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Cardiac sarcoidosis – silent destroyer

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Abstract. We report a case of histologically proven pulmonary sarcoidosis and cardiac involvement in a 53-year old woman with progression leading to the heart failure documented in cardiovascular magnetic resonance studies. *(Sarcoidosis Vasc Diffuse Lung Dis 2016; 33: 175-177)*

Key words: sarcoidosis, cardiac involvement, magnetic resonance

Sarcoidosis is a multi-organ disease predominantly affecting lungs and lymph nodes, with a generally good prognosis, high remission rate and low mortality (1). However cardiac involvement alters this status for those who are affected. Cardiac sarcoidosis (CS) is the leading cause of death due to sarcoidosis in Japan and the second in Western Europe and the United States (2). In majority cases it is clinically asymptomatic, which delays the diagnosis and treatment. Symptoms, if present, are non-specific. In a few cases, the first manifestation is sudden death from ventricular arrhythmia or complete heart block. Although death can occur at any stage of the disease, it is more common in cases with extensive myocardial damage. Early treatment with corticosteroids improves prognosis; possibly reverses cardiac involvement; and substantially prolongs survival (3-5).

We now report a case of histologically proven pulmonary sarcoidosis and cardiac involvement in a 53-year old woman. She was diagnosed in 2006 because of abnormal x-chest ray (performed routinely in her workplace). A Holter monitor ECG revealed ventricular arrhythmias, with occasional bigemini, trigemini, steam, and volleys, but the patient did not have palpitations. Galium-67 scintigraphy detected possible active heart involvement. Cardiovascular magnetic resonance (CMR) performed before treatment revealed global cardiac edema (Figure b), enlargement of the left ventricle, LVEF of 38% (a), with thinning and akinesis of basal anteroseptal, mid-cavity inferolateral and inferior segments. Delayed enhancement (DE) predominantly involved subendocardial layers of akinetic segments and partially papillary muscles but did not involve coronary arteries (Figures c and d).

After 3 weeks of treatment with prednisone (1 mg/kg per day), we observed improvement in cardiac tests. A repeat Holter ECG showed fewer arrhythmias and a repeat echo showed improved LVEF (from 43 to 65%). However, due to side-effects of prednisone, the patient requested dose reductions, and the dose was tapered to none after 12 months of treatment. However, lower daily prednisone doses were accompanied by increases in ventricular arrhythmias. In 2008, a repeat CMR showed improved LV contraction, LVEF was 45% (e), edema resolved in all segments except mid-cavity inferolateral seg-

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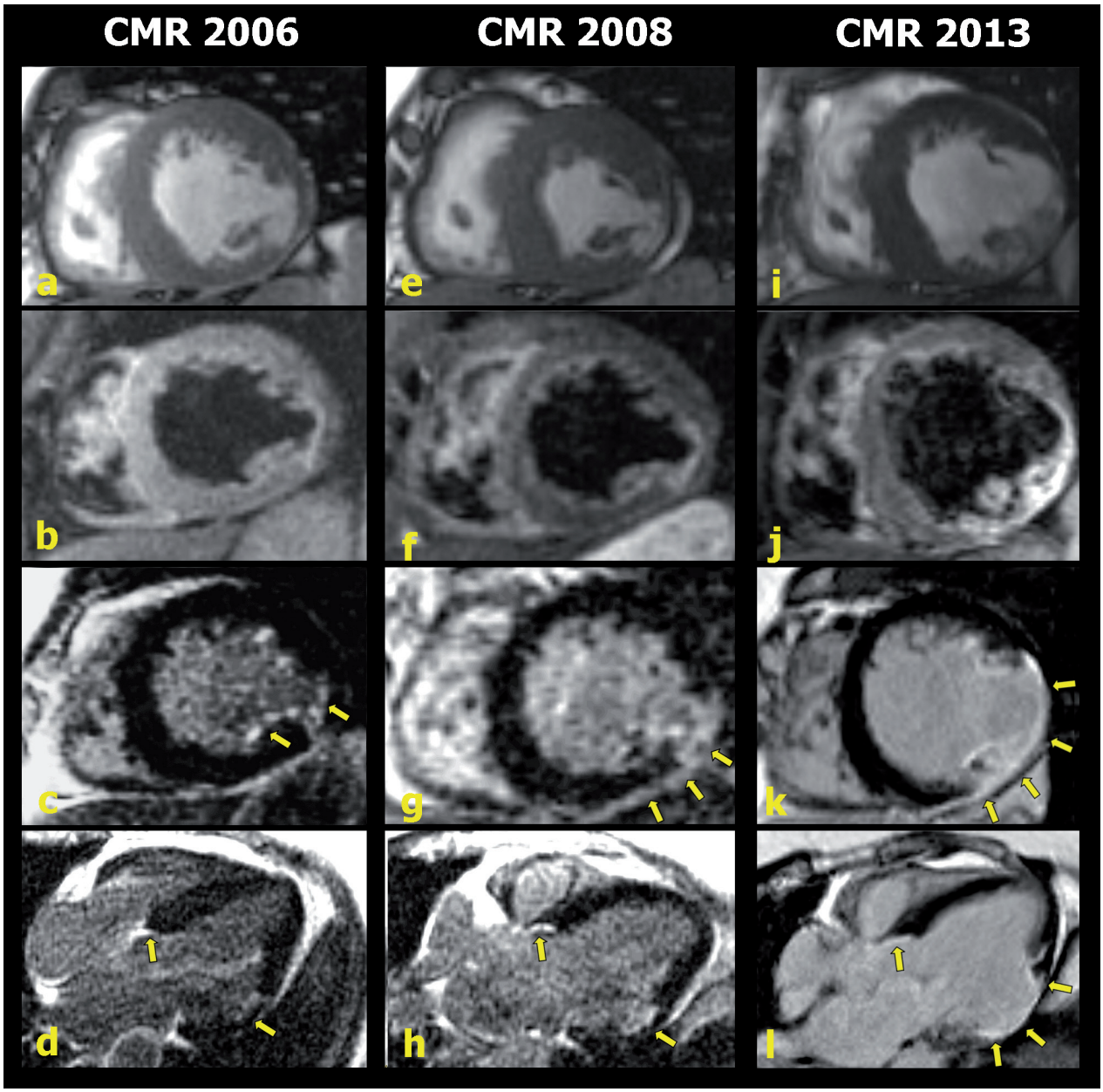

