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Cardiac Involvement in Sporadic Inclusion-Body Myositis

Wolfgang Utz, MD; Saskia Schmidt, MD; Jeanette Schulz-Menger, MD;
Friedrich Luft, MD; Simone Spuler, MD

A 36-year-old man came to the emergency department with acute onset of exertional chest pain. He had had no recent infections, and no cardiovascular risk factors were present. However, the patient had used a wheelchair since his mid-20s because of sporadic inclusion-body myositis, as established by muscle biopsy. There were no other physical findings. Inflammatory markers and troponin T were normal, and the creatine kinase was elevated 2-fold with a significant muscle brain fraction. Chest roentgenogram was normal, but ECG showed a normal sinus rhythm at 62 bpm with deep Q waves, tall R waves in the right precordial leads, interventricular conduction delay, and T wave inversion in the left lateral leads (Figure 1). Echocardiography did not reveal any abnormalities. Cardiovascular magnetic resonance (CMR) examination was next performed.

Two-chamber (Movie I of the online-only Data Supplement) and 4-chamber (Movie II of the online-only Data Supplement) cine images were obtained by balanced, steady-state free precession cine sequences. Hypokinetic wall motion was detected in the midventricular to apical region of the lateral and anterior walls of a left ventricle (LV) of normal size. High spatial resolution turbo spin

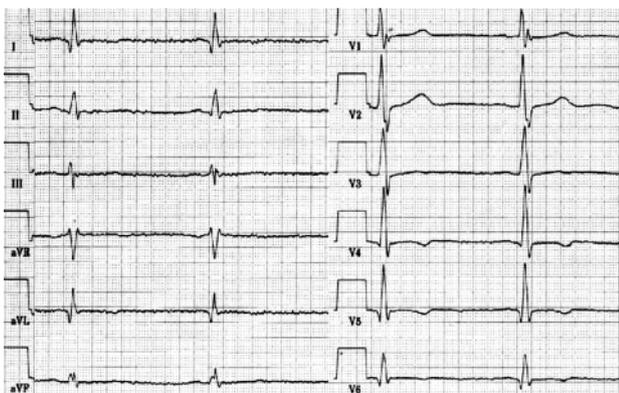


Figure 1. Twelve-lead electrocardiogram demonstrating deep Q waves, tall R waves in the right precordial leads, interventricular conduction delay, and T inversion in the left lateral leads.

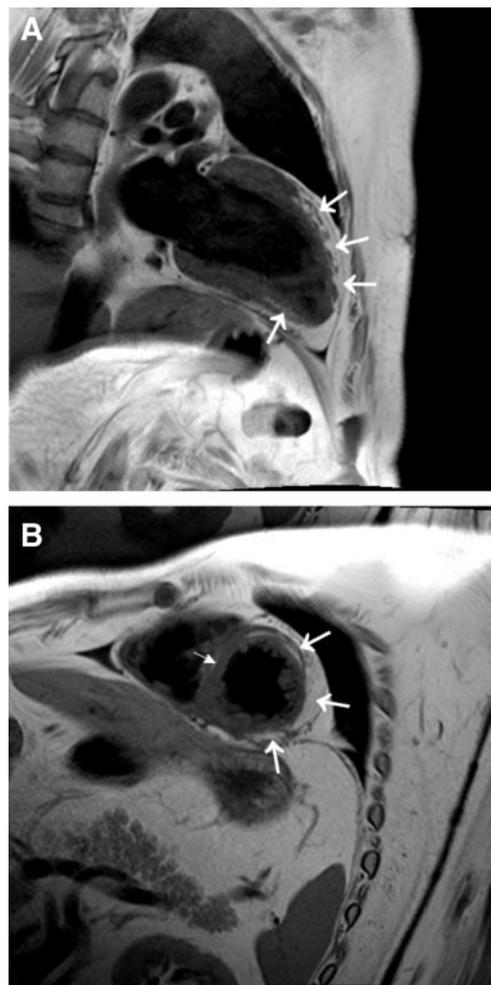


Figure 2. Turbo spin echo images showing extensive epicardial fat with fatty replacement of subepicardial layers of the myocardium (large arrows) and suspected intramural fat in the septum (small arrow) in the 2-chamber view (A) and a midventricular short axis (B).

echo imaging revealed massive fatty replacement of skeletal musculature of trunk which was expectable from upper leg muscle biopsy. Furthermore, extensive epicardial fat was found with regional fatty replacement of subepicardial

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The online-only Data Supplement is available with this article at <http://circ.ahajournals.org/cgi/content/full/121/5/706/DC1>.

