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Case Report

A rare case of eosinophilic granulomatosis with polyangiitis complicated with progressive pericardial effusion

Toyonori Arinaga (MD)[a,1], Tomo Komaki (MD)[b,c,1], Shin-ichiro Miura (MD, FJCC)[c,d,*], Makito Futami (MD)[c], Joji Morii (MD)[c], Makoto Sugihara (MD)[c], Keijiro Saku (MD, FJCC)[c,d]

[a] Postgraduate Clinical Training Center, Fukuoka University Hospital, Fukuoka, Japan
[b] Department of Laboratory Medicine, Fukuoka University School of Medicine, Fukuoka, Japan
[c] Department of Cardiology, Fukuoka University School of Medicine, Fukuoka, Japan
[d] Department of Molecular Cardiovascular Therapeutics, Fukuoka University School of Medicine, Fukuoka, Japan

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ABSTRACT

A 44-year-old woman had a 15-year history of asthma and had recently been treated for sinusitis and unidentified limb eruption. She had presented at a nearby clinic with inspiratory chest pain one week before hospitalization, and was diagnosed as having eosinophilic pneumonia based on peripheral blood eosinophilia and ground glass opacities in the right lung field, without pericardial effusion, as detected by chest computed tomography. She additionally presented with a feeling of chest tightness, and extensive pericardial effusion appeared within a week. She developed heart failure on admission, and we performed pericardiocentesis. We gave a clinical diagnosis of acute probable myopericarditis as the cause of pericardial effusion based on pleuritic chest pain, pericardial effusion, and elevation of cardiac enzymes, as well as eosinophilic granulomatosis with polyangiitis (EGPA) based on eosinophilia, her history of sinusitis, asthma, and migratory pulmonary opacities. We initiated oral prednisone 25 mg daily and pericardial effusion disappeared. In patients with EGPA, cardiac involvement is more serious than the involvement of other organs, and is associated with a poor prognosis. In this report we describe a rare case of EGPA complicated with progressive pericardial effusion and discuss the importance of the early diagnosis and treatment of EGPA.

<**Learning objective:** Eosinophilic granulomatosis with polyangiitis (EGPA), or Churg Strauss syndrome, is a multisystem disorder. It is important for patients with EGPA complicated with cardiac involvement to be diagnosed and treated early because cardiac involvement may lead to a fatal outcome.>

Introduction

Eosinophilic granulomatosis with polyangiitis (EGPA), or Churg Strauss syndrome, is a multisystem disorder that is characterized by allergic rhinitis, asthma, and prominent blood eosinophilia [1–5]. The prevalence of EGPA in Japan has been reported to be 17.8/1,000,000. The commonly involved organs are the peripheral nervous system and lungs, followed by the skin. However, EGPA can affect any organ system, including the cardiovascular, gastrointestinal, renal, and central nervous systems. In general, most patients with EGPA achieve remission with glucocorticoid therapy alone in the absence of factors associated with a poor prognosis, including cardiac, renal, and/or central nervous system involvement [6]. Cardiac involvement is more serious in patients with EGPA, and this case report highlights the importance of early diagnosis and treatment. We describe here a rare case of EGPA complicated with progressive pericardial effusion and discuss the importance of the early diagnosis and treatment of EGPA.

Case report

A 44-year-old woman presented at a nearby clinic with inspiratory chest pain one week before hospitalization. She had a 15-year history of bronchial asthma (with allergic rhinitis) which was well-controlled by inhaled corticosteroid. She was treated for sinusitis which was diagnosed based on fever, purulent nasal drainage, and facial pain 7 months before hospitalization, and was also treated with steroid ointment for unidentified limb eruption

* Corresponding author at: Department of Cardiology, Fukuoka University School of Medicine, 7-45-1 Nanakuma, Jonan–ku, Fukuoka, 814-0180, Japan.
Fax: +81 91 865 2692.
E-mail address: miuras@cis.fukuoka-u.ac.jp (S.-i. Miura).
[1] Toyonori Arinaga and Tomo Komaki contributed equally to this work.

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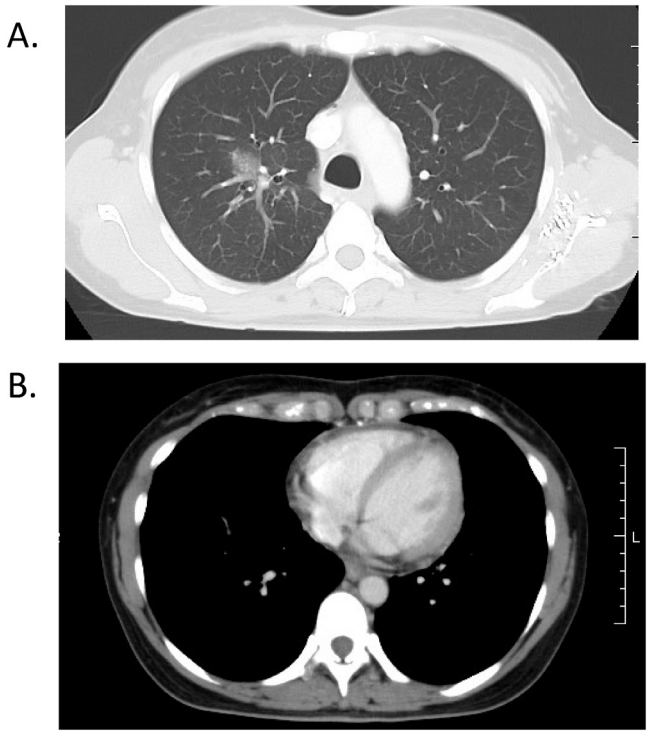

Fig. 1. Ground glass opacities in the right upper lobe lung field (A) and without pericardial effusion (B).

