A Rare Case of Polyneuropathy

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PRESENTATION OF CASE

A 46 years of rt handed Hindu grocer male with history of nasal polyp, allergic rhinitis for 12 years and off and on cough was admitted at Tripura Medical College & Dr. B.R.A.M. teaching hospital with chief complaints of tingling and numbness of extremities for 45 days and weakness of the lower limbs for 30 days. Forty five days before hospitalization he developed decreased sensation over both feet which progressed up to both legs below knee level over few days and was associated with burning, tingling, pins and needle sensation. Simultaneously he developed weakness of the lower limb, unable to maintain slippers, difficulties in standing up from squatting and required assistance of family member to walk. From the day of admission he developed decreased sensation over left hand then right hand with similar upward progression. He developed weakness of upper limbs in holding cup & raising hand arm above head. There was no history suggestive of involvement of any cranial nerves.

There was no history of convulsion, loss of consciousness, headache, neck pain, radiculopathy, stiffness, itching, joint aches, swellings. In his past he was treated with montelukast, inhaled bronchodilators, and intranasal corticosteroids of and on for 08 years. He was on self-medication with Montelukast for last 10 years. There was No history of diabetes, hypertension, and tuberculosis. He had no history of any high-risk behaviour, no addiction. His bowel & bladder habit was normal. He has four family members all are healthy. He is allergic to dust and had off and on malaise, feverishness with intermittent non pruritic rash over legs. He is alert, co-operative, well oriented with time, place & person. Having BMI (21.08). His vitals were stable.

Central nervous system examination shows he is conscious, GCS scale 15/15, MMSE- 30/30. Appearance, behaviour, speech, language is normal. No evidence of hallucination, delusion. All twelve pairs of cranial nerves examination were found to be normal. Muscle bulk symmetrical on either side. No evidence of atrophy, wasting, fasciculation, tremors, no involuntary movement. Tone decreased in all group of muscles in both upper & lower limbs. Power around shoulder, elbow are 4+/5 and power around the hip joint, knee joint, ankle joint respectively are 4/5, 4/5, 3/5. Small muscles of hand & foot are both weak. Superficial reflexes are normal. Planter reflex is bilateral flexor. Deep tendon reflexes in upper limb triceps, biceps & supinator are 1+/4, 1+/4 & 0 bilaterally. In lower limb knee reflexes are bilaterally reduced to 1/4. Ankle reflexes were absent bilaterally. No evidence of presence of any primitive reflexes. There was evidence of loss of pain, touch, temperature up to knee & wrist. Vibration & join position was lost up to knee & wrist. There was no evidence of cerebellar involvement. Gait was stepping gait. Romberg's sign was positive. Cranium & spine were normal and there were no signs of meningeal irritation.

In respiratory system examination nasal polyp was seen in left side & there was paranasal sinus tenderness over left maxillary sinus. All quadrant of lung were equally resonant. In auscultation vesicular prolonged expiration was heard in all quadrants of lung. Bilateral polyphonic wheeze heard. Cardio vascular system and gastro intestinal were essentially normal.

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DOI: 10.18410/jebmh/2020/110

Financial or Other Competing Interests: None.

How to Cite This Article:
Debnath G, Chakraborty A, A Rare Case of Polyneuropathy. J. Evid. Based Med. Healthc. 2020; 7 (10), 508-510. DOI: 10.18410/jebmh/2020/110

Submission 10-02-2020,
Peer Review 16-02-2020,
Acceptance 25-02-2020,
Published 09-03-2020.

CLINICAL DIAGNOSIS

Acute Sensory Motor Polyneuropathy with Bronchial Asthma and Rhinosinusitis.

DIFFERENTIAL DIAGNOSIS

- Chronic inflammatory demyelinating neuropathy
- Vasculitis
- Toxic/ drug induced
- Paraneoplastic

PATHOLOGICAL DISCUSSION

Complete blood count shows haemoglobin 12 gm%, TLC 28200/cumm. Eosinophil 51%. Absolute eosinophil count 14382/cmm. ESR 43 mm 1st hour. Blood sugar fasting & post-prandial are respectively 110 & 140 & HbA_1c 5.5%. Liver function test with in normal limit. Urine examination shows protein 1 +, 24-hour urinary protein 450 mg. HBsAg, Anti HCV, ICTC is negative. ANA is negative. Chest x ray is with in normal limit. Pulmonary function test shows obstructive pattern with reversibility of 18%.