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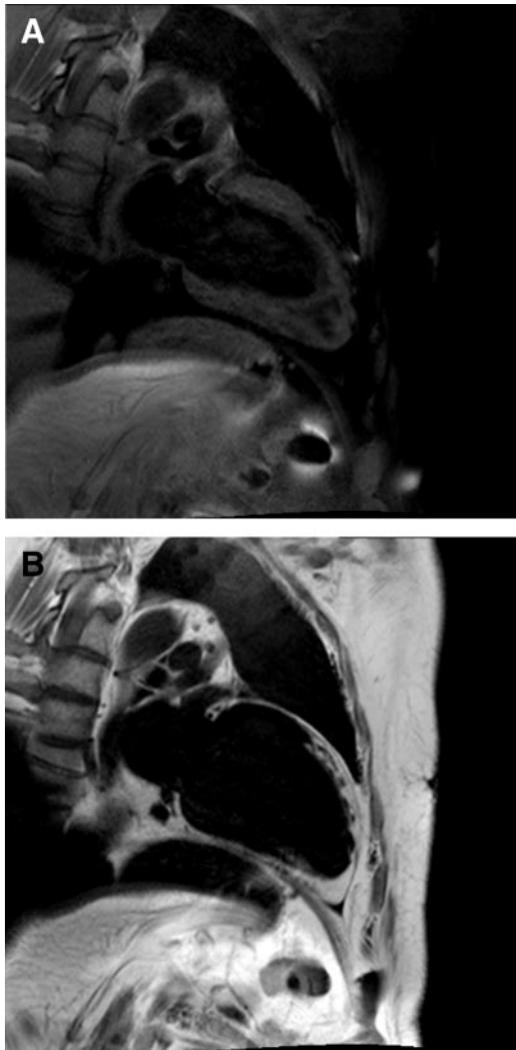


Figure 3. Fat- (A) and water- (B) saturated turbo spin echo images of the 2-chamber view, giving evidence of fatty replacement.

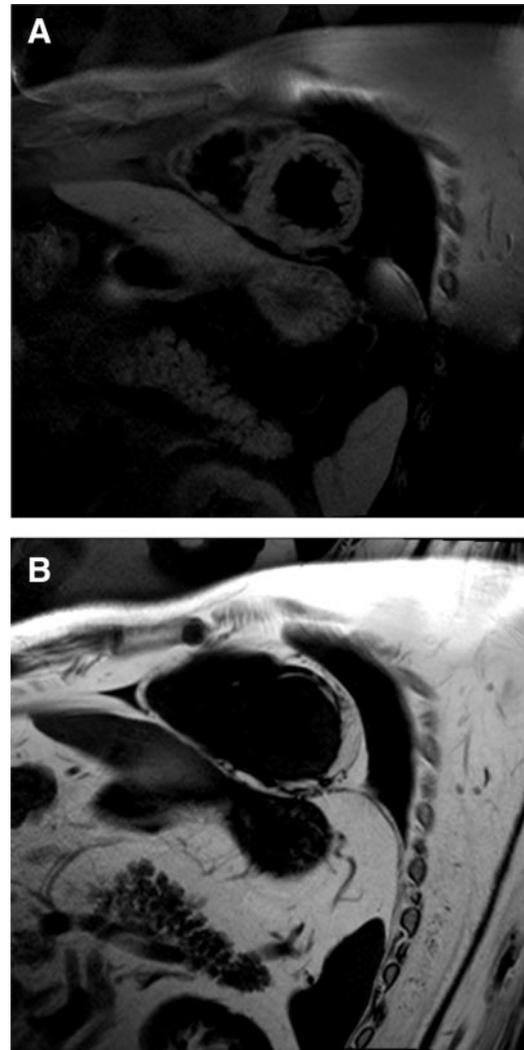


Figure 4. Fat- (A) and water- (B) saturated turbo spin echo images of the midventricular short axis, giving evidence of fatty replacement.

layers of the LV myocardium (Figure 2). This finding was confirmed by spectrally fat- or water-suppressed turbo spin echo images (Figures 3, 4). T2-weighted short tau inversion recovery imaging excluded myocardial edema and thereby concomitant inflammation (Figure 5). Because of limitations in specificity of fat-suppressed CMR imaging, we next tested for suspected subvoxel fatty infiltrations in the LV septum using dually gated ^1H spectroscopy. Quantification of spectral lines of myocardial lipids derived from a voxel, which was placed centrally in the septum, yielded a relative fat content of 8.9% (Figure 6). This amount is almost 10-fold higher than intramyocellular lipids that have been reported in metabolic disorders like obesity or diabetes mellitus.¹ Additional fibrotic replacement in myocardial areas other than those with fatty replacement was excluded by late Gadolinium enhancement imaging (Figure 7).

Cardiac involvement is a well-known finding in primary myopathies and may lead to life-limiting heart failure and conduction abnormalities.² Replacement of myocardium

by connective tissue and fat can frequently be found spreading from the lateral free wall of the LV. Sporadic inclusion-body myositis is a rare idiopathic inflammatory myopathy initially affecting the proximal muscles of the lower extremity and the distal limb muscles with characteristic findings in muscle biopsy. The findings include inflammation, myopathic changes, rimmed vacuoles, and cellular aggregates similar to those observed in amyloidosis. However, myocardial involvement has so far been unknown, to our knowledge.

