

Cardiac Imaging

The Right Ventricle in Systemic Sclerosis: Insights from Cardiac Magnetic Resonance Imaging

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Right ventricular (RV) dysfunction in systemic sclerosis (SSc) may be secondary to pulmonary arterial hypertension (PAH), or it may occur independently of PAH and be related to myocardial fibrosis. The type and exact prevalence of RV dysfunction in these patients are not known, partly because of the lack of sensitive diagnostic tests and the complex RV geometry. Delayed-enhanced cardiac magnetic resonance imaging (MRI) detects functional and morphological abnormalities in the majority of patients with SSc.^{1,2}

A 49-year-old woman with a 20-year history of diffuse scleroderma along with Raynaud's phenomenon, esophageal and lung involvement was further evaluated for possible myocardial involvement with delayed-enhanced cardiac MRI (Figure 1). The clinical presentation of this patient included persistent palpitations and dyspnea on exertion. The left ventricular ejection fraction was 70%; the end-diastolic and end-systolic volumes were 99 cm³ and 30 cm³, respectively. The RV ejection fraction was 45%, with end-diastolic and end-systolic volumes of 98 cm³ and 60 cm³, respectively. Apart from the aneurysmal area, the inversion recovery sequence showed evidence of fibrosis

in the lower RV insertion point and the anteroseptal medial segment of the left ventricular myocardium (Figure C, arrowheads).

Heart pathophysiology in SSc is still unclear, and cardiac MRI can help because it offers functional and tissue characterization information using steady-state free precession and inversion recovery sequences, respectively. Cardiac MRI in SSc is a useful modality for assessing cardiac function and detecting myocardial fibrosis,³ which cannot be studied in detail using echocardiography. Delayed-enhanced cardiac MRI is uniquely able to provide a more "realistic" visualization of the right ventricle. Because of its higher contrast and spatial resolution, MRI can also detect isolated RV wall motion abnormalities more accurately than other conventional imaging methods. Further electrophysiology studies may be warranted in order to evaluate cardiac involvement in this particular group of patients.

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Figure 1. Four-chamber (A) and short-axis views (B) of cine MR images show thinning of the right ventricular (RV) free wall, with aneurysm formation (arrows). Mild flattening of the interventricular septum is also present. Linear enhancement (C, arrows) corresponding to the aneurysmal area, as well as subtle patchy enhancements (arrowheads) in the lower RV insertion point and the anteroseptal medial segment of the left ventricular myocardium on the delayed-enhancement MR image. The fibrosis in the RV insertion point was suggestive of pulmonary arterial hypertension. Right ventricular end-diastolic volume was markedly increased, while RV systolic function was impaired, with an RV ejection fraction of approximately 45%. Left ventricular systolic function was normal.

References

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