CLINICAL IMAGE

Acute myocarditis with heart failure in the course of eosinophilic granulomatosis with polyangiitis in a patient on maintenance hemodialysis

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Eosinophilic granulomatosis with polyangiitis (EGPA, formerly known as Churg–Strauss syndrome) is a rare autoimmune vasculitis with an incidence of less than 3 cases per million. Heart involvement is observed in 27% to 47% of the patients and remains the main predictor of poor long-term prognosis and premature mortality, although it is not included in the diagnostic criteria of EGPA developed by the American College of Rheumatology (ACR).[1]

We present the results of cardiac magnetic resonance imaging (MRI) in a patient on chronic hemodialysis (end-stage renal disease [ESRD] secondary to reflux nephropathy followed by nephrectomy) who developed progressive heart failure due to EGPA.

A 34-year old man was referred to a renal clinic with advanced chronic kidney disease (estimated glomerular filtration rate, 26 ml/min/1.73 m^2). He was diagnosed with vesicoureteral reflux soon after birth. Because of complications, he underwent right-side nephrectomy in early childhood and received conservative treatment for the next 3 years until ESRD developed. Owing to nonspecific symptoms from the upper respiratory tract and occasional eosinophilia (7%–24%), he was consulted by a pulmonologist and diagnosed with asthma. He received fluticasone/salmeterol and levocetirizine. At the age of 37, soon after hemodialysis was started, he was admitted to the Department of Nephrology with fever and acute dyspnea. Echocardiography revealed large thrombi in both ventricles and infiltrative lesions in the endomyocardium. N-terminal-pro-B-type natriuretic peptide levels increased to 29 451 pg/ml (upper reference limit, 125 pg/ml) and eosinophilia reached 64.5%. Cardiac MRI (considered an excellent tool to diagnose heart involvement in EGPA)[2] revealed lesions corresponding with endomyocardial inflammatory infiltration and thrombosis in both ventricles (FIGURE 1AB). Perinuclear antineutrophil cytoplasmic antibody (pANCA) titer was positive (1:80). Based on the clinical picture and test results, we suspected EGPA. The patient was treated with methylprednisolone (3 intravenous pulses of 0.5 g followed by 32 mg/d orally) and with subcutaneous enoxaparin followed by warfarin. His general status improved within a few days, and pANCA titer decreased to 1:10 after 3 months. Methylprednisolone was tapered to a maintenance dose of 4 mg/d. Echocardiography was performed regularly during follow-up to monitor regression of cardiac lesions. Follow-up cardiac MRI was repeated at 3 months and revealed complete resolution of infiltrative and thrombotic lesions (FIGURE 1CD). Currently, the patient is in an excellent general condition and is awaiting a kidney transplant.

Patients with ESRD caused by small vessel vasculitis are carefully monitored because life-threatening nonrenal flares (including cardiac manifestations) are common (especially in granulomatosis with polyangiitis).[3,4] However, in our patient, the early (prodromal) symptoms of the disease during a predialysis period were overlooked (probably because another cause of ESRD was established, and no other ACR criteria were fulfilled at the onset of the disease). The patient presented with a fulminant, life-threatening flare while already on hemodialysis.

