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Case report

Eosinophilic granulomatosis with polyangiitis (formerly known as Churg-Strauss syndrome) as a differential diagnosis of hypereosinophilic syndromes

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ABSTRACT

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg-Strauss syndrome, is a rare systemic disease situated between primary small vessel vasculitides associated with antineutrophil cytoplasmic antibodies (ANCAs) and hypereosinophilic syndromes (HES). Here, we present a case of EGPA in a 38-year-old male, with a previous diagnosis of asthma, who presented with fever, migratory lung infiltrates and systemic eosinophilia that was refractory to previous courses of antibiotics. This case highlights the importance of the primary care physician understanding the differential diagnosis of pulmonary eosinophilic syndromes.

1. Introduction

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg-Strauss syndrome (CSS) [1], was first described in 1951 by Churg and Strauss as a rare disease characterized by disseminated necrotizing vasculitis with extravascular granulomas occurring exclusively among patients with asthma and tissue eosinophilia [2–4]. EGPA is a disease situated between primary systemic vasculitides [1] and hypereosinophilic disorders [5,6]. Within this dual categorization, EGPA is classified among small-vessel vasculitides associated with antineutrophil cytoplasmic antibodies (ANCAs) and hypereosinophilic syndromes (HESs) [5], which are syndromes with accompanying hypereosinophilia [6]. Both vessel inflammation and eosinophilic proliferation have been proposed to contribute to organ damage, but the clinical presentations are heterogeneous, and the respective roles of vasculitis and hypereosinophilia in the disease process are not well understood [3].

Here, we present a case of EGPA in a 38-year-old male, with a previous diagnosis of asthma, who presented with fever, migratory lung infiltrates and systemic eosinophilia that was refractory to previous courses of antibiotics.

2. Case report

A 38-year-old male, non-smoker, with a previous diagnosis of asthma from childhood, presented with fever, productive cough with haemoptoic sputum and upper airway respiratory symptoms. He reported that his asthma had worsened in previous years and

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was refractory to medications. With an exception of budesonide/formoterol (400/12 μg twice daily in the last 6 months) and prednisone (10 mg once daily in the last 2 months), he denied using other medications. One month prior to admission, he had intermittent fever and a worsening of the cough, which became more intense and productive and was associated with dyspnoea. He had received multiple antibiotic courses (azithromycin for 10 days, levofloxacin for 10 days and amoxicillin-clavulanate for 14 days) without success.

A chest computed tomography (CT) showed diffuse ground-glass opacification (GGO) (Fig. 1), and a paranasal sinuses CT revealed opacification of the frontal and ethmoidal sinuses (Fig. 2). Pulmonary function tests indicated a severe obstructive pattern and no post-bronchodilator response (forced vital capacity (FVC) = 59% predicted; forced expiratory volume in 1 s (FEV_1) = 35% predicted; FEV_1/FVC = 50%; total lung capacity (TLC) = 89% predicted; residual volume (RV) = 177% predicted; RV/TLC = 191% predicted; diffusing capacity for carbon monoxide (DLco) = 69% predicted). Laboratory examinations demonstrated leucocytosis (14400/mm^3) with marked eosinophilia (3168 mm^3/22%) and normal renal function, and a urine dipstick test revealed urinary occult blood (2+) without erythrocyte dysmorphism, elevated immunoglobulin (Ig)E and a reactive p-ANCA (myeloperoxidase) on two separate occasions. Stool microscopy did not identify any ova, cysts or parasites, and serum antibody tests for the parasites *Fasciola hepatica*, *Strongyloides* spp., *Trichinella* spp., *Taenia solium*, *Schistosoma mansoni* and *Toxocara canis* were negative. Antigen-specific IgE antibody test to *Aspergillus fumigatus* was also negative. Despite the negative results of the serologies, it was opted for the administration of albendazole 400 mg once daily for three days, as antiparasitic prophylaxis. An open lung biopsy was performed and demonstrated intense perivascular eosinophilic inflammatory infiltrate (Figs. 3 and 4), confirming the diagnosis of EGPA [4,7,8] (Table 1). Posteriorly, a transthoracic echocardiogram and electroneuromyography for complementary investigation were performed and produced normal results.