Figure 1. X-Ray PNS Waters View Shows Left Nasal Polyp

Figure 2. CECT Thorax Shows Peribronchial Infiltrate and Inter Alveolar Thickening

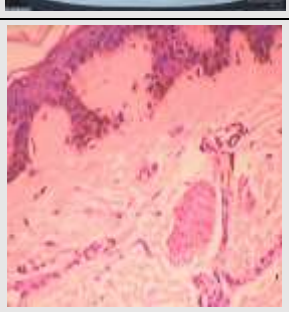

Figure 3. Skin Biopsy

Skin biopsy shows unremarkable epidermis. Dermis shows prominence of blood vessels with perivascular infiltrate of eosinophils with few polymorphs &lympho-histocytes. No necrotizing granuloma seen. Extravasated RBC seen around the blood vessels. Serum IgE was 1801.1/ml, pANCA was positive. NCV all four limbs show Motor sensory polyneuropathy.

DISCUSSION OF MANAGEMENT

In view of evidence of vasculitis with high IGE, marked eosinophilia, PANCA positive diagnosis of Eosinophilic granulomatosis with polyangiitis was made. As the patient showed evidence of progressive peripheral neuropathy, aggressive immunosuppressive in the form of pulse prednisolone & cyclophosphamide was planned.

Treatment given-

- Tab prednisolone 1 mg/kg/day given.
- Cyclophosphamide 2 mg/kg/day given for 3 months then 1.5 mg/kg/day given for 3 to 6 months.

Patient responded to treatment, his eosinophilia and hyper IgE subsided as do his neuropathy. After six months he had minimal weakness and could do all activities of his own without assistance.

DISCUSSION

Formerly known as the Churg–Strauss Syndrome. It is a specific variant of small and medium sized systemic blood vessels characterized by the coexistence of asthma, rhino sinusitis, and the presence of peripheral eosinophilia.[1] It was first described in 1951 By Dr. Jacob Churg & Dr. Lotte Strauss as a syndrome consisting of asthma eosinophilia, fever and accompanying vasculitis of various organ systems.[1] EGPA is diagnosed with the presence of at least four out of six below mentioned criteria as per American college of rheumatology in 1990.[2]

1. Asthma
2. Eosinophilia(>105 Of DLC)>1500/cumm
3. Presence of mononeuropathy or polyneuropathy
4. Transient pulmonary infiltrates
5. Presence of paranasal sinus abnormalities
6. Histological evidence of extravascular eosinophils.

Neuropathy in EGPA is evident in the form of wrist or foot drop resulting from mononeuritis multiplex, or mixed sensory-motor peripheral neuropathy is reported. Common peroneal and internal popliteal nerves are most commonly involved.[3] Peripheral neuropathy can be present in 75% to 80% cases.[4] Peripheral blood eosinophilia (greater than 1500/dl) is the best-known lab hallmark of the disease. Elevated serum IgE is also found in 75% of patients.[5] ANCA positivity in EGPA is seen in approximately 40% of cases. The common pattern is pANCA[6] Patients with EGPA are treated with:[7] Oral prednisolone 1 mg/kg daily then taper, or intravenous methylprednisolone pulse followed by oral prednisolone, Cyclophosphamide 2 mg/kg for 3 to 6 months, Cyclophosphamide pulses 600 mg/m^2. Other medications include Methotrexate, Cyclosporin A, Azathioprine and Mepolizumab.

Our patient fulfilled all the six criteria of EGPA including peripheral sensory motor neuropathy. We are reporting this case because of its rarity with only few case reports of neuropathy in EGPA.

FINAL DIAGNOSIS

Eosinophilic Granulomatosis with Polyangiitis (EGPA) with Peripheral Neuropathy.

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