Extensive atrial fibrosis in a patient with systemic lupus erythematosus and atrial fibrillation

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Introduction
The study of left atrial (LA) fibrosis with delayed enhancement–magnetic resonance imaging (DE-MRI) is an established technique that provides a noninvasive means of detecting the electroanatomic substrate of atrial fibrillation (AF).1 We present a case of a middle-aged woman with systemic lupus erythematosus (SLE) and persistent AF who had extensive atrial fibrosis as detected by DE-MRI before the ablation procedure.

Case Report
In October 2010, a 46-year-old woman was admitted to our cardiology department for an ablation procedure of symptomatic persistent AF. She had SLE since 1990, which started as articular and then became renal involvement (diffuse proliferative glomerulonephritis, class IV) and was treated with steroid therapy, cyclosporine, and cyclophosphamide bolus. All therapies were discontinued in 2006 because of a complete remission of the disease. No cardiovascular risk factor was present, in particular no family history of cardiomyopathy. She was symptomatic for impaired functional status, and she was refractory to pharmacological treatment with antiarrhythmic drugs of class 1C. The echocardiogram revealed normal dimensions of the left ventricle with overall normal systolic function. The LA was moderately enlarged, and the right atrium was only slightly enlarged. Mild mitral regurgitation was found.

Before the ablation procedure, she underwent MRI with atrial fibrosis detection (DE-MRI), as reported previously.1 DE-MRI was performed with a 1.5-T scanner (MAGNETOM Avanto, Siemens Medical Systems, Erlangen, Germany) 15 minutes after a bolus injection of 0.1 mmol/kg of gadobenate dimeglumine (MultiHance, Bracco Spa, Milan, Italy) using a 3-dimensional (3D) inversion recovery prepared, respiration navigated, ECG-gated, gradient echo pulse sequence with fat saturation (Gradient Echo 3-dimensional Inversion Recover Turbo Flash). The axial DE-MRI images revealed massive delayed enhancement in different regions of the LA (see the Online Video), including anterior wall (Figure 1A), posterior wall, and septum (Figure 1B). The percentage of fibrosis relative to the volume of the LA wall2 was 40%. Furthermore, a significant homogeneous thickening distribution of the atrial wall was present (measured thickness 6.8 mm). The maximum LA volume and LA emptying fraction3 were 38 ml/mq and 47%, respectively. The DE imaging performed using a 2D inversion recovery sequence after contrast injection demonstrated a small area of fibrosis in the inferolateral mid-wall ventricular myocardium.

During the electrophysiological study (CARTO3, Biosense Webster, Inc., Diamond Bar, CA), the endocardial electroanatomic map of the LA confirmed large areas of low bipolar voltage (<0.05 mV) in the anterior wall and septum (Figure 1C) and an area of electrically abnormal tissue (0.05 mV < bipolar voltage < 1 mV) in the posterior wall and roof (Figure 1D). The percentage of fibrosis relative to the LA surface estimated by using the CARTO3 system was 52%.

The ablation procedure was performed without complications; nevertheless, during follow-up, the patient had recurrences of atrial tachycardia.

SLE is an inflammatory autoimmune disease that can virtually affect all organ systems.5 Cardiovascular involvement occurs frequently (58%–77%).5 The most common cardiac manifestations include pericarditis (~25% of all patients with SLE),6,7 myocarditis,8 coronary artery disease,8 valvular lesion such as valvular thickening and regurgitation, and Libman-Sacks endocarditis (prevalence 11%–74%).10 In the context of cardiac arrhythmias, sinus tachycardia (50% of patients), AF, and atrial ectopic beats are the most frequent.11 All types of atrioventricular blocks, intraventricular conduction disturbances, and sick sinus syndrome are also observed.12,13

KEYWORDS Atrial fibrillation; Atrial fibrosis; Systemic lupus erythematosus; Magnetic resonance imaging
ABBREVIATIONS 3D = 3-dimensional; AF = atrial fibrillation; DE-MRI = delayed enhancement–magnetic resonance imaging; LA = left atrium/atrial; SLE = systemic lupus erythematosus

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Very little is known about the pathogenesis of cardiac manifestations in SLE. Histological features of SLE cardiomyopathy include initially inflammatory cell infiltrates and, as the disease advances, myocardial necrosis, fibrotic replacement, and scarring. Cases of intramural and subepicardial ventricular fibrosis detected by DE-MRI have been described in the literature. In addition, in other inflammatory autoimmune and vasculitic diseases, DE-MRI most frequently detected a midwall ventricular fibrosis, probably owing to a combination of subclinical inflammatory and immunological processes, while a mosaic patchy distribution of myocardial fibrosis is a pathognomonic feature of systemic sclerosis and recently all ventricular patterns (subendocardial, subepicardial, intramural, and transmural) are observed in these diseases. Nevertheless, to our knowledge, atrial fibrosis has not been previously reported in SLE or other autoimmune diseases. However, the study of atrial fibrosis using DE-MRI is recent and not widely implemented in the clinical setting of patients with AF.

Herein, we also highlight the following peculiar features of this case:

1. The occasional feedback of a significant thickening of the atrial wall in a patient with SLE and AF, a finding of DE-MRI that is frequently detected in storage disorders, such as amyloidosis. Delayed enhancement–magnetic resonance imaging helps to make a differential diagnosis.
2. The reduced atrial functionality could be interpreted as a predictor of cardiovascular events.

These findings are matched to a given reduced atrial functionality that could be interpreted as a predictor of cardiovascular events.
In conclusion, in a patient with SLE with no other cardiovascular risk factors, the presence of an uncommon thickening of the LA wall associated with an extensive atrial fibrosis could, in our opinion, justify the refractoriness of AF. The important question that needs to be answered is whether SLE leads to atrial fibrosis, and other studies are necessary to confirm a correlation between them.

Appendix

Supplementary data

Supplementary material cited in this article is available online at http://dx.doi.org/10.1016/j.hrcr.2015.02.014.

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