Sarcoidosis is a multisystemic granulomatous disease of unknown etiology. Granulomas that infiltrate the heart exist in up to 27% of sarcoidosis patients at autopsy, but the clinical manifestations of cardiac sarcoidosis are present in merely 5% of cases. Patients present with a cardiomyopathy if the myocardial infiltration is extensive. The conduction system is frequently affected. These alterations can cause brady- and tachyarrhythmias that can result in palpitations, syncope, or sudden death. Treatment is aimed at improving symptoms, controlling the inflammation, and preventing the deterioration of left ventricular function and sudden death.

Case

A 31-year-old man experienced 6 months of malaise and fatigue. Approximately 9 years earlier, he was diagnosed with neurosarcoidosis and underwent resection of a right cerebral granulomatous mass. At that time, there was no evidence of cardiac involvement, and steroids were not initiated. In subsequent years, he developed no signs of recurrence.

On physical examination, he was afebrile. A macular lesion was detected in the right side of the nose, measuring 2 cm in diameter. A 1.5-cm mobile and painless lymph node was palpated in the left groin. The patient's blood pressure was 120/60 mmHg, his pulse was 52 beats per minute (bpm), and his respiratory rate was 18 breaths per minute. His heart rhythm was irregular, with a variable S1, a midsystolic 1/6-ejection murmur in the mitral area, and a normal S2. The remainder of the physical exam was normal.

No disclosures.

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Arrhythmias in a Patient With Sarcoidosis

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Arrhythmias in a Patient With Sarcoidosis. Sarcoidosis is a multisystemic granulomatous disease of unknown etiology; up to 27% of cases entail cardiac involvement. Conduction abnormalities and ventricular tachycardia are the most common arrhythmias and can cause sudden death. We describe a patient who developed cardiac sarcoidosis 9 years after undergoing surgery for neurosarcoidosis. He presented with 2:1 second-degree atrioventricular block. Ventricular tachycardia with 3 morphologies was induced by exercise stress test. A DDD pacemaker/implantable cardioverter defibrillator (ICD) was implanted, which prevented exercise-induced ventricular tachycardia in a follow-up stress test. Treatment with steroids was initiated. The AVB disappeared, and no further arrhythmias were documented at the 1-year follow-up. (J Cardiovasc Electrophysiol, Vol. 22, pp. 1387-1390, December 2011)

An electrocardiogram showed sinus rhythm at a rate of 90 bpm. There was a 2:1 second-degree atrioventricular block (AVB) with right bundle branch block (RBBB). The chest X-ray was normal. Blood tests, including liver and renal function tests, ionic calcium, sedimentation rate (2 mm/h), troponine T (0.01 ng/mL), and angiotensin-converting enzyme levels (45 µ/L), were normal. Viral serology was negative, and the echocardiogram was normal.

By high-definition multidetector thoracic and abdominal computed tomography, we noted central and peripheral peribronchial and perivascular nodular lesions; superior pretracheal and perivascular mediastinal calcified lymph nodes, and abdominal periaortie and enlarged inguinal lymph nodes. The spleen harbored multiple nodular lesions. A biopsy of the inguinal lymph node revealed noncaseating granulomas with lymphocytes, epithelioid and multinucleated giant cells (Langhan’s type with asteroid and Schaumann bodies). A cardiac magnetic resonance image revealed delayed gadolinium uptake in the anterobasal segment of the interventricular septum (Fig. 1). The left ventricular size and function were preserved. Radionuclide imaging with SESTAMIBITc 99 showed basal anteroseptal hypoperfusion at rest that improved slightly after exertion.

During an exercise stress test the baseline 2:1 AVB and RBBB persisted until the sinus rate reached 125 bpm, when a high-grade AVB with an idioventricular escape rhythm and left bundle branch block (LBBB) morphology developed at a rate of 55 bpm (Fig. 2). The patient went into ventricular tachycardia (VT) with an LBBB morphology and frontal left superior axis (VT # 1) and a progressively increasing heart rate, from 75 to 100 bpm (cycle length 800–600 ms). Then, a second type of VT developed with RBBB morphology and frontal left superior axis (VT # 2) that alternated with the previous VT at similar heart rates (cycle length 520–560 ms; Fig. 3). At maximal exertion, a third type of VT emerged (VT # 3) with RBBB morphology, a frontal right superior axis, and cycle lengths between 500 and 550 ms (Fig. 4). The patient achieved 12 Mets and a maximum blood pressure of 170/85 mmHg. He remained asymptomatic during the entire procedure.
In the recovery phase, he experienced the same cardiac rhythms that developed during exertion but in reverse order, returning to baseline sinus rhythm with 2:1 AVB.

