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CASE REPORT
Inherited cardiac conditions/cardiomyopathies

Atrioventricular nodal re-entrant tachycardia unmasking cardiac sarcoidosis: a clinical case report

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Background Sarcoidosis is a rare disease, and cardiac involvement is seen in the minority of patients. The clinical symptoms depend on the location of the noncaseating granulomas in the heart and vary from asymptomatic to atrioventricular (AV) conduction block, ventricular arrhythmia, heart failure, and sudden cardiac death. Clinically manifest cardiac sarcoidosis seldomly presents with supraventricular tachycardia.

Case summary We present a case where a female patient presented with AV nodal re-entrant tachycardia as an uncommon initial presentation of cardiac sarcoidosis. Her resting electrocardiogram showed a complete left bundle branch block and first-degree AV conduction block. During hospitalization, there was continuous switching between sinus rhythm with first-degree AV block, 2:1 AV block, and AV nodal re-entrant tachycardia.

Discussion It is important to be aware that cardiac sarcoidosis can rarely present with supraventricular tachycardia as initial symptom. Given the elevated risk of sudden cardiac death, early detection is crucial and all patients who require permanent pacing should be considered for implantable cardioverter-defibrillator implantation.

Keywords Cardiac sarcoidosis • Atrioventricular nodal re-entrant tachycardia • Atrioventricular block • Sudden cardiac death • Case report

ESC curriculum 2.3 Cardiac magnetic resonance • 5.1 Palpitations • 5.5 Supraventricular tachycardia • 5.10 Implantable cardioverter defibrillators • 6.5 Cardiomyopathy

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Learning points

- Sarcoidosis is a rare disease, and cardiac involvement is seen in the minority of patients. The clinical symptoms depend on the location of the noncaseating granulomas in the heart and vary from asymptomatic to atrioventricular conduction block, ventricular arrhythmia, heart failure, and sudden cardiac death.
- Clinically manifest cardiac sarcoidosis seldom presents with supraventricular tachycardia. However, in young patients with new onset supraventricular tachycardia, cardiac sarcoidosis should be in the differential diagnosis, especially when the resting electrocardiogram shows signs of slow conduction.
- Given the elevated risk of sudden cardiac death, early detection is crucial and all patients who require permanent pacing should be considered for implantable cardioverter-defibrillator implantation.

Introduction

Cardiac involvement is observed in the minority of patients with sarcoidosis. The clinical symptoms depend on the location of the noncaseating granulomas and the extent of the subsequent inflammation, oedema, and tissue scarring in the heart. The presentation varies from asymptomatic to atrioventricular (AV) conduction block, ventricular arrhythmia, heart failure, and sudden cardiac death. Patients with cardiac sarcoidosis, who present with high-grade AV block (AVB) with or without ventricular tachycardia or left ventricular dysfunction, have worse prognosis because of a greater risk of life-threatening ventricular arrhythmia and sudden cardiac death.[1] Given this elevated risk of sudden cardiac death, early detection is crucial. Once diagnosed, treatment with corticosteroids and/or other immunosuppressants is recommended. In cardiac sarcoidosis, all patients who require permanent pacing should be considered for implantable cardioverter-defibrillator (ICD) implantation.

Clinically manifest cardiac sarcoidosis seldom presents with supraventricular tachycardia, and atrial arrhythmias occurred in a minority of patients at follow-up.[2] In the present case however, an AV nodal re-entrant tachycardia (AVNRT) was the presenting symptom that led to the diagnosis of cardiac sarcoidosis. The mechanism behind this is thought to be an acquired functional slow pathway due to interfering granulomas and its subsequent oedema. This in turn was the substrate for the development of an atypical AVNRT of the slow–slow subtype.

