Case Report

Hypereosinophilic Atopic Transverse Myelitis

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Atopic transverse myelitis is a rare disorder that is defined as a localized myelitis of an unknown cause in patients with either high immunoglobulin E (IgE) level or mite-specific IgE or coexistent atopic disease. It is a cause of intramedullary cord lesions, but its diagnosis does not need tissue confirmation. We report a case of a patient who presented with bladder and anal incontinence, paresthesia, and lower limb weakness. Neither IgE level nor mite-specific IgE level could be determined due to lack of fund; however, magnetic resonance imaging (MRI) of the thoracolumbar region showed hypotense-isotense lesion within the spinal cord at T4 vertebral level, suggestive of transverse myelitis. Blood cell count showed hypereosinophilia. Therefore, a diagnosis of atopic transverse myelitis based on high eosinophil count and MRI was made. Patient was commenced on prednisolone and had good response to treatment. Complete blood count is a cheap simple diagnostic tool in resource-poor country to distinguish atopic transverse myelitis from other causes of intramedullary cord lesions.

Keywords: Atopic transverse myelitis, blood count, eosinophilia

INTRODUCTION

The diagnosis of the different intramedullary cord lesions appears very difficult for the neurosurgeon.¹ The availability of simple diagnostic procedures to rule out some of these intramedullary cord lesions will preclude the need for tissue confirmation which could be invasive and predispose the patient to complications.

Transverse myelitis is referred to as a group of intramedullary inflammatory disorders of the spinal cord, which could be of acute or subacute onset. The etiology is principally immune based.¹ Diagnosis is based on the results of spinal cord magnetic resonance imaging (MRI), cerebrospinal fluid cell count, and protein concentration, electrophysiological studies which include somatosensory evoked potentials and motor evoked potentials in the upper and lower limbs and immunological studies; total and allergen-specific IgE levels in the serum.² The frequency of eosinophilia in the peripheral blood of patients with atopic myelitis was reported to be at least 4 times what is observed in other myelitis due to other causes.¹

Eosinophil is one of the polymorphonuclear granulocytes that are derived from stem cells in the bone marrow. They contain cytoplasmic granules which when released may provoke toxic damage to adjacent tissue.¹⁻⁴ Peripheral blood eosinophilia can be caused by numerous allergic, infectious, and neoplastic disorders, which require a variety of different treatments. Eosinophilia is a condition, in which there is increase in number of eosinophils in the peripheral blood.⁴ The absolute eosinophil count is between 40 and 440/mm³. The degree of eosinophilia is rarely helpful for identifying the cause, except at extremes of eosinophil counts; however, we describe a case of atopic transverse myelitis with eosinophilia.⁵

CASE REPORT

A 15-year-old male patient presented with 3 weeks history of fever, bladder and anal incontinence, paresthesia, and worsening lower limb weakness. Fever was high grade

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How to cite this article: Fasola FA, Aworanti OW. Hypereosinophilic atopic transverse myelitis. Niger J Clin Pract 2018;XX:XX-XX.
Fasola and Aworanti: Eosinophilia in atopic myelitis

and persistently high until presentation. No associated headache, however, there was malaise, chills and rigors. Two days after the onset of fever, the patient initially had difficulty with passing stool and urine, despite the urge to do so and later developed bladder and anal incontinence. He also had low back pain with initial numbness in the lower limbs and paresthesia on the sole of the feet and subsequently developed bilateral lower limb weakness. There was no history of fall or trauma to the back, no associated cough, or contact with someone with chronic cough. No significant drug history or history of skin rashes and rhinitis.

Examination at presentation showed an anxious young man, not pale, anicteric but febrile with temperature of 39.8°C without significant peripheral lymphadenopathy. Chest, cardiovascular and abdominal examination did not reveal significant findings. Central nervous system examination showed no cranial nerve abnormality, no sign of meningeal irritation. The pupils were 3 mm dilated bilaterally and reactive. There were hypotonia and hyporeflexia in the right and left lower limbs. The power was one over five in both lower limbs with flat plantar response; the sensory level in the patient was T6 vertebral level. Allergy was not reported by the patient.

