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Case Report

Isolated cardiac sarcoidosis requiring open-chest myocardial biopsy for differentiation from malignant lymphoma

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ABSTRACT

A 68-year-old woman with a history of hypertension was admitted to our hospital because of dyspnea during physical exertion. Echocardiography demonstrated impaired left ventricular systolic function, and her ejection fraction was reduced to 30%. Coronary angiography did not show significant stenosis. Endomyocardial biopsy showed only nonspecific findings without noncaseating granulomas. Cardiac magnetic resonance (CMR) imaging showed transmural late gadolinium enhancement on the basal part of the left ventricle. 18F-Fluorodeoxyglucose positron emission tomography (18F-FDG PET) showed abnormal focal uptake specific to the left ventricle; no abnormal manifestations in other organs were observed. The CMR and 18F-FDG PET features could not rule out either sarcoidosis or malignant lymphoma. Therefore, we conducted open-chest myocardial biopsy to differentiate between the two possible diseases. Histopathological findings showed noncaseating epithelioid cell granuloma, confirming isolated cardiac sarcoidosis. This is an example of a challenging case of diagnosing isolated cardiac sarcoidosis.

<**Learning objective:** We describe a case of "isolated" cardiac sarcoidosis that is of special interest as no abnormal manifestations of sarcoidosis were noted in organs other than the heart; moreover, the condition could not be diagnosed with transvenous endomyocardial biopsy, but was finally confirmed by open-chest myocardial biopsy.>

Introduction

Cardiac involvement is an important manifestation of sarcoidosis; it can cause heart failure and sudden death, and affects the prognosis of sarcoidosis. Early diagnosis of cardiac sarcoidosis is important to improve the prognosis of patients with sarcoidosis because steroid therapy is an effective treatment. However, the histological diagnostic yield of cardiac sarcoidosis by transvenous endomyocardial biopsy is around 20–30% [1,2]. Therefore, in the absence of extracardiac lesions of sarcoidosis, it is difficult to diagnose cardiac sarcoidosis. Here we present a case of "isolated" cardiac sarcoidosis that could not be diagnosed by transvenous endomyocardial biopsy, but was confirmed by open-chest myocardial biopsy.

Case report

A 68-year-old woman was admitted to our hospital because of dyspnea during physical exertion. She had a history of

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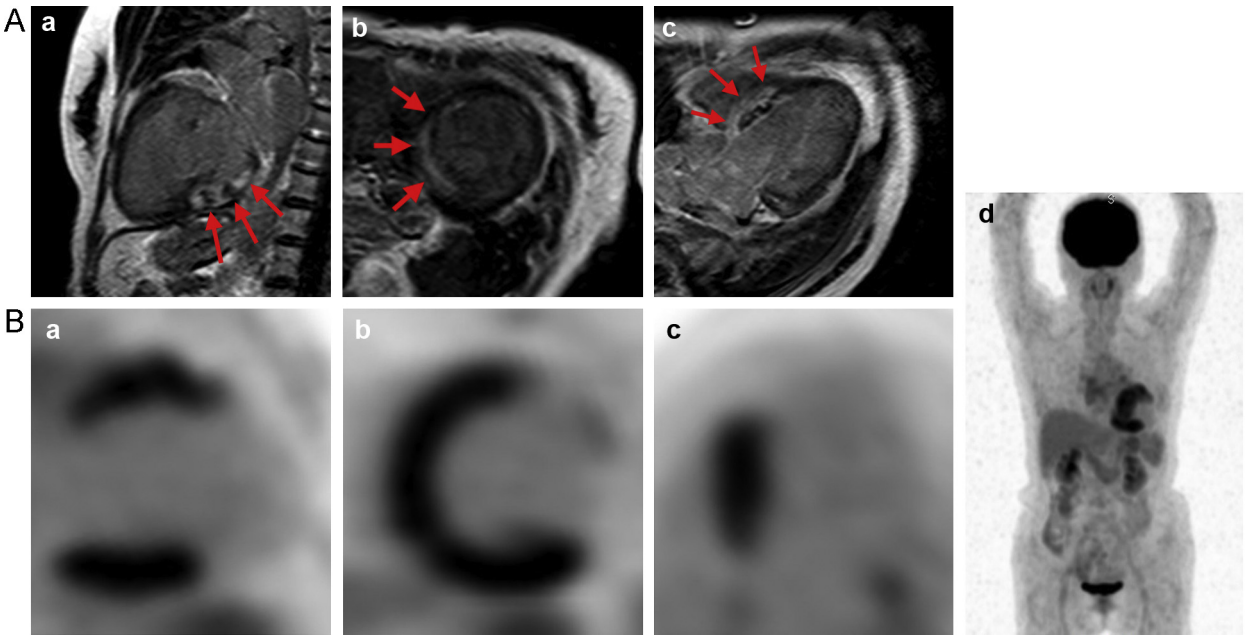

