Beyond The Common Ground: The Unusual Suspects in Late Gadolinium Enhancement

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Learning objectives

1. To briefly describe the technique of delayed enhancement cardiac magnetic resonance (CMR) imaging.

2. To discuss pathophysiology and patterns of late gadolinium enhancement (LGE) in a myriad of uncommon non-ischemic cardiomyopathies, congenital cardiac conditions, as well as inflammatory and infiltrative myocardial disease. These include endocardial fibroelastosis, non-compaction cardiomyopathy, collagen vascular diseases such as scleroderma, muscular dystrophy, Fabry's disease, as well as conditions causing pulmonary arterial hypertension, both idiopathic and those related to congenital heart disease.

Background

CMR permits optimal differentiation between normal and diseased myocardium with the use of gadolinium-based contrast agents and specific pulse sequences. First described more than 10 years ago [1], delayed enhancement CMR has now become the standard of reference for evaluation of myocardial scar due to infarction. Delayed myocardial enhancement is, however, not specific for myocardial infarction (MI) and can occur in a variety of non-ischemic cardiac conditions. Differential myocardial contrast enhancement forms the basis of tissue characterization and is seen in many pathophysiologic scenarios other than MI: increased volume of contrast material distribution in inflammatory or infectious conditions, retention of contrast material by fibrous tissue in infiltrative and cardiomyopathic conditions. Because it allows differentiation among diagnoses based on the different patterns of delayed enhancement, LGE is being increasingly used for the assessment of non-ischemic cardiac conditions, such as dilated cardiomyopathy (DCM) and hypertrophic cardiomyopathy (HCM). However, LGE is also a feature of a number of rare cardiomyopathic conditions and infiltrative/inflammatory myocardial diseases.

Imaging findings OR Procedure details

The technique for LGE-CMR involves intravenous administration of gadolinium chelate contrast material (0.05-0.2 mmol/kg) followed 8-30 minutes later by a cardiac-gated T1-weighted pulse sequence. The typical LGE pulse sequence is a cardiac-gated segmented inversion-recovery-prepared fast gradient-echo sequence: an inversion recovery preparatory pulse nulls normal myocardium, followed by a segmented k-space gradient-echo acquisition. Retention of contrast material results in T1 shortening and

thus increased signal intensity on T1-weighted images relative to that of the normal myocardium. Typical values for inversion time (TI) are 150-400 milliseconds (ms), varying from patient to patient based on their cardiac and renal function. Phase-sensitive inversion recovery (PSIR) reconstruction algorithm can also be applied to reduce the effects of dynamic changes in TI, resulting in consistently good image quality [2]. Unlike in ischemic heart disease, delayed enhancement in non-ischemic myocardial disease generally does not correspond to any particular coronary artery distribution and is often midwall rather than subendocardial or transmural [3, 4].

In advanced cardiac *sarcoidosis*, LGE can be seen, as areas of focal, patchy hyperenhancement, usually located subepicardial or in the midwall (Figures 1-3). *Fabry Disease*, an X-linked disorder of lysosomal metabolism, is a relatively common cause of left ventricular (LV) hypertrophy in middle-aged men. Fibrosis from an unclear cause results in patients with severe disease showing focal inferolateral midwall LGE (Figure 4) [5]. Systemic sclerosis, a subtype of *scleroderma,* can result in myocardial fibrosis, likely throughfibroblast proliferation and collagen accumulation [6]. Midwall enhancement of predominantly the basal and midcavity segments of left ventricle (LV) is the most commonly described LGE pattern (Figures 5-7) [6].

Non-compaction cardiomyopathy is a complex, not fully understood, entity characterised by the presence of numerous and prominent trabeculations together with deep intertrabecular recesses in a portion of the ventricular wall, thought to be a result of a congenital anomaly of endomyocardial development [7]. LGE is seen in areas of myocardial fibrosis (Figure 8).

LGE-CMR also has an important role in other congenital cardiac conditions. Delayed enhancement of the right ventricle (RV) in association with reduced function or an aneurysm is highly suggestive of *arrhythmogenic right ventricular cardiomyopathy (ARVC)* in patients with non-sustained ventricular tachycardia and left bundle-branch block [8] (Figures 9-12). LGE has also been described in *muscular dystrophy* cases, where cardiac myocyte dystrophin deficiency leads to fibre necrosis causing replacement of morbid myocardium with connective tissue and fat [9]. This manifests as high signal intensity on LGE-CMR imaging, predominantly in the midwall (Figures 13-15) [9].

In patients with suspected *endocardial fibroelastosis*, accurate identification has previously required endomyocardial biopsy, though LGE-CMR, with its ability to demonstrate subendocardial fibrosis and mural thrombus (Figure 16), has become relevant to diagnosis and prognosis [10].

Images for this section:

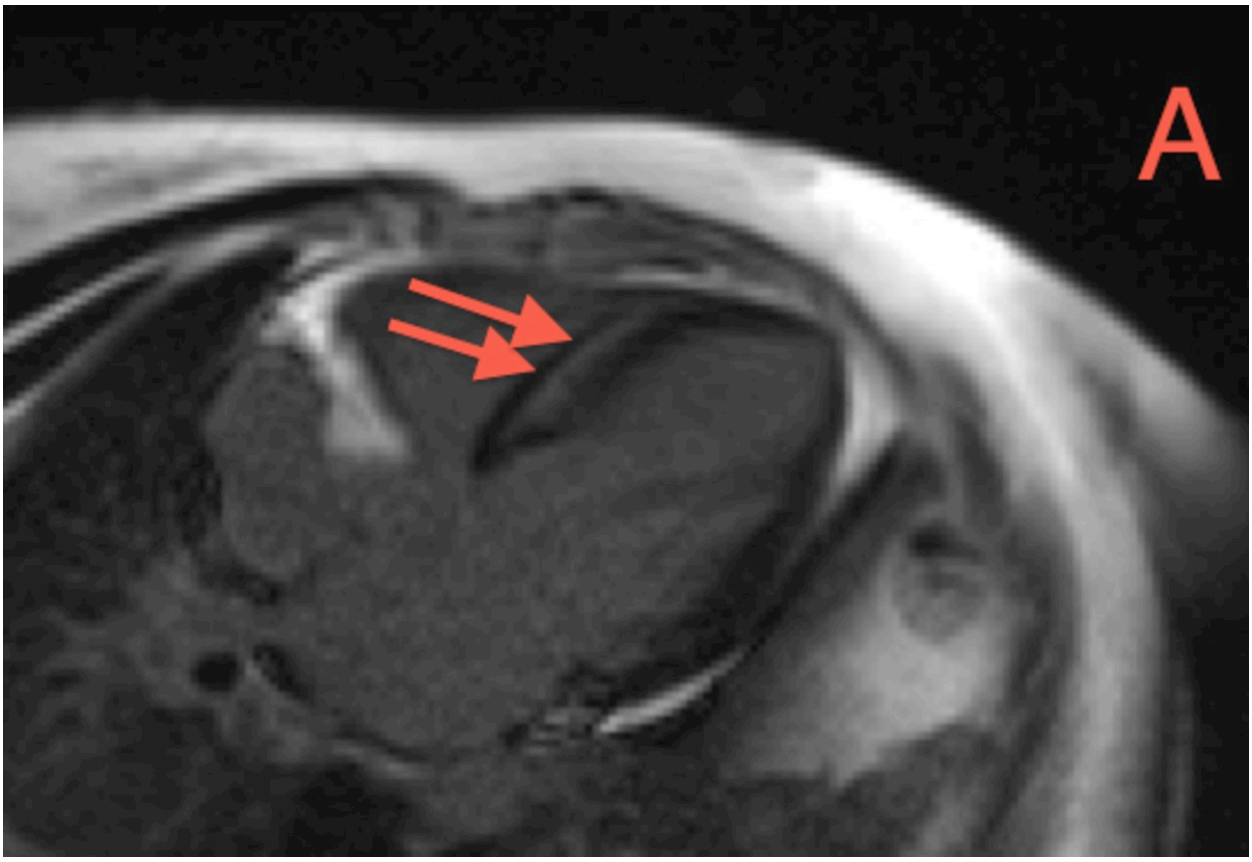

Fig. 1: Sarcoidosis. Mid-wall LGE (arrows) involving septal (A) and inferoseptal (B) LV walls.