The patient received prednisone 40 mg once daily (0.5 mg/kg/day) for four months leading to a resolution of respiratory and

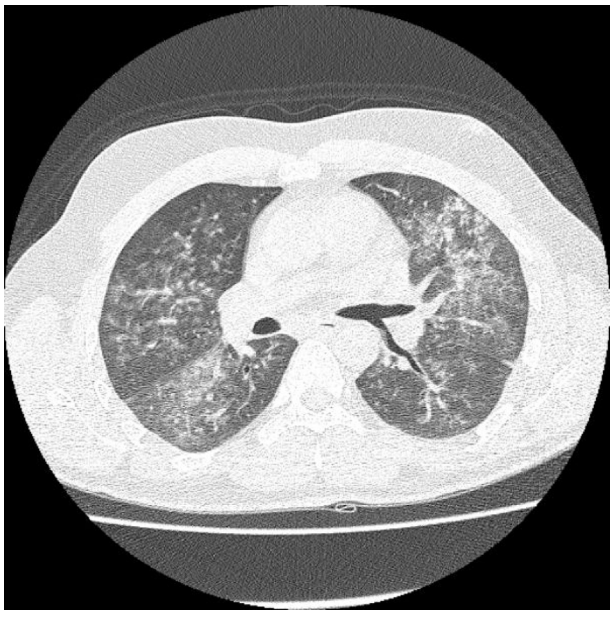

Fig. 1. Chest computed tomography showing diffuse ground-glass opacities.

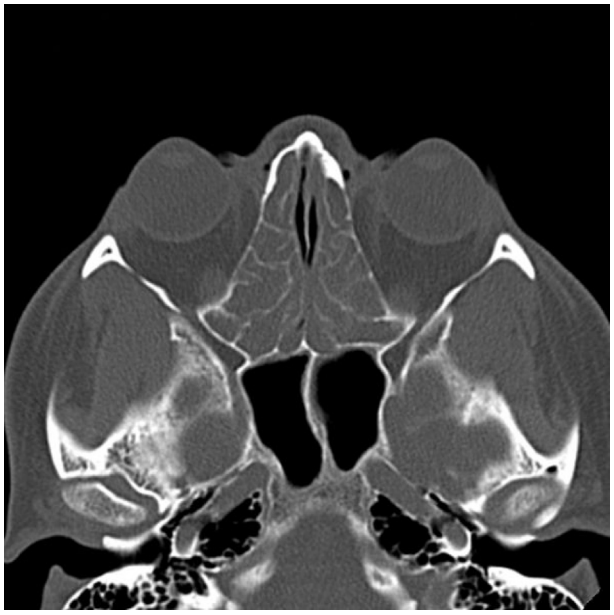

Fig. 2. Paranasal sinuses computed tomography showing opacification of ethmoidal sinuses.

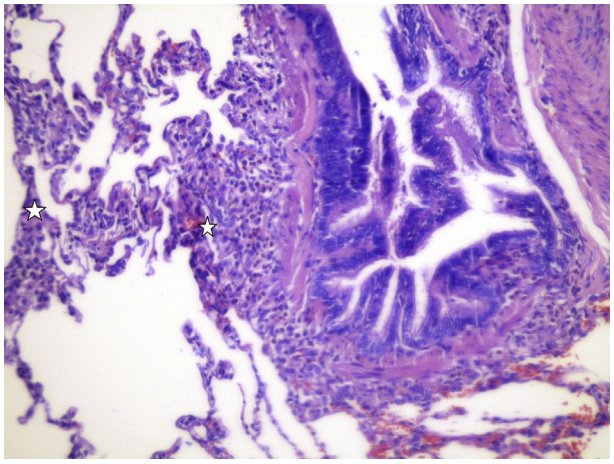

Fig. 3. A diffuse chronic inflammatory infiltrate with a marked presence of eosinophils (star) is found near the bronchioles.

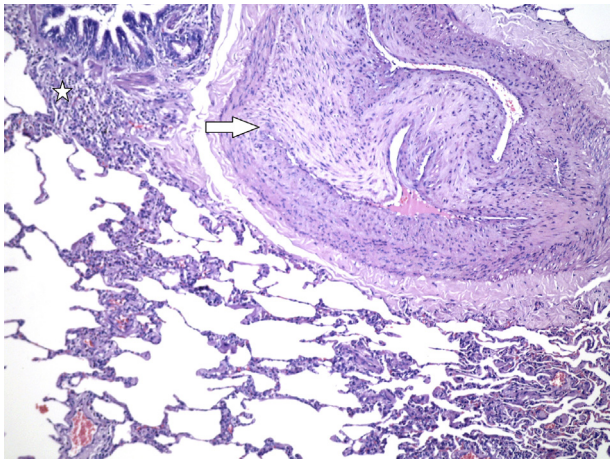

Fig. 4. Arterial vessels present thickening of the wall (arrow) due to muscular hypertrophy and fibrosis of the intima, in a plexiform arrangement that appears to present several vascular lights. Note the eosinophil infiltrate near the bronchiole (star).