Atypical presentation of cardiac sarcoidosis: role of multimodality imaging

SUMMARY

Sarcoidosis is a rare multi-system, granulomatous disorder of unknown cause, which has a variable clinical presentation that depends on organ involvement. Mediastinal and pulmonary sarcoidosis is reported in 95% of cases. While cardiac sarcoidosis is much rarer. Myocardial involvement is associated with poor prognosis, and can lead to sudden cardiac death caused by ventricular arrhythmia and conduction system disease. The diagnosis of sarcoidosis is challenging given the varied clinical spectrum and imperfect diagnostic techniques. It is supported by a compatible clinical and radiographic presentation, and histological evidence of non-caseating granulomas on biopsy. Several diagnostic criteria have been proposed. We report the case of mediastinal and cardiac sarcoidosis diagnosed in a 32-year-old man with no medical history after his first episode of syncope. Cardiac sarcoidosis was initially suspected by Magnetic resonance imaging and confirmed by histological evidence.

KEYWORDS

Cardiac sarcoidosis; Syncope; Magnetic resonance imaging.

RÉSUMÉ

MOTS-CLÉS

Correspondance

INTRODUCTION

Sarcoidosis is a multi-system, granulomatous disorder that has been first described by Jonathan Hutchinson in 1869 as a dermatological condition (1). It has a variable clinical presentation that depends on organ involvement and the severity of involvement , the lungs and the lymph nodes are mainly affected , while cardiac sarcoidosis (CS) is much rarer (2). Myocardial involvement can precede, follow or occur concurrently with involvement of other organs, it is being increasingly recognised and is associated with poor prognosis and can lead to sudden cardiac death caused by ventricular arrhythmia and conduction system disease. The diagnosis of sarcoidosis is challenging given the varied clinical spectrum and imperfect diagnostic techniques.

CASE REPORT

A 32-year-old man presented to the emergency department after his first episode of syncope. He was an active tobacco smoker; his medical history was unremarkable. He did not have any symptoms before the onset of the syncope.

On physical examination, the patient appeared calm, had a blood pressure of 120/ 72 mmHg, had a heart rate of 67 beats per minute, had a respiratory rate of 15 per minute and had an oxygen saturation of 98% on room air. Thoracic and cardiac auscultation was unremarkable as were the abdominal and neurological examination. No skin lesions were noted and in a thorough external examination, no peripheral lymphadenopathy was found.

Blood counts, cardiac enzymes, inflammatory markers, renal and liver were all within the normal range. On further examination, laboratory results detected no abnormalities of serum angiotensin converting enzyme levels and calcium levels. Electrocardiogram showed sinus rhythm with no major abnormalities.

The patient's initial evaluation included a transthoracic echocardiography that revealed normal function of left ventricle (LV) without regional wall motion abnormalities and the global longitudinal strain (GLS) was normal (-21%), no valvular disease, right ventricular dilatation with a normal function and an estimated pulmonary artery systolic pressure of 24 mmHg.

Twenty-four-hour cardiac monitoring revealed very frequent bimorphic short coupled premature ventricular contractions with triplets, couplets, bigeminy, and trigeminy patterns, without sustained or non-sustained ventricular tachycardia.

Cardiac magnetic resonance (CMR) imaging revealed late gadolinium enhancement (LGE) in the anterior wall of right ventricle (RV) (Figure 1) associated with mediastinal bilateral lymphadenopathy (Figure 2).

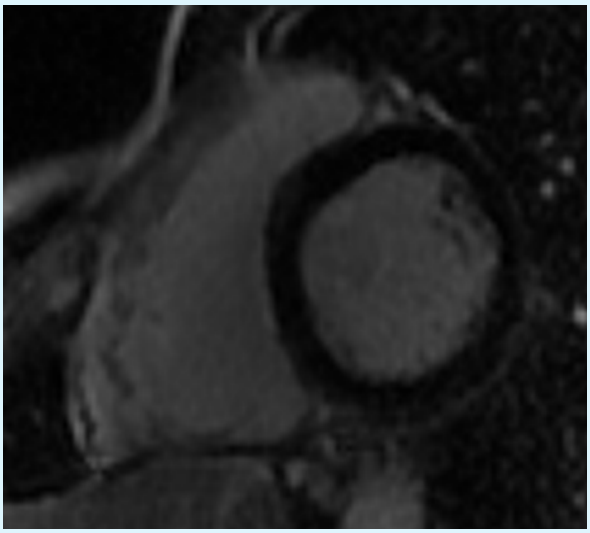

Figure 1. Cardiac magnetic resonance showing Late gadolinium enhancement in the anterior right ventricular wall in the short-axis view(arrow).

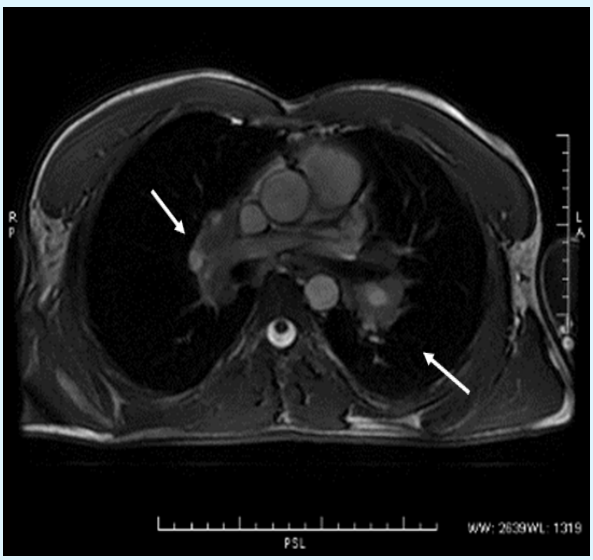

Figure 2. Mediastinal bilateral lymphadenopathy (arrows)

A chest computed tomography CT scan performed as a complement of CMR revealed bilateral mediastinal and hilar symmetric and non-compressive lymphadenopathy without parenchymal involvement (Figure 3).