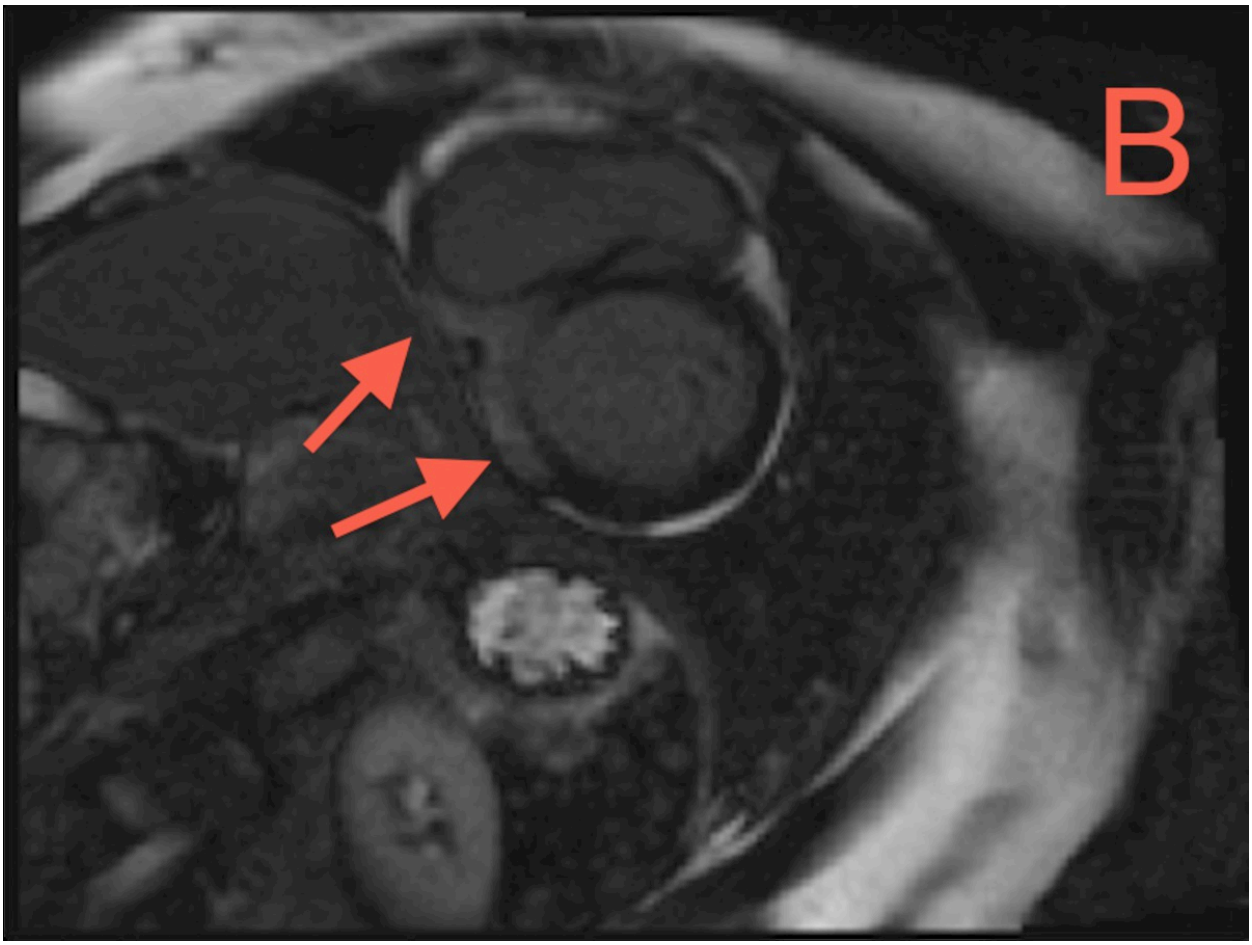

Fig. 2: Sarcoidosis. Mid-wall LGE (arrows) involving septal (A) and inferoseptal (B) LV walls. Mid-wall LGE of small portion of the inferior RV wall (B) is also present.

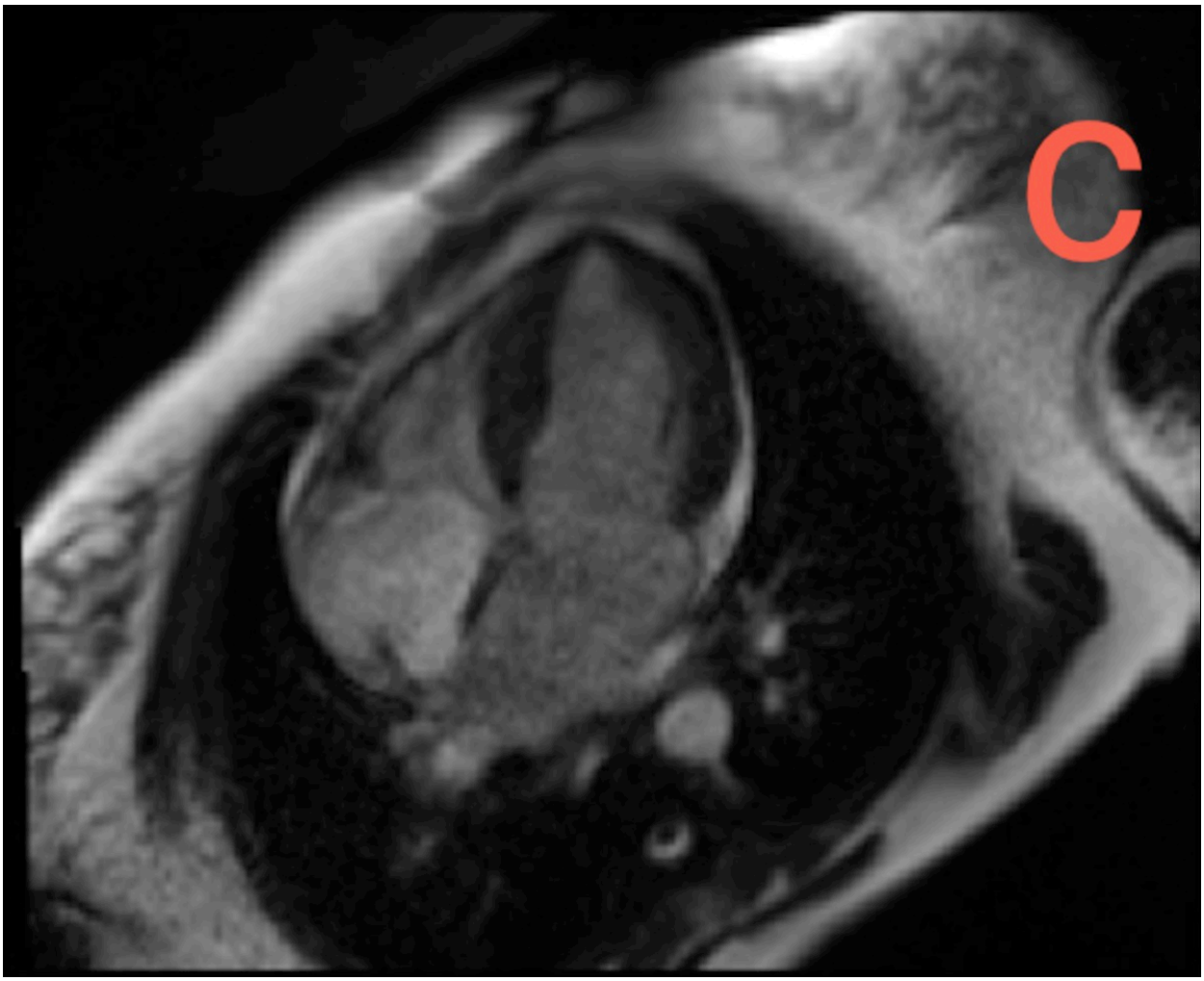

Fig. 3: Sarcoidosis. Four-chamber steady state free precession (SSFP) image (C) shows a mildly thickened LV with apical thinning.

