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Original Research Paper

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A CASE OF EOSINOPHILIC GRANULOMATOSIS WITH POLYANGITIS IN A 32 YEARS OLD MALE AT TERTIARY CARE CENTRE

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ABSTRACT The author presents a case of Churg-strauss syndrome with an exuberant clinical picture in a 32 year old male. He showed the following diagnostic criteria: asthma, polyneuropathy, rhinitis, marked eosinophilia and skin histopathology results suggestive of vasculitis with eosinophilia. There was a good response to prednisolone and cyclophosphamide.

Eosinophilic granulomatosis with polyangitis (Churg-strauss), abbreviated EGPA, which was previously called the churg-strauss syndrome (CSS) or allergic granulomatosis and angitis, is a multisystem disorder characterized by chronic rhinosinusitis, asthma and prominent peripheral blood eosinophilia(1,2,3,4).EGPA is classified as a vasculitis of the small and medium size arteries, although the vasculitis is often not apparent in the initial phase of the disease.

The most commonly involved organ is lung, followed by skin. EGPA, however can affect any organ system, including the cardiovascular, gastrointestinal, renal, and central nervous systems. Vasculitis of extra pulmonary organs is largely responsible for the morbidity and mortality associated with EGPA.

KEYWORDS :

Case Report

A 32 year male was admitted to CHA trauma center with chief complain of black stool since 2 days and development of macular erythematous rash involving bilateral lower limbs and abdomen since 1 day.

The patient was known case of bronchial asthma since childhood and patient was put on MDI with long acting inhaled beta-agonist and steroids since 20 years. Patient was also taking oral steroids since 20 years which he had stopped since 1 year.

On examination his vitals were normal, respiratory examination revealed scattered rhonchi over both lungs, macular erythematous rash on bilateral lower limb and abdomen and rest of examination was unremarkable.

Initial blood investigation revealed : hemoglobin-12.4 g/dl, lecukocyte-49000cells/mm^3 with absolute eosinophil count->32000 cells/mm^3,urea-28 mg/dl, creatinine-0.84 mg/dl, ESR-105 mm/hr, Liver function Test-normal, urine analysis was normal with no cast, active sediments and no proteinuria,stool routine micro was normal and stool occult blood- positive. His ultra sonogram for abdomen and pelvis was normal. He was tested negative for cytoplasmic-antineutrophil cytoplasmic antibody(c-ANCA) and perineuclear-antineutrophil aytoplasmic antibody (p-ANCA).

Chest X-ray was suggestive of prominent bronchovascular markings.On HRCT