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Case Report

Suppression of Ventricular Tachycardia Associated with Cardiac Sarcoidosis by Steroid Therapy

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In patients with cardiac sarcoidosis, ventricular tachycardia (VT) is observed in some cases. However, effective therapies for the VT are still unknown.

Case: A 50-year old female with cardiac sarcoidosis underwent DDD pacemaker implantation for a high degree atrioventricular block with symptoms of faintness and shortness of breath. One month after the surgery, she was admitted for frequent episodes of non-sustained VT. In the electrophysiologic study (EPS), sustained monomorphic VT and ventricular fibrillation were induced; therefore pacemaker was replaced with implantable cardioverter-defibrillator (ICD). Amiodarone was started orally but it couldn't suppress frequent VT episodes, and frequent ICD shocks were delivered. When the oral steroid therapy was initiated for the cardiac sarcoidosis, it not only suppressed the frequent VT but also improved the atrioventricular nodal dysfunction.

In conclusion, steroid therapy might be an option to consider in cardiac sarcoidosis with refractory VT.
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Key words: Cardiac sarcoidosis, Ventricular tachycarida, Steroid therapy

Introduction

In patients whose cardiac dysfunction was the result of cardiac sarcoidosis, sudden death is the most common cardiac manifestation.[1)] Ventricular tachycardia (VT) and complete heart block are among the causes, and are frequently observed. For the treatment of VT associated with cardiac sarcoidosis, the implantable cardioverter-defibrillator (ICD) is necessary to prevent the arrhythmic events.[2–4)] However the best way of bailing out the frequent VT and ICD shocks has not been established. In this article, we report a case with cardiac sarcoidosis in which VT was suppressed and atrioventricular nodal function was improved by oral steroid therapy.

Case Report

A 50-year-old woman was hospitalized for symptoms of faintness and shortness of breath. She was diagnosed as cardiac sarcoidosis 4 years ago from

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the left bundle branch block in electrocardiography (ECG), diffuse left ventricular hypocontractility with an ejection fraction (EF) of 26 percent in echocardiography, lung and liver nodules, and the accumulation of Gallium-67 in the heart. Histologic diagnosis was not made because she did not consent to the biopsy. A dose of oral prednisolone 60 mg every other day effectively improved the ECG finding of intraventricular conduction disorder and the accumulation of Gallium-67 disappeared, but the left ventricular contraction improved only slightly. Other medications including digoxin, spironolactone and carvedilol were also effective and she had no acute exacerbation of heart failure and sarcoidosis for 4 years. Dose of steroid was tapered to 2 mg per day. But this time of admission 12-lead ECG showed type II second degree atrioventricular block, left anterior hemiblock and non-specific ST-T change (**Figure 1**) for the first time for 4 years. A dual chamber pacemaker was implanted.

One month after pacemaker implantation, chest X ray revealed the enlargement of cardiac silhouette with cardiothoracic ratio increasing from 46 to 50 percent and she was hospitalized and treated with diuretics. During this hospitalization no arrhythmia event was demonstrated, but episodes of non-sustained VT (NSVT) were documented at the pacemaker clinic. She had late potentials on the signal averaged ECG (total QRS 184 ms, HFLA 71 ms, RMS40 12 μV) in sinus rhythm, and EF was 18.7 percent in quantitative gated myocardial single photon emission computed tomography. There thought to be the arrhythmogenic scar in the heart and electrophysiologic study was performed to evaluate the inducibility of ventricular arrhythmias.

Electrophysiologic Study

The electrophysiologic study was undertaken while the patient was sedated. Sustained monomorphic VT (VT1) was reproducibly induced by programmed stimuli at the right ventricular apex (**Figure 2a**). VT1 changed to clinical VT (VT2) after direct current shock with 300 J (**Figure 2b**), and degenerated into ventricular fibrillation with rapid right ventricular outflow tract pacing. The morphology of both VT1 and VT2 was left bundle branch block with a superior axis, the cycle length of which was 280 ms and 360 ms, respectively. Though detailed mapping was not performed, estimated exit sites of both VTs were the left ventricular mid–septum or the right ventricular mid-region.[5] The left ventriculography showed diffuse severe hypokinesis with EF of 7 percent and severe mitral regurgitation but coronary angiography exhibited no significant coronary stenosis. Subsequently, an ICD was implanted for primary prevention of sudden cardiac death and pacemaker and ventricular lead was taken out.