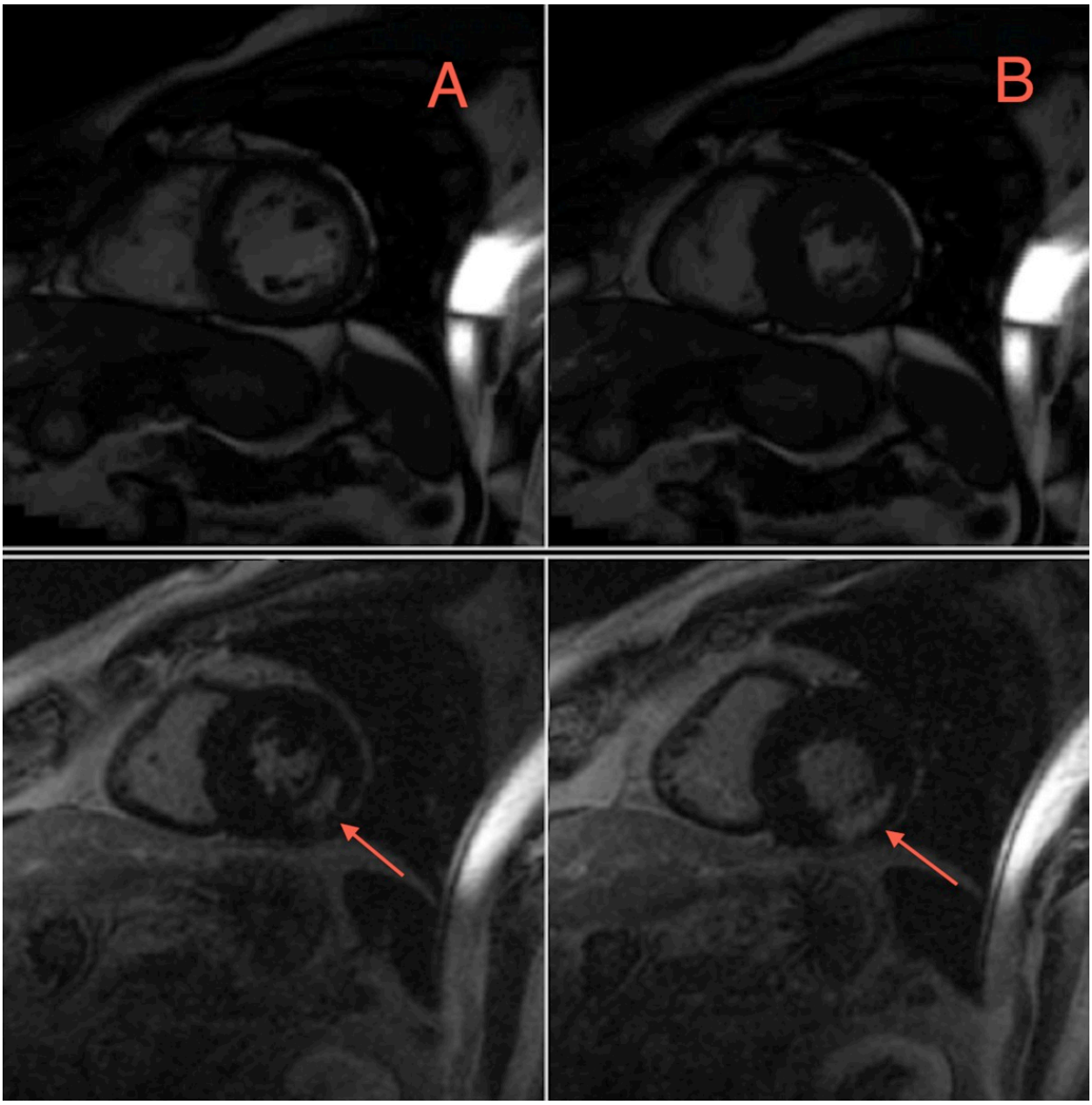

Fig. 4: Fabry disease. Top row: Two-chamber SSFP images in diastole (A) and systole (B) show concentric LV hypertrophy. Bottom row: Focal mid-wall LGE (arrows) involving the infero-lateral LV wall.

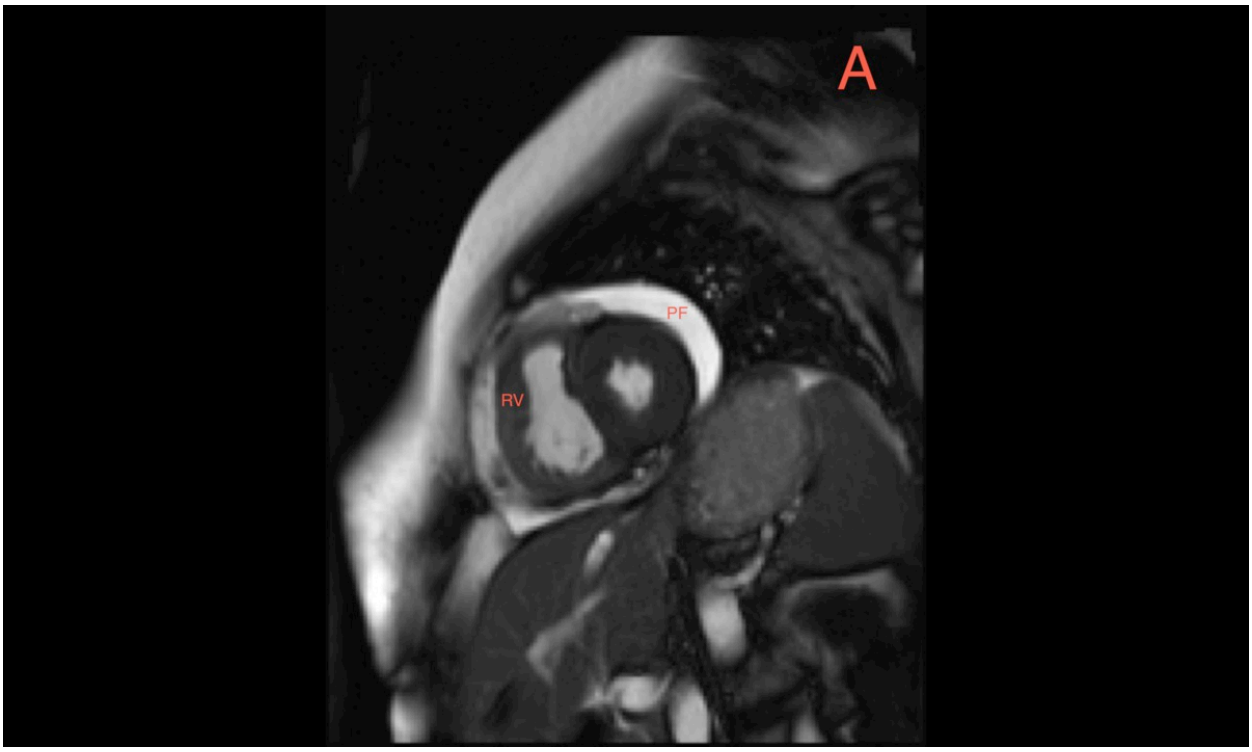

Fig. 5: Scleroderma. Short-axis SSFP image in systole (A) demonstrates RV hypertrophy secondary to pulmonary hypertension in this patient with scleroderma. PF = pericardial fluid. Straightening of the inter-ventricular septum suggests increased RV pressure.

