CLINICAL IMAGE

A rare case of complex cardiac involvement in granulomatosis with polyangiitis

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We present a case of a 68-year-old woman with a 12-year history of limited form of granulomatosis with polyangiitis (GPA). During the course of the disease, she suffered from recurrent granulomatous inflammation in the subglottic area, bloody nasal discharge, chronic sinusitis, sensorineural hearing loss in the right ear, scleritis, and nasal bridge collapse. Antineutrophil cytoplasmic antibodies (ANCAs) were assessed several times in the course of the disease but were always within normal limits. The patient underwent 6 upper respiratory tract surgeries due to subglottic stenosis. Unfortunately, histopathological examinations of the resected inflammatory tissue did not reveal any histological evidence of GPA. The diagnosis of GPA was made on the basis of typical clinical manifestations and exclusion of other diseases. The patient was treated with immunosuppressive therapy for several years. After obtaining clinical and laboratory remission (Birmingham Vasculitis Activity Score = 0), immunosuppressive therapy was discontinued.

Five months later, complete heart block with macrophage activation syndrome occurred, requiring pacemaker implantation. The patient also complained of dyspnea on moderate exertion. An echocardiographic examination demonstrated an echo-dense mass in the left atrium, attached to the interatrial septum and posterior wall of the ascending aorta, as well as thickening of the mitral valve anterior leaflet and anteromedial papillary muscle with concomitant moderate mitral regurgitation (FIGURE 1A). The clinical examination was unremarkable, with no symptoms of heart failure or other cardiac abnormalities. A computed tomography scan revealed involvement of the cardiac structures similar to the echocardiographic images with good delineation of affected tissues. Laboratory tests revealed signs of inflammation with an elevated erythrocyte sedimentation rate (54 mm/h), increased C-reactive protein levels (34 mg/dl), and anemia (hemoglobin levels, 9.8 mg/dl).

Taking into account the previous diagnosis of GPA and strong suspicion of cardiac involvement with granulomatous tissue, we decided to initiate immunosuppressive treatment with cyclophosphamide and methylprednisolone pulses. A partial reduction of the cardiac infiltration extent and normalization of inflammation parameters were achieved after 7 months of therapy (FIGURE 1B). The patient is now on a maintenance dose of prednisone and methotrexate.

To date, the reported prevalence of cardiac involvement in GPA remains highly variable, ranging from 6% to 86%.[1] In a prospective cohort study of 41 patients with GPA, electrocardiogram and echocardiography demonstrated cardiac abnormalities in 46% of the patients. On additional cardiac magnetic resonance imaging, the prevalence of cardiac involvement increased to 61%.[1] With respect to ANCA detection, cardiac involvement was equally frequent in ANCA-negative as in ANCA-positive patients. The majority of patients do not show cardiac symptoms, so heart involvement is likely to be underdiagnosed. It most often includes pericarditis (50%), coronary arteritis (50%), and myocarditis (25%).[2] Approximately 15% of patients with cardiac manifestations of GPA have conduction system abnormalities, including complete heart block as a very rare manifestation. Granulomatous inflammation in the course of GPA may also occur as a tumor-like mass located in different organs and uncommonly including left- and right-sided cardiac chambers.[2-4] Valve involvement is a very rare complication of GPA, and especially mitral valve involvement has been reported only a few times.[5,6] The increase in the prevalence of heart involvement in GPA may be attributed to the routine use of cardiac imaging

