Spontaneous Development of Left Ventricular Aneurysm in a Patient With Untreated Cardiac Sarcoidosis

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A young adult patient with untreated sarcoidosis spontaneously developed a left ventricular (LV) aneurysm in the anterolateral free wall. Single-photon emission computed tomography (SPECT) using Gallium-67 clearly demonstrated widespread abnormal uptake, including the LV aneurysm. Thallium-201 SPECT revealed a perfusion defect in the anterolateral wall, and abnormal uptake of technetium-99m pyrophosphate was seen, especially in the borders of the defect lesion. (Circ J 2002; 66: 519–521)

Key Words: Cardiac sarcoidosis; Gallium-67 single-photon emission computed tomography (SPECT); Left ventricular aneurysm

Sarcoidosis is a systemic granulomatous disorder of unknown origin with a poor prognosis in patients with cardiac involvement because of the associated sudden death, fatal cardiac tachyarrhythmia, conduction block, and heart failure.1–5 Therefore, early diagnosis and corticosteroid therapy are important in patients with cardiac sarcoidosis1 but clearly establishing cardiac involvement can be difficult in the early stages of the disease. The cardiac lesions are usually focal and scattered, mostly in the interventricular septum (IVS) and left ventricular wall.1–3 Extension of the ventricular lesions can lead to aneurysm formation and these have been reported as ‘healed granuloma’ after corticosteroid therapy.1 We report here a case of cardiac sarcoidosis in which a left ventricular (LV) aneurysm developed spontaneously, not after corticosteroid therapy.

Case Report

A 27-year-old man was referred for evaluation of an abnormal chest shadow detected in a routine X-ray examination. There were diffuse infiltrative shadows in both lung fields, no hilar lymphadenopathy and a normal cardiac shadow. The serum concentration of angiotensin-converting enzyme (ACE) was increased (32.0 U/L; normal range, 7.0–25.0 U/L), as was the concentration of lysozyme (16.9 μg/ml; normal range, 5.0–10.0 μg/ml). Other laboratory findings were normal. An electrocardiogram (ECG) showed right axis deviation, poor progression of the R wave in leads I and aVL, and incomplete right-bundle branch block (Fig 1A). Echocardiography revealed mild hypokinesis in the LV anterior wall with normal thickness of the IVS (7 mm) and posterior wall (8 mm). The LV ejection fraction (LVEF) was 60%. The ratio of CD4/CD8 cells in bronchoalveolar lavage fluid was increased to 11.6. Noncaseating sarcoïd granuloma was confirmed by transbronchial lung biopsy (TBLB). Although abnormal uptake of gallium-67 (67Ga) was seen in both lung fields and the right paratra-

Fig 1. Electrocardiogram at the age of 27 years (A) and after 18 months of follow up (B). An abnormal R wave in the I and aVL leads gradually developed.

Fig 2. Left ventriculography show an aneurysm (triangle) in the anterolateral wall.
The abnormal uptake of $^{67}\text{Ga}$ by the cardiac system was still widespread, including the LV aneurysm. In contrast, $^{67}\text{Ga}$ SPECT indicated that the abnormal uptake was widespread, including the borders of the defect lesion. $^{67}\text{Ga}$ uptake can localize the cardiac involvement and indicate the likely response to corticosteroid therapy. The present case demonstrates the difficulty in establishing cardiac involvement in the early stage of sarcoidosis. There were no symptoms of heart failure or ventricular arrhythmia and cardiac involvement was suspected on the basis of trivial ECG changes and LV functional abnormality on echocardiography. The abnormal cardiac uptake seen on planar $^{67}\text{Ga}$ scintigraphy was equivocal, and $^{67}\text{Ga}$ SPECT could only just confirm cardiac involvement.

It is rare for a LV aneurysm to develop without concomitant coronary artery disease, and if normal cardiac function is present, the prognosis for an aneurysm caused by myocarditis is not poor. In contrast, the prognosis for aneurysm caused by cardiac sarcoidosis is uncertain because of its rarity. Fibrosis and degeneration of the ventricle can predict it, the sensitivity and specificity are relatively low. Therefore, myocardial perfusion imaging has been used. $^{201}\text{Tl}$ SPECT can detect myocardial defects and can be used to evaluate the size of the lesion. An abnormal $^{67}\text{Ga}$ uptake can localize the cardiac involvement and indicate the likely response to corticosteroid therapy. The present case demonstrates the difficulty in establishing cardiac involvement in the early stage of sarcoidosis.

There was no histological evidence of sarcoid granuloma in the endomyocardial biopsy of the right ventricle, and we did not take a biopsy. However, based on the perform results of TBLB and the clinical course, we believe that the LV aneurysm was a clinical manifestation of cardiac sarcoidosis. Therefore, it is a dilemma when administration of corticosteroids should be started. There was no uptake of $^{99m}\text{Tc}$-PYP, especially in the borders of the defect lesion observed on $^{201}\text{Tl}$ SPECT, before steroid therapy was begun in the present case. Accumulation of $^{99m}\text{Tc}$-PYP reflects the absence of calcium and the presence of denatured proteins in myocardial cells indicative of an active aneurysm. In contrast, $^{67}\text{Ga}$ SPECT indicated that the abnormal uptake was widespread, including the LV aneurysm. Thus, the LV aneurysm may have contained an active sarcoid granuloma with scar tissue. After steroid therapy, there was no uptake of $^{99m}\text{Tc}$-PYP or $^{67}\text{Ga}$ in the cardiac lesion, and although the perfusion defect detected on $^{201}\text{Tl}$ was still present, it had not extended. Corticosteroid therapy prevented expansion of the LV aneurysm, but further careful follow-up is needed for this patient.

References
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