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Case Report

Markedly high peripheral blood eosinophilia: A rare manifestation of Eosinophilic granulomatosis with polyangiitis

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Abstract

Eosinophilic granulomatosis with polyangiitis (EGPA) is a systemic necrotizing multi-system vasculitis. EGPA presents with severe eosinophilia is rare. Here we report a case of EGPA, presenting with markedly high eosinophilia with tissue infiltrates. It was successfully treated with immunosuppressive medications.

Keywords

EGPA, Late-onset asthma, Peripheral neuropathy, Markedly high eosinophilia

Introduction

EGPA is a multi-system disorder, predominantly affecting small to medium vessels and is associated with allergic rhinosinusitis, late onset poorly controlled asthma, blood and tissue eosinophilia, peripheral neuropathy, migratory lung opacities, vascular and extravascular granuloma (1). This could be fatal if left untreated. Treatment is primarily with corticosteroids and, severe disease is managed with the addition of other immunosuppressants (2).

Case presentation

A 57-year-old previously healthy female has presented with chronic cough with non-purulent sputum and wheeze for more than six months. She also had exertional breathlessness of mMRC Grade 3, and chest tightness. She also reported anorexia and unintentional weight loss of 5kg over the same duration. She didn't recall a history of fever or night sweats. There was no history of atopy. She was a non-smoker. There were no focal symptoms of malignancy such as altered bowel habits, hemoptysis, hematuria, or abdominal pain. Her BMI was 18kg/m^2. There was a polyphonic wheeze in both phases of respiration. She developed progressive, asymmetrical sensory-motor neuropathy involving both upper and lower extremities manifested as right-sided foot drop and left-sided ulnar claw hand. Cranial nerves were normal. The rest of the examination was normal.

Her laboratory data revealed leukocytosis (50,780/microL) with markedly high eosinophilia (74%-37,580/microL) with high ESR (88mm/1st hour) and CRP of 24 mg/L. Her renal and liver function tests, urinalysis, Chest X-ray, Ultrasonography of the abdomen and pelvis, and transthoracic echocardiogram were all normal. Further, her contrast-enhanced computed tomography of Chest-Abdomen-Pelvis, upper and lower gastrointestinal endoscopic examinations did not reveal any significant pathology. Her blood picture revealed marked eosinophilia and bone marrow biopsy showed active marrow with granulocytic hyperplasia with predominant eosinophil lineage. Histology of combined sural nerve and muscle biopsy showed tissue eosinophilia. Her pANCA was positive with negative cANCA. Diagnosis of EGPA was made as she was having unexplained eosinophilia, late onset of asthma, mononeuritis multiplex and extravascular eosinophil accumulation on biopsy with positive pANCA.

She was initiated on treatment with oral Prednisolone 1mg/kg/day for 6 weeks and pulse therapy of IV Cyclophosphamide 15mg/kg monthly for 3 months with MESNA. After the induction of remission, a maintenance dose of azathioprine was commenced with low-dose prednisolone. Her respiratory and neurological symptoms improved with therapy and follow-up at the clinic.

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Discussion

EGPA, previously known as Churg-Strauss syndrome is a rare, progressive small to medium vessel vasculitis that can lead to death due to multi-organ failure. It manifests into a unique sequence of prodromal, eosinophilic, and vasculitic phases. The prodromal phase is usually adult onset and is characterized by atopy, allergic rhinitis, and bronchial asthma. The eosinophilic phase is distinguished by peripheral blood eosinophilia with organ infiltration, especially lung and gastrointestinal tract. Subsequently patient develops life life-threatening systemic vasculitic phase, often associated with vascular and extra-vascular granulomatosis and constitutional symptoms, especially fever, fatigue and weight loss. Asthma is the cardinal feature of EGPA. Patients can have life-threatening cardiac, skin, nervous and gastrointestinal manifestations (3).

Most commonly used criteria for diagnosis is the American College of Rheumatology (ACR). It includes the following six criteria: asthma, eosinophilia of more than 10% (more than 1500/microL), mono or polyneuropathy, transient or migratory pulmonary opacities, paranasal sinus abnormality and extravascular eosinophils accumulation on biopsy. The presence of at least four criteria confirms EGPA firmly. Our patient fulfilled 4 criteria. Patients with EGPA often have eosinophilia of 5000-9000/microL. However, our patient had eosinophilia of 37,580/microL. Markedly high eosinophilia is described in eosinophilic leukemia and idiopathic as well as a malignancy-related hypereosinophilic syndrome (4). 30 to 60 % of patients have positive ANCA with a majority (70 to 75 %) of pANCA.

Commonly using a scoring system to assess the disease activity is the "Five-Factor Score" (FFS), used to guide initial therapy and prognosis. Age >65, cardiac insufficiency, gastrointestinal involvement, renal insufficiency, absence of ear, nose and throat manifestations are considered in FFS. This score ranges from 0 to 2. A score of 0 when all factors are absent, a score of 1 for one factor, and a score of 2 for two or more factors. (5) (6). Our patient's FFS score is 1.

Early initiation of immunosuppressive therapy is mandatory for good clinical outcomes. EGPA is treated with Prednisolone doses of 0.5 to 1 mg/kg/daily for 6 to 12 weeks, or until remission. Glucocorticoid is combined with Cyclophosphamide for induction of severe disease (FFS >2), FFS of 1 (especially with cardiac or central nervous system manifestations) or FFS of 0 with positive ANCA as these patients tend to develop multi-organ failure. After induction of remission, Prednisolone was gradually tapered, and maintenance therapy with Azathioprine was commenced as a steroid-sparing agent and continued for 12 to 18 months. The disease has a risk of relapse hence close follow-up is necessary (6).

Conclusion

This case illustrates the fact that treating clinicians to always consider atypical presentations of rare diseases to avoid diagnostic delays of fatal disorders.

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