Summary figure

Day 1	A 36-year-old female patient, with a history of Löfgren disease, presented at the emergency department with dyspnoea and palpitations. Resting ECG shows an AVNRT (*Figure 1A*) for which she was treated with adenosine, resulting in restoration of sinus rhythm.
Day 2	Her ECG in sinus rhythm shows a first-degree AVB and left bundle branch block (LBBB) (*Figure 1B*). She is discharged with an ambulatory appointment for cardiac MRI.
Day 5	She returned to the emergency department with several episodes of presyncope. Resting ECG shows a 2:1 AVB (*Figure 1C*).
Days 6–12	During monitoring in the cardiac intensive care unit, episodes of 2:1 AVB alternated with episodes of supraventricular tachycardia. Cardiac MRI and PET–CT are both suggestive of cardiac sarcoidosis. The final diagnosis was confirmed through lymph node biopsy and histopathology. Start therapy with high-dose intravenous corticosteroids and methotrexate (10 mg).
Day 13	Implantation of an implantable cardioverter-defibrillator (ICD).
Day 68	No recurrence of high degree AVB and no registrations of transient supraventricular tachycardia. The PR interval normalized and QRS duration decreased.

Case report

A 36-year-old female patient, with a history of Löfgren disease, presented to the emergency department with dyspnoea and palpitations. Physical examination showed regular tachycardia on auscultation and no signs of right-sided or left-sided fluid overload. The electrocardiogram (ECG) revealed the presence of an AVNRT (*Figure 1A*) for which she was treated with adenosine, resulting in restoration of sinus rhythm. Since her resting ECG revealed a first-degree AVB and left bundle branch block (LBBB), a beta-blocker was not initiated (*Figure 1B*).

Five days later, she returned to the emergency department with several episodes of presyncope. The ECG this time showed a 2:1 AVB (*Figure 1C*). During monitoring in the cardiac intensive care unit, episodes of 2:1 block alternated with episodes of supraventricular tachycardia. Exercise stress testing revealed a 2:1 AVB at peak exertion, while during recovery, the rhythm changed to a supraventricular tachycardia with late retrograde P-waves, indicative of an atypical AVNRT.

Given her history of Löfgren syndrome, additional cardiac magnetic resonance imaging (MRI) and positron-emission tomography-computed tomography (PET–CT) scans were performed. The MRI identified multifocal myocardial involvement with severe regional dysfunction and clear signs of myocardial fibrosis, suggestive of cardiac sarcoidosis. There was overall preservation of global systolic function, but reduced longitudinal strain (left ventricular ejection fraction 57%, global longitudinal strain −11.4%) (see Supplementary material online, *Video S1*). Additionally, ‘dark-paps’ were detected on cardiac MRI (*Figure 2*). This is a hypo-intense aspect of the papillary muscles that is being recognized as an independent predictor associated with an increased risk of ventricular arrhythmia.[3] Positron emission tomography–computed tomography showed the presence of hypermetabolic hilar nodes and increased tracer uptake in the spleen and myocardium. The final diagnosis of sarcoidosis was confirmed through lymph node biopsy and histopathology.

