MRI of Sarcoid Heart Disease

Saurabh Jha MBBS,
University of Pennsylvania,
Philadelphia

Objectives

- To appreciate the pathophysiology and clinical presentation of myocardial sarcoid
- To understand the role of imaging, specifically MRI, in sarcoid heart disease
- To understand the rationale for the imaging protocol and the imaging findings in myocardial sarcoid
- To be cognizant of the differential diagnosis of the imaging findings in cardiac sarcoid

Sarcoidosis

- Multi-system disease characterized by non-caseating granulomas
- Affects predominantly the lungs, lymph nodes, skin and eyes
- More common in African Americans
- Clinical course and presentation variable – indolent to aggressive
- Treatment with immunomodulators such as steroids

Cardiac Sarcoidosis

- Many more with sarcoidosis have myocardial involvement at autopsy (25%) than are symptomatic from cardiac sarcoid (5%)
- Responsible for 13-25% of deaths from sarcoidosis in the US (higher in Japan)
- Prognosis is worsened by myocardial involvement but is not as dismal as previously thought
- Treatment with steroids, pacemaker/ AICD, cardiac transplantation

Pathophysiology of cardiac Sarcoidosis

Cardiac dysfunction:

a) Direct granulomatous involvement (basal septum, lateral LV, free wall of RV)

b) Secondary to pulmonary hypertension (cor pulmonale) – right-sided failure

Cardiac involvement can be defined in 3 stages:
1. Edema
2. Granulomatous infiltration
3. Fibrosis
Axial b-SSFP (a) and axial double IR (b) shows pulmonary sarcoid and enlarged pulmonary arteries due to pulmonary hypertension. Short axis mid cavity IR-prepped gradient echo (c) shows a focus of delayed enhancement at the insertion point of the RV on to the septum – a finding associated with elevated right-sided pressures. The elevated right-sided pressures and a bundle branch block leads to abnormal septal motion (d).

Clinical features of cardiac sarcoid

- Cardiac failure – may be bi-ventricular
- Restrictive cardiomyopathy – granulomatous infiltration causes diastolic dysfunction
- Arrhythmias/ sudden death – depends on the proximity of the granulomas to the conduction system. Can cause complete heart block. Granulomas can be a pivot for re-entry tachyarrhythmias
- Pericardial effusion
- Acute chest pain, elevated troponin

Diagnostic strategies for myocardial sarcoid

A) EKG
- RBBB, PVCs, T wave changes. The findings are non-specific but if new are highly suggestive of myocardial sarcoid involvement

B) Biopsy

C) Imaging
- MRI
- Nuclear medicine
- Coronary angiography – exclusion of revascularizable coronary lesion

Purposes of Imaging

- Diagnosis
- Prognosis
- Direction and monitoring of response to therapy
- Immunomodulators work best when there is “active” sarcoid. The corollary is that successful treatment reduces the activity of sarcoid. This can be assessed by both MRI and nuclear medicine. In MRI, the myocardial edema and the degree and extent of MDE reduces with successful steroid therapy
Purposes of Imaging

• Avoidance/ direction of endomyocardial biopsy

Strong imaging findings of cardiac sarcoid coupled with histological diagnosis of extra-cardiac sarcoid obviates the need for endomyocardial biopsy. In equivocal cases, the pre-procedural location of sarcoid involvement can reduce the incidence of sampling error/ false negative biopsy.

• Offering of alternative explanations for myocardial dysfunction in patients with sarcoidosis

Patients with sarcoid may have coronary artery disease which would change the management strategy.

Key point

The systemic toxicity of high doses of immunomodulators to treat cardiac sarcoid mandates that the diagnosis of myocardial sarcoid be reliably made.

Cardiac MRI protocol - principles

To conceive the appropriate protocol imaging the pathophysiology should be visited. Imaging must be able to detect:

a) Acute inflammation/ edema - fluid-sensitive sequence (triple inversion recovery)
b) Granulomas – early post gadolinium T1 weighted sequence
c) Chronic inflammation/ fibrosis – inversion recovery myocardial delayed enhancement (MDE)
d) Segmental myocardial dysfunction – cine bright blood sequence
e) Restrictive cardiomyopathy/ altered chamber size – black blood sequence

Cardiac MRI protocol

• 3-plane bright blood localizers - balanced steady state free precession (b-SSFP)
• EKG – triggered axial and short axis double inversion recovery single shot fast spin echo (black blood), and at least one plane triple inversion recovery
• VLA and 4 chamber b-SSFP cine.
• Perfusion using fast gadolinium injection and a low flip angle gradient echo with pre-pulse to achieve T1 weighting
• Early post gadolinium short axis and axial 2D gradient echo T1
• Short axis b-SSFP cine
• Myocardial delayed enhancement – inversion recovery-prepped EKG-triggered

Findings on MRI

• Myocardial thickening disproportionate to the degree of contraction
• Mural edema
• Wall thinning
• Disproportionate atrial dilatation (if diastolic dysfunction)
• Segmental wall motion abnormality not in an epicardial coronary artery territory
• Enhancing mural nodules
• Myocardial delayed enhancement

The presence of mediastinal or pulmonary sarcoid adds specificity to the above findings.