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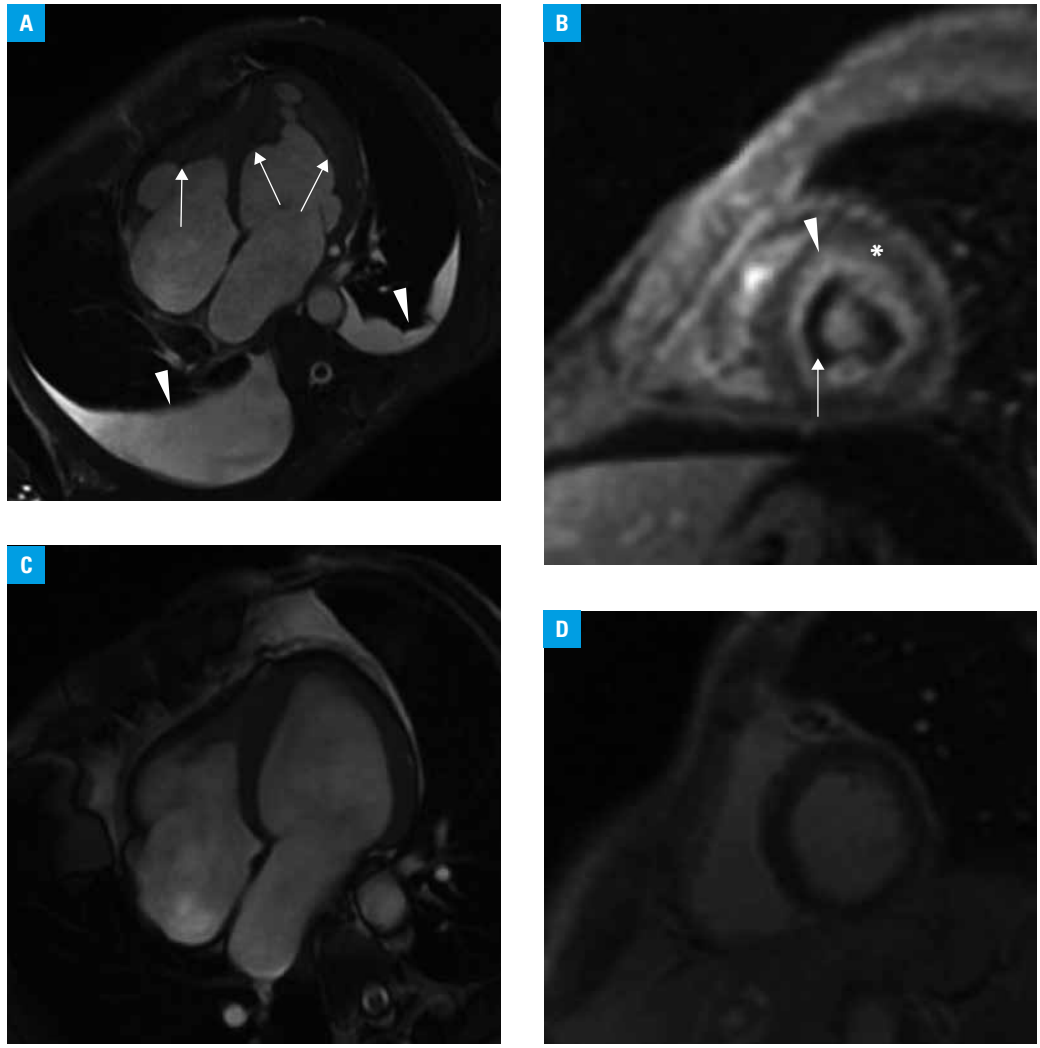

FIGURE 1 **A** – organized thrombi filling apical and midventricular segments of the left and right ventricle lumen, tightly attached to the ventricular walls (arrows); a large amount of fluid in the pleural cavities (arrowheads); 4-chamber view, FIESTA-SSFP (GE Signa HDx equipment); **B** – short-axis view of midventricular segments; typical "triple layer" appearance of the left ventricular wall including (center to outwards): hypointense thrombus (arrow), hyperintense inflammatory lesions within the endocardium (arrowhead), and normal signal of myo- and epicardium (asterisk); late gadolinium enhancement imaging; **C** – complete resolution of intraventricular thrombi, pleural and pericardiac effusion; 4-chamber view, FIESTA-SSFP; **D** – complete resolution of endocarditis and intraventricular thrombi observed previously; late gadolinium enhancement imaging

Atypical clinical manifestations of EGPA (ie, not fulfilling at least 4 of 6 ACR criteria) are quite common; for example, crescentic glomerulonephritis with eosinophilic infiltrations (typical for EGPA) was described in a patient without previous or concomitant symptoms of asthma.[5] ANCA seropositivity is observed in up to 50% of the patients with EGPA; in these cases, more than 70% of the antibodies are pANCA. Hemodialysis itself does not seem to interfere with disease activity and the risk of flare. A new onset of antiphospholipid antibodies has been described in patients with and without systemic lupus who have been dialyzed with bioincompatible (ie, cuprophane) membranes. This seems to be the only known type of an autoimmune reaction triggered by hemodialysis itself.

REFERENCES

1 Mahr A, Moosig F, Neumann T, et al. Eosinophilic granulomatosis with polyangiitis (Churg-Strauss): evolutions in classification, etiopathogenesis, assessment and management. Curr Opin Rheumatol. 2014; 26: 16-23.

2 Marmursztejn J, Guillevin L, Trebossen R, et al. Churg-Strauss syndrome cardiac involvement evaluated by cardiac magnetic resonance imaging and positron-emission tomography: a prospective study on 20 patients. Rheumatology. 2013; 52: 642-650.

3 Romeu M, Couchoud C, Delarozière JC, et al. Survival of patients with ANCA-associated vasculitis on chronic dialysis: data from the French REIN registry from 2002 to 2011. QJM. 2014; 107: 545-555.

4 Merino JL, Galeano C, Espejo B, et al. A retrospective study on outcome of microscopic polyangiitis in chronic renal replacement therapy. Nephrol Dial Transplant. 2011; 26: 1360-1366.

5 Próchnicka A, Żelichowski G, Wojtecka A, et al. Atypical clinical presentation of Churg-Strauss syndrome with rapidly progressive glomerulonephritis: case report. Pol Arch Med Wewn. 2009; 119: 830-833.