A permanent DDDR pacer/ICD was inserted. A follow-up stress test showed a baseline sinus-sensed ventricular paced rhythm. With exertion, the heart rate increased to 150 bpm without evidence of ventricular arrhythmia.

Two weeks after the pacemaker/ICD was inserted, treatment with prednisone 1 mg/kg/day was initiated. During the next 14 months, the patient remained asymptomatic, without experiencing arrhythmias or ICD events. The conduction through the AV node recovered, and RBBB disappeared.

Discussion

Cardiac sarcoidosis can range from a benign subclinical condition to a life-threatening disorder. Although autopsy-based studies have demonstrated that cardiac granulomas exist in as many as 27% of sarcoidosis patients, clinical manifestations are present in merely up to 5% of cases.\(^2\) Granulomas have a patchy distribution, lying predominantly in the free wall of the left ventricle and basal septum.\(^1\) When the right ventricle is involved, it can mimic the clinical presentation of arrhythogenic right ventricular dysplasia.\(^5,6\) They can also affect the pericardium. Cardiomyopathy with congestive heart failure can occur when the myocardial infiltration is extensive.
Granulomas frequently affect the atrioventricular node and bundle of His, inducing AVB and various intraventricular conduction defects.7,8 These lesions can be silent or can cause syncope. Sarcoid granulomas in the myocardium can also affect abnormal automaticity and alter conduction and refractoriness, inducing reentrant ventricular arrhythmias, which exist in as many as 23% of patients with demonstrated cardiac involvement.4,9,10 Sudden death due to ventricular arrhythmias or conduction abnormalities accounts for 25–65% of mortalities in patients with cardiac sarcoidosis.11,12

There is limited evidence regarding the most effective treatment for patients with cardiac sarcoidosis.1 Treatments should improve symptoms, control the inflammation, and prevent the deterioration of left ventricular function and sudden death.3 Steroids are believed to be capable of halting or slowing the inflammation and fibrosis, which might explain their efficacy in reverting conduction abnormalities and improving alterations in myocardial perfusion.

Small retrospective studies have suggested that steroids improve long-term survival,13 but their effectiveness against ventricular arrhythmias has not been properly studied. Due to the high rate of recurrence of VT in patients who are treated with antiarrhythmic agents, some physicians recommend ICD placement in these patients.3,14-16 In cases of advanced AVB, a permanent pacer is recommended to improve the prognosis.3
We present a case of systemic sarcoidosis with many of the arrhythmias that are commonly described in cardiac sarcoidosis. The baseline electrocardiogram showed 2:1 second-degree AVB and RBBB. An exercise stress test induced VT with 3 morphologies. By cardiac MRI and radionuclide imaging, we noted the involvement of the anterobasal segment of the interventricular septum, and the biopsy of an inguinal lymph node revealed noncaseating granulomas.

Based on the revised 2006 Guidelines for diagnosing cardiac sarcoidosis from the Japan Society of Sarcoidosis and Other Granulomatous Disorders, our patient met the diagnostic criteria for cardiac sarcoidosis and, therefore, we felt that an endomyocardial biopsy was unnecessary. The extent of the granulomatous infiltration was insufficient to reduce left ventricular systolic function but was likely responsible for the conduction abnormalities and the exercise-induced VT. During exertion, the altered atrioventricular conduction and increased sympathetic drive likely facilitated reentry at the septum, predisposing the patient to the development of VT. The modulations in VT morphology were likely related to disparate reentry loops in the same area or the same basic loop with progressive changes in its own circuit or exit site. We speculate that the absence of VT during the follow-up stress test, without changes in anatomical substrate, was attributed to a progressive increase in ventricular pacing during exertion, preventing the reentry mechanism.

Treatment with steroids was effective in reverting AVB. The degree of anatomical resolution of the anteroseptal granuloma was unknown because a follow-up cardiac MRI and radionuclide imaging were not performed.

In conclusion, this case illustrates the spectrum of arrhythmias that can be observed in patients with cardiac sarcoidosis. Pacing avoided ventricular bradycardia due to AVB during exertion, preventing exercise-induced VT and ICD events during follow-up. Treatment with steroids improved AV nodal conduction abnormalities.

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