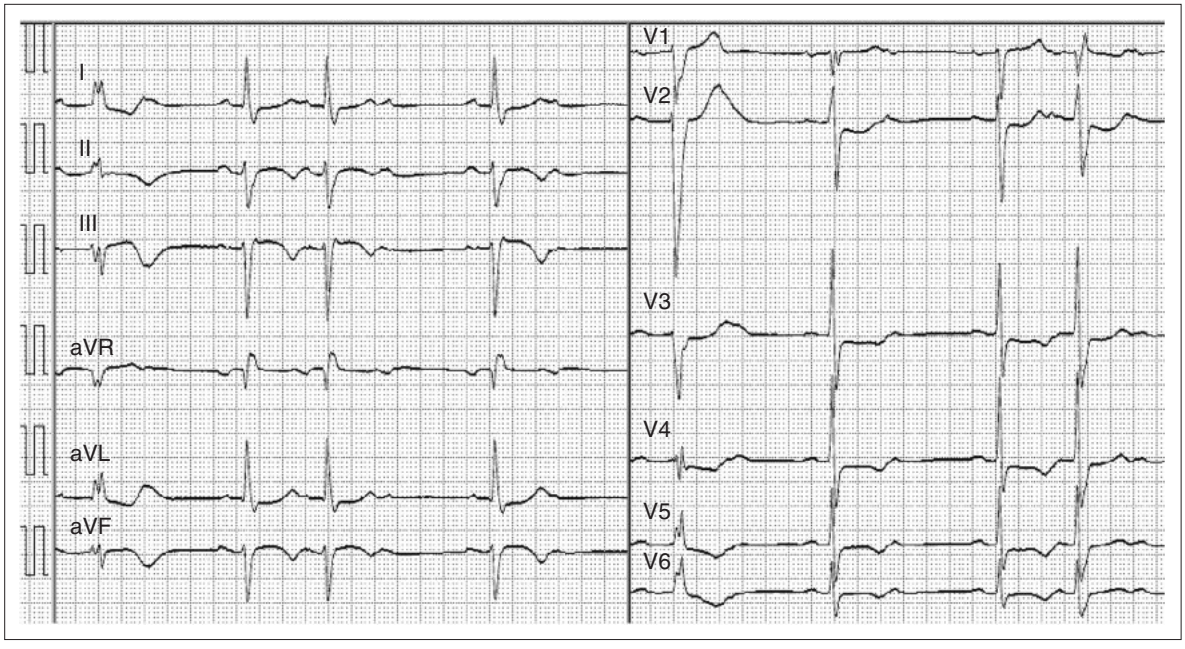

Figure 1 Surface twelve-lead ECG.
Surface twelve-lead ECG revealed type II second degree atrioventricular block, left anterior hemiblock and non-specific ST-T change.

