Response to steroid therapy in cardiac sarcoidosis: insights from myocardial strain

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Sarcoidosis is a multisystem granulomatous disease of unknown aetiology and cardiac involvement can occur. Echocardiographic abnormalities, such as left ventricular dysfunction, segmental wall thinning, ventricular aneurysm, or valvular abnormalities are often subtle until the later stages of the disease. However, sarcoid has a predilection to cause ventricular arrhythmias or conduction system abnormalities in the early stages and hence may develop palpitations, syncope, or sudden death before structural abnormalities are detected. If patients with early cardiac sarcoid are identified, they respond well to corticosteroid therapy, and defibrillator implantation may reduce the risk of sudden death from malignant arrhythmias. We present a case of a patient with cardiac sarcoid, manifesting as conduction system disease. Cardiac magnetic resonance imaging demonstrated a sarcoid granuloma within the LV septum. Although the standard echocardiographic evaluation was unremarkable, this area corresponded to abnormal strain imaging on echo. Similarly, while conventional echocardiographic measurements failed to demonstrate a response to steroid therapy, strain imaging showed improved regional myocardial function. This coincided with improvement in the conduction abnormalities. This case study showed the potential of strain imaging in demonstrating cardiac involvement from sarcoidosis and in assessing the therapeutic response to corticosteroid therapy.

Keywords
Sarcoidosis • Cardiac MRI • Strain imaging • heart block

Case report
A 59-year-old man was admitted following loss of consciousness. A bilateral inflammatory arthropathy was evident. Electrocardiogram demonstrated complete atrioventricular (AV) block with pauses >4 s. Biopsy of a skin nodule confirmed sarcoid.

Echocardiography showed normal left ventricular (LV) size and systolic function with an ejection fraction of 55%.
Figure 3  Strain curves for the basal (green), mid- (yellow), and apical (red) septum before the commencement of thrombolytic therapy. Note that the basal septum demonstrated abnormal systolic lengthening (positive strain—above the zero baseline). The systolic strain of the mid-septum was reduced.

Figure 4  Strain curves for the basal (green), mid- (yellow), and apical (red) septum at 6 months after the commencement of corticosteroid therapy. Note that the basal septum demonstrated normal systolic shortening (negative strain—below the zero baseline). The systolic strain of the mid-septum has greatly improved.
Figure 5  (A) Strain curves for the basal septum (yellow) and basal anterolateral wall (green) before the commencement of thrombolytic therapy. Note that the basal septum demonstrated abnormal systolic lengthening (positive strain—above the zero baseline). (B) Strain curves for the basal septum (yellow) and basal anterolateral wall at 6 months after the commencement of corticosteroid therapy. Note that the basal septum demonstrated normal systolic shortening (negative strain—below the zero baseline). The systolic strain basal anterolateral wall has greatly improved.
Anteroseptal thickness was moderately increased (15.5 mm). A small echogenic area was shown at the basal anteroseptum (Figure 1). Late gadolinium magnetic resonance imaging (MRI) showed a corresponding area of enhancement (Figure 2) consistent with sarcoid granuloma at the level of the AV node.

Tissue velocity imaging-derived longitudinal strain was measured from the apical four-chamber view at the basal, mid-, and apical segment of the ventricular septum. The basal septum demonstrated positive strain (+7%), indicating dyskinesis in this segment (Figure 3). Corticosteroid therapy (oral prednisone at 25 mg/day following intravenous therapy) was commenced.

A follow-up echocardiogram performed 6 months later showed unchanged LV systolic and diastolic function. The echogenic area at the basal septum remained. However, the strain measurements at the basal and mid-ventricular septum (Figure 4) and lateral wall (Figure 5A and B) had improved. The basal ventricular septum now demonstrated a negative strain. The strain of the mid-septum and lateral wall also improved. Strain assessment could play an important role for the serial assessment of cardiac sarcoid patients, as implantable cardioverter defibrillator implantation precludes the assessment of a therapeutic response by MRI.

Strain imaging is an important tool in the early characterization of a therapeutic response to corticosteroid therapy.