



Case Report

Eosinophilic endomyocardial disease



Victor Y Liu *, Raymundo A Quintana-Quezada, L Maximilian Buja

Department of Internal Medicine, McGovern Medical School, The University of Texas Health Science Center at Houston, 6431 Fannin, MSB 1.150, Houston, TX 77030, United States.

ARTICLE INFO

Article history:

Received 20 December 2016

Received in revised form 7 January 2017

Accepted 9 January 2017

Available online xxxx

Keywords:

Eosinophilia

Vasculitis

Cardiac Pathology

Biopsy

ABSTRACT

Eosinophilic granulomatosis with polyangiitis is the rarest of the antineutrophil cytoplasmic antibody-associated vasculitides. We describe the case of a 58-year-old woman with a history of asthma found to have a left ventricular apical thrombus. Cardiac catheterization with endomyocardial biopsy was consistent with eosinophilic endomyocardial disease. The disease affects several organ systems and shares many features with more common conditions, making it a difficult diagnosis.

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A 58-year-old woman presented to the hospital with 4 days of worsening dyspnea, cough, and pleuritic chest pain. She had a history of asthma diagnosed in her twenties, progressively worsening in the last year. Her medications included inhaled corticosteroids and albuterol. She had lived in rural Texas her entire life. Review of symptoms was negative for fever, weakness, paresthesias, hematuria, or rashes. On presentation, she had a temperature of 97.6°F, heart rate of 102, blood pressure of 116/77, and oxygen saturation of 90% on room air. Chest auscultation revealed diffuse crackles bilaterally, more prominent in the upper lobes, without wheezing. Normal strength and sensation were appreciated throughout. Laboratory evaluation revealed leukocytosis of 27,600 cells/ml with 66% eosinophils. Computed tomography (CT) of the chest with contrast showed bilateral patchy pulmonary infiltrates and small pleural effusions (Fig. 1A). The patient immediately received empiric antibiotics and high-dose oral steroids for presumed asthma exacerbation in the setting of pneumonia. She was subsequently found to have an electrocardiogram without ST-segment abnormalities but an elevated troponin of 5.55 ng/ml. Echocardiogram showed ejection fraction of 30%–35%, abnormal relaxation pattern, and a left ventricular (LV) apical thrombus; the patient was heparinized. Cardiac magnetic resonance (CMR) confirmed the presence of an LV thrombus (Fig. 1B) and showed multiple areas of subendocardial delayed gadolinium enhancement. Antinuclear antibody was positive at a titer of 1:160. Antineutrophil cytoplasmic antibody (ANCA), anti-myeloperoxidase, anti-proteinase 3, rheumatoid factor, and *Aspergillus* and *Strongyloides* serologies were all negative. Cardiac catheterization ruled out coronary artery disease.

Endomyocardial biopsy of the right side of the interventricular septum showed an organizing mural thrombus adjacent to a plaque-like thickening of the endocardium consisting of advanced granulation tissue containing lymphocytes, macrophages, eosinophils, and moderately dense collagen. The underlying myocardium showed mild interstitial fibrosis and vacuolar change of myocytes. These findings were consistent with intermediate-stage eosinophilic endomyocardial disease of the right ventricle, indicating biventricular involvement (Fig. 2A–F). The patient was diagnosed with eosinophilic granulomatosis with polyangiitis (EGPA), and antibiotics were discontinued. In addition to her high-dose steroids, she began monthly intravenous cyclophosphamide. The patient's symptoms subsided, complete blood count normalized, and subsequent chest X-ray showed resolution of pulmonary infiltrates. Repeat echocardiogram found an ejection fraction of 40%–45% with abnormal relaxation pattern. She was discharged home on daily oral prednisone with appropriate rheumatology follow-up.