Fig. 1. (A) Cardiac magnetic resonance imaging showed transmural late gadolinium enhancement on the basal part of inferior and septal wall of the left ventricle (arrows): (a) Vertical long-axis view; (b) short-axis view; (c) horizontal long-axis view. (B) 18F-fluorodeoxyglucose positron emission tomography showed abnormal uptake in only the left ventricle: (a) vertical long-axis view; (b) short-axis view; (c) horizontal long-axis view; (d) whole body.

hypertension for 5 years and colon cancer 4 years previously. There was no history of drinking and no family history of heart diseases. On physical examination, her blood pressure was 140/58 mmHg, her pulse rate was 80 beats/min, and her oxygen saturation was 98%. Cardio-respiratory examination revealed bilateral moist rales and normal heart sounds. Superficial lymph nodes were not swollen. No obvious pretibial edema was noted. Chest X-ray showed a cardio-thoracic ratio of 61% with pulmonary congestion and pleural effusion. A 12-lead electrocardiogram showed normal sinus rhythm, first-degree atrioventricular (AV) block (PQ interval: 240 ms), incomplete right bundle branch block (QRS width: 110 ms), and left anterior hemiblock. Electrocardiographic monitoring revealed transient complete AV block at night. Echocardiography showed diffuse hypokinesis of the left ventricular wall with akinesis of the basal intact ventricular septum, and her ejection fraction was reduced to 30%. The laboratory data were as follows: serum brain natriuretic peptide level was increased to 458.0 pg/mL, and soluble interleukin-2 receptor (sIL-2R) level was increased to 1166 U/mL. Plasma angiotensin-converting enzyme activity and other tumor markers were all within normal limits.

Her symptoms promptly improved after medication with diuretics. Several examinations were conducted to diagnose the underlying disease. Coronary angiography did not show significant stenosis, while right ventricular endomyocardial biopsy showed nonspecific findings without noncaseating granulomas. Cardiac magnetic resonance (CMR) imaging showed transmural late gadolinium enhancement on the basal part of the inferior and septal wall at the left ventricle (Fig. 1A). The wall thickness was considerably increased in comparison with normal myocardium; it appeared to have been replaced by different tissue. Thoracic computed tomography (CT) image showed no swollen mediastinum lymph nodes, and no abnormal findings in the lung field. Gallium-67 scintigraphy and ^{18}F-fluorodeoxyglucose positron emission tomography (^{18}F-FDG PET) showed abnormal uptake only in the left ventricle, consistent with the abnormalities found by CMR; no abnormal manifestations were observed in other organs (Fig. 1B). There were no abnormal findings suggesting sarcoidosis in dermatology and ophthalmology. Isolated cardiac sarcoidosis was suspected, but cardiac malignant lymphoma could not be ruled out. Therefore, we conducted open-chest myocardial biopsy to refine the diagnosis. The procedure was carried out by a cardiovascular surgeon after obtaining informed consent from the patient. Macroscopic findings of epicardial surface were normal. Biopsy sites were determined at the basal part of the septal wall, and we approached from the anterior right ventricle under a beating heart. We used a straight needle designed for prostate biopsy (Bard Monopty Biopsy Needle; CR Bard, Inc., Covington, GA, USA) and resected three specimens guided by transesophageal echocardiography (TEE) (Fig. 2). Histopathological findings showed noncaseating epithelioid cell granuloma, suggesting cardiac sarcoidosis (Fig. 3).

Prednisolone was initiated to prevent the progression of sarcoidosis. The initial dose was 30 mg, and was gradually tapered. One year after starting prednisolone, the maintenance dose was 10 mg. Advanced AV block was improved, and ejection fraction was slightly improved in echocardiography.

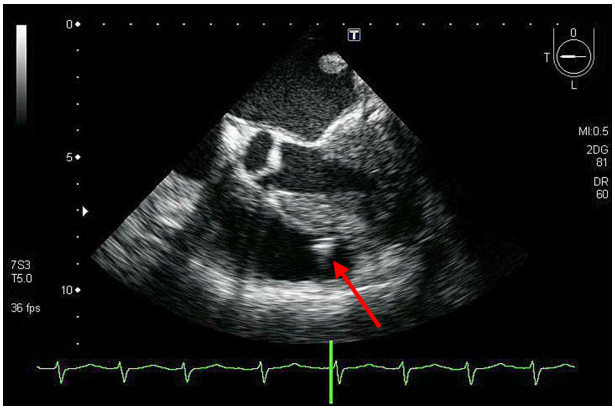

Fig. 2. Transesophageal echocardiography during surgery, showing a straight needle to biopsy sites (arrow).