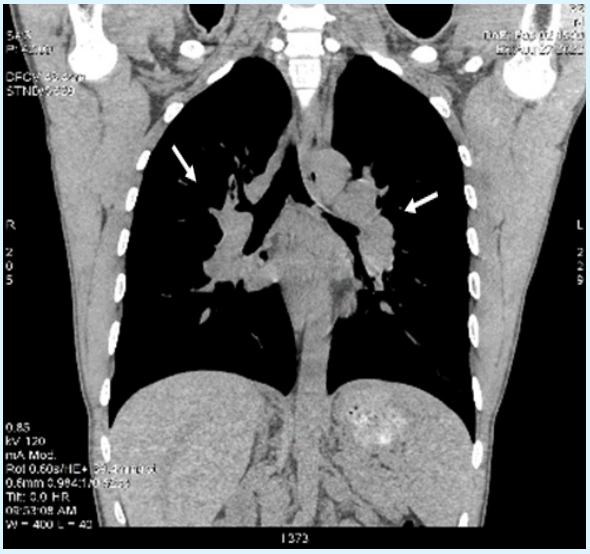

Figure 3. Bilateral mediastinal and hilar symmetric and non-compressive lymphadenopathy without parenchymal involvement on mediastinal window on CT scan (arrows).

Bronchoscopy with endotracheal and trans bronchial lung biopsy was normal, Broncho alveolar lavage showed lymphocytosis and CD4/CD8 ratio more than 3.5, pulmonary function testing and Diffusing capacity of the lung for carbon monoxide were within normal ranges and six-minute walk test distance was normal.

The patient was diagnosed systemic sarcoidosis based on non-caseating granuloma with Langhans type giant cell by the biopsy of the affected lymph nodes sampled via mediastinoscopy.

DISCUSSION

The clinical manifestations of cardiac sarcoidosis ranges from having no symptoms to severe consequences such as sudden cardiac death, ventricular arrhythmia, atrial arrhythmia, other conduction system disease, congestive heart failure, papillary muscle dysfunction, mitral insufficiency, myocardial infarction (due to Coronary vasculitis), ventricular aneurysm, and pericardial effusion (1,3) and that depend on the location and extent of involvement.

The diagnosis of CS, relies heavily on imaging modalities, the transthoracic echocardiography result is often abnormal, some results are considered as major and minor criteria in various diagnostic guidelines despite not being pathognomonic. Findings including isolated non-coronary distribution wall motion abnormalities, LV and/or RV diastolic and systolic dysfunction, impaired LV longitudinal strain, regional ventricular wall thickening (especially septal thinning), dilation of the left ventricle and ventricular aneurysms may be highly suggestive of CS.

CMR has become an important diagnostic and prognostic modality in the evaluation for cardiac involvement, it identifies areas of myocardial damage including oedema and scar, primarily via the LGE technique, The JMHW criteria consider LGE on CMR a minor criterion, while the recently published HRS criteria have included specific LGE patterns on CMR and cardiac uptake on fluorodeoxyglucose-PET as major criteria. LGE is mostly seen in basal segments, particularly of the septum and lateral wall, and usually in the mid-myocardium and sub-epicardium, RV free wall may also be involved (4) . In addition, CMR is considered as a valuable tool for risk stratification, in fact Patel et al (5) demonstrated that LGE may independently predict future adverse events (ventricular tachycardia, sudden cardiac death, atrioventricular block), furthermore Smedema et al believe that involvement of the RV in addition to the LV increases the risk of worse outcomes and all-cause mortality (3). Apart from LGE, CMR can detect morphologic abnormalities such as increased myocardial wall thickness, or increased intra-myocardial signal intensity on T2-weighted images and aneurysms

The CMR superiority lies in its ability to identify myocardial fibrosis, without any electrical or functional abnormality on conventional testing, allowing detection of «silent" CS (6).

Although FDG-PET is not very accessible in our country it is currently the best clinical tool for the assessment of myocardial inflammation in CS enabling the diagnosis in different stages of the disease from the early stage (no or mild perfusion defect with increased corresponding FDG uptake), to the stage

of fibrosis (severe perfusion defect with minimal or no FDG uptake as illustrated) therefore it serves as an excellent tool for an early diagnosis, an early treatment and thus a better prognosis (6,7).

CMR and FDG-PET scans can also be used for evaluating the efficacy of immunosuppressive therapy in patients with CS (8).

CONCLUSION

Sarcoidosis is a multisystem inflammatory disorder with an unclear aetiology and a variable clinical presentation. CS is often underdiagnosed, but the diagnosis has become significantly more facilitated in recent years thanks to new non-invasive imaging modalities (CMR and FDG-PET scan).

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