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Eosinophilic Granulomatosis with Polyangitis (Churg Strauss Syndrome) presenting as Guillain Barre Syndrome (GBS)- A Case Report

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Abstract
Churg Strauss Syndrome (CSS) is a rare ANCA Associated Vasculitis (AAV) causes disseminated necrotizing vasculitis in patients with eosinophilia and Asthma. Initial presentation as Neuropathy is quite rare (15%).Usually Mono neuritis multiplex is the common presentation of Vasculitic neuropathy. Acute Neuropathy with GBS like initial presentation is extremely rare. There are only 6 case reports in literature. Here we present a 57 yr old male with history of asthma presenting with acute onset and progressive weakness of lower limbs followed by upper limbs with painful tingling and paresthesia over 3 weeks duration. On examination there was areflexic flaccid weakness of lower limbs and upper limbs both proximal and distal with distal sensory loss without any cranial nerve involvement. Investigations showed severe eosinophilia with proteinuria and urinary casts. Vasculitic workup showed high titre P-ANCA positivity. Nerve Conduction Studies showed severe axonal and demyelinating type of neuropathy effecting all four limbs. In view of vasculitis involving target organs patient was treated with pulse steroids and cyclophosphamide according to EUVAS Regimen. Patient showed good improvement in neurological deficits and proteinuria and is under regular follow up.
Keywords: *Churg Strauss Syndrome GBS ANCA Associated Vasculitis Nephritis Asthma.*

Introduction

Churg-Strauss syndrome (CSS) or Eosinophilic Granulomatosis with Polyangitis (EGPA) is a rare form of systemic vasculitis occurring in patients with asthma and eosinophilia. It causes vasculitis effecting small, medium vessels and venules with multisystem involvement. It is classically associated with antineutrophil cytoplasmic antibodies (ANCAs), was first described in 1951 by Churg and Strauss. EGPA can affect multiple organ systems, including the cardiac, pulmonary, renal, nervous, and vascular systems.

Pheripheral Nervous system involvement is noted in about 60% of patients, mainly in the form of mononeuritis multiplex or symmetric mild sensory axonal neuropathy.[1,2,3-6] The neuropathy is caused mainly by nerve ischemia due to occlusion of vasa nervorum.

The American College of Rheumatology (ACR) has proposed six criteria for the diagnosis of Churg Strauss syndrome[7]. The presence of four or more criteria yields a sensitivity of 85% and a specificity of 99.7%. These criteria include (1) asthma (wheezing, expiratory rhonchi), (2) eosinophilia of more than 10% in peripheral

blood, (3) paranasal sinusitis, (4) pulmonary infiltrates (may be transient), (5) histological proof of vasculitis with extravascular eosinophils, and (6) mononeuritis multiplex or polyneuropathy During the vasculitic phase, besides renal and pulmonary disease, central and peripheral nervous system involvement may occur, with Polyneuro-pathy, radiculopathy, and mononeuritis multiplex being common presentations 8. Differentiating between GBS and EGPA-associated vasculitis Polyneuropathy is important, considering the different management strategies.

Case Report

57 year old male, with history of well controlled Bronchial Asthma presented with 3 weeks history of progressive weakness of both lower and upper limbs. Weakness started in the lower limbs distally 3 weeks back spread proximally over a week. Since last week patient developed weakness of upper limbs distally .This was associated with tingling and numbness and paresthesia of both lower and upper limbs. Weakness was severe enough to make him bed bound. There was no associated Neck pain, bladder or cranial nerve symptoms. There was history of on and off fever, along with weight loss for one month. No history of recent infections, vaccination, drug or toxin exposure. On examination he had grade 2 power of both lower limbs both proximally and distally. Upper limb had gr 3 power distally and 4-proximally. All reflexes were lost with distal sensory loss in lower limbs upto knees diffusely. No muscle tenderness. Cranial nerves including fundi were normal. Thus a possibility of Acquired acute Neuropathy like Guillain Barry Syndrome was considered. In view of systemic symptoms secondary causes like Vasculitis or paraneoplastic etiology was also kept in the differentials. Investigations revealed total count of Hb-10.2gm/dl 46,100 with eosinophils (65%), Absolute eosinophil count was 29965, ESR-70 mm hr, Renal functions were mildly deranged-B Urea-63,S.Creatinine-1.7.S.CPK was normal. Urine routine examination showed RBC and granular casts.24 Hour Urine protein was 1744.8mg/day. Viral Markers like Hbsag, HIV and antiHCV was negative. Nerve conduction study was done which showed severe axonal and demyelinating type of motor and sensory Neuropathy effecting both upper and lower limbs.CSF Study was done which was normal and did not reveal any albumino-cytological dissociation. Vasculitic and connective tissue disease workup was done, ANA Profile was negative, C –ANCA was negative. P-ANCA was high titre positive >100U/ml. S.IGE was negative. Paraproteinemia workup was negative. CT Chest was done in view of history of asthma and suspected vasculitis which showed patchy lower lobe lung infiltrates. In view of Eosinophilia, History of Asthma, Renal involvement ANCA strongly positive and lung infiltrates a diagnosis of Churg Strauss syndrome was made. Sural nerve biopsy and Renal biopsy was not done as the patient and relatives were not willing for the same. The Neuropathy was a manifestation of Vasculitic Neuropathy related to the systemic ANCA Associated vascultits. In view of major organ threatening damage the patient was treated with Pulse Methyl Prednisolone for 3 days along with cyclophosphamide .Patient was followed up with Pulse cyclophosphamide every 3 weeks. There was significant improvement in weakness over initial 2 weeks, Patient could walk with support. Total WBC count reduced to 14300 with 1% eosinophils. Urine routine examination became normal. Systemic symptoms subsided in 1 week. Repeat CT Chest done after 2 weeks showed complete disappearance of the chest infiltrates.

