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CASE REPORT

Rare presentation of sarcoidosis: Multimodal imaging diagnosis of cardiac involvement

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KEYWORDS
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Abstract We report a case of sarcoidosis with an unusual presentation, initially manifesting as bilateral pulmonary embolism and then as a cardiac form of the disease with an ominous clinical event consisting of sustained ventricular tachycardia. The diagnosis was established by clinical and magnetic resonance criteria despite normal conventional echocardiographic study. Detailed functional assessment provided by tracking techniques (speckle tracking echocardiography and cardiac magnetic resonance tissue tracking) enabled the detection of regional deformation abnormalities, indicating prominent circumferential strain and epicardial layer alterations, partly matching the structural changes depicted by distribution of delayed enhancement.

We find this case notable for various issues it raises concerning diagnosis and management of cardiac sarcoidosis. These are mainly related to recent developments in imaging modalities that enable non-invasive identification of structural and functional abnormalities in this condition early, before overt deterioration in left ventricular ejection fraction. Information from different imaging modalities and tools provide information that could potentially assist preclinical diagnosis, with possible prognostic implications.

PALAVRAS-CHAVE
Sarcoidose cardíaca;
Embolia pulmonary;
Speckle tracking;
Tissue tracking

Apresentação rara de sarcoidose: diagnóstico de envolvimento cardíaco por imagem multimoda

Resumo Apresenta-se o caso de um doente de 44 anos com o diagnóstico prévio de sarcoidose pulmonar, admitido consecutivamente no serviço de urgência por embolia pulmonar e taquicardia ventricular sintomática. Embora o estudo ecocardiográfico mostrasse normal fração de ejeção ventricular esquerda, o diagnóstico de sarcoidose cardíaca foi estabelecido por elementos de ordem clínica, em conjugação com os achados de ressonância magnética

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cardíaca. Quando se efetuou estudo funcional detalhado do ventrículo esquerdo por *speckle* (ecocardiografia) e *tissue tracking* (ressonância magnética), detetaram-se alterações regionais da deformação miocárdica, parcialmente coincidentes com a distribuição do realce tardio por ressonância. Para além da apresentação pouco habitual sob a forma de evento embólico, este caso de sarcoidose com subsequente documentação de envolvimento cardíaco, permitiu a aplicação de modalidades de imagem cardiovascular avançadas, colocou em evidência a afeção estrutural e funcional miocárdica, na presença de normal fração de ejeção ventricular esquerda.

Introduction

Sarcoidosis is a multisystem inflammatory disease of unknown etiology, with heterogeneous presentation and possible cardiovascular involvement. Associated mortality is mainly due to respiratory, neurological or cardiac complications. Myocardial involvement, which may manifest as heart block, ventricular arrhythmias or heart failure from both systolic and diastolic dysfunction, may have life-threatening consequences, and cardiac sudden death may be the first presentation of cardiac sarcoidosis, occurring independently of pulmonary or other organ involvement. When sarcoidosis is fatal, cardiac involvement is a frequent cause of death[1,2]. Recently, an association between sarcoidosis and pulmonary embolism has been described, which is explained by inflammatory and other biochemical mechanisms[3,4]. This could worsen the prognosis of both pulmonary and cardiovascular disease and makes it important to seek early identification of patients at risk.

It has become clear that asymptomatic cardiac involvement is far more prevalent that previously thought. Nevertheless, there is a lack of consensus as to the diagnostic and cardiovascular imaging modalities to be used and their relative accuracy in identifying the presence of preclinical cardiac disease.

Conventional two-dimensional (2D) echocardiography is recommended for assessing sarcoidosis patients with suspicion of cardiac involvement. However, morphological changes and overt global ventricular dysfunction, as assessed by left ventricular ejection fraction (LVEF), probably occur simultaneously with already established clinical cardiac manifestations.

Speckle tracking echocardiography (STE) is a valuable tool for the quantitative assessment of regional myocardial function. As in its early stages cardiac sarcoidosis does not affect the myocardium uniformly or globally, the disease could theoretically be identified by this technique, possibly before overt deterioration in LVEF[2]. In keeping with the mainly regional nature of the disease, cardiac magnetic resonance (CMR) is currently one of the advanced high-resolution imaging techniques of choice in the assessment of sarcoidosis, enabling rapid, accurate, and non-invasive diagnosis. Some studies have set out to find correlations between functional myocardial changes as assessed by STE and scar distribution on contrast-enhanced (CE) CMR[5]. Furthermore, as tissue regions are identified by individual anatomical features in CMR, feature- and tissue-tracking CMR has been explored in 2D cine image stacks, and a series of deformation parameters describing myocardium mechanics can also be derived[6].

We present a case report of cardiac sarcoidosis with a rare presentation, preceded by extensive pulmonary embolism. Although the diagnosis was made by CE-CMR, we proceeded with further segmental functional analysis by STE, investigating possible effects on regional function. We also sought to assess myocardial deformation by tissue-tracking CMR and the extent of its agreement with STE.

Case report

A 44-year-old black male with pulmonary sarcoidosis diagnosed at the age of 39, under irregular corticosteroid therapy, presented to the hospital with sudden onset of effort dyspnea. On admission the patient was in no distress, with tachycardia (100 beats/min) and normal blood pressure and oxygen saturation (ambient air). Physical examination was negative except for the presence of bilateral inspiratory crackles in the middle third of both lung fields. The electrocardiogram (ECG) revealed no changes other than sinus tachycardia, and blood analysis was remarkable for D-dimer elevation with normal BNP levels. The patient underwent pulmonary computed tomography (CT) angiography, which revealed bilateral pulmonary embolism in addition to parenchymal nodular infiltration, interstitial ground glass pattern and bilateral mediastinal lymphadenopathy with several lymph node conglomerates (Figure 1A). Transthoracic echocardiography was normal, with no signs of acute right-sided pressure overload or of right ventricular dysfunction. Lower limb Doppler ultrasound was positive for partial right popliteal vein thrombosis. The patient was started on oral anticoagulation with rivaroxaban in addition to corticotherapy; hospital stay was uneventful and he was discharged.

Six weeks after admission he returned to the emergency department complaining of sudden dizziness. He was