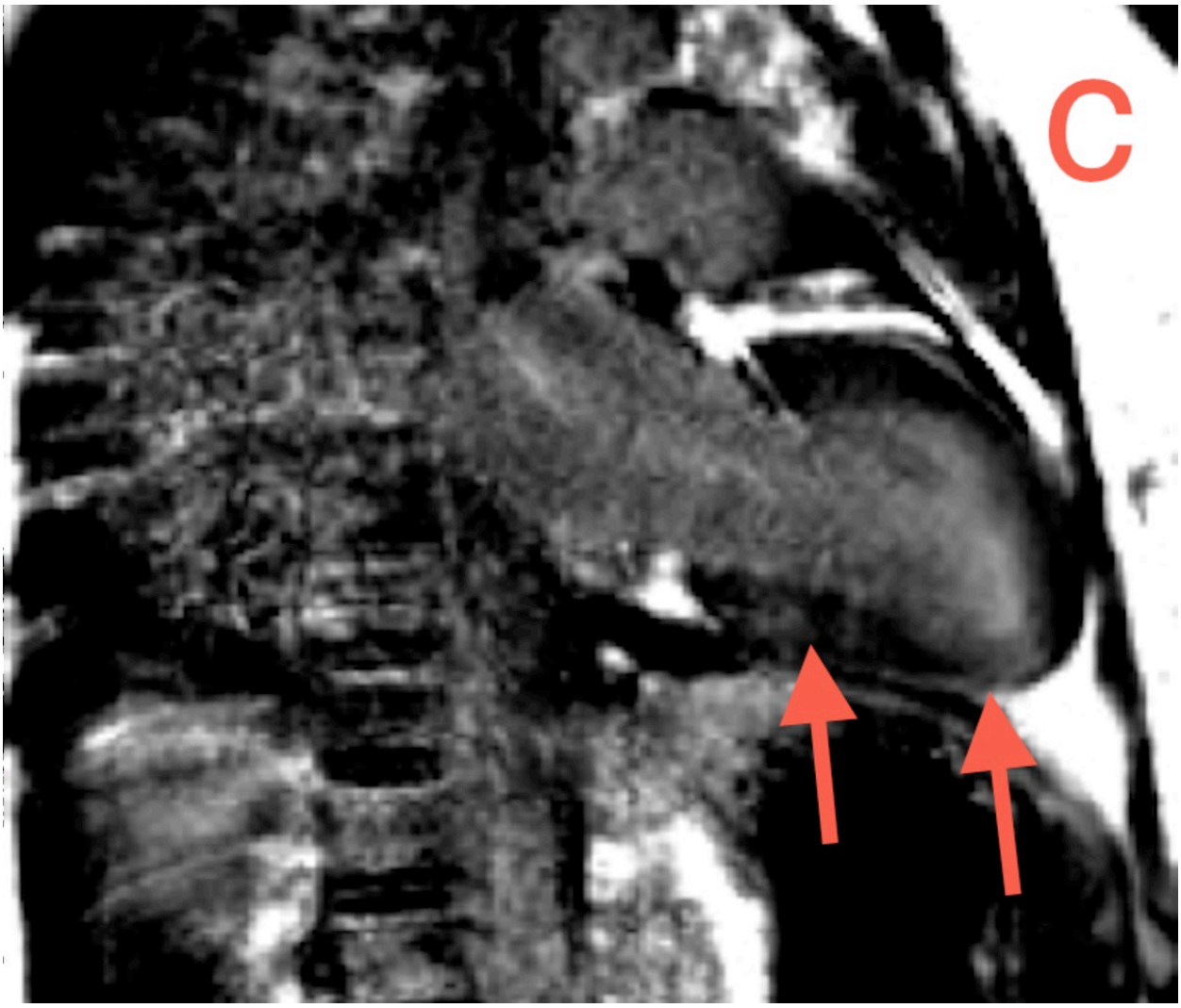

Fig. 7: Scleroderma. Short-axis (B) and long-axis (C) images demonstrate focal mid-wall LGE (arrows) in basal and mid-cavity inferoseptal LV wall and inferior RV wall.

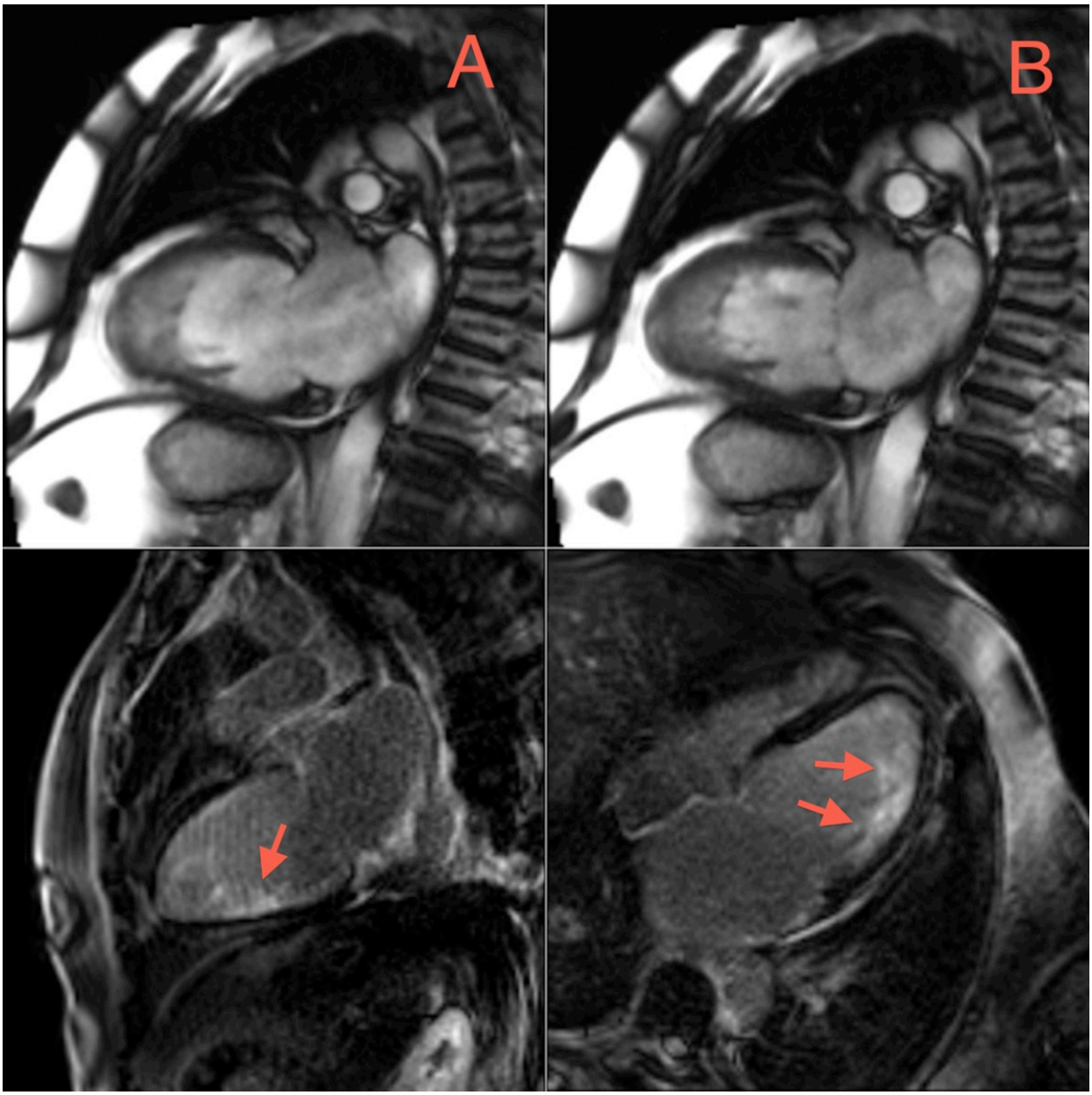

Fig. 8: Non-compaction cardiomyopathy. Top row: Two-chamber SSFP images in diastole (A) and systole (B) demonstrate extensive non-compacted left ventricular myocardium and apical trabeculation. Bottom row: LGE (arrows) involving the trabeculae from mid-ventricle to apex.

