Systemic sclerosis: Detection of myocardial fibrosis by magnetic resonance imaging.

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Introduction

Progressive systemic sclerosis (PSS) represents a complex disorder of obscure etiology which affects the skin as well as various organs. Depending on subtypes and different clinical stages of sclerosis, patients exhibit a variety of clinical symptoms including myositis, erosive arthropathy, esophageal dysmotility, scleroderma renal crisis, gastrointestinal and pulmonary fibrosis, pulmonary hypertension, and cardiac involvement. Cardiac manifestations of PSS may result in pericardial disease, vavular diseases, conduction system abnormalities and arrhythmias; however myocardial fibrosis is the hallmark with mayor impact on treatment and patients' prognosis. Early myocardial involvement is characterized by interstitial inflammatory alterations leading to patchy areas of fibrosis. Our current study aimed to assess the potential of contrast enhanced MRI for early detection of cardiac involvement in patients with systemic sclerosis.

Material and methods

The open study was performed in accordance with the regulations of the local ethics committee and included 36 patients (31 female, 5 male, mean age of 53 ± 14 years) suffering from severe systemic sclerosis. Patients with known coronary artery disease (CAD) or a history of myocardial infarction were excluded from the study. All examinations were performed on a 1.5T MR scanner equipped with high performance gradients (Magnetom Avanto, Siemens medical solutions, Erlangen, Germany). The MRI protocol included Steady State Free Precession (SSFP, TR 3ms, TE 1.5ms, FA 60°) cine sequences in long and short axis orientation for the assessment of the myocardial function. Based on these measurements left ventricular volumes and ejection fraction were calculated and all myocardial segments were characterized as normo-hypo-, a- or dyskinetic. For assessment of myocardial edema fat-suppressed T2-weighted turbo spin echo (TR 2 heart beats, TE 49ms, FA 180°) sequences were performed in standard orientations. Additionally, an inversion recovery fast low angle shot sequence (IR-turboFLASH: TR 8.0ms, TE 4.0ms, TI 180-240ms, FA 20°) was acquired in short and long axis views 10 min after injection of a 0.2 mmol/kg bodyweight of Gd-DTPA (Schering AG, Berlin, Germany). To distinguish areas of late enhancement (LE) from artifacts only areas of LE that could be detected in two orthogonal views were considered as pathologic findings.

Results

Diagnostic image quality could be achieved in all but one patient due to severe claustrophobia. Cardiac pathologies were detected in 51 % of our patients. Evaluation of the left ventricular function revealed a mean normalized enddiastolic volume of 64 ± 18ml, a mean normalized endsystolic volume of 27 ± 16 ml and a mean left ventricular ejection fraction of 60 ± 10 %. A reduced ejection fraction (defined as an ejection fraction <55%) was observed in 23% (8 of 35). 7 of 35 patients (20%) showed a pericardial effusion (maximal thickness of the effusion 8 mm). Mitral valve prolaps was observed in 9, low grade aortic valve regurgitation in 3 and tricuspidal valve rerurgitation in 1 patient. No patient showed a myocardial edema on T2-w images. The analysis of the contrast enhanced images showed late enhancement (maximum extent 15% of the LV mass) in 5 of 36 patients (15 %). The area of late enhancement was detected in the mid-myocardial layer, showing either an ill-defined, diffuse (Fig. 1a) or sharply margined, spotted (Fig. 1b) distribution.

Discussion

The concept of myocardial late enhancement has recently been established for the assessment of myocardial viability. In patients with a history of myocardial infarction the accumulation of gadolinium-based contrast reflects irreversible damage and scar formation. However, whereas LE is highly sensitive in characterizing myocardial scarring, it is not specific for ischemic damage since contrast agents generally accumulate in tissues with an increased interstitial space or areas with cell membrane damage. Thus, LE also occurs in myocardial areas of inflammation, edema, as well as fibrosis (Hunold, AJR 2005), and contrast enhanced MRI must be considered as imaging modality of first choice for the detection of small areas of myocardial fibrosis in-vivo. Our data show that late enhancement as a marker of myocardial fibrosis can be detected in 15% of PSS patients with no clinical evidence of myocardial involvement. Therefore, contrast enhanced MRI seems to be well suited for screening of myocardial fibrosis, monitoring the progression and possibly evaluating therapeutic effects. However, long-term follow-up studies are mandatory to investigate the impact of late enhancement on patients' prognosis.

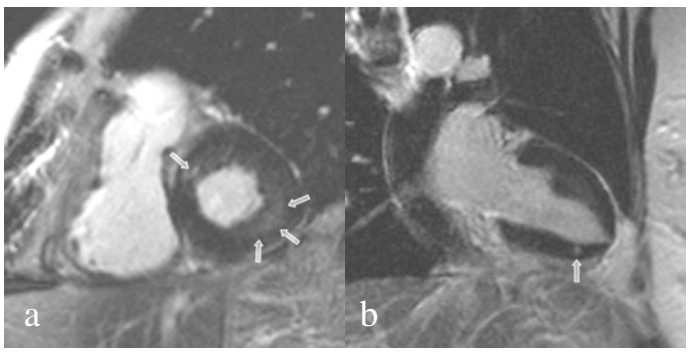

Fig 1 a,b: Diffuse (a) and focal (b) delayed-enhancement in patients suffering from severe PSS.