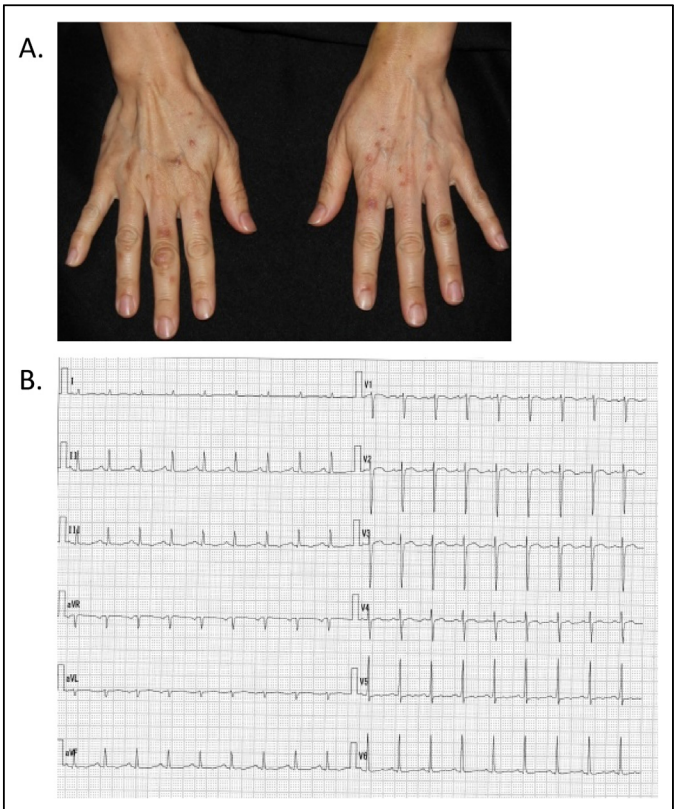

Fig. 2. Crusted eruption on the dorsal side of the hands (A) and electrocardiogram (B) on admission.

with itching, painfulness, and partial purulence 4 months previously. At the clinic, she had peripheral blood eosinophilia (eosinophil 1029/μL) and ground glass opacities (GGO) in the right upper lobe lung field, without pericardial effusion, by chest computed tomography (CT) (Fig. 1A and B). She was diagnosed as having eosinophilic pneumonia. She additionally complained of a feeling of chest tightness, and when she returned to the clinic, cardiomegaly and extensive pericardial effusion, as detected by chest radiography and echocardiogram, respectively, had developed. She was referred to our hospital for the treatment of extensive pericardial effusion. On admission, her level of consciousness was clear, blood pressure was 96/65 mmHg, pulse rate was 103 beats per minute, temperature was 37.8 °C, respiratory rate was 22 breaths per minute, and oxygen saturation (measured by pulse oximetry) was 94% while breathing room air. In the physical examination, distended jugular veins, distant heart sounds, and bilateral leg edema were present. Additionally, crusted eruptions which were treated with steroid ointment 4 months previously were seen on the dorsal side of the hands (Fig. 2A). The electrocardiogram showed flat T wave in the inferior (II, III, and aVf) and lateral walls (I, aVL, and V5-6) (Fig. 2B). Chest radiography showed cardiomegaly (cardiothoracic ratio 59%) and left pleural effusion (Fig. 3A). Transthoracic echocardiogram showed extensive pericardial effusion (front of the right ventricle: 17.8 mm, back of the left ventricle: 29.6 mm, at systole, respectively) which developed progressively within one week and dilatation of the inferior vena cava without asynergic motion, left ventricular hypertrophy, or valvular disease (Fig. 3B and C). With regard to biochemical parameters in blood, the creatine phosphokinase level, the creatine phosphokinase MB isoenzyme and renal,

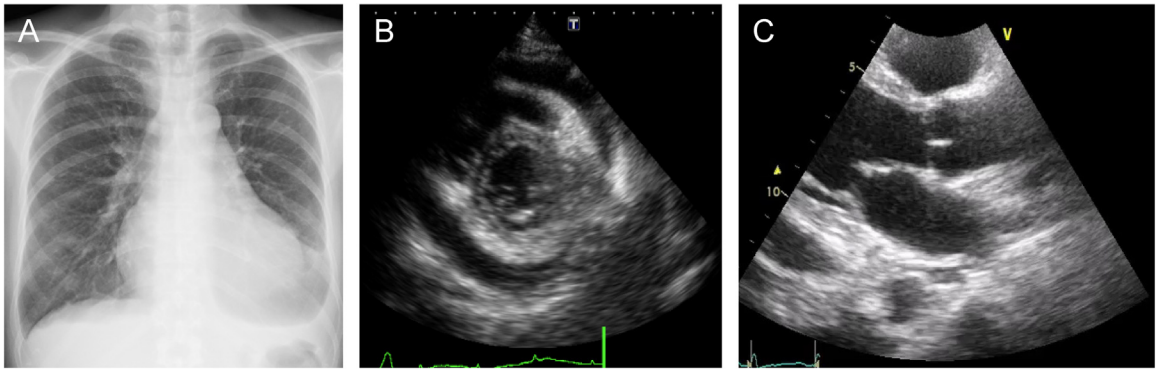

Fig. 3. Chest radiography (A) and transthoracic echocardiogram (B and C) on admission. Ejection fraction was 67%, the thickness of interventricular septum and left ventricular posterior wall was 8.6 mm and 8.6 mm, respectively, and the dimension of left ventricular end-diastole and end-systole was 44.6 mm and 28.2 mm, respectively.

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