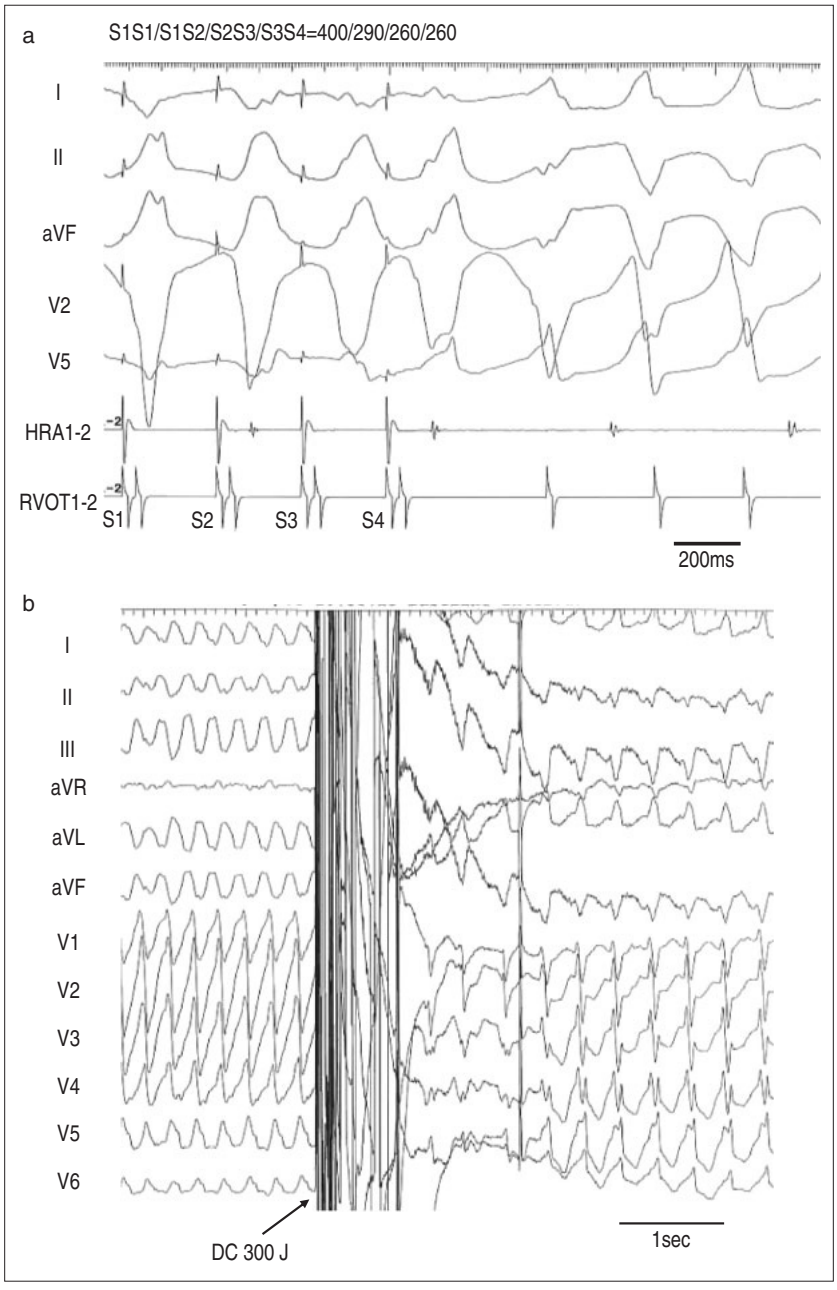

Figure 2
a: Programmed electrical stimulation. Monomorphic ventricular tachycardia (VT1) was reproducibly induced at 3 extrastimuli from the site of the outflow tract of the right ventricle. S1S1: basic stimulation. S1S2: coupling interval of first extrastimulus. S2S3: coupling interval between first and second extrastimlus. S3S4: coupling interval between second and third extrastimulus. HRA: high right atrium. RVOT: outflow tract of the right ventricle.
b: VT1 changed into VT2. VT1 changed into VT2 which was clinically demonstrated by direct current shock with 300 joules. DC: direct current. J: joule

Clinical Course

After the implantation of ICD, the value of BNP and cardiothoracic ratio got worse from 236 pg/ml to 370 pg/ml and 46 percent to 53 percent, respectively, which was thought to be due to the frequent PVCs, NSVT and dyssynchronized pacing. The dosage of the diuretics was increased and the both values got improved. Though the status of heart failure got improved, the cardiac function in echocardiography did not get better and the frequency of NSVT (**Figure 3**), sustained VT (**Figure 4**) and ICD shocks became increased. Oral amiodarone and beta blockers failed to suppress them. The physical findings and the laboratory data, including the value of Ca, showed no abnormalities with the exception of slight elevation of lysozyme and the serum protein fractionation of γ globulin (9.4 and 24.4 percent, respectively). The level of ACE remained normal range with the value of 13.5 IU/l, which was slightly higher than her control level of about 5 IU/l. The 24 hours Holter ECG detected 3633 PVCs, 129 couplets