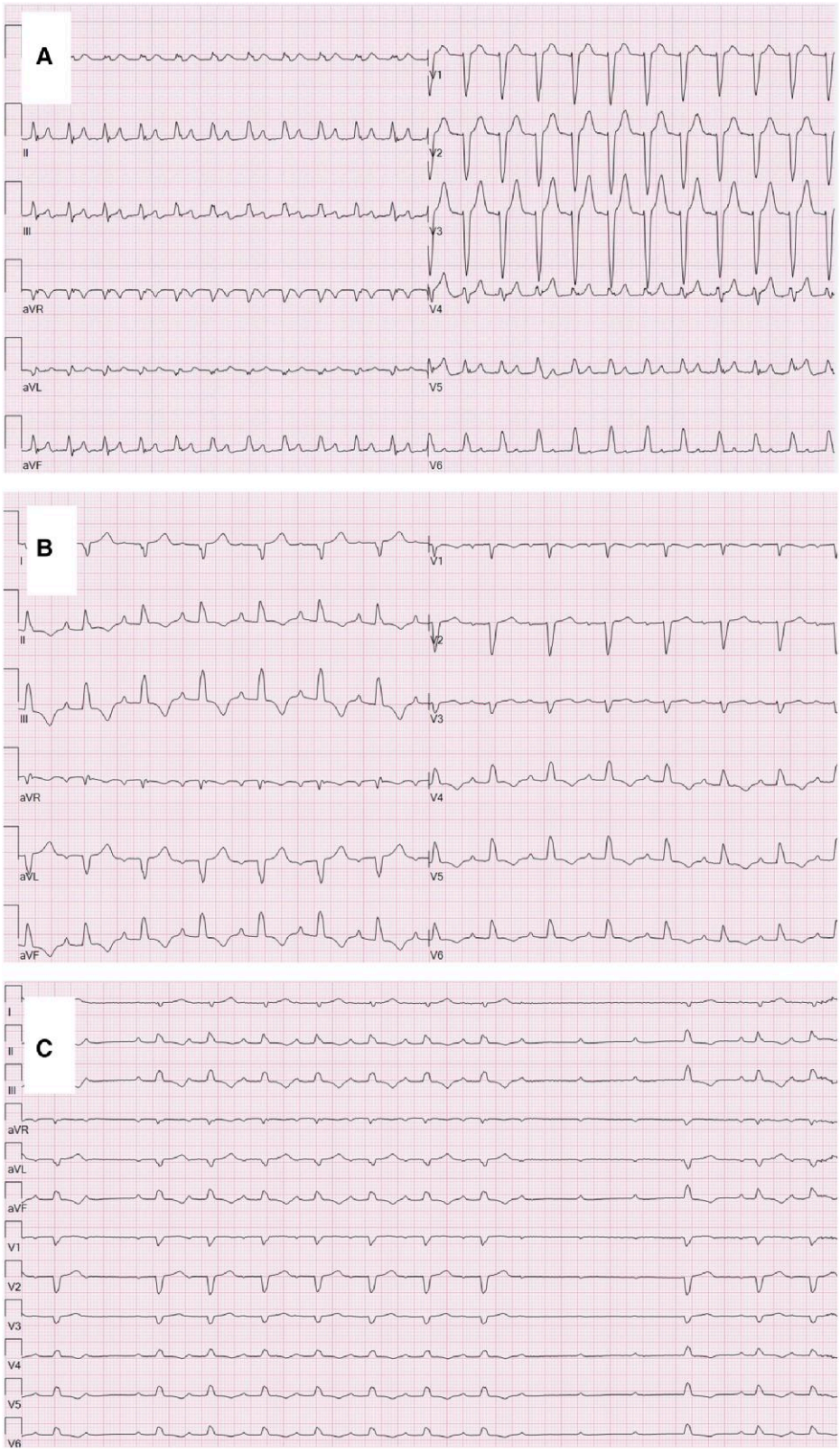

Figure 1 (*A*) The resting electrocardiogram at initial presentation revealed the presence of an atrioventricular nodal re-entrant tachycardia for which she was treated with adenosine (6–12 mg). (*B*) The resting electrocardiogram in sinus rhythm after treatment with adenosine. (*C*) The resting electrocardiogram at the second presentation revealed the presence of a high-grade atrioventricular block.

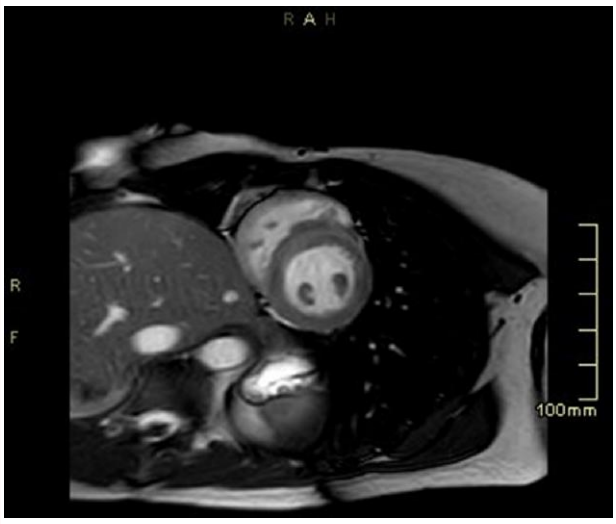

Figure 2 The image of 'dark-paps' on cardiac MRI. This is a hypo-intense aspect of the papillary muscles that is being recognized as an independent predictor associated with an increased risk of ventricular arrhythmia.[3]

Following the diagnosis, high-dose intravenous corticosteroids were initiated, accompanied by methotrexate (10 mg). A gradual tapering schedule was provided. Given the progressive conduction abnormalities and the associated risk of malignant ventricular arrhythmias, an ICD was implanted.

