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Case Report #5

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Case Title

On Pins And Needles: Recognizing Neuropathy As First Clinical Manifestation Of Eosinophilic Granulomatosis With Polyangiitis

Summary

Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare form of necrotizing vasculitis that affects small and medium sized vessels and typically develops in patients with preexisting asthma and allergic rhinosinusitis. While neurologic complications of EGPA are common, it remains a challenge to suspect and diagnose EGPA if neuropathic complications dominate the clinical picture and precede the onset of other vasculitic symptoms. We present a case of ANCA-negative EGPA that manifested as neuropathy and was successfully treated with cyclophosphamide and prednisone.

Patient Presentation

47-year-old male with asthma presented with complaints of pain and weakness in his legs. Symptoms started 4 months prior as numbness and tingling in his right lower extremity, and then gradually progressed to burning pain and foot drop bilaterally. Findings of initial physical examination revealed diminished sensation and weakness of the flexors/extensors of the feet.

Diagnosis

Initial work-up was positive for blood eosinophilia of 33% (3,125 cells/mcL) and elevated ESR of 45 mm/hr. Additional work-up showed positive RF, negative ANA, negative c-ANCA and p-ANCA, and negative CXR. Nerve conduction study (NCS) was not revealing of axonal or demyelinating process. While awaiting sural nerve biopsy, the patient had worsening of the weakness in his right foot and developed a purpuric rash on his lower extremities. He was admitted to the hospital and the skin biopsy was performed. ESR increased to 91 mm/hr and eosinophilia progressed to 49% (8,550 cells/mcL). A constellation of symptoms of asthma, eosinophilia, neuropathy, and purpuric rash led to establishing a diagnosis of EGPA and initiation of methylprednisone 1 g daily for 3 days followed by oral prednisone and cyclophosphamide.

Testing

Skin biopsy showed leukocytoclastic vasculitis and tissue eosinophilia, confirming the diagnosis.

Treatment

Over the next month the patient remained on 50 mg of cyclophosphamide twice daily and 40 mg of prednisone once daily which resulted in improvement in sensory deficits. His ESR decreased to 10 mm/h and percentage of eosinophils normalized to 1.4% (158 cells/mcL).

Patient Outcomes

He was then continued on induction therapy of cyclophosphamide and prednisone for additional 2 months until improvement in the weakness was achieved. For the maintenance therapy, we are planning to start mepolizumab and methotrexate.

Lessons Learned

EGPA is a rare vasculitis of small and middle size vessels. It tends to progress in 3 stages: prodromal, eosinophilic and vasculitis phase. In its prodromal phase, symptoms of asthma and rhinosinusitis predominate. Eosinophilic phase is characterized by blood eosinophilia and eosinophilic infiltration of organs, most commonly the lungs, resulting in transient pulmonary infiltrates. In its vasculitic phase, the disease becomes systemic and can affect virtually any organ but typically involves skin, kidneys, and nervous system. The multisystemic nature of this disease was acknowledged in Lanham diagnostic and later on in ACR classification criteria. ACR model remains the preferred tool for the diagnosis of EGPA and requires the presence of at least 4 criteria in a patient with histologically proven vasculitis: asthma, eosinophilia > 10% of total WBC count, mononeuropathy or polyneuropathy, paranasal sinuses involvement, transient pulmonary infiltrates, and evidence of extravascular eosinophilia on tissue biopsy. However, relying on ACR criteria can be challenging when vasculitis is limited to one organ system only while signs and symptoms of preceding prodromal and eosinophilic phases are mild or absent. Our patient never had rhinosinusitis or pulmonary infiltrates on chest computed tomography. His asthma was stable and well controlled on a combination of fluticasone furoate and vilanterol. More importantly, the neuropathy heavily dominated the clinical picture and preceded the onset rash by several months. In addition, his ANCA was negative. This case illustrates the importance of recognizing EGPA as a possible cause of newly developed neuropathy in a patient with asthma and eosinophilia, in the absence of other symptoms of EGPA. Our observation that neuropathy in EGPA started as painful sensory mononeuropathy and then progressed to mixed symmetric polyneuropathy correlated with results of previous studies and such clinical course appears to be typical of EGPA neuropathy