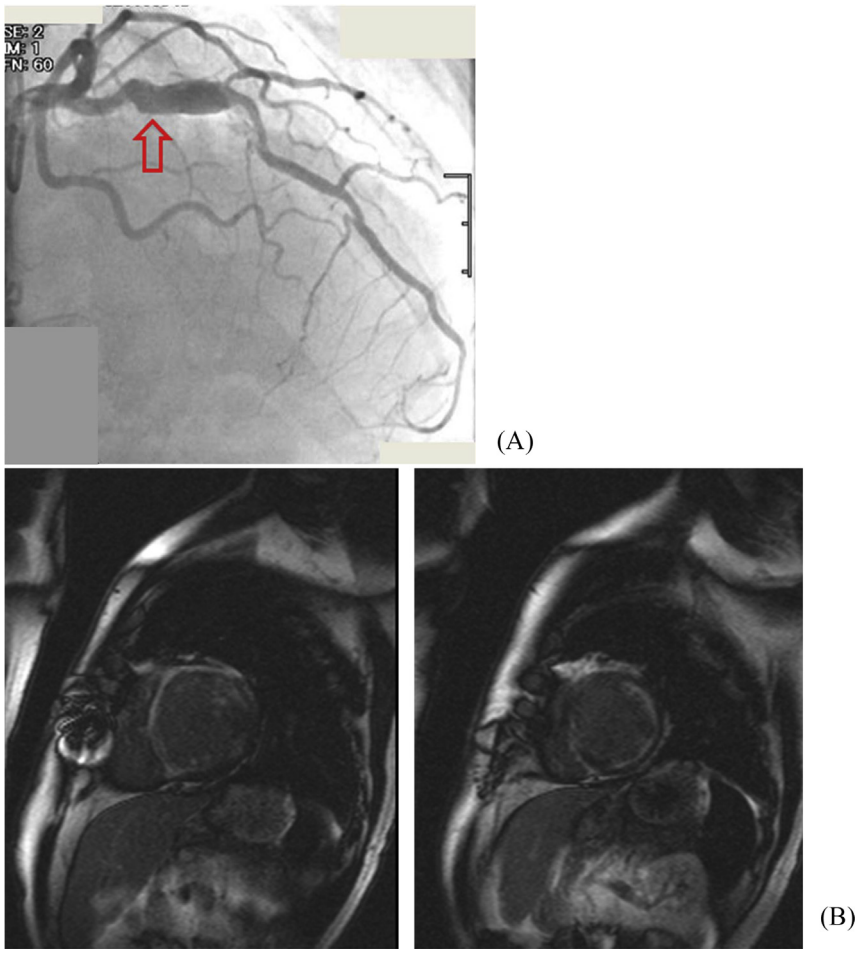

Fig. 1. (A) Coronary angiography shows coronary artery aneurysm at left anterior descending artery #6. (B) Cardiac magnetic resonance imaging with gadolinium transmural and patchy-delayed enhancement in diffuse left ventricle myocardium.

month later, symptoms spontaneously resolved. At the age of 48 years, he was diagnosed with Kawasaki disease based on his past medical history at the age of 20 years and coronary angiography (CAG) showing coronary artery aneurysm with severe calcification on the proximal portion of the left anterior descending artery (Fig. 1A). At the age of 55 years, he was found to have complete right bundle branch block and first-degree atrioventricular block on an electrocardiogram (ECG) tracing, but his left ventricular function was preserved at that time [left ventricular ejection fraction (LVEF): 57%]. The next year, he developed heart failure with left ventricular dysfunction (left ventricular diastolic dimension/systolic dimension: 61/47 mm, LVEF: 36%, CAG: #1 100%, #6 aneurysm, #7 75%) and severe mitral regurgitation. He underwent coronary bypass graft surgery (a graft using left internal thoracic artery to left anterior descending artery; sequential grafts using radial artery to first diagonal branch and #4 posterior descending artery), mitral valve repair, and tricuspid annuloplasty. Despite successful surgical procedure, left ventricular function did not improve except for some decrease in the left ventricular diastolic dimension (diastolic dimension/systolic dimension: 56/47 mm, LVEF: 30%).

Six months after the cardiac operation, he developed dyspnea. On admission, inspiratory rales were heard at both lungs with oxygen saturations of 90% on room air. There were a protodiastolic gallop and grade 2 systolic murmur in the 4th left intercostal space. He was diagnosed with acute decompensated heart failure [New York Heart Association (NYHA) class IV] in the emergency room, and was immediately treated with oxygen and intravenous diuretics. Blood sample investigations revealed that the serum level of B-type natriuretic peptide was 1284 pg/mL (normal: <18.4 pg/mL), and troponin T 0.044 ng/mL (normal: <0.014 ng/mL). ECGs showed a tri-fascicular block (left axis deviation, first-degree atrioventricular block, and complete right bundle branch block), and wide QRS duration (170 ms). A monitor ECG revealed non-sustained ventricular tachycardia. A transthoracic echocardiogram showed severely reduced left ventricular function (LVEF: 17%) and abnormal septal wall thinning (wall thickness <6 mm). CAG proved patency of bypass graft. Dyspnea improved 3 days after admission. However, he suffered from paroxysmal dyspnea, and an ECG monitor tracing showed an advanced atrioventricular block at 12th day after hospitalization. He received a transvenous temporary pacemaker.

The serum level of angiotensin-converting enzyme was 31.3 U/L (normal: 7.7–29.4 U/L). Tuberculin test turned negative, and there was no obvious sign of uveitis. Computed tomography showed bilateral hilar lymphadenopathy. A cardiac magnetic resonance imaging showed a broad damage of the cardiac tissue, which did not follow the coronary artery distribution (Fig. 1B). 67Gallium scintigraphy and fluorodeoxyglucose (FDG)-positron emission tomography (PET) revealed broad inflammation of the cardiac tissue (Fig. 2A and B). FDG-PET also showed bilateral hilar lymphadenopathy (Fig. 2C). He underwent a right ventricular endomyocardial biopsy. The pathological analyses proved fibrotic changes in myocardium, but no non-caseating epithelioid cell