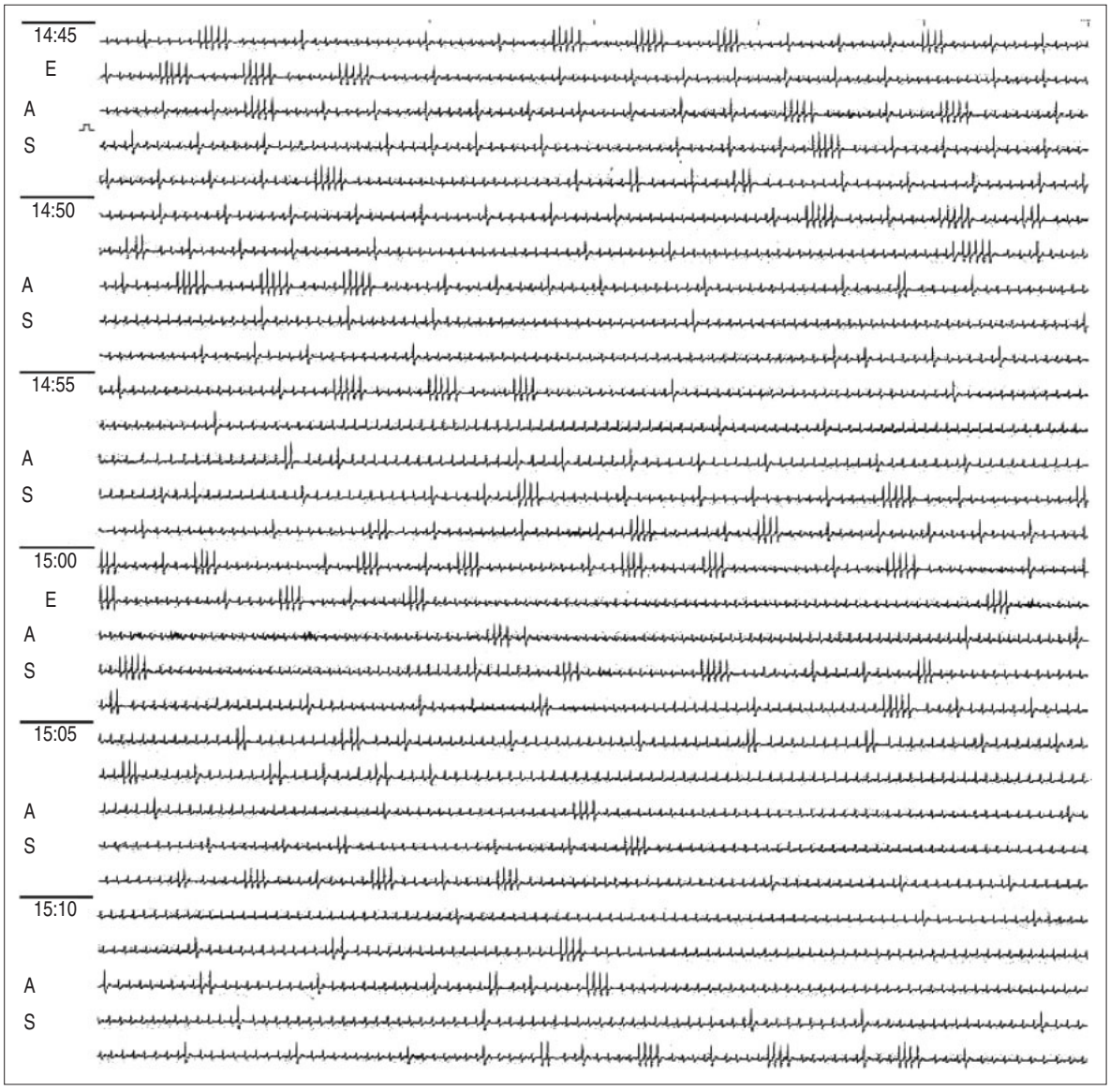

Figure 3 24 hour Holter ECG.
24 hour Holter ECG revealed frequent NSVTs. Shown is the CC5 recording. NSVT occurred frequently during the daytime.

and 268 triplets or NSVTs of more than 10 beats. We diagnosed that the cardiac sarcoidosis was active based on the cardiac arrhythmia and abnormal laboratory findings of lysozyme and the serum protein fractionation of γ globulin, and 60 mg of oral prednisolone every other day was started. After the initiation of the steroids, the value of ACE returned to her control level of 5.5 IU/l and the 24 hours Holter ECG detected 1261 PVCs without NSVTs per day, which was remarkably fewer than before. Moreover, ICD shocks were suppressed and the NSVT were terminated spontaneously within 4 or 5 beats, and atrioventricular nodal function improved as well (**Figure 5**). During the admission period, echocardiographic evaluation was repeatedly performed, but it was limited by the poor view. Though detailed evaluation had been difficult, left ventricular diastolic diameter and EF remained about 62 mm and 30 percent, respectively.