The following were the results of investigations carried out: complete blood count; hematocrit (hct) –37%, white cell count (WCC) –29,200/mm³, and platelet count (Plt ct) –511,000/mm³. White cell differentials were as follows: neutrophil; 47.4%, eosinophil; 32.6%, lymphocyte; 14.2%, and monocyte; 5.7%. The absolute eosinophil count was 9,344/mm³. The absolute value of other cells is as shown in Table 1. Review of peripheral blood film showed moderate microcytosis, mild macrocytosis, moderate hypochromia, leukocytosis with eosinophilia, and adequate platelet count of approximately 325,000/mm³. The erythrocyte sedimentation rate (ESR) was 84 mm/h⁻¹ (Westergreen), and coagulation profile was essentially within normal range. The patient was nonreactive to human immunodeficiency virus 1 and 2 and also negative for hepatitis B surface antigen and anti-hepatitis C virus. Antinuclear antibody was negative. Stool microscopy and blood culture did not yield any organism.

MRI of the thoracolumbar region showed hypotense-isointense lesion of about 0.8 cm in diameter within the spinal cord in contrast at the T4 vertebral level, which was suggestive of transverse myelitis.

An assessment of atopic transverse myelitis was made, the patient was asked to do further tests including serum immunoglobulin E (IgE) level, but he could not do most of the investigations because of cost. The patient was empirically treated with antibiotic and antimalarial. The patient did not receive anthelminthic because stool microscopy did not identify any parasite. He was commenced on tablet prednisolone at 60 mg/day into two divided doses. On Day 5 on prednisolone, the repeated complete blood count showed a reduced eosinophil percentage of 20% of the total WCC, other differentials are as shown in Table 1. At this time, most of the abnormal neurological presentations were resolving – bladder, and anal continence was restored while power in the lower limbs has improved to 3/5. On day 10 on prednisolone, the power had increased to 5/5 in the lower limbs, and the eosinophil percentage was 08%, other cell differential is as shown in Table 1. Figure 1 shows the trend of response of eosinophil percent while the patient was on prednisolone. The prednisolone was tapered down to 30 mg/day and the patient was discharged home but lost to follow-up.

**DISCUSSION**

This case report aims to underscore the benefit of simple routine complete blood count in aiding diagnosis and

![Figure 1: Response of eosinophil percent to prednisolone use in the patient with atopic myelitis](http://www.njcponline.com)
reducing cost in a resource-poor country. Eosinophilic or atopic transverse myelitis is a rare disorder with most reports from Japan. It is defined as localized myelitis of an unknown cause in patients with either high IgE level and mite-specific IgE or coexistent atopic disease. Working within the context of this definition, the diagnosis of this disease entity in our patient could not have been established if not for the presence of eosinophilia which is a normal immune response to allergy and parasitic infections and other suggestive clinical features. At the time of presentation, the patient did not have coexistent atopic disease. By the time, the diagnosis of atopic myelitis was considered, the patient had exhausted fund to determine IgE level; however, this diagnosis was upheld since there has been report of atopic myelitis in the absence of a typical history of atopy and normal total IgE levels. A combination of clinical and laboratory findings are required to make a diagnosis.