Raised levels of cardiac troponin T have been reported in patients with sporadic inclusion-body myositis presenting without clinical signs or symptoms of a diseased heart; however, this observation has been attributed to reexpression of cardiac troponin T in the regenerating muscle fiber.³ In contrast to inflammatory reactions of the heart, such as in polymyositis,⁴ fatty replacement prevailed in this patient with a long-lasting history of inclusion-body myositis. Early identification of morphologic and

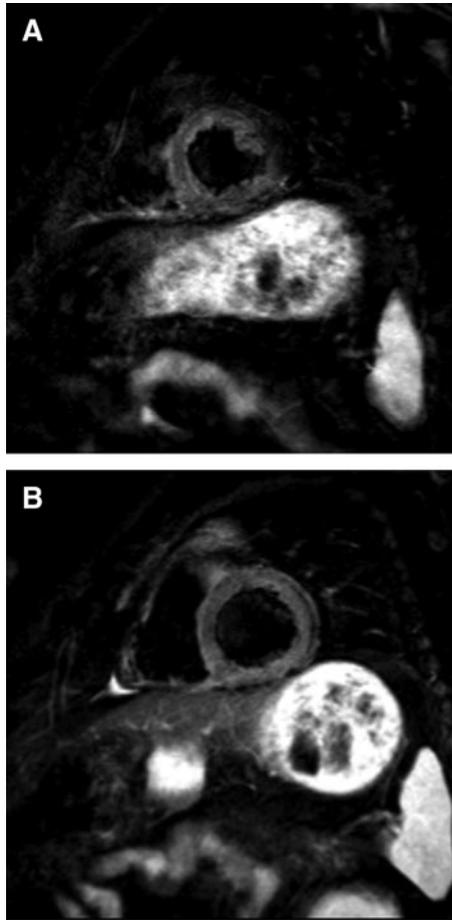


Figure 5. Myocardial edema indicating concomitant inflammation was not detectable in T2-weighted short-axis images: mid-ventricular (A) and basal (B).

functional alterations of the heart may be of prognostic value and guide initiation of therapeutic interventions. In this regard, CMR seems to be the imaging method of choice. Furthermore, CMR spectroscopy allows detection

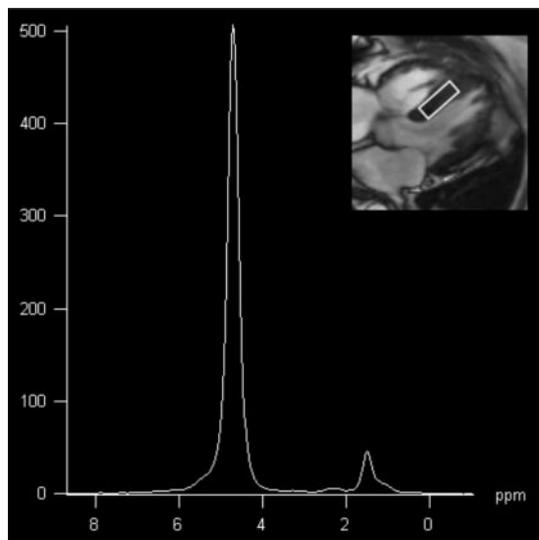


Figure 6. CMR proton spectrum of a 4-mL septal voxel showing a significantly increased amount of myocardial lipids (peak at 1.6 ppm) in relation to water (peak at 4.7 ppm). Inlay panel: position of voxel.

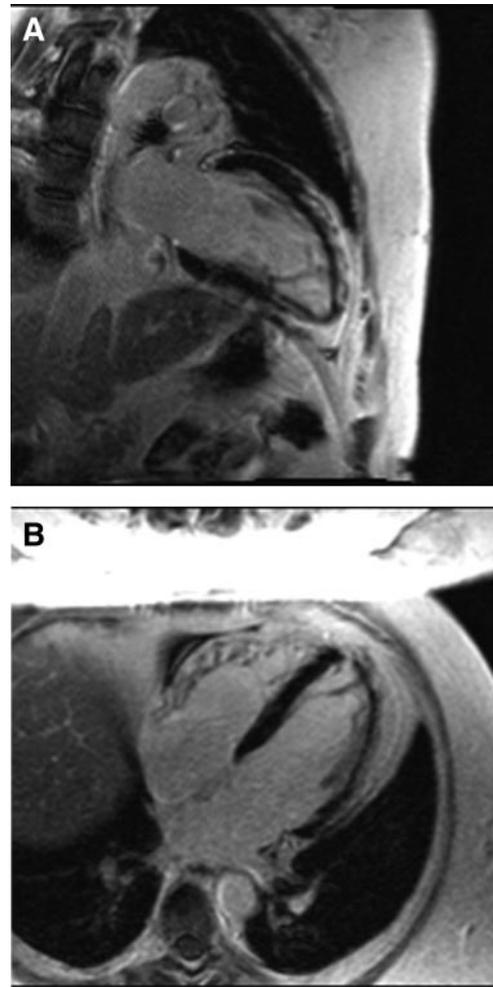


Figure 7. Late gadolinium enhancement imaging of 2-chamber view (A) and 4-chamber view (B). Comparison with the turbo spin echo images shows that myocardial fibrosis is absent in areas not affected with gross fatty replacement. Subvoxel fibrotic contributions cannot be excluded in the affected areas.

and quantification of subvoxel fatty infiltration in the septum.

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Disclosures

None.

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