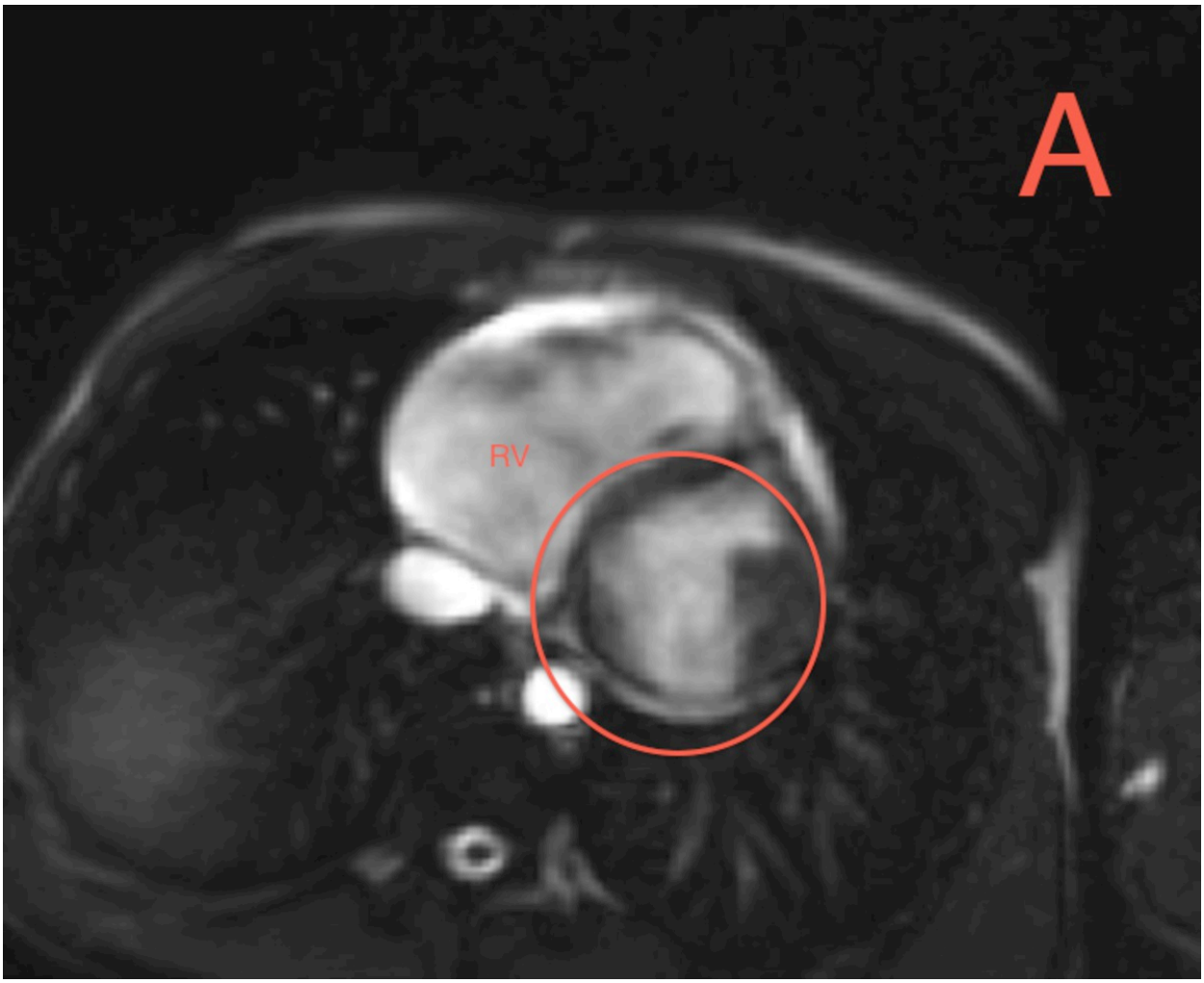

Fig. 9: ARVC. Short-axis SSFP image in diastole (A) demonstrates a dilated RV, which was hypo-functioning on the dynamic examination. A basal infero-lateral apical LV aneurysm (circle) is also demonstrated.

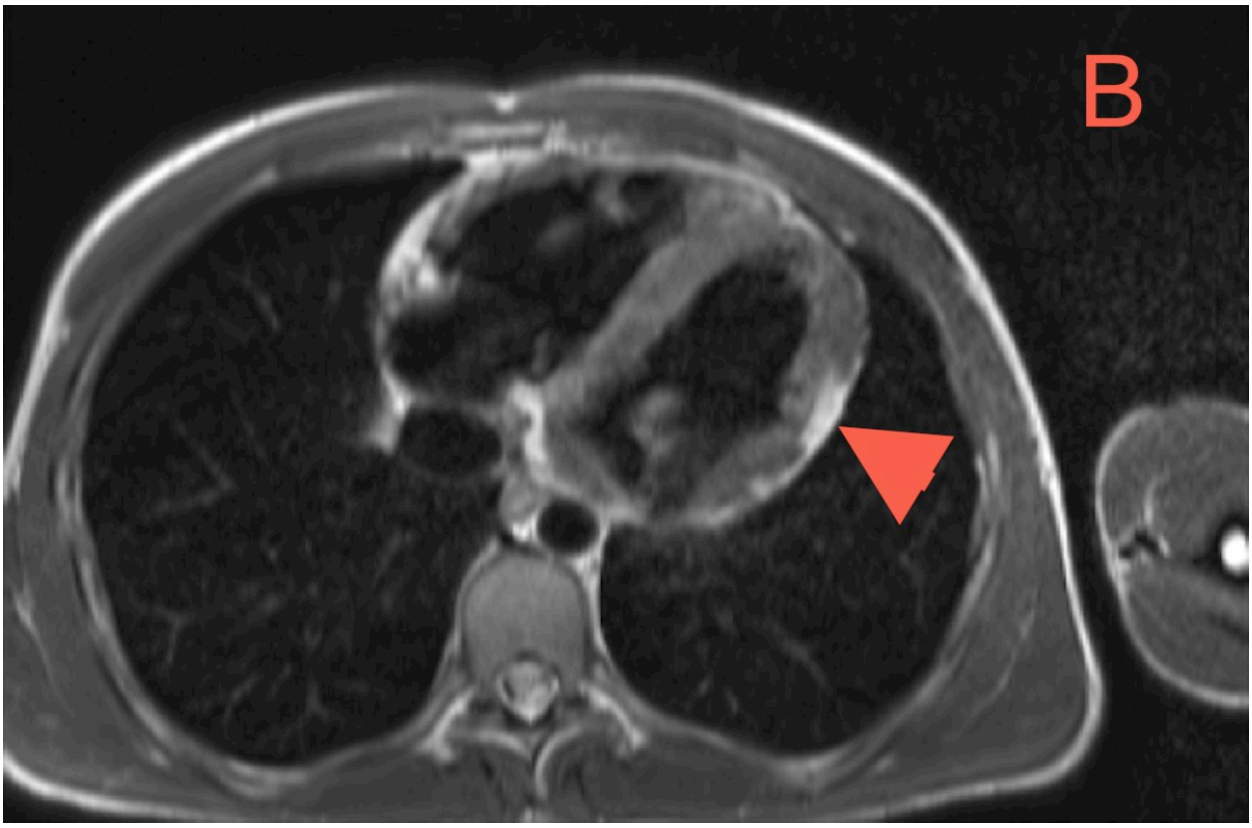

Fig. 10: ARVC. Axial T1-weighted black blood image (B) demonstrates fat deposition (arrowhead) in the LV free wall.

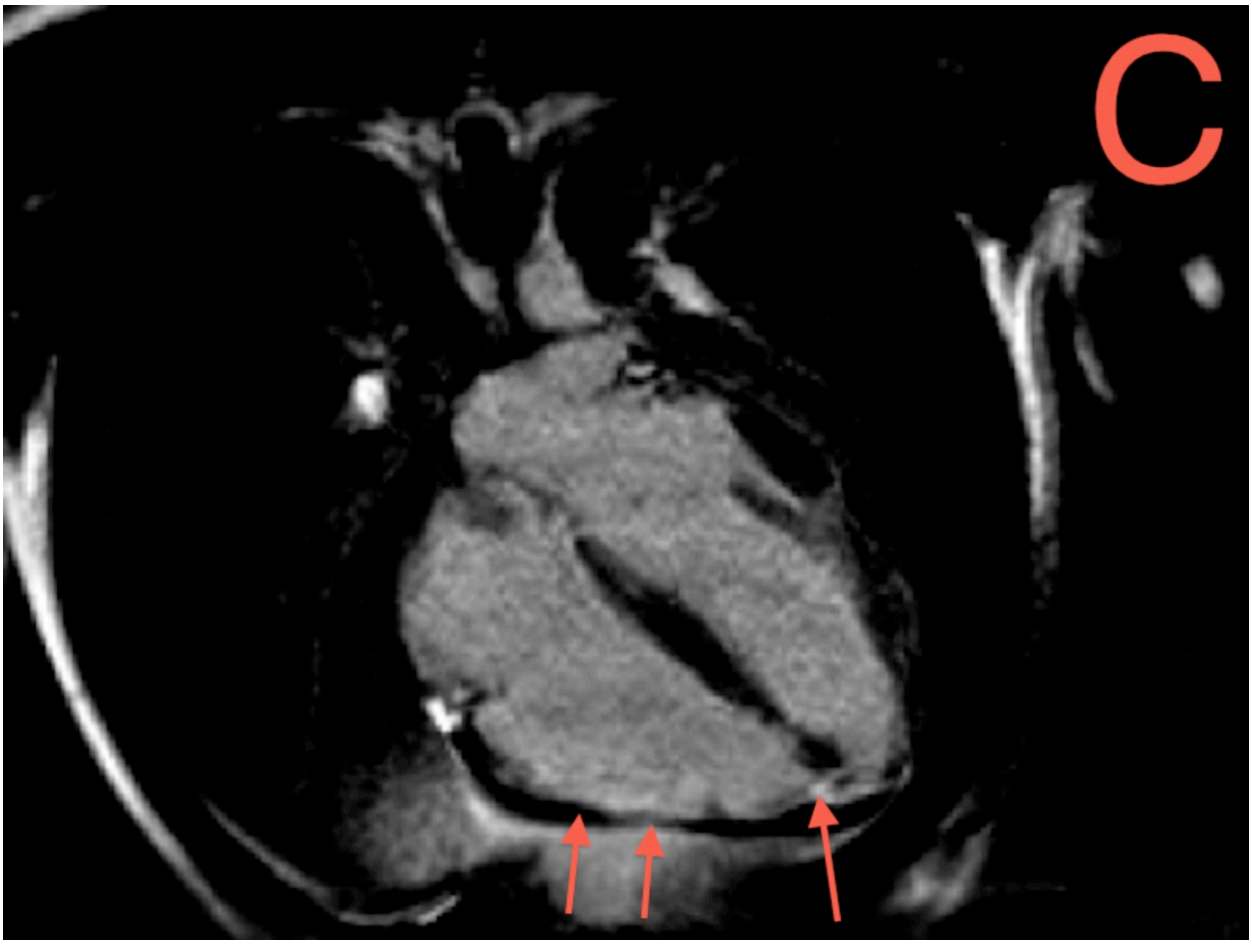

Fig. 11: ARVC. Four-chamber (C) and short-axis (D) images demonstrate apical RV and apical and inferolateral LV subendocardial LGE (arrows).