EGPA is a rare ANCA-associated vasculitis with an incidence and prevalence of 0.5 to 3.7 and 2 to 22 per million, respectively. The most commonly affected organs are the lungs and skin. Heart involvement is responsible for 50% of deaths due to EGPA; patients within this subgroup have higher eosinophil counts and are less likely to be ANCA-positive [1,2]. Thrombus visualized on echocardiography prompted endomyocardial biopsy, which confirmed cardiac involvement [3,4]. Typical findings on CMR with contrast are contractile dysfunction, subendocardial late enhancement, and mural thrombus [5]. The worsening of previously controlled asthma in an adult and prominent eosinophilia in a patient with a systemic illness are important clues that should make us consider EGPA as a potential culprit. This disorder is associated with a very high mortality if left untreated. Glucocorticoids are the mainstay of induction therapy for all patients with EGPA, and cases with cardiac involvement additionally require cyclophosphamide. Eosinophilic endomyocardial

* Corresponding author at: Department of Internal Medicine, McGovern Medical School, The University of Texas Health Science Center at Houston, 6431 Fannin, MSB 1.150, Houston, TX 77030, United States.

E-mail address: victor.y.liu@uth.tmc.edu (V.Y. Liu).

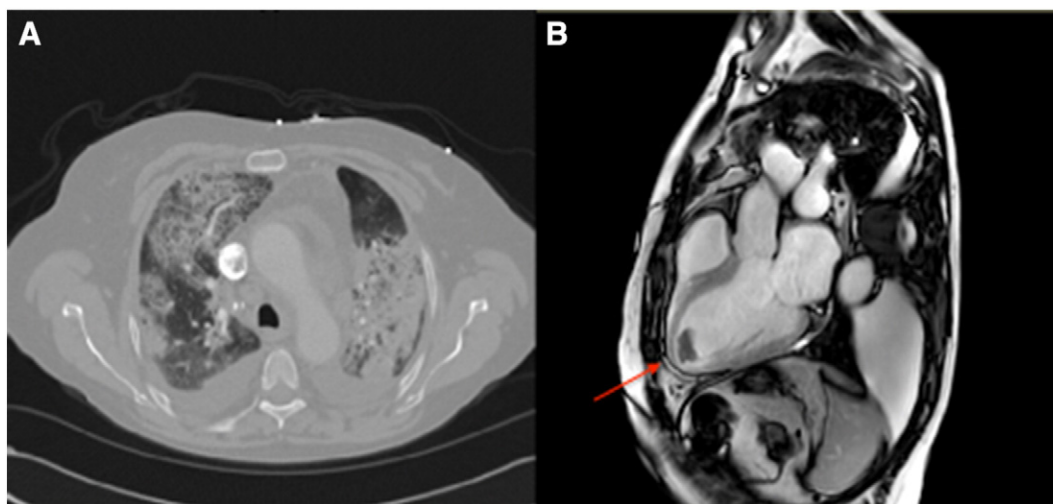


Fig. 1. CT of the chest with contrast revealing bilateral patchy pulmonary infiltrates more prominent in the upper lobes and small pleural effusions (A). CMR with contrast showing a 2.5-cm thrombus (arrow) in the left ventricular apex (B).

disease should be distinguished from tropical endomyocardial fibrosis, which can present similarly but is endemic only to tropical climates.

