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Letter to the Editor

Cardiac sarcoidosis complicated with atrioventricular block and wall thinning, edema and fibrosis in left ventricle: Confirmed recovery to normal sinus rhythm and visualization of edema improvement by administration of predonisolone

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#### ABSTRACT

A 65 year-old female had a node of some kind in her right leg five years ago and was diagnosed with sarcoidosis by gallium scintigraphy. Serum angiotension-converting enzyme levels had gradually increased, and three months ago she felt palpitations and dizziness when standing. On electrocardiogram, 2:1 atrioventricular (AV) block was observed. On transthoracic echocardiogram, the basal portion of the interventricular septum (IVS) revealed wall thinning with dyskinetic motion and lack of systolic thickening, and low attenuation. The basal portion of the left ventricular (LV) posterior inferior wall revealed mild wall thickening with low attenuation. Enhanced multislice-CT revealed a thickened LV posterior wall and thinned basal portion of IVS with interstitial change suggesting presence of fibrosis or edema. Late enhancement was also observed in the basal portion of the LV posterior inferior wall and basal IVS in T1 weighted magnetic resonance imaging (MRI); in addition, an area, the center of which indicated low attenuation surrounded by high attenuation, was observed in the basal portion of the LV posterior inferior wall in T2 weighted MRI. Positron emission tomography (PET) imaging using F-18 fluoro-deoxyglucose with the subject fasted for 6 h beforehand, revealed strong uptake in the basal portion of IVS and a thickened LV posterior wall, suggesting the presence of inflammation. Administration of predonisolone was started before pacemaker implantation and clinical symptoms immediately disappeared; in addition AV block recovered to normal sinus rhythm. On a repeat MRI performed four months later, the late enhancement in T1 weighted MRI and the high attenuation surrounding low attenuation in the basal portion of the LV posterior inferior wall in T2 weighted MRI both disappeared, and we confirmed that temporary edema had also disappeared.

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## 1. Introduction

If cardiac sarcoidosis can be diagnosed in the early phase, administration of predonisolone can lead to a good prognosis [1–3]. But if it is not diagnosed early, wall thinning of interventricular septum (IVS) may occur, followed by occurrence of complete atrioventricular (AV) block; dizziness may result and subjects may often require implantation of cardiac pacemakers [4,5]. Therefore establishment of non-invasive and accurate diagnosis of cardiac sarcoidosis would be desirable.

Recently we encountered a subject with cardiac sarcoidosis complicated with AV block and wall thinning, edema or fibrosis in left ventricle (LV). By administration of predonisolone, we confirmed recovery from AV block to normal sinus rhythm and visualized improvement of edema.

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# 2. Case

A 65 year-old female was diagnosed with uveitis seven years previously. Five years ago, on physical examination, there was a node of some kind in her right leg; she was referred to another hospital and was diagnosed with sarcoidosis by gallium scintigraphy. Serum angiotensin converting enzyme levels had gradually increased (21.8I U/L on this admission); three months ago she experienced palpitations and dizziness when standing and was referred to our hospital. Chest X ray revealed a markedly increased bilateral hilar shadow (Fig. 1). On electrocardiogram, 2:1 AV block was observed (Fig. 2). On transthoracic echocardiogram (TTE) (IE33, Philips), the basal portion of IVS revealed wall thinning (minimum wall thickness 7 mm) with dyskinetic motion, lack of systolic thickening, and low attenuation. The basal portion of the LV posterior inferior wall revealed mild wall thickening (12 mm) with low attenuation. Enhanced multislice computed tomography (CT) (Light Speed Ultra 16, GE) (Fig. 3), revealed a thickened LV posterior wall and thinned basal portion of IVS with strong interstitial change suggesting the presence of fibrosis or edema. Late enhancement was also observed in the basal portion of the LV posterior inferior wall and basal IVS in T1 weighted magnetic

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**Fig. 1.** Chest X ray revealed a markedly increased bilateral hilar shadow with cardiothoracic ratio of 51%. No lung congestion was observed.

resonance imaging (MRI) (Intera Achieva; Philips) (Fig. 4); in addition, an area, the center of which indicated low attenuation surrounded by high attenuation, was observed in the basal portion of the LV posterior inferior wall in T2 weighted MRI. Wall thickness of the basal portion of IVS became reduced and reduced systolic wall thickness was evident by MRI.

To evaluate the presence of inflammation, positron emission tomography (PET) (Advance NXi, GE) (Fig. 5) was performed using F-18 fluorodeoxyglucose (FDG) with the subject fasted for 6 h beforehand. The PET image revealed strong uptake in the basal portion of IVS (arrows, A) and thickened LV posterior wall (arrowheads, B and C), a markedly increased multiple hilar shadow (arrows in upper portion, C) and right femoral (arrows in lower portion, C), suggesting the presence of inflammation. Myocardial single-photon emission computed tomography images obtained using rest iodine-123 15-(p-iodophenyl)-3-(R, S) methylpentadecanoic acid (BMIPP) and test thallium-201 showed no abnormalities.

As wall thickness had already become reduced, implantation of a pacemaker was considered for 2:1 AV block in this subject. However, T2 weighted MRI and PET images suggested the presence of temporary edema and before pacemaker implantation, administration of predonisolone was started. Weekly administration of 40 mg predonisolone was started involving titration downwards of the dose by 5 mg every two weeks (i.e., 40 mg, 40 mg, 35 mg, 35 mg, 30 mg, 30 mg,...). After administration of predonisolone, clinical subjective symptoms such as palpitation or dizziness immediately disappeared and 24 h monitoring showed reduced frequency of AV block and finally AV block recovered to normal sinus rhythm on electrocardiogram (Fig. 6).

On repeat performance of an MRI four months later (Fig. 7), late enhancement in T1 weighted MRI and the high attenuation surrounding low attenuation in the basal portion of the LV posterior inferior wall in T2 weighted MRI had disappeared.

In contrast to the MRI findings, repeated CT examination performed four months after administration of predonisolone, revealed some late enhancement in the basal portion of the LV posterior inferior wall and basal IVS (data not shown).

We conclude that administration of predonisolone led to disappearance of temporary edema.

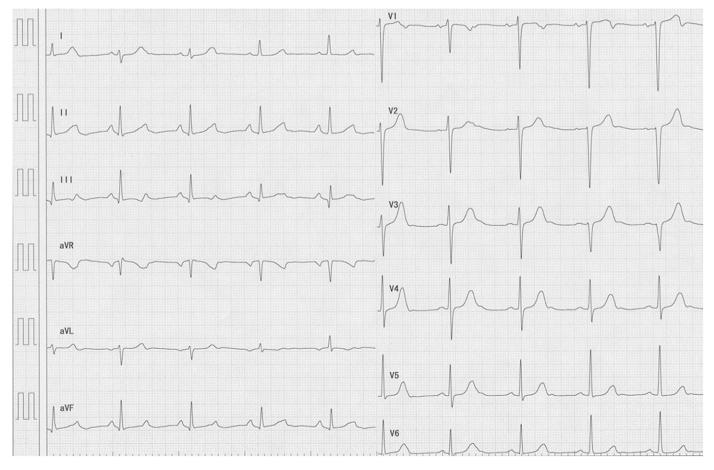


Fig. 2. Electrocardiogram at admission revealed 2:1 atrioventricular block with a ventricular rate of 57 bpm.

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