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Case Report

Successfully treated eosinophilic granulomatosis with polyangiitis relapse presenting as myocarditis and followed by multimodality imaging

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ABSTRACT

A 60-year-old man with eosinophilic granulomatosis with polyangiitis (EGPA), which was diagnosed 12 years earlier and managed with prednisolone, was admitted to our hospital because of dyspnea and paresthesias in both hands. Laboratory test revealed peripheral eosinophilia along with elevated troponin T and brain natriuretic peptide (BNP). The patient's clinical picture was consistent with myocarditis and relapse of EGPA. Endomyocardial biopsy showed marked infiltration of eosinophils in myocardium, which confirmed relapse of EGPA with myocarditis. Thallium-201 and iodine-123-beta-methyl iodophenyl pentadecanoic acid dual single-photon emission computed tomography (TL-BMIPP SPECT), as well as cardiac magnetic resonance imaging (CMR), also confirmed cardiac involvement. The patient was treated with methylprednisolone and improved dramatically. CMR and TL-BMIPP SPECT performed after discharge showed improvement of abnormal lesions, while anomalies detected by these modalities remained. This is a case of EGPA relapse presenting as myocarditis despite treatment with prednisolone.

<**Learning objective:** Cardiac involvement and relapse are frequent in eosinophilic granulomatosis with polyangiitis patients. Patients should be screened and monitored carefully for cardiac involvement during follow-up by multimodality imaging.>

Introduction

Eosinophilic granulomatosis with polyangiitis (EGPA) is a systemic, small-to-medium vessel necrotizing vasculitis characterized by asthma, eosinophilia, and eosinophil infiltration of various organs. Cardiac involvement is not rare and is associated with poor prognosis. In addition, relapse after remission remains frequent, despite taking immunosuppressants. Here we report a case of successfully treated EGPA relapse presenting as myocarditis and followed by multimodality imaging.

Case report

A 60-year-old man with EGPA was admitted to our hospital because of gradually progressive dyspnea [New York Heart Association (NYHA) class III] and weakness. Two weeks before admission, he noted dyspnea with exertion, weakness, and paresthesias in both hands. These symptoms worsened over the following weeks.

A diagnosis of EGPA was made 12 years earlier based on findings of numbness in legs, asthma, elevated peripheral blood eosinophilia (17,998/μl), positive myeloperoxidase antineutrophil cytoplasmic antibody (MPO-ANCA) of 212 U/ml (normal range, <3.5 U/ml), and eosinophilic infiltrates in nerve biopsy specimen. After the treatment with prednisolone (60 mg daily), he was considered in remission with no symptoms of numbness and normal eosinophils

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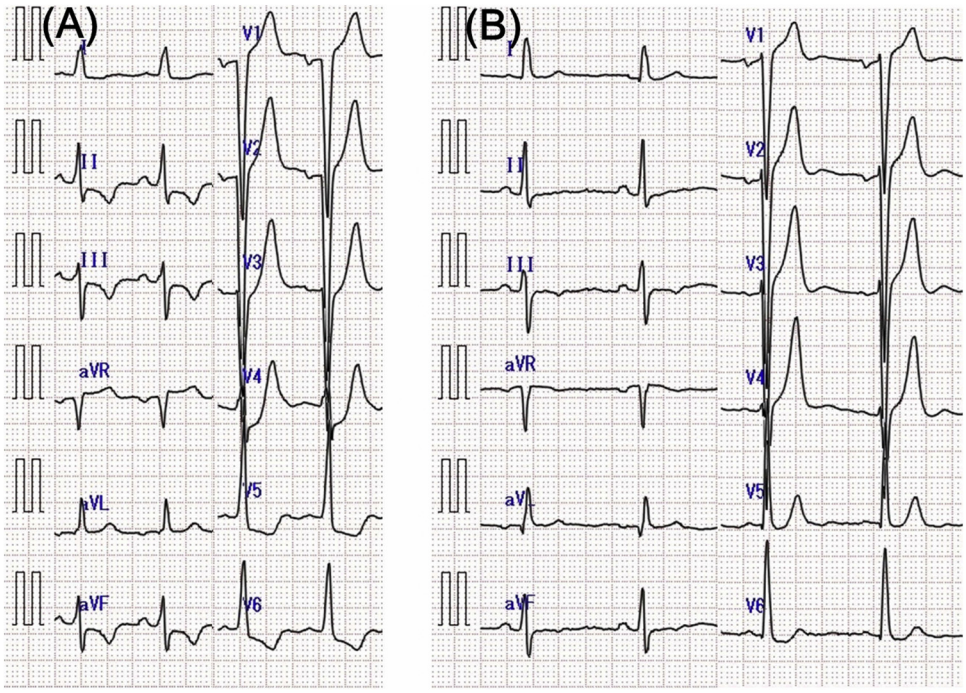

