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Case Report

A case of idiopathic giant cell myocarditis with a past history of sarcoidosis

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ABSTRACT

A 70-year-old woman with back pain and breathlessness was referred to our hospital for suspected myocardial infarction. Coronary angiogram was normal and endomyocardial biopsy showed inflammatory cell infiltrates consisting of eosinophils and multinucleated giant cells. The clinical course was hemodynamically fulminant, but steroid therapy improved the cardiac function. Interestingly, this patient had a past history of sarcoidosis. We diagnosed this case with idiopathic giant cell myocarditis (IGCM) from its clinical course. However, whether IGCM and cardiac sarcoidosis belong to the same histological entity has been debated. This case is important with respect to the pathogenic association between these two disorders.

<**Learning objective:** Both idiopathic giant cell myocarditis and cardiac sarcoidosis are known to show multinucleated giant cell infiltration in the myocardium histologically. In particular, idiopathic giant cell myocarditis is a severe and fulminant disease, making its early diagnosis and treatment important. Although it is difficult to diagnose these diseases, endomyocardial biopsy is useful to decide the treatment strategy in such disorders that may assume a fulminant course.>

Introduction

Myocarditis exhibits a wide spectrum of signs and symptoms, and clinical outcomes range from asymptomatic to fatal [1]. Of these, idiopathic giant cell myocarditis (IGCM), which is a rare condition characterized by multinucleated giant cell infiltration and extensive myocardial cell necrosis, has an especially poor prognosis. Cooper et al. reported that the median transplant-free survival from symptom onset of this disease was only 5.5 months [2]. Meanwhile, cardiac sarcoidosis (CS) is known to also show multinucleated giant cell infiltration in the myocardium histologically [3], and therefore there has been discussion as to whether or not these two diseases belong to the same entity [1]. We experienced a patient who had a past history of sarcoidosis, and in whom IGCM was strongly suspected early in the clinical course.

Case report

A 70-year-old woman experienced sudden thoracodorsal pain 10 days before presentation and complained of gradually worsening breathlessness. Vomiting and dyspnea on effort had worsened beginning 3 days before presentation. She consulted her family physician who then referred her to our hospital after a diagnosis of heart failure due to myocardial infarction. Bilateral hilar lymphadenopathy was observed on chest plain computed tomography 10 months prior to the onset of the current illness (Fig. 1A), and prior to that an ophthalmologist had noted uveitis. From both of these conditions, she was thought to suffer from sarcoidosis (extracardiac), and was followed up conservatively. The lymph nodes appeared to shrink without any medication (Fig. 1B). Her height was 144 cm, weight 40 kg, and there were neither risk factors for coronary artery disease nor any other relevant past history. On physical

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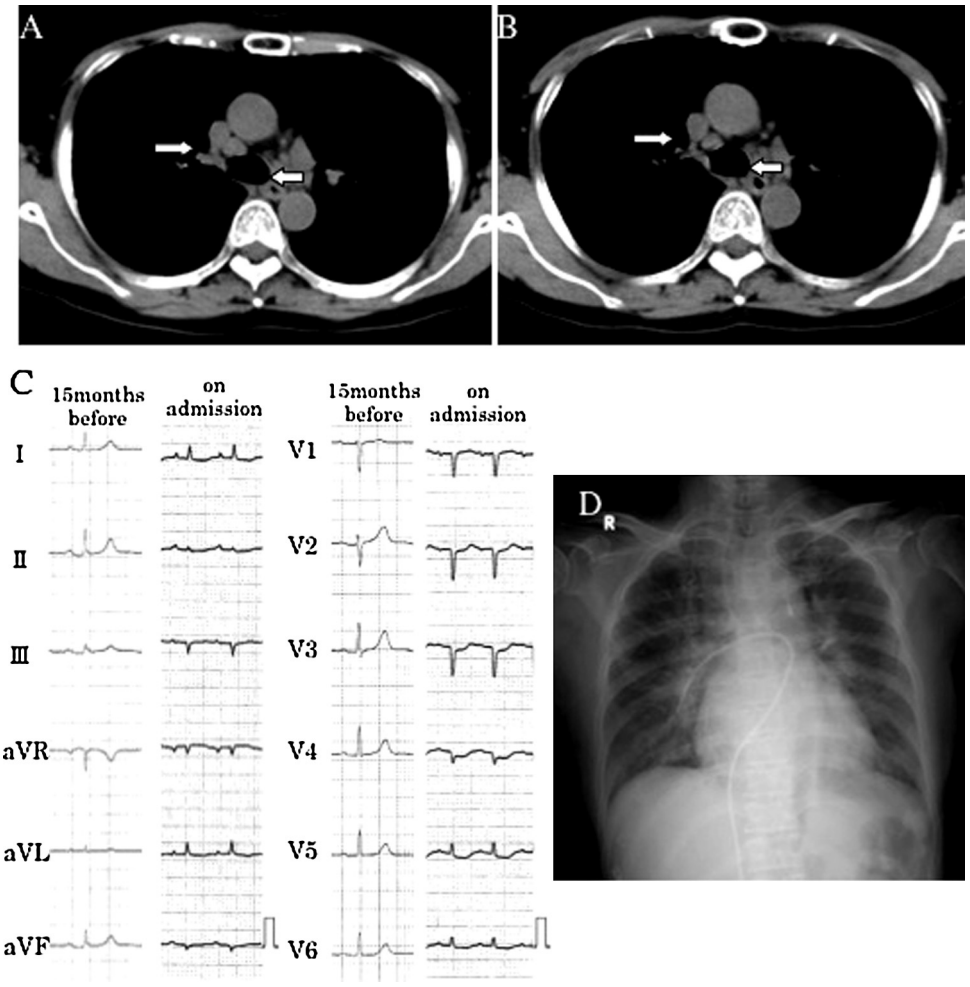

