Lower Extremity Presentation of Eosinophilic Granulomatosis with Polyangiitis (EGPA)

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Purpose

Eosinophilic granulomatosis with polyangiitis (EGPA), formally known as Churg Strauss Disease, is an extremely rare condition that involves allergic rhinitis and asthma, eosinophilia, and small vessel vasculitis. There are often lower extremity findings including peripheral neuropathy and rash[3]. The infrequent presentation of this disease often leads to delay in diagnosis and treatment[5]. This case study highlights the presentation of EGPA in a 48-year-old female at our hospital.

Literature Review

EPGA is a disorder primarily involving necrotizing small vessel vasculitis with associated respiratory manifestations and eosinophilia[3]. The American College of Rheumatogy classified EPGA in 1990 by most common signs and symptoms (Table 1)[4]. The guidelines for diagnosis were modified in 2012 by the International Chapel Hill consensus conference to include the presence of anti-neutrophil cytoplasmic antibody (ANCA), which is most common in the setting of glomerulonephritis. It typically presents in three sequential phases[6]. The first phase, the allergic phase, is characterized by the presence of asthma, allergic rhinitis, and sinusitis. The second phase is the eosinophilic phase, which typically involves an increase in eosinophil count in the lungs, heart, and gastrointestinal count. The third and final phase is termed the vasculitic phase as the patient presents with symptoms of necrotizing vasculitis, including the lower extremity symptoms of neuropathy and purpura[6]. Cutaneous manifestations are seen in 40%-81% of EPGA. It is the presenting complaint in 14% of patients. Cutaneous manifestations include most commonly palpable purpura and urticaria, less commonly papular/nodular lesions, livedo reticularis, ulcerations, bullous lesions, and cutaneous infarcts are also seen[1]. The second notable lower extremity manifestation is peripheral neuropathy. Neuropathy affects 70% of patients with EPGA. Mononeuropathy is slightly more common in EPGA and frequently involves the peroneal, tibial, ulnar, and median nerves. Mononeuropathy can be complicated by foot drop. Symmetric or asymmetric polyneuropathy, sensory disturbances, and neuropathic pain are also frequently seen[2].

Case Study

A 48 year old female presented with lower extremity cellulitis in the setting of a lower extremity purpuric rash. She has a past medical history significant for juvenile rheumatoid arthritis, diffuse ground glass opacities treated with prednisone, cardiomyopathy, and pulmonary hypertension. Over the past year she developed severe asthma like symptoms as well as venous thromboembolic event. Twenty days prior to admission the patient lower extremities erupted with a purpuric rash with several loose bulla. Prior to admission to our hospital she was found to have eosinophilia of 17,404/µl, erythrocyte sedimentation rate of 97 mm/hr. She had been treated with 20mg prednisone daily. She reported worsening of paresthesia throughout her hospital stay. At time of admission she was found to have multiple purpura on bilateral lower extremities extending to the thigh and multiple bulla in various stages of healing. There was cellulitis of the left lower extremity. At this time there was improvement of eosinophilia to 1,800/µl. Her erythrocyte sedimentation rate increased to >120 mm/hr. ANCA was found to be positive. She was treated with bilateral zinc impregnated gauze wraps, intravenous antibiotics, and an increase in prednisone to 60mg daily for the prevention of peripheral neuropathy progression.

Image 1: Palpable purpura present on day of admission

Results and Discussion

EPGA is an uncommon disorder that is often met with a delay in diagnosis and a delay in appropriate treatment. Oiwa and colleagues showed that there is often a greater than 14-day physician delay. During this time patients are often inappropriately treated[5]. They suggest in patients with asthma and sudden sensory disturbance EPGA should be considered[5]. Patients diagnosed with EPGA must be monitored closely for lower extremity manifestations in order to provide appropriate treatment. Cutaneous lesions should be treated with supportive care until clearance occurs[1]. Neuropathy has more lasting morbidity than cutaneous lesions. Mononeuropathy, while often painless, poses a greater risk for disability than polyneuropathy[2]. The first line treatment is glucocorticoid therapy during acute phase disease followed by maintenance therapy with azathioprine or methotrexate[3]. Recent analyses demonstrate good mortality rates. In a review of 118 patients 91.5% of patients went onto complete remission at long term follow up. Typical relapse symptoms included pulmonary manifestations, return or worsening of neuropathy, and ear nose and throat symptoms[3].

Table 1: American College of Rheumatogy Classification criteria (1990)*

Asthma
Eosinophilia (>10% of total WBC)
Neuropathy
Pulmonary infiltrates non-fixed
Paranasal sinus abnormalities
Extravascular eosinophils

* four or more of these six criteria yielded a sensitivity of 85% and a specificity of 99.7% for the classification of vasculitis as EGPA

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