Patient is under follow up for past 3 months and has shown significant improvement in his systemic, Neurological and renal parameters.

Discussion

The clinical features of Churg-Strauss Syndrome (CSS) typically develop in several sequential phases, although these phases are not always clearly distinguishable.[9-10] The prodromal phase is characterized by airway inflammation

presenting with allergic rhinitis and asthma. The second phase is marked by the presence of Eosinophilic infiltrative disease which include peripheral blood eosinophilia and Eosinophilic infiltration of multiple organs, especially the lung and gastrointestinal tract.

Finally, the third and final phase, which can be life threatening, consists of systemic medium- and small-vessel vasculitis with granulomatous inflammation which manifests with Polyneuropathy and strokes. The vasculitic phase may be heralded by nonspecific constitutional symptoms and signs, especially fever, weight loss, malaise, and lassitude. This phase usually develops within 5 years of the onset of asthma, although it may be delayed for several decades. Pathological findings of Vasculitic neuropathy are characterized by axonal degeneration of nerve fibers caused by vasculitis-induced ischemia.

Our patient had history of asthma but was exceptional because the presenting manifestation was Vasculitic Neuropathy and subclinical Renal involvement. However the presence of systemic features and history of asthma with severe eosinophilia led us to investigate the patient for vasculitis leading to the diagnosis of CSS.

Our patient presented with acute ascending neuropathy mimicking GBS like presentation. This is a rare situation with only 6 case reports mentioning such presentation. EGPA and other vasculitides should always be part of the differential diagnosis of GBS, as the first line treatments may differ. While steroids are of no use (and may even be harmful) in GBS, they are the mainstay of treatment in EGPA[(8)]. Adding cyclophosphamide could reduce recurrence of EGPA and may be life-saving when there is multiple organ involvement. On the other hand, IV immunoglobulin and plasmapheresis maybe effective in both EGPA and GBS .Thus a diagnosis of vasculitis early in GBS like presentation is vital in starting initial treatment to prevent major organ damage and also planning of long term therapy and prognosis.

Generally, systemic symptoms and the involvement of organ systems in EGPA are mediated by three major mechanisms: the presence of autoantibodies within organ systems, specific or nonspecific organ damage by inflammatory mediators, and antiphospholipid-related hypercoagulability and thrombosis [11-13].

Peripheral neuropathy is quite common in patients with EGPA. Pathological findings of vasculitic neuropathy are characterized by axonal degeneration of nerve fibers caused by vasculitis-induced ischemia. In contrast to lung or renal involvement, peripheral neuropathy alone is rarely life threatening but does significantly affect quality of life.

Treatment regimen is based on Five Factor Score (FSS) highlighting organ involvement[(14)]. FFS includes the following five factors: 1. Cardiac involvement, 2. Gastrointestinal (GI) disease (bleeding, perforation, pancreatitis), 3. Renal insufficiency (Creatinine > 1.6 mg/dl), 4. Proteinuria (> 1gm/day) and 5. Central nervous system (CNS) involvement (mononeuritis, polyneuropathy). An FFS of 0 connotes a 12% five year mortality rate; FFS of 1, a 26% five-year mortality rate and FFS >3, 46% five-year mortality rate.Our patient had FFS 3 thus high mortality rate. Hence required cyclophosphamide pulse along with Intravenous Methyl Prednisolone pulse 1Gm/day for 3 days followed by oral Prednisolone 1 gm/day to be continued for 2-3months followed by gradual taper. Initial Induction therapy is for 3-6 months and consists of steroids with or without oral cyclophosphamide 2mg/kg or Intravenous cyclophosphamide 15mg/kg every 2-3 weeks for 3-6 months. This is followed by maintainenece therapy which consists of Immunosuppressive agents like Azathioprine/ Methotrexate(1^{st} line) or Mycophenolate/ cyclosporine(2^{nd} line) for 18-24 months. Oral steroids may be stopped or continued in lowest possible dosage.

Our patient improved very well in neurological functions and was able to walk without support in 3 weeks time .His renal parameters improved as

well as the urinary casts and proteinuria disappeared in 3-4 weeks. Eosinophilia and systemic features started to disappear in a week's time. Thus timely diagnosis could lead to a good outcome with reversal of major organ damage.

Conclusion

Churg Strauss syndrome can present with GBS like illness. Early diagnosis of underlying vasculitis in GBS like presentation can help in initiation of early and appropriate intensive immunosuppressive therapy and can lead to good outcomes.

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