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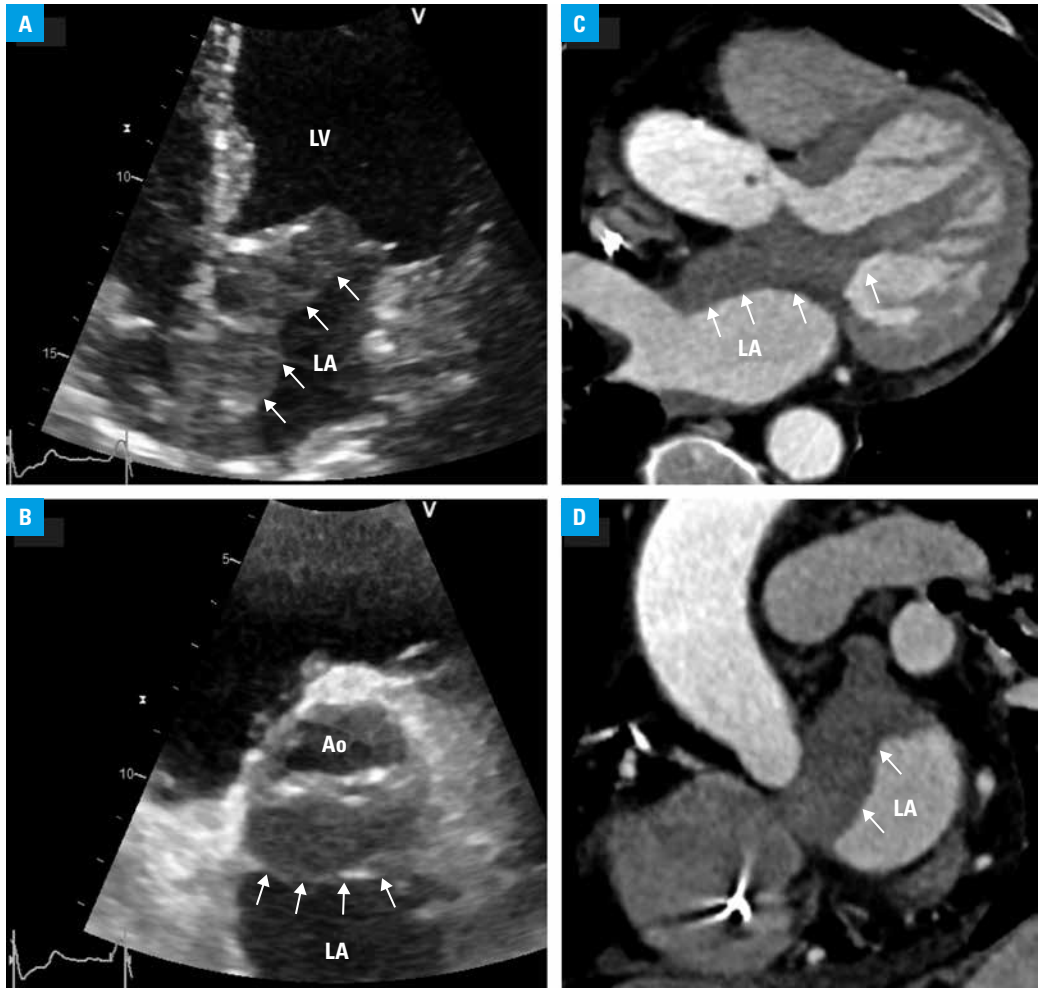

FIGURE 1 Echocardiographic and cardiac computed tomography (CT) images on admission in a patient with granulomatosis with polyangiitis; **A** – echocardiogram, 4-chamber view: note the echo-dense mass on the anterior mitral leaflet and thickening of the interatrial septum (arrows); **B** – echocardiogram, short-axis view: note the mass in the left atrium, adjacent to the bulb of the aorta (arrows); **C** – CT multiplanar reconstruction reformat (MPR) image demonstrating a mass in the left atrium, thickening of the anterior mitral leaflet and anterolateral papillary muscle; **D**– CT MPR image demonstrating a mass in the left atrium, adjacent to the aorta
Abbreviations: LA – left atrium, Ao – aorta, LV – left ventricle

modalities such as echocardiography, computed tomography, and magnetic resonance.

We present a case of limited GPA with exceptionally rare coexistence of cardiac complications: complete heart block, mitral valve involvement, and tumor-like mass located in the left atrium. Our patient was successfully treated with pacemaker implantation followed by immunosuppressive therapy, and inflammatory infiltration was significantly reduced. Echocardiography and computed tomography allowed an accurate and complementary assessment of cardiac involvement and monitoring of disease regression.

REFERENCES

1 Hazebroek MR, Kemna MJ, Schalla S, et al. Prevalence and prognostic relevance of cardiac involvement in ANCA-associated vasculitis: eosinophilic granulomatosis with polyangiitis and granulomatosis with polyangiitis. Int J Cardiol. 2015; 199: 170-179.

2 Harris JG, Salvay DM, Klein-Gitelman MS. Asymptomatic intracardiac mass in a 14-year-old girl with granulomatosis with polyangiitis: case report. Pediatr Rheumatol. 2012; 10: 9.

3 Singh R, Rosen S. Tumor of the heart in a young woman; a rare manifestation of Wegener granulomatosis. Hum Pathol. 2012; 43: 289-292.