Fig. 1. Electrocardiogram at admission (A) showing ST-segment depressions in leads II, III, aVf, V4-6, and increase in QRS duration compared with that of 1 month earlier (B).

in peripheral blood. The dose of prednisolone was tapered intermittently at maintenance dose of 10 mg daily. One year before admission, surgical replacement of aortic valve with a mechanical valve was required because of aortic regurgitation which was diagnosed at 45 years of age and developed gradually. He had no symptoms (NYHA I), but serial echocardiography showed severe aortic regurgitation with mild left ventricular (LV) systolic dysfunction [ejection fraction (EF) 48%] and progressive severe LV dilatation with LV end-systolic diameter (LVESD) and LV end-diastolic diameter (LVEDD) of 54 mm and 65 mm, respectively. Aortography at the aortic root and transesophageal echocardiography revealed severe aortic regurgitation without aortic valve prolapse, aortic annulus dilatation, and ascending aortic dilatation. Histological examination of aortic valve specimen obtained at surgery demonstrated myxoid and hyalinized change with no evidence of infiltration of eosinophils. Postoperative echocardiography revealed reduction in LVESD and LVEDD to 40 mm and 54 mm, respectively, with mild LV systolic dysfunction (EF 48%). After surgery, he had been well until 2 weeks before this admission under treatment with medications including warfarin and prednisolone (10 mg daily).

On examination, blood pressure was 96/62 mmHg, pulse rate 92 bpm, and body temperature 37.7 °C. Examination of the heart sound showed metallic closing sound without an abnormal regurgitant murmur.

Full blood count showed leukocytosis (25,100/μl) with eosinophilia (15,310/μl). Serum biochemistry revealed elevated levels of troponin T at 1.67 ng/ml (normal range, <0.014 ng/ml), brain natriuretic peptide (BNP) 818 pg/ml, C-reactive protein 3.49 mg/dl, and IgE 2600 IU/ml (normal range, <170 IU/ml). Creatine phosphokinase (CPK), aspartate aminotransferase (AST), lactate dehydrogenase (LDH) were also elevated at 850 U/l, 88 U/l, and 1347 U/l, respectively. The plasma level of creatinine was 0.9 mg/dl (normal, <1.06 mg/dl) and MPO-ANCA was negative (2.7 U/ml).

The electrocardiogram showed ST-segment depressions in leads II, III, aVf, V4-6, and QRS duration broadened compared with that of 1 month earlier (Fig. 1). Chest X-ray demonstrated mild bilateral congestion. Echocardiography showed reduced LVEF of 40%, mild LV enlargement (LVESD 41 mm, LVEDD 52 mm), thickened ventricular wall (interventricular septum 12 mm, posterior LV wall 12 mm), and no valvular dysfunction or vegetations (Fig. 2).

Because of high suspicion of relapse mainly presenting as myocarditis, coronary angiography including endomyocardial biopsy was performed on the 2nd hospital day, which revealed normal coronary angiogram and diffuse infiltration of eosinophils in myocardium (Fig. 3). Based on these findings, relapse was confirmed. He was given three pulses of methylepredonisolone (1000 mg/day) on the 2nd hospital day, followed by maintenance dose of 1 mg/kg/day.

One day after treatment with methylepredonisolone, fatigue, paresthesias in both hands, and dyspnea improved. One week after the onset of treatment, eosinophils had dropped to 329/μl and C-reactive protein was 0.2 mg/dl. The levels of IgE had dramatically decreased (400 U/ml). Oral azathioprine (100 mg daily) was added for maintenance therapy to prevent relapse and allow steroid tapering.

Cardiac magnetic resonance imaging (CMR) performed on the 3rd hospital day demonstrated extensive subendocardial late gadolinium enhancement (LGE) and high intensity T2-weighted signaling of apex, basal inferior, and posterior segment, suggesting myocardial edema, and decreased LV systolic function of 27% (Fig. 2). Thallium-201 (^{201}TL) and iodine-123 (^{123}I)-beta-methyl iodophenyl pentadecanoic acid (BMIPP) dual single-photon emission computed tomography (TL-BMIPP SPECT) also performed on the 4th hospital day showed severe defect in the inferior and posterior regions [TL defect score (TLDS), 12; BMIPP defect score (BMDS), 17; summed mismatch scores (sumMS), 5] (Fig. 2).

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