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Case Report

A Curious Case of Churg-Strauss

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Abstract

Eosinophilic granulomatosis with polyangiitis (EGPA), formally known as Churg-Strauss, is a rare multisystem vasculitis characterized by asthma, rhinosinusitis and eosinophilia. The lungs are the most common organ system affected, followed by the skin. Extrapulmonary organ involvement, specifically cardiac involvement, is the leading cause of morbidity and mortality associated with the disease [1]. Disease remission is usually achieved with high dose corticosteroids or immunosuppressive therapy. The clinical features of EGPA typically evolve over several stages, which include a prevasculitic or prodromal stage, an eosinophilic stage, and finally, a life-threatening vasculitic stage. Historically, EGPA was a pathologic diagnosis, meaning that overt vasculitis had to be present in order to establish a diagnosis; however, nearly forty-percent of patients with EGPA present with asthma, pulmonary infiltrates and eosinophilia prior to the development of vasculitis [1-3]. The American College of Rheumatology (ACR) implemented a diagnostic criterion that encompasses a clinical approach, which allows for the diagnosis of EGPA to be made, even when there is no pathologic evidence of vasculitis. This allows for early recognition of disease and prompt initiation of treatment, which may lead to improved outcomes. We introduce a patient who was misdiagnosed with bacterial pneumonia on three separate occasions, but was later found to have EGPA despite lacking pathologic evidence of vasculitis.

Keywords: *Churg-Strauss; Eosinophilic Granulomatosis with Polyangiitis; Eosinophilia; Vasculitis; Asthma*

Introduction

Eosinophilic granulomatosis with polyangiitis (EGPA), formally known as Churg-Strauss, is a rare multisystem vasculitis characterized by asthma, rhinosinusitis and eosinophilia.

Case Presentation

A fifty-two year old Japanese male with past medical history significant for diabetes and asthma presented for evaluation of cough and shortness and breath over a one-month period. He was previously evaluated at an urgent care and was started on Doxycycline, Levaquin and oral steroids for suspected community acquired pneumonia (CAP). He improved initially, but his symptoms returned after completion of therapy prompting a second round of antibiotic treatment. Unfortunately, he developed an allergic reaction and therapy was discontinued. He subsequently presented to the hospital due to persistent shortness of breath, sinus congestion, skin rash, and dysphagia. Additionally, he complained of a 40lb unintentional weight loss over a 1-month period. Upon evaluation of the patient, he was tachycardic, tachypneic and hypoxic requiring 5L of supplemental oxygen. A maculopapular rash was noted over his forehead, elbows, buttocks and flanks. He had a leukocytosis of 19.5k but remained afebrile and normotensive. He was admitted for presumed community acquired pneumonia and was started on IV Azithromycin, Vancomycin, Zosyn, Fluconazole and oral steroids. Work-up revealed multifocal bilateral airspace disease on chest x-ray, moderate bilateral groundglass opacities on CTPA, hypereo-

sinophilia with eosinophil percentage of 22.6%, and elevated IgE levels (1351). His CRP was elevated to 90 mg/dL and his ESR was 47 mm/hr. Patient had negative ANA and ANCA levels. No blood, protein or casts were noted on urine analysis and his kidney function was normal. His infectious work-up was negative including viral respiratory panel, Legionella, Aspergillus, Histoplasmosis, HIV, blood cultures, fungal cultures and respiratory cultures, thus antibiotic and antifungal therapy was discontinued.

On hospital day 3, patient was started on IV steroids, which drastically improved his symptoms. Due to the non-specific CT chest findings, patient underwent bronchoscopy, which demonstrated eosinophilic infiltrates on lung biopsy; however, no eosinophils were found in the BAL fluid. Additionally, gastroenterology evaluated the patient for dysphagia with EGD findings grossly consistent with eosinophilic esophagitis; however, biopsy showed only scant eosinophils. A punch biopsy was obtained to further investigate diffuse skin rash, results indicated eosinophils without granulomas. The patient continued to improve over the course of his hospitalization with one-gram IV solumedrol and his eosinophil count normalized. Although the patient did not have evidence of vasculitis, he was ultimately diagnosed with EGPA given the systemic eosinophilia, uncontrolled asthma, pulmonary infiltrates, skin rash, elevated IgE and his remarkable response to high dose steroids. He was discharged with 60 mg Prednisone for 30 days followed by a steroid taper with plans to follow-up with rheumatology on an outpatient basis. Per chart review, patient was completely tapered off of prednisone and has remained symptom free for over six-weeks.

Discussion

EGPA is not only rare, but has proven itself to be difficult to diagnose, and our case was no exception. Our patient presented with a constellation of symptoms that were initially and understandably dismissed as an infectious process despite failed antibiotic therapy. After spending six-days in the hospital and undergoing an extensive work-up, the diagnosis of EGPA was finally made. Our case of EGPA is interesting in several ways. First, the patient had eosinophils seen on lung biopsy, but no eosinophils noted on bronchoalveolar lavage. Second, the patient's EGD was grossly consistent with eosinophilic esophagitis; however, only scant eosinophils were seen on biopsy. Third, the patient's skin rash appeared to be consistent with a vasculitic rash; however, his biopsy showed no evidence vasculitis. Finally, the patient's ANCA was negative. In the past, this patient would not have met diagnostic criteria for EGPA as he had no sign of vasculitis on biopsy. The ACR's diagnostic criteria of EGPA includes asthma, paranasal sinus abnormalities, eosinophilia > 10%, neuropathy, pulmonary infiltrates and biopsy containing a blood vessel with extravascular eosinophils. Patients must have at least 4 out of the 6 criteria in order to make a diagnosis [1]. By utilizing this criterion, the diagnosis of EGPA was established and the patient could be treated with high dose steroids early on in the disease. With that said, the early course of steroid therapy may explain our patient's diagnostic findings and inconsistency with peripheral eosinophilia as steroid therapy suppresses the disease. Another interesting finding in this case is the fact that his ANCA was negative. Nguyen and Guillevin [4] reported that ANCA negativity was associated with eosinophilic infiltrates and higher rates of cardiac involvement; whereas, ANCA positivity was associated with frequent vasculitis-associated symptoms, renal involvement and mononeuritis. ANCA positivity can be found in thirty to sixty percent of EGPA patients but is not needed to confirm diagnosis. Our patient's negative ANCA and its association with eosinophilic infiltrates is consistent with Nguyen's and Guillevin's report. This particular case illustrates how EGPA can easily be overlooked in the early stages of disease if we rely solely on pathologic evidence of vasculitis to establish diagnosis [5].

Conclusion

Although rare, EGPA is an important consideration when managing patients with asthma, eosinophilia, pulmonary infiltrates, neuropathy, paranasal sinus disease, and extravascular eosinophils. It is important to work-up patients thoroughly and rule out infection and other causes of hypereosinophilia. Early detection of EGPA and prompt initiation of high dose steroid therapy is critical, as earlier stages of disease tend to have a better response to steroids with possible resolution of symptoms when compared to later stages of disease.

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