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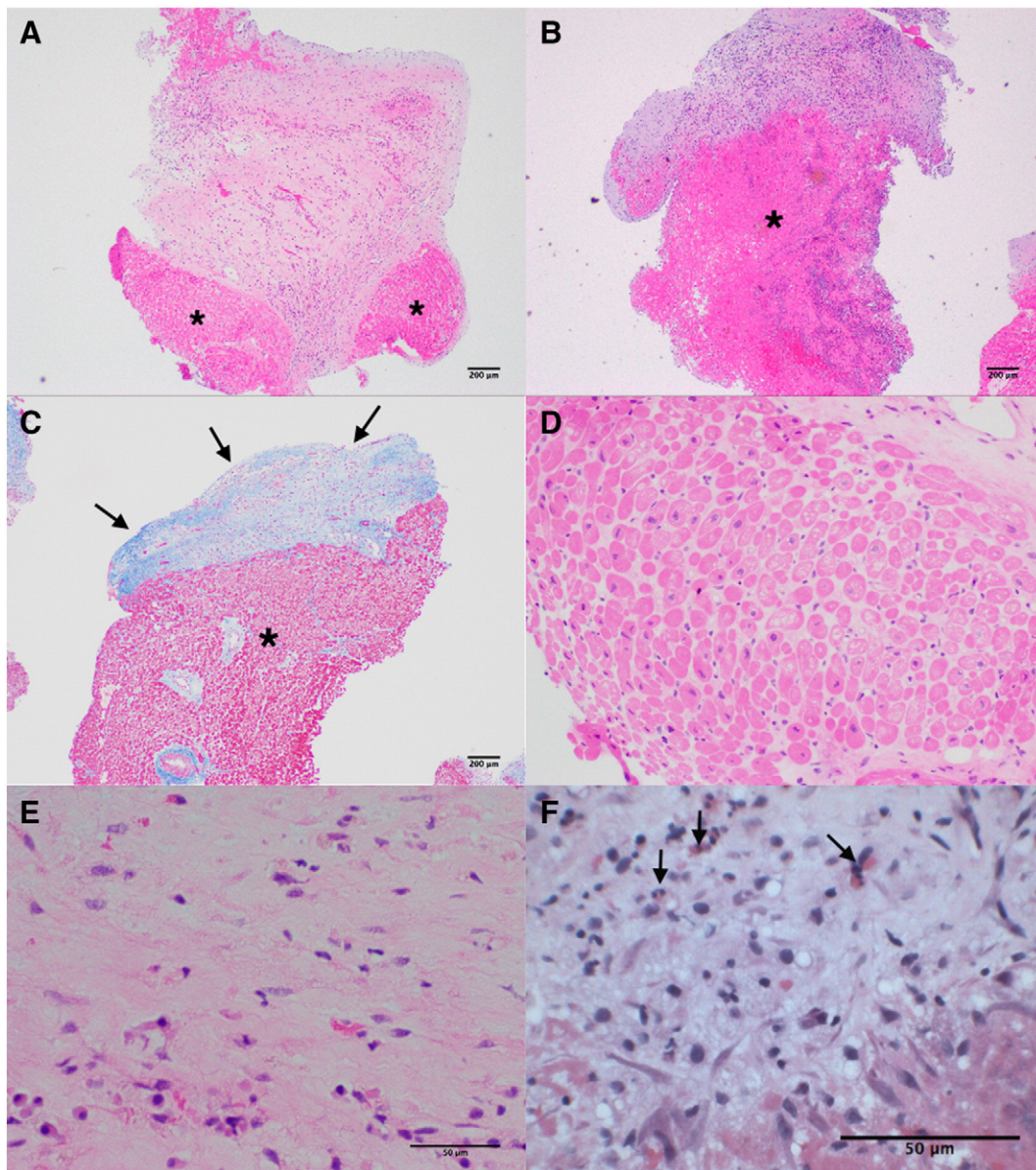


Fig. 2. Endomyocardial biopsies. Endocardial plaque containing collagen and inflammatory cells. Small portions of myocardium (asterisk) are shown (hematoxylin–eosin [H&E] stain, 40 $\times$) (A). Organizing thrombus (asterisk) attached to the endocardial plaque is shown (H&E, 40 $\times$) (B). Masson's Trichrome stain confirming the presence of abundant collagen fibers in the fibrous endocardial plaque (arrows). Underlying myocardium (asterisk) is also seen (40 $\times$) (C). Myocytes with vacuolar change and interstitial fibrosis (240 $\times$) (D). Endocardium with areas of loose collagen with fibroblasts (400 $\times$) (E) and inflammatory infiltrates containing numerous eosinophils (arrows) (650 $\times$) (F).