Fig. 1. Chest plain computed tomography 10 months before admission shows swollen mediastinal lymph nodes (white arrows) (A). Their size subsequently decreased spontaneously without medication (B). Comparison of electrocardiogram (ECG) on admission with that of 15 months earlier (C) and chest radiography on admission (D). ECG on admission shows low voltage in the limb leads and QS pattern in leads III, aVF, and V1–4, indicating extensive myocardial damage. Chest radiography obtained after cardiac catheterization showed bilateral pulmonary congestion and cardiomegaly.

examination, her complexion was pale and she was afebrile. Her blood pressure was 122/69 mmHg, and she was tachycardic with a pulse of 132 bpm. On chest auscultation, coarse crackles were noted in the lung fields. There were no murmurs. Blood tests revealed elevated levels of troponin I (6.197 ng/mL), white blood cells (WBC: 8340/μL), lactate dehydrogenase (916 IU/L), aspartate aminotransferase (152 IU/L), creatine kinase (2924 IU/L), and creatine kinase-MB (80.9 ng/mL). The eosinophil count was not increased (1.1% of WBC). Brain natriuretic peptide was 4500 pg/mL. Angiotensin-converting enzyme (7.2 U/L), IgG (1427 mg/dL), IgA (260 mg/dL), and IgM (59 mg/dL) were not elevated. A 12-lead electrocardiogram (ECG) showed sinus tachycardia, with low voltage in the limb leads, and a QS pattern in leads III, aVF, and V1–4 (Fig. 1C). Chest radiography showed bilateral pulmonary congestion and cardiomegaly (cardiothoracic ratio 58%) (Fig. 1D). Both ventricles had diffuse hypokinetic motion (ejection fraction of left ventricle was 20%) with no mitral regurgitation and respiratory variation of inferior vena cava on ultrasound cardiography. Emergent coronary angiography was performed due to suspected acute coronary syndrome, but revealed normal coronary arteries, while a left ventriculogram showed diffuse severe hypokinesis. The mean pulmonary capillary wedge pressure was 31 mmHg and the cardiac index was 1.5 L/min/m^2. Under a provisional diagnosis of acute myocarditis associated with severe heart failure, an endomyocardial biopsy was performed from the posterior wall of the left ventricle, after which intra-aortic balloon pumping was initiated. Although her appearance and symptoms improved dramatically for a short while with intra-aortic balloon pumping support, a pale complexion, coldness of the extremities, and severe nausea gradually became evident. On the 3rd day ventricular tachycardia occurred frequently, and it became difficult to maintain her blood pressure, and so percutaneous cardiopulmonary support was commenced. Taking into consideration the possibility of fulminant viral myocarditis, gamma globulin (1 g/day) was administered for 4 days beginning on day 3 (total, 4 g). On the 5th day, an ECG showed advanced atrioventricular block and disappearance of the QRS complex. The results of endomyocardial biopsy obtained on the same day verified the presence of inflammatory cell infiltrates consisting mainly of lymphocytes, eosinophils, and multinucleated giant cells in the myocardial tissue. Marked fibrosis was also observed (Fig. 2A, B). Immunostaining with *Propionibacterium acnes* specific monoclonal antibodies (PAB antibody) of this biopsy specimen was negative for inflammatory lesions (Fig. 2C). From the clinical course and histological findings, IGCM, CS, and eosinophilic myocarditis were considered as the differential diagnosis, and steroid pulse therapy (methylprednisolone; mPSL 1 g/day) was started. Immediately thereafter restoration of sinus rhythm on the ECG monitor and left ventricular contraction on the echocardiogram were seen. After 3 days of steroid pulse therapy, steroid administration was continued (mPSL 40 mg/day