Our patient falls within the age group of reported cases and presented with a range of neurological signs and symptoms similar to that which has been reported. Atopic myelitis is more common in male sex such as found in our patient. He, however, presented within 3 weeks, which is against the other cases reported in which the duration of illness before diagnosis was between 3 and 13 months. Rather than the subacute/chronic onset of the disease features reported in literature, our patient presented with an acute disease. Acute onset has been reported in 24.3% of patients. Though fever is an uncommon feature in patients, it has also been reported. In the absence of an identifiable organism from blood culture, the fever could have been part of an immune response to an allergic process. The finding on MRI of the thoracolumbar region which was suggestive of transverse myelitis in addition to the finding of blood eosinophilia was highly suggestive of atopic myelitis despite not being able to determine the IgE level. A lesion at the T4 vertebral level is in support of previous reports in which the cervical and thoracic spinal cords were the most affected though the observed cauda-equine like presentation is not expected in a T4 lesion. The cauda equine-like presentation could be as a result of spinal/neuronal shock considering the early presentation of the patient. The simultaneity of development of neurological symptoms and fever in a patient with eosinophilic leucocytosis suggested a common mechanism. Attesting to this is the observed dramatic reduction in blood eosinophil count with the use of steroid and corresponding rapid improvement in clinical status of the patient as demonstrated by normal neurological status within 10 days of treatment. The improvement is consistent with the response in patients described by Osogawa et al. and Lee JY et al.

Parasitic infection, fungal infection, sarcoidosis, and Churg-Strauss syndrome are thought to be associated with eosinophilic myelitis. The pathophysiology of the neurological disorder is not very clear; however, Osogawa et al. observed eosinophil infiltration on biopsy and postulated that inflammation and infiltrated eosinophil contribute to anatomic and functional neural damage. The etiology of the disease could not be ascertain in our patient due to the limitation of fund, a search should have been made of mite-specific anti-IgE.

Blood eosinophilia signifies either a cytokine-mediated reactive phenomenon (secondary) or an integral phenotype of an underlying hematological neoplasm (primary). Secondary eosinophilia is usually associated with parasite infection in the third world countries and allergic conditions in the West. The percentage eosinophil in this patient was as high as 32% of the total white blood cell count of 29,200/mm³, far higher than the percentage eosinophil reported in other cases which ranged between 3% and 5.9%. High blood eosinophil counts in humans are detected in approximately 10% of the population. Eosinophilia is operationally classified into secondary, clonal, and idiopathic types. Causes of secondary eosinophilia which is cytokine-mediated reactive phenomenon include parasite infections, allergic or vasculitis conditions, autoimmune process, drugs, and nonmyeloid malignancies. Clonal eosinophilia is distinguished from idiopathic eosinophilia by the presence of histologic, cytogenetic, or molecular evidence of an underlying myeloid malignancy.

Distinguishing the benign secondary causes from the rare but more serious idiopathic hypereosinophilic syndrome (HES) and malignant disease could be a difficult task. This patient had hypereosinophilia. Hypereosinophilia is defined as moderate-to-severe eosinophilia (i.e., ≥1500 eosinophils/μL). A secondary cause could not be completely eliminated within the limit of available resources since the patient did not have chest radiography, antiment specific IgE, serologic tests for suspected pathogens (Strongyloides stercoralis, Schistosoma spp, Toxocara species, and filarial) as stool microscopy for ova and parasite could not detect all parasites. Within the context of available information on clinical features, idiopathic eosinophilia is an unlikely diagnosis. Idiopathic eosinophilia is defined by the presence of a peripheral blood eosinophil count of 1.5 × 109/L or greater for at least 6 months. The widespread increase in all other inflammatory cells suggests an ongoing inflammatory process which may not necessarily be infective. HES is a subclass of idiopathic eosinophilia with eosinophil-mediated organ damage. The elevated ESR further suggested an inflammatory process in the patient.
The gradual decline in eosinophil reduced from 32% to 20% in the first 5 days, and to 8% within 10 days, is consistent with the results obtained by Osoegawa et al. and Lee JY et al.[6,7] for patients on steroid therapy.

**CONCLUSION**

As part of the workup for patients with transverse myelitis, a complete blood count with peripheral blood film review is imperative; this is to aid the diagnosis of eosinophilic or atopic transverse myelitis to avert the need for invasive cord biopsy.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**