Fig. 1. *Left column:* Baseline CMR performed before treatment (in 2006). a) SSFP image in short axis mid-cavity orientation (end-systolic phase) showing enlargement of the LV and thinning of inferolateral and inferior segments. b) T2-weighted TSE image with fat saturation in short axis mid cavity orientation. Demonstration of significantly increased signal intensity of the LV myocardium. c) Delayed enhancement image in short axis mid-cavity orientation showing predominantly subendocardial enhancement of the inferolateral segment and papillary muscles (arrows). d) Delayed enhancement image in 4-chamber orientation demonstrating additional involvement of basal septum (arrow). *Middle column:* CMR performed after treatment in 2008. e) SSFP image in short axis mid-cavity orientation (end-systolic phase) showing significant improvement of LV contractility in all segments except infero-lateral segment. f) T2-weighted TSE image with fat saturation in short axis mid cavity orientation. Edema resolved in all segments except mid-cavity inferolateral. g) Delayed enhancement image in short axis mid-cavity orientation. Enhancement regions remain stable. h) Delayed enhancement image in 4-chamber orientation. Enhancement regions remain stable. *Right column:* CMR performed in 2013. i)SSFP image in short axis mid-cavity orientation (end-systolic phase) revealing severe LV dilation with thinning and dyskinesis of mid-cavity inferolateral segment, hypokinesis of anterior and anteroseptal segments. j) T2-weighted TSE image with fat saturation in short axis mid cavity orientation showing global edema. k) Delayed enhancement image in short axis mid-cavity orientation revealing significant progression with transmural enhancement of akinetic and dyskinetic segments of LV (arrows). l) Delayed enhancement image in 4-chamber orientation revealing significant progression with transmural enhancement of akinetic and dyskinetic segments of LV (arrows)

ment (f), and DE remained stable (Figures g and h). During prednisone treatment, regression and stabilization of pulmonary involvement was observed. Despite periodic inspection of outpatient cardiac and signs of active CS, the patient, who had no cardiac symptoms, refused additional prednisone therapy. A CMR performed five years later (2013) revealed severe LV dilation with thinning and dyskinesis of mid-cavity inferolateral, akinetic basal inferoseptal segments, as well as hypokinesis of mid-cavity anterior and anteroseptal segments (Figure i). Her LVEF decreased to 35%. DE (k, l) and edema (j) significantly increased, involving akinetic and dyskinetic segments of LV. Three episodes of ventricular tachycardia were observed during repeat Holter ECG monitoring. An implantable cardiac defibrillator was recommended. Despite this, patient still denied proposed treatment.

Fortunately, the disease shows no further activity, the patient is still alive, and heart failure is stable.

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