CLINICAL IMAGE

Cardiac involvement as a fatal complication of granulomatosis with polyangiitis

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Granulomatosis with polyangiitis (GPA) is a necrotizing vasculitis with granuloma formation and is associated with antineutrophil cytoplasmatic antibodies (cANCA) with specificity against proteinase 3 (PR3).[1] As opposed to the upper respiratory tract (92%), lungs (85%), and kidneys (77%), cardiac involvement is a rare manifestation of GPA (3%–44%).[1,2] Pericarditis is the most common cardiac disease (50%) in the GPA population with cardiac involvement, followed by myocarditis (25%), valvular involvement (21%), conduction block (17%), and myocardial infarction (11%). Cardiac lesions in GPA are likely underdiagnosed.[3]

A 41-year-old man with no medical history was admitted to a district hospital due to sudden hearing loss, arthralgia with joint stiffness, feet paresthesia, and petechiae on the skin of the lower limbs. Urine test revealed proteinuria (2 g/24 h) and erythrocyturia. Peripheral blood eosinophil count was within the reference range (0.27 G/l; reference range, 0.02–0.5 G/l). Mucosal lesions in the sphenoid sinus together with ground-glass lesions and nodules in the lungs were noted on computed tomography. After the initiation of treatment with 3 pulses of methylprednisolone (1500 mg), the patient was transferred to our department.

On admission, his general condition was good, cANCA titer was 1:320, and the concentration of PR3 antibodies was 55.08 RU/ml. Histological examination revealed pauci-immune crescentic glomerulonephritis. The Birmingham Vasculitis Activity Score for GPA was 9 points. As a remission induction therapy, 5 monthly pulses of cyclophosphamide (3064 mg/m^2 in total) and prednisone (0.8 mg/kg) were administered with good clinical response.

On admission to our outpatient clinic on schedule for the sixth pulse of cyclophosphamide, the patient unexpectedly complained about a significant deterioration in exercise tolerance, class II according to the New York Hear Association classification, elevated heart rate at rest (110–120 bpm), as well as edema in both legs. Blood tests showed increased concentrations of creatinine (1.7 mg/dl; reference range, 0.7–1.3 mg/dl), brain natriuretic peptide (3353 pg/ml; reference range, <73 pg/ml), and high-sensitivity troponin I (0.055 ng/ml; reference range, <0.03 ng/ml). The level of creatine kinase–myocardial band was not elevated. Electrocardiography documented sinus tachycardia.

Echocardiography showed left ventricular ejection fraction of 40%, severe aortic and moderate mitral regurgitation, and hypokinesia of the inferior wall of the heart with diffuse segmental dysfunction of cardiac muscle contractility. The patient was immediately admitted to the Department of Cardiology and underwent transesophageal echocardiography which revealed no signs of infective endocarditis; blood culture test was negative as well. Coronary angiography did not show signs of coronary artery disease. Tissue characterization with magnetic resonance imaging revealed subtle diffuse active myocarditis most likely associated with GPA (FIGURE 1A–1C).

Due to high disease activity (12 points in the Birmingham Vasculitis Activity Score), 7 therapeutic plasma exchanges were performed, followed by 3 weekly infusions of rituximab (375 mg/m^2), which caused partial improvement of the patient's general condition.

Despite the treatment, the patient suddenly died at home in anticipation of a fourth pulse of rituximab and heart valves replacement.

We present the case to emphasize the need for cardiac monitoring (electrocardiography and echocardiography) in the entire GPA population. However, the most accurate noninvasive technique to diagnose cardiac lesions in GPA patients