Short axis (a) and (c) and VLA inversion recovery MDE shows a pattern that can be described as “non-typical” for ischemic heart disease. Note the patchy involvement, with sparing of the sub-endocardium. The distribution is just as important as morphology – this is not in a coronary artery territory.
Short axis and VLA cine images correlate the wall motion of the segment with the MDE. Note that the contraction is relatively preserved for the degree of MDE, a finding unlikely to be present with MI.

Axial DIR at the level of the main pulmonary artery (a) shows mediastinal adenopathy. Axial double IR at the level of the RA (b) shows a subtle high signal along the RV free wall which maintains its high signal on the triple IR (c), indicating that it is fluid not fat.

4 chamber cine shows subtle abnormality along the septal wall (a) confirmed by the short axis grid tagging (b) where the deformation of the grids is greater along the lateral than the septum. Note the myocardial edema (c) on the triple inversion sequence. Images (d), (e) and (f) show MDE in the basal septal wall in this patient with suspected cardiac sarcoid.

The patient with neurosarcoidosis presented with chest pain and elevated troponins. Coronary angiogram was negative. Axial (a) and short axis (b) double inversion recovery shows myocardial edema confirmed on the short axis triple inversion recovery. This suggests an acute non ischemic process which could be myocarditis or sarcoidosis, but the latter is favored because of the presence of systemic sarcoidosis.

**Myocardial delayed enhancement in sarcoid**

A. Distribution
   - Patchy
   - Does not conform to an epicardial coronary artery territory
   - Sub-epicardial
   - Basal septum
   - More intense on the RV side of the septum

B. Morphology
   - Nodular

   Lacks specificity without the clinical context

**Key point**

The pattern of MDE may sometimes be highly suggestive of sarcoid, but more often is not a signature pattern.
Short axis PSIR shows mid myocardial MDE in the inferior wall in a patient with potential cardiac sarcoid. This pattern is not pathognomonic for any entity and may be due to myocarditis. The 4 chamber view shows MDE involving the papillary muscle. This patient has histologically proved extra-cardiac sarcoid and a diagnosis of cardiac sarcoid was made.

**Differential diagnosis of cardiac sarcoidosis**

A) Non-ischemic myocardial processes such as myocarditis  
B) Hypertrophic cardiomyopathy  
C) Ischemic heart disease

**Alternative imaging strategy to MRI**

Contraindications to MRI – pacemakers, risk of nephrogenic systemic dermopathy may require alternatives.  

**Nuclear medicine**  
a) Thallium-Gallium combination  
   - Gallium is only taken up by active lesions.  
   - Thallium shows reduced perfusion that worsens with exercise/vasodilators (reverse redistribution)  
b) FDG-PET  
Nuclear medicine lacks the spatial resolution and multi-planar capabilities of MRI. High on radiation dose.

**Cardiac CT**  
Lacks the soft tissue contrast of MRI. High radiation, particularly if both the arterial and the delayed acquisitions are EKG-synchronized.

**MRI in difficult circumstances**

**Patient cannot breath hold**

a) Decrease the imaging time - reduce the phase matrix, increase the views per segment, use a single shot sequence instead of a fast spin echo, use non cartesian k space traversals, parallel imaging.  
b) Respiratory synchronization, navigator gating  

**Patient has an arrhythmia**

a) Increase the arrhythmia rejection window in cine imaging  
b) Real time sequences such as non cartesian MRI

**Patient has a pacemaker**  
Previously a taboo, now done in extremely controlled situations with favorable risk benefit ratio. Important to lower the SAR, appropriate EP personnel and support, familiarity with PM settings

Short axis cine (a) using b-SSFP with retrospective gating. The images are blurred due to inability to suspend respiration as the patient had obstructive sleep apnea. Short axis cine (b) using retrospectively gated b-SSFP with a radial k space traversal shows less blurring due to respiratory motion. Note the presence of radial “spokes” that represent artifact of this technique

**Key points**

- Myocardial involvement is a common and potentially fatal outcome of sarcoidosis  
- Early and accurate diagnosis is important to direct therapy  
- MRI is sensitive in the diagnosis of early sarcoidosis  
- Myocardial edema is indicative of active sarcoid  
- Myocardial delayed enhancement indicates granulomatous infiltration/ fibrosis
References

1. Doughan AR, Williams BR. Cardiac sarcoidosis. Heart. 92: 2006; 282–8
2. Olivier Vignaux, Cardiac Sarcoidosis: Spectrum of MRI Features AJR, Jan 2005; 184: 249 – 254

Acknowledgement

Harold Litt MD PhD, University of Pennsylvania