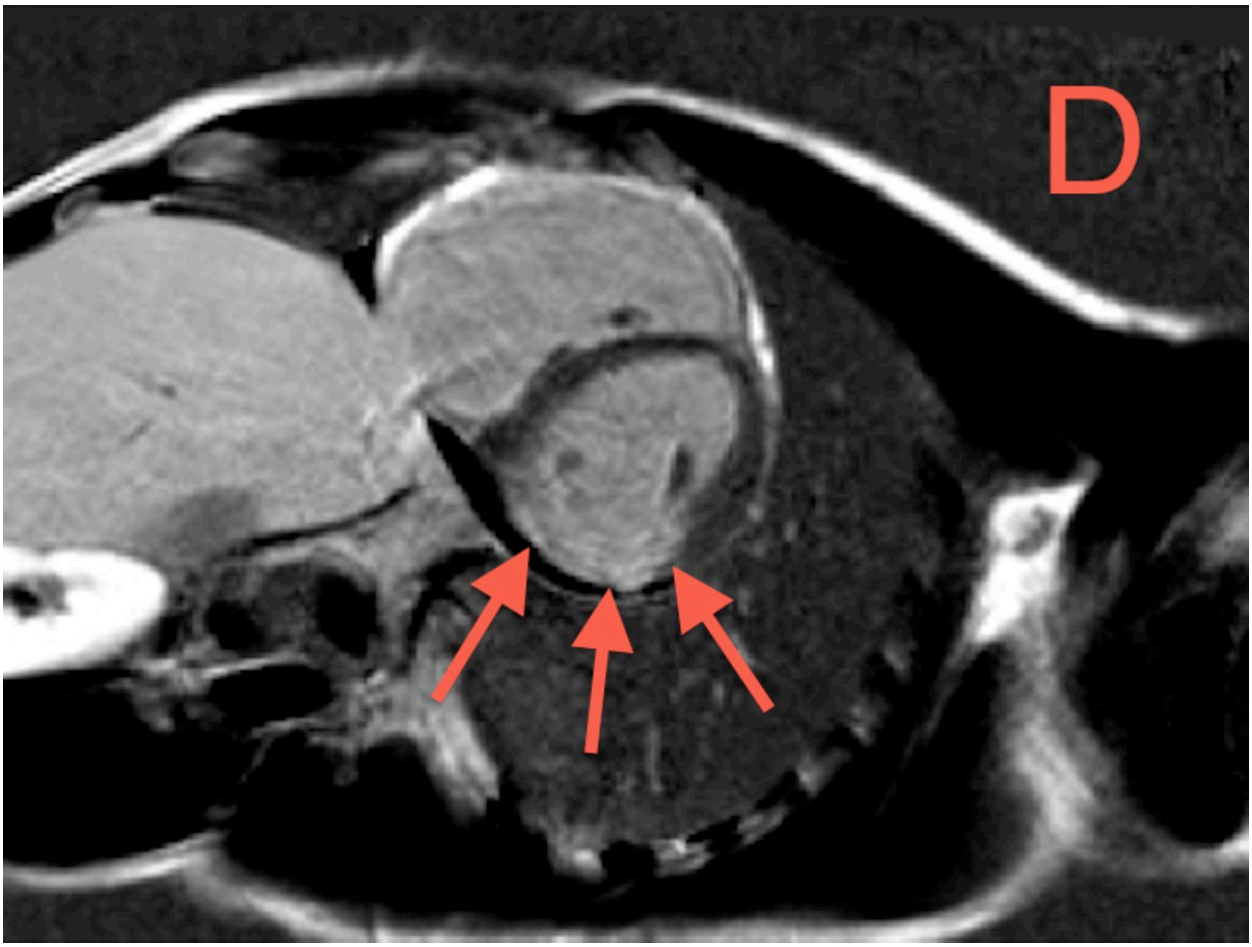

Fig. 12: ARVC. Four-chamber (C) and short-axis (D) images demonstrate apical RV and apical and inferolateral LV subendocardial LGE (arrows).

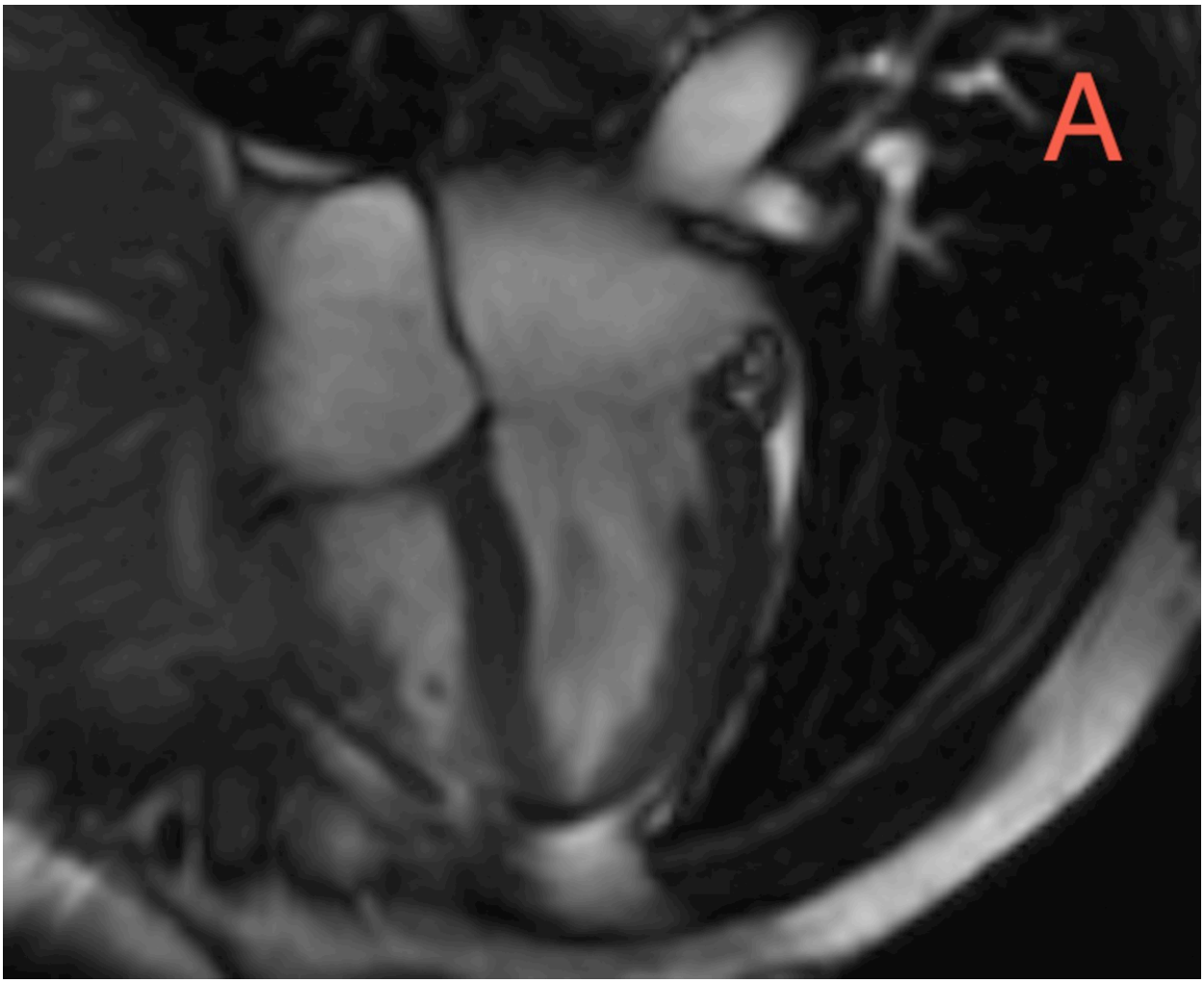

Fig. 13: Muscular Dystrophy. Four-chamber SSFP image in systole (A) demonstrates mild decrease in LV muscle mass (44 grams/metre).