In the following months, as her corticoid therapy was tapered and methotrexate dose increased, there was no recurrence of high-degree AVB and there were no registrations of transient supraventricular tachycardia. Furthermore, the PR interval normalized and QRS duration decreased.

Discussion

To the best of our knowledge, this is the first case of cardiac sarcoidosis presenting with AVNRT. A cardiac electrophysiology study was not performed in our patient. Therefore, it is not possible to confirm the diagnosis of AVNRT with certainty. However, several characteristics of the case indicate that AVNRT is the most likely underlying substrate for the patient's episodes of narrow QRS tachycardia. Her presenting ECG and the restoration of sinus rhythm after administration of adenosine are both hallmark characteristics of AVNRT. Although some forms of ectopic atrial tachycardia occasionally terminate after adenosine administration, no ectopic atrial activity or episodes of atrial tachycardia with 2:1 or higher degree AVB were seen during ECG monitoring. In order for AVNRT to develop, the patient must exhibit dual AV nodal physiology in which, in addition to conduction over the so-called fast pathway, conduction is also possible over a slow pathway, which has a slower conduction velocity and a shorter refractory period. Why some people exhibit such dual AV nodal physiology and others do not is not known, as it appears to be a common variant of AV conduction solely via the fast pathway and is subject to variations in autonomic tone. While the presence of dual AV nodal pathways as a predisposing factor for AVNRT before the onset of cardiac sarcoidosis in this patient cannot be excluded, we hypothesize that the conduction disturbances induced by cardiac inflammation were the main trigger for the development of AVNRT in our patient. The main argument for this is the absence of any episodes of AVNRT/palpitations in her past medical history, and the development of AVNRT episodes in parallel with clear AV conduction disturbances (first degree AVB and LBBB). A similar mechanism of AVNRT induction caused by iatrogenic slowing of the AV nodal conduction was described by Roggen *et al.*[4] in a congenital heart disease patient who experienced a mechanical block of the AV nodal conduction system during balloon angioplasty of the pulmonary artery.

We suspect that a similar mechanism is responsible for the development of a re-entrant tachycardia in this particular case. The presence of the granulomas and their ensuing oedema likely interfered with the AV conduction system, creating a region of slow conduction. This acquired mechanical slow pathway gave rise to a dual conduction circuit, forming an anatomical substrate for an AVNRT. Given the ECG findings, including a long RP interval and negative P-wave in lead II, we hypothesize that this was an AVNRT of the slow–slow subform.[5] The resolution of AVNRT episodes after treatment of the cardiac sarcoidosis provides a further argument for our hypothesis, although definitive proof would have required an electrophysiology study, which was not performed given the favourable clinical course after immunosuppressive treatment. Although AVNRT due to altered AV nodal conduction properties in cardiac sarcoidosis has not been described, the presence of conduction abnormalities, such as first degree AVB, is one of the diagnostic hallmarks of cardiac sarcoidosis and can provide the conduction slowing required as a prerequisite for re-entry. Of course, once AV conduction further deteriorates during the course of (untreated) disease, the occurrence of AVNRT is no longer possible due to higher degree AVB.[6]

In conclusion, this case illustrates an AVNRT as an uncommon initial presentation of cardiac sarcoidosis. It is important to be aware that cardiac sarcoidosis can rarely present with supraventricular tachycardia as an initial symptom.

Lead author biography

Dr Laura Casteur is a last year resident in cardiology, mainly focusing on non-invasive cardiac imaging and heart failure. She graduated as MD at KU Leuven in Belgium in 2018. She is currently working in the University Hospital of Leuven, Belgium.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Consent: The authors confirm that written consent for submission and publication of this case report, including images and associated text, has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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Data availability

All data are incorporated into the article and its online Supplementary material.

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