Discussion

In patients with cardiac sarcoidosis, the mechanism of VT is thought to be reentry or abnormal automaticity.[4] Usually the steroid therapy halts the progression of cardiac disease and improves survival, but it is not the enough treatment to control the ventricular arrhythmias.[3] Antiarrhythmic agents and

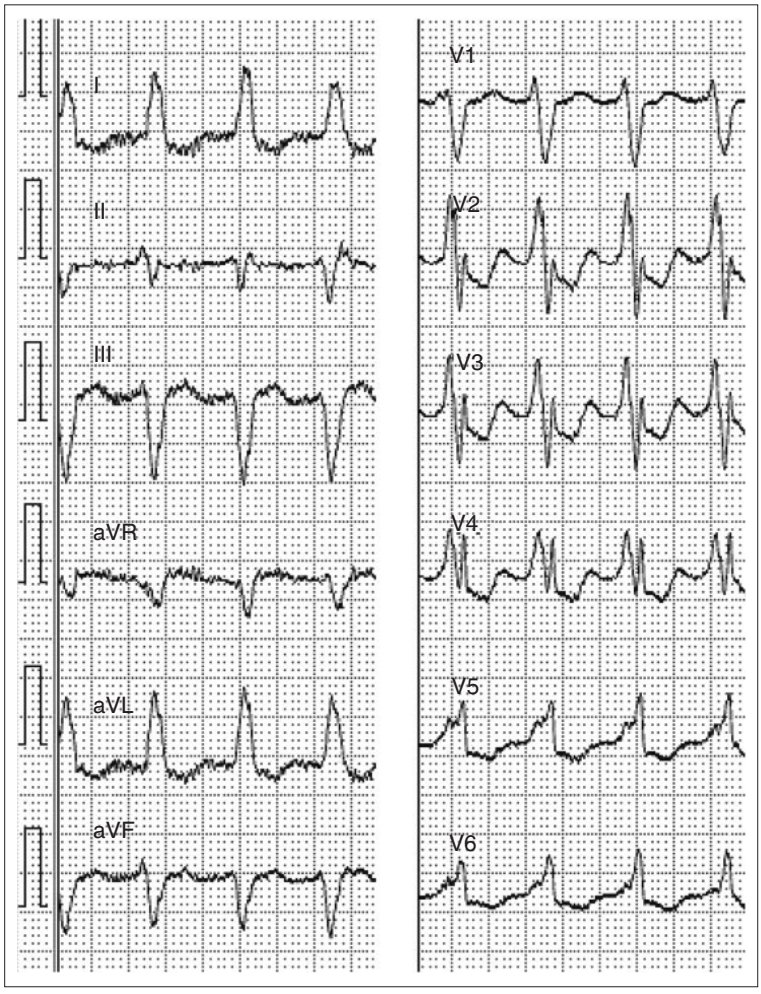

Figure 4 Surface twelve-lead ECG.
Surface twelve-lead ECG revealed VT with the morphology of right bundle branch block and superior axis and tachycardia cycle of 450 ms.

beta blockers are often used together with steroid, but these agents still can not control the arrhythmia completely, even when guided with EPS. So it is recommended to place the ICD for the ventricular tachycardia.[3,6] To evaluate the arrhythmic risk, programmed ventricular stimulation is useful to predict the future arrhythmic event.[6] Aizer et al. reported that 10 of 12 patients who underwent ICD insertion received appropriate ICD shocks.

Although the mechanism of steroid treatment in cardiac sarcoidosis is unknown, it is reported that steroid therapy can cause healing of cardiac granulomas, which is a possible cause of the cardiac aneurysms.[1] Regarding the cardiac function, Chiu CZ et al. reported that patients with an ejection fraction of over 30 percent and under 55 percent benefited from steroid therapy.[7]

In this patient, entrainment phenomenon was not demonstrated because of the degeneration into VF by rapid ventricular pacing, but programmed ventricular stimulation reproducibly induced the VT. Though it is difficult to determine the mechanism of VT, ICD was implanted, and it delivered the appropriate shocks.