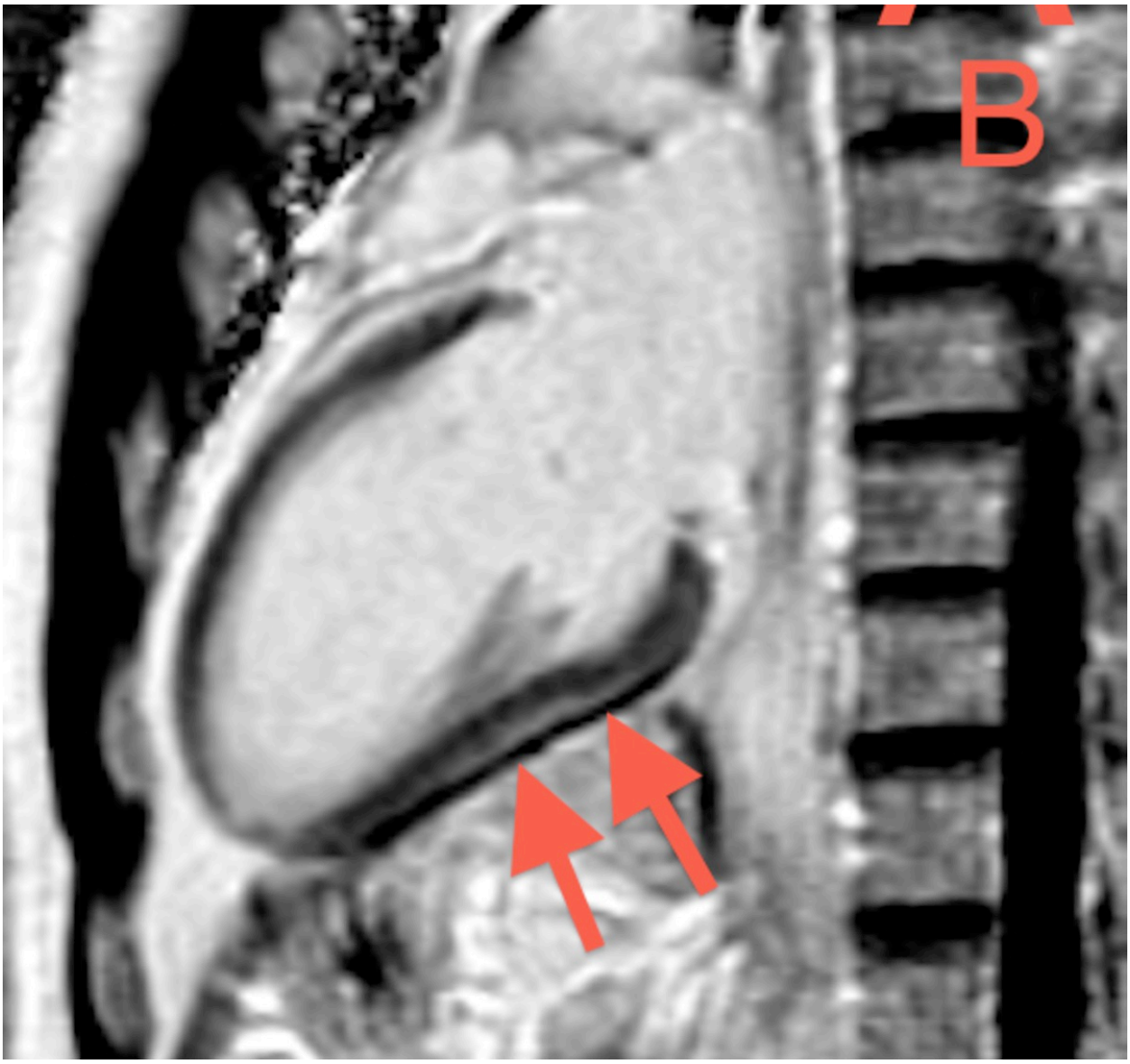

Fig. 14: Muscular Dystrophy. Long-axis (B) and short-axis (C) images demonstrate mid-wall LGE in the basal infero-lateral LV wall (arrows).

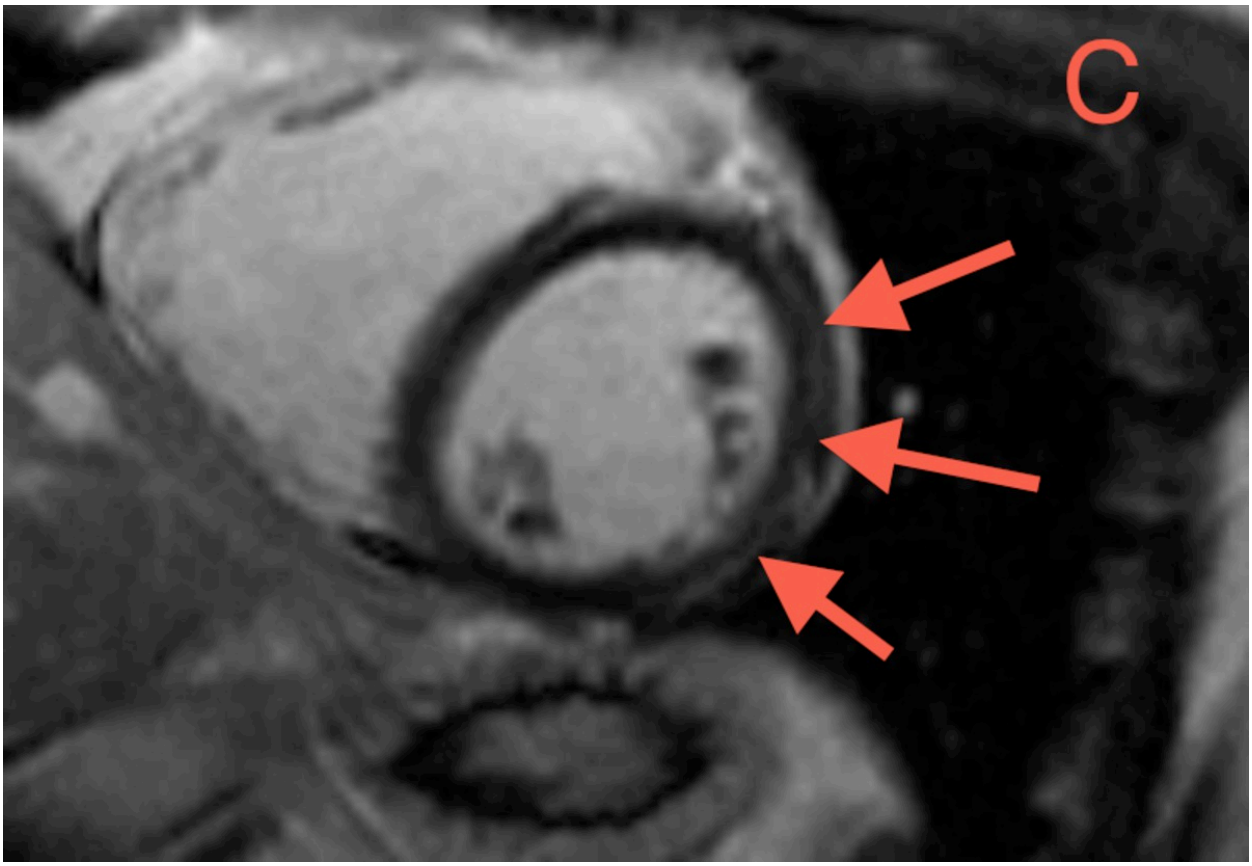

Fig. 15: Long-axis (B) and short-axis (C) images demonstrate mid-wall LGE in the basal infero-lateral LV wall (arrows).

