Cardiac Sarcoidosis Mimicking Right Ventricular Dysplasia

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A 59-year-old woman with skin sarcoidosis was admitted to hospital for assessment of complete atrioventricular block. Cross-sectional echocardiography showed that the apical free wall of the right ventricle was thin and dyskinetic with dilation of the right ventricle. Thallium-201 myocardial imaging revealed a normal distribution. Both gallium-67 and technetium-99m pyrophosphate scintigraphy revealed no abnormal uptake in the myocardium. Right ventriculography showed chamber dilation and dyskinesis of the apical free wall, whereas left ventriculography showed normokinesis, mimicking right ventricular dysplasia. Cardiac sarcoidosis was diagnosed on examination of an endomyocardial biopsy specimen from the right ventricle. A permanent pacemaker was implanted to manage the complete atrioventricular block. After steroid treatment, electrocardiography showed first-degree atrioventricular block and echocardiography revealed an improvement in the right ventricular chamber dilation. Reports of cardiac sarcoidosis mimicking right ventricular dysplasia are extremely rare and as this case shows, right ventricular involvement may be one of its manifestations. (Circ J 2003; 67: 169–171)

Key Words: Atrioventricular block; Cardiac sarcoidosis; Endomyocardial biopsy; Right ventricle; Right ventricular dysplasia; Steroids

Sarcoidosis is a granulomatous disease of unknown cause that involves multiple organ systems, including the heart. Although cardiac sarcoidosis can lead to heart failure or sudden death, it can also remain clinically silent despite extensive involvement. Left ventricular wall motion abnormalities of varying degrees are common manifestations1-2 and wall thinning and focal asynergy in the basal portion of the interventricular septum are features of this disease! According to recent reports, hypertrophic cardiomyopathy may be a manifestation of sarcoidosis3-4. Here we describe an unusual case of cardiac sarcoidosis associated with right ventricular abnormalities that mimicked right ventricular dysplasia (RVD).

Case Report

A 59-year-old woman was admitted to Meijishinkyu University Hospital because of general fatigue. She had had a skin eruption 8 years ago, which was diagnosed as sarcoidosis, confirmed by skin biopsy. She had no past history of cardiovascular disease. The results of physical examination were unremarkable, except for bradycardia (heart rate, 45 beats/min). Routine blood test results were normal, except for an erythrocyte sedimentation rate (ESR) of 43 mm in the first hour. The serum level of angiotensin-converting enzyme (ACE) was 22.3 IU/L, slightly above the upper limit of normal (normal range, 8.3–21.4 IU/L). A chest radiograph showed right hilar lymphadenopathy and enlargement of the cardiac silhouette. An ECG taken 4 years prior to admission was normal (Fig 1A), whereas that taken on admission (Fig 1B) showed complete atrioventricular...

Fig 1. Twelve-lead ECG from health screening 4 years before admission was within normal limits (A). On admission (B), complete atrioventricular block and abnormal Q waves in leads V1-V3 were recorded. After 2 days of steroid treatment, the atrioventricular block improved from third-degree to first-degree (PR 0.32 s) (C).

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ular block and Q waves in leads V1–3. After a temporary pacemaker was implanted, she was transferred to Kyoto Prefectural University Hospital for evaluation and treatment of the complete atrioventricular block.

Two-dimensional transthoracic echocardiography revealed that the apical free wall of the right ventricle was thin and dyskinetic with dilation of the right ventricle and a small amount of pericardial effusion (Fig 2). Tricuspid insufficiency was not evident. In contrast, the left ventricle showed normal contractions. Thallium-201 (201Tl) myocardial imaging revealed normal perfusion. Both gallium-67 (67Ga) and technetium-99m pyrophosphate (99mTc-PYP) scintigraphy showed no abnormal uptake in the myocardium. Coronary angiography was normal, and left ventriculography showed normokinesis. However, right ventriculography showed chamber dilation, fissures in the anterior wall, and dyskinesis in the apical free wall, findings initially considered consistent with RVD (Fig 3). Non-caseating epithelioid granuloma with multinuclear giant cells was detected on the right ventricular endomyocardial biopsy (Fig 4). No fatty degeneration was observed. On the basis of these histopathologic findings, cardiac sarcoidosis was diagnosed, and the patient was begun on prednisolone, 60 mg every other day. A permanent pacemaker was implanted. After 2 days of steroid treatment, the complete atrioventricular block resolved, and first-degree block was resolved.
observed (Fig 1C). The dilated right ventricle gradually decreased; however, echocardiography revealed aneurysmal changes in the apical free wall, which had not changed since admission. Right hilar lymphadenopathy decreased, and the serum ACE level and ESR became normal.

Discussion

The echocardiographic manifestations of presumed or proved cardiac sarcoidosis include chamber dilatation, systolic dysfunction, and regional wall-motion abnormalities of the left ventricle and wall thinning of the basal interventricular septum is a well-known feature of this disease. On the other hand, RVD is a primary heart muscle disease affecting right ventricle and is characterized pathologically by extensive fatty replacement of the myocardium and clinically by dilation and impaired contraction of the right ventricle, with little or no impairment of the left ventricle. To our knowledge, this is the first report to document a case of cardiac sarcoidosis involving mainly the right ventricle.

The present patient may have been in the early phase of cardiac sarcoidosis, because of the rapid response to steroid treatment. Although early diagnosis is an important determinant of outcome, cardiac sarcoidosis involving chiefly the right ventricle is liable to be overlooked or misdiagnosed as other types of myopathy, such as RVD. In addition, neither perfusion abnormalities on 201Tl imaging nor abnormal 67Ga and 99mTc-PYP myocardial uptake was observed in this case, although radionuclide studies are considered helpful for detecting cardiac involvement and monitoring the response to steroid treatment in patients with sarcoidosis. The poor distribution of sarcoid lesions in the left ventricle may be the reason why the scintigraphic abnormalities were not detected in this patient.

Cardiac sarcoidosis is rarely diagnosed histologically because the sarcoid granulomas are distributed locally within the myocardium. In the present case, only 1 of the 4 specimens obtained by right ventricular endomyocardial biopsy was useful for diagnosis. Correct histological diagnosis is less likely in patients who have presumed cardiac sarcoidosis with conduction disturbances, but with well-preserved left ventricular systolic function (6.7%), than in those with severe left ventricular impairment (36.4%). Therefore, patients with presumed cardiac sarcoidosis should be treated as such, even when the results of myocardial biopsy are negative.

Steroid therapy is expected to change the course of the disease if started early but may encourage the development of a ventricular aneurysm because steroids promote replacement of the granulomas by fibrous tissue. Therefore, echocardiography should be performed at regular intervals in order to assess the aneurysmal change of the right ventricular wall in our case. In addition to echocardiographic examination, serum ACE, and interleukin-10 also may be indicators of disease activity.

The present case demonstrates that cardiac sarcoidosis can affect mainly the right ventricle and cause morphological changes mimicking RVD.

References