Newly Diagnosed EGPA Presenting as Epiglottic Ulceration and Severe Acute Renal Failure

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INTRODUCTION

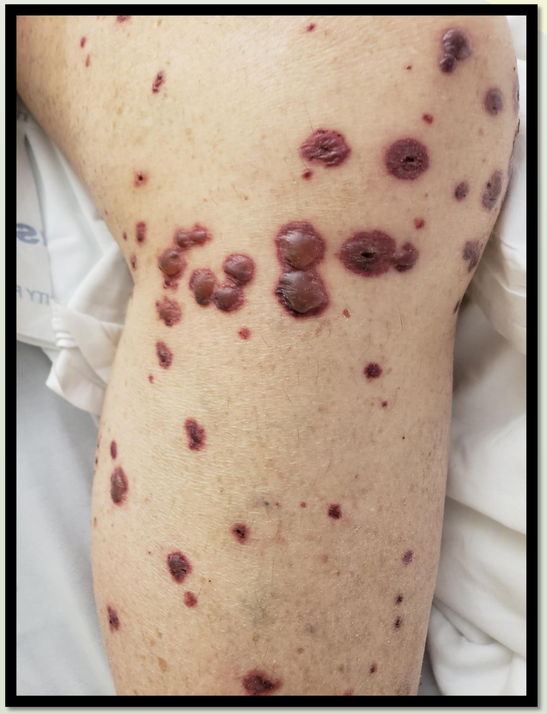
Eosinophilic granulomatosis with polyangiitis (EGPA), previously called Churg-Strauss syndrome, is a multisystem disorder characterized by chronic rhinosinusitis, asthma, and prominent eosinophilia. It is classified as a vasculitis of the small and medium-sized arteries. EGPA frequently involves the respiratory tract with the lungs being the most commonly involved organ but the involvement of epiglottis has not been documented. Additionally, initial presentation as severe acute renal failure is uncommon. Here we are reporting a newly diagnosed EGPA presenting with severe acute renal failure and epiglottic ulceration.

THE CASE

Our patient is a 67-year-old male with a past medical history of obesity, asthma, peripheral neuropathy, chronic back pain, and radiculopathy, who came to the emergency department (ED) due to swelling of his hands and legs for 1 week which has been extending proximally. The patient also reported productive cough, weight loss secondary to poor intake due to throat pain, chronic nasal congestion, and acute worsening of chronic right-sided hearing loss since the last few months. The patient reported worsening shortness of breath despite increasing his usage of an albuterol inhaler.

HOSPITAL COURSE

In the ED patient was found to be hypoxic on room air and tachypneic with wheezes heard on auscultation. On examination, a shallow ulcer was seen in the left buccal mucosa with a few purpuric lesions in the upper lip. He was also found to have moderate-sized purpuric lesions across bilateral lower extremities from knees to toes, a few of which had scabbed over and were crusted with blood.



WORK UP

Leukocytosis of 15,000 cells/mm3 with 36% eosinophils

Creatinine at 5.69mg/dl.

CRP and ESR: Elevated

MPO-ANCA positive and Pr3-ANCA negative

CT Chest: A 2.3cm x 1.6cm oval focal area of pneumonitis

CT Face: Pansinusitis with possible nasal polyposis

Lower Extremity Venous Doppler study: Right lower extremity DVT

Flexible laryngoscopy: Mildly erythematous epiglottis with ulceration of proximal lingual surface.

Pt was started on a heparin drip for DVT. He was initiated on hemodialysis for the worsening acute kidney injury. The patient underwent a kidney biopsy and it revealed acute tubulointerstitial inflammation with eosinophils, acute tubular necrosis, and sclerosed glomerulus. The patient was positive for 6 of the 6 ACR diagnostic criteria for EGPA including asthma, eosinophilia, neuropathy, radiologically detected pulmonary opacity, paranasal sinus abnormality, and renal biopsy with eosinophils. He was started on treatment with cyclophosphamide and steroids as his five-factor score was 2. His renal function and odynophagia improved, and lower extremity lesion began to dry up and heal with no new lesions. He remained stable off dialysis and was discharged on steroids.

CONCLUSIONS

EGPA is a rare disease with a prevalence of 5-17 per million adults. It is characterized by blood/tissue eosinophilia, asthma, extravascular granuloma formation, and vasculitis of multiple organ systems. The mean age of diagnosis of EGPA is 50 years and it is an uncommon form of vasculitis in people aged over 65 years.

ENT manifestations are common in EGPA and these usually involve allergic rhinitis, nasal polyposis, sensorineural hearing loss, and nasal crusting. Besides unilateral hearing loss and chronic nasal congestion, our patient interestingly also had odynophagia due to ulceration of epiglottis. Though uncommonly Wegener's Disease has been shown to cause epiglottis involvement, this is an extremely rare finding in EGPA.

Renal abnormalities are present in about one-quarter of patients with EGPA, however severe acute renal failure is relatively rare with an incidence of only about 4.3%.

This case report is directed at increasing awareness among medical providers about the widely varied presentations of EGPA. The rarity of this disease makes it important to bring to light the diverse ways in which it manifests.