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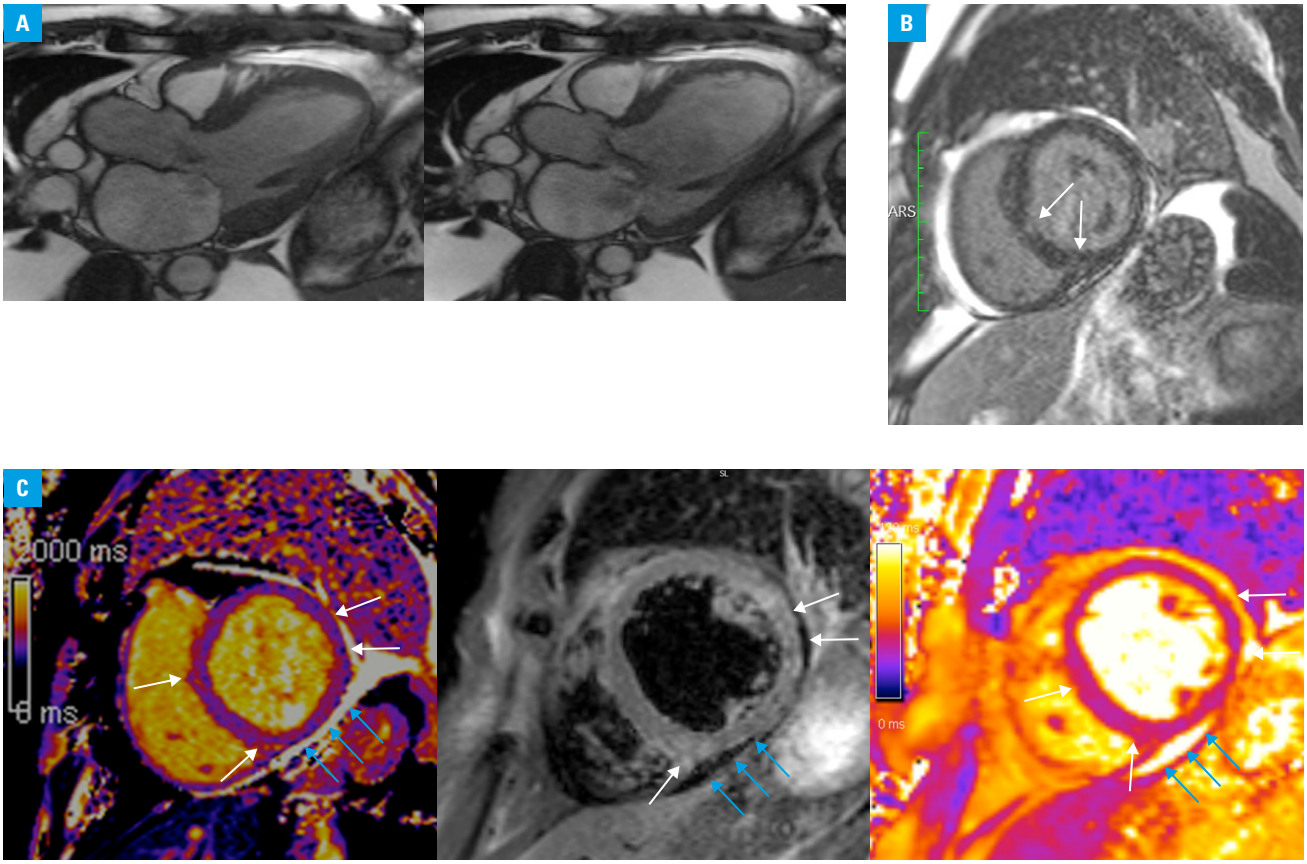

FIGURE 1 Cardiac magnetic resonance imaging with a Siemens Aera 1.5T device (Erlangen, Germany); **A** – long axis systolic (left) and diastolic (right) still frames from routine cine balanced steady-state free precession sequence showing slight left atrial and marked left ventricular enlargement (left ventricular end-diastolic volume index of 169 ml/m^2), with severe aortic insufficiency (registration volume in a phase-contrast sequence: 65 ml/cycle) and moderately reduced left ventricular ejection fraction (45%); **B** – late gadolinium enhancement; phase-sensitive inversion recovery sequence: minimal subendocardial/intramural late gadolinium enhancement in mid-infero-septal and inferior segments (arrows); **C** – parametric mapping showing inhomogeneous myocardium with local areas of increased native T1 relaxation time (white arrows on the left panel; local native T1 increases up to 1110 ms; reference range, 951–1035 ms; basal short axis slice, images acquired with a modified Look-Locker inversion recovery sequence) and locally increased signal in the T2 weighted short tau inversion recovery sequence pointing to patchy edema (white arrows on the middle panel; mid-ventricular slice), matching the areas of locally elevated T2 time predominantly in the septal and infero-lateral segments (white arrows on the right panel; local increases up to 54 ms; reference range, 39–49 ms; images acquired with a balanced steady-state free precession sequence). This is suggestive of inhomogeneous diffuse acute injury due to myocardial involvement. Small pericardial effusion is apparent in all 3 images (blue arrows).

is magnetic resonance imaging.[4] The probable cause of the patient's death was arrhythmia or complete atrioventricular heart block, which was previously reported by Colin et al.[5] Pacemaker implantation might have prevented fatal complication of GPA in our case. Moreover, immediate aortic valve replacement should also have been taken into consideration. Guidelines for the detection and management of cardiac involvement in GPA are lacking, and further research in this area is required.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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