4 Mortazavi M, Nasri H. Granulomatosis with polyangiitis (Wegener's) presenting as the right ventricular masses: A case report and review of the literature. J Nephropathol. 2012; 1: 49-56.

5 Goodfield NER, Bhandari S, Plant WD, et al. Cardiac involvement in Wegener's granulomatosis. Br Heart J. 1995; 73: 110-115.

6 Attaran S, Desmont M, Ratnasingham J, et al. Mitral valve involvement in Wegener's granulomatosis. Ann Thorac Surg. 2010; 90: 996-997.

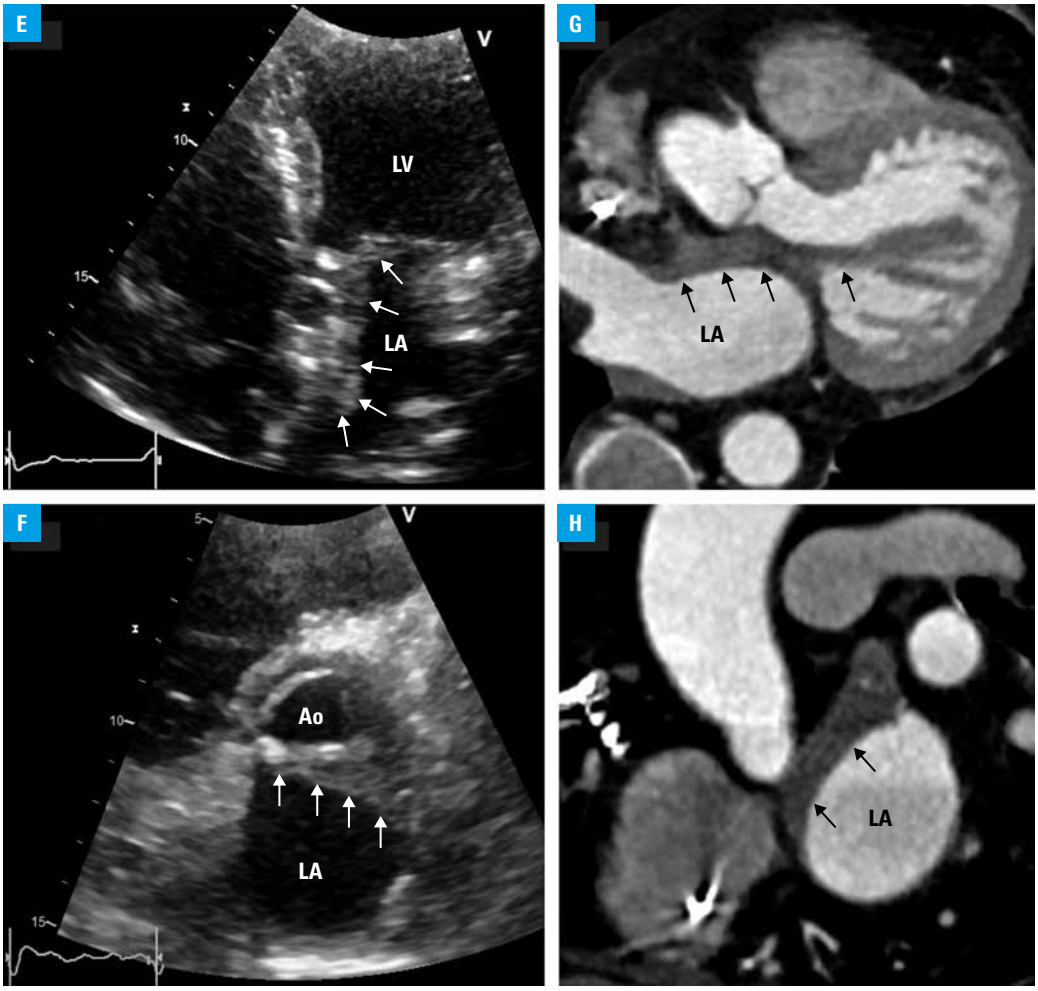

FIGURE 1 **E–H** – echocardiographic and CT MPR images after 7 months of immunosuppressive therapy, showing a significant regression of cardiac infiltration at the end of the treatment (**E**, **F**, **G**, and **H** correspond to **A**, **B**, **C**, and **D**, respectively).
Abbreviations: see **FIGURE 1A–1D**