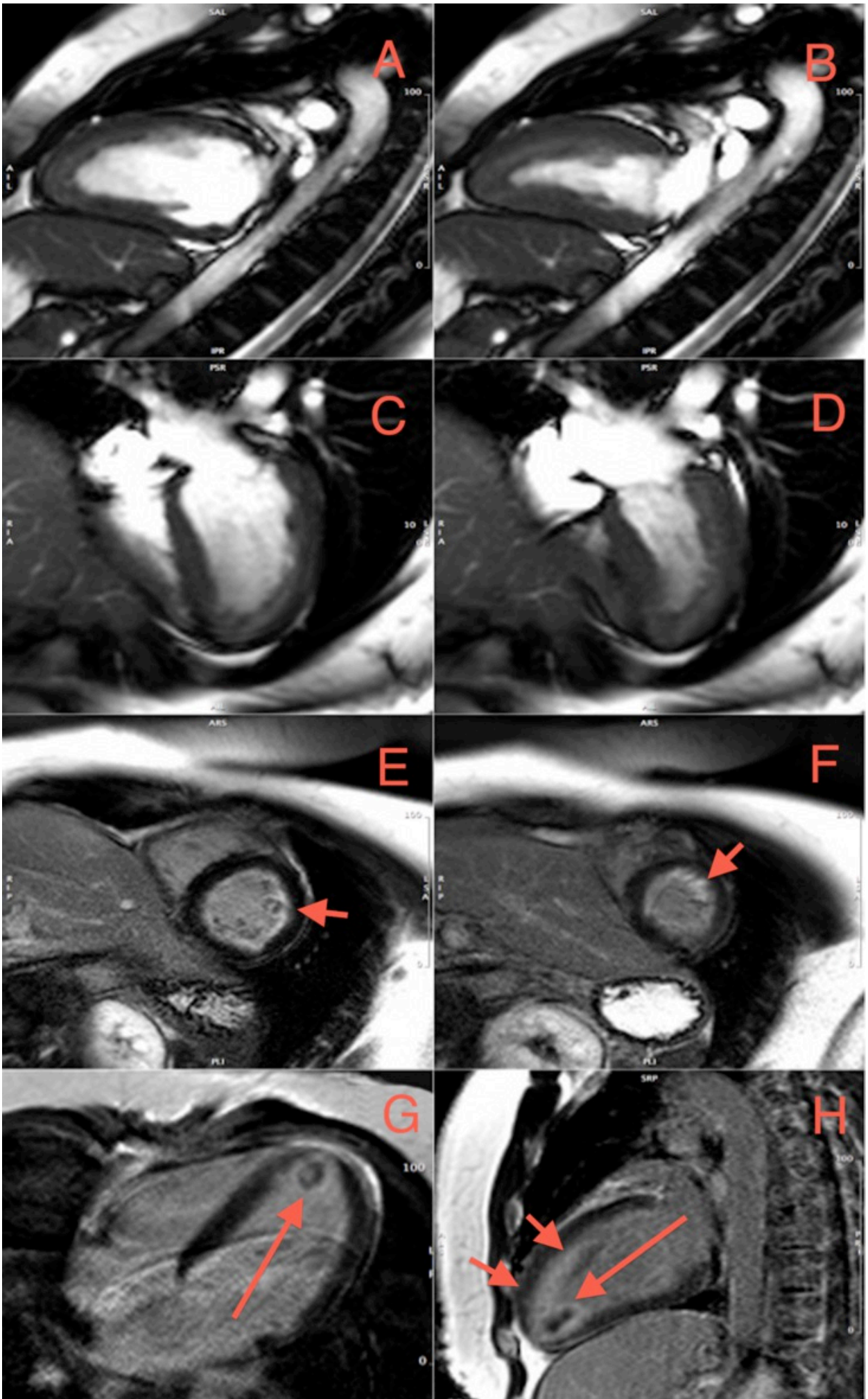
A
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Fig. 16: Endocardial Fibroelastosis. Two-chamber diastole (A) and systole (B) and four chamber diastole (C) and systole (D) SSFP images demonstrate abnormally increased signal throughout the thickened LV sub-endocardium as well as LV apical hypkinesis. Short axis basal (E) and mid (F) LV cavity and two-chamber (H) images demonstrate subendocardial LGE (short arrows) consistent with fibrosis. Focal mural thrombus (long arrow) demonstrated within the apex of the left ventricle on the four-chamber (G) and two-chamber (H) LGE images.

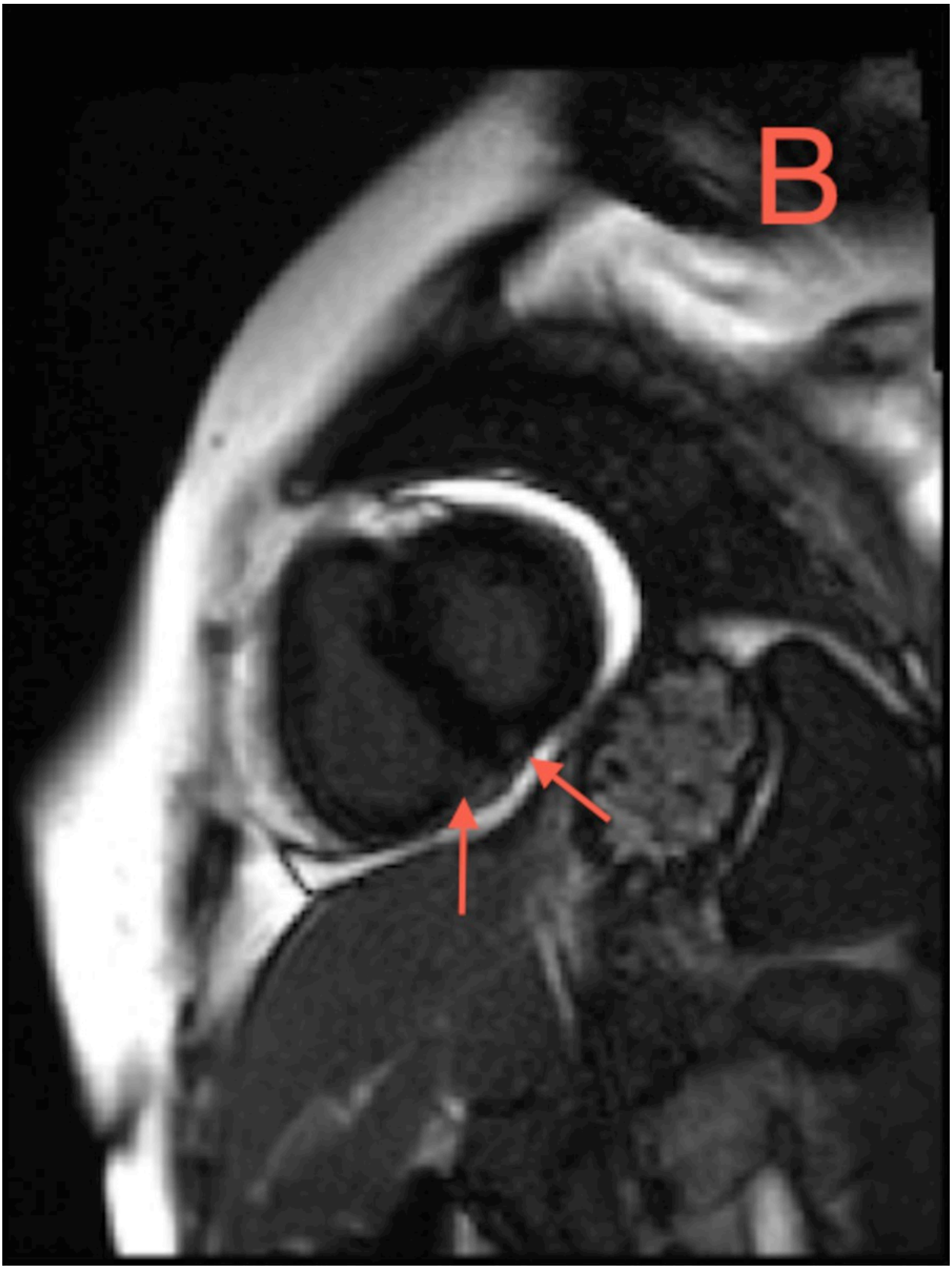
B

Fig. 6: Scleroderma. Short-axis (B) and long-axis (C) images demonstrate focal mid-wall LGE (arrows) in basal and mid-cavity inferoseptal LV wall and inferior RV wall. Straightening of the inter-ventricular septum is suggestive of elevated RV pressure.

Conclusion

Due to its superior spatial resolution as well as its capabilities for cardiac anatomical and physiological assessment, LGE-CMR imaging has become the preferred tool to determine the presence and extent of myocardial disease. While LGE CMR has become fairly well established for imaging of acute and chronic MI and relatively more common non-ischemic cardiomyopathies such as HCM and DCM, delayed myocardial enhancement can be seen in many other lesser known cardiac pathologic conditions. Hence, it behoves the cardiac imager to be aware of appearance and implications of LGE in these more uncommon cardiomyopathic processes.

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