In this case there thought to be myocardial scars as well as active sarcoid granulomas in the heart. Reproducible inducibility of VT suggests that the mechanism of VT may be reentry, but healing of the granulomas by steroid therapy might eliminate the focus of the automaticity. The frequency of VT and PVCs increased after improvement of heart failure, which suggests that the arrhythmic event was caused by active cardiac sarcoidosis. After steroid therapy, though no improvement or deterioration of the EF and the left ventricular diameter was revealed in echocardiography, VT was suppressed. The frequency of PVCs was also remarkably decreased. Based on these finding, the suppression of VT might be caused by decreased PVCs by steroid therapy, even though there may remain the increased myocardial scars which cause ventricular arrythmias.

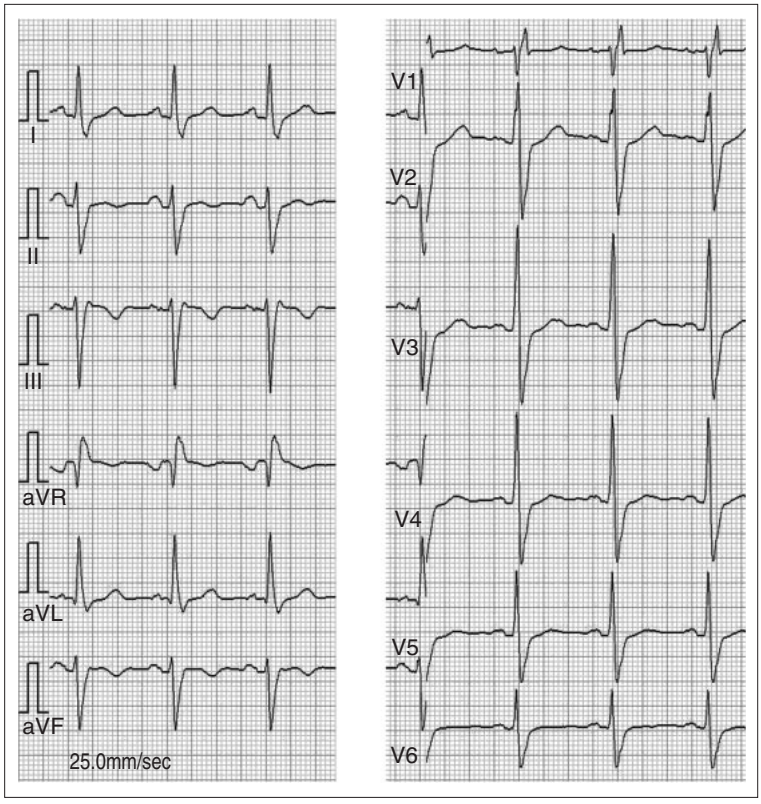

Figure 5 Surface twelve-lead ECG after the administration of steroid therapy.
Surface twelve-lead ECG revealed left anterior hemiblock and right bundle branch block.

Roberts et al. reported that in the majority of patients with cardiac sarcoidosis causing cardiac dysfunction, there was no clinical evidence of the dysfunctions of other organ systems other than the heart.[1)] This time the systemic examination including Gallium-67 scintigraphy could not reveal any typical abnormality. Though in some cases the value of ACE or Gallium-67 scintigraphy is useful to detect the activity of the disease,[8)] these examinations disclosed no abnormal findings in this patient. The number of PVCs in Holter ECG and the value of ACE, even if it remained the normal range, might be useful to speculate the activity of sarcoidosis and determine the doses of steroid in this case. And if the activity of sarcoidosis is controlled, then tapering the dose of prednisolone had better been attempted gradually to 10–15 mg per day over a period of 6 months.[8)]

In cardiac sarcoidosis, ICD implantation is recommended for the VT, and when VT and ICD shocks occur frequently, it is necessary to evaluate the disease activity. In the patients with active sarcoidosis, there might be some cases which steroid therapy is effective to suppress the arrhythmic events. In the case of VT without active sarcoidosis, though the efficacy is not established, catheter ablation rather than higher doses of steroid might be needed.

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