Diagnosis and follow-up of Wegener’s granulomatosis by cardiac magnetic resonance

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A 46-year-old woman was referred to our institution for chest pain and dyspnea. Wegener’s granulomatosis (WG) had recently been diagnosed based on typical chest and sinus CT findings.

Clinical examination was normal. ECG disclosed an acute episode of atrial fibrillation. Troponin level was moderately increased at 3.25 μg/L. There was a marked inflammatory syndrome (Erythrocyte sedimentation rate 116 mm; C-reactive protein 161 mg/mL). Trans-thoracic echocardiography was normal.

First, cardiac MRI was performed to rule out cardiac involvement, after resolution of the acute atrial fibrillation event. Cine sequences revealed a normal LVEF (66%), normal left ventricular volumes, and segmental wall motion. A small pericardial effusion was present. Fat-suppressed T2-weighted sequence showed an increased signal of the lower third of the septum consistent with myocardial oedema (Panel A, arrow). A four-chamber plane inversion recovery sequence demonstrated a linear, transmural, delayed enhancement in the septum (Panel B), confirmed by an orthogonal short-axis phase-sensitive inversion recovery sequence (Panel C, arrow). Pulmonary nodules related to WG were also noted (Panels A and B, arrowheads).

Wegener’s granulomatosis-related myopericarditis was diagnosed, and corticosteroid therapy was started in association with an immunosuppressive treatment (cyclophosphamide).

Three months later, the symptoms and inflammatory syndrome had clearly improved. A second MRI was then performed. Septal oedema had become hardly visible (Panel D) whereas late enhancement was unchanged (Panels E and F). This was interpreted as a decreased local inflammation and scar formation. A significant regression of the pulmonary nodules was also demonstrated by follow-up cardiac MR (Panels D and E).

Cardiac involvement in WG is considered to be rare, but it could be underestimated due to the lack of sensitivity of conventional techniques (ECG, TTE, and scintigraphy). The most frequent cardiac manifestations in patients with WG are pericarditis, coronary arteritis, and focal myocarditis while valvulitis, endocarditis, conduction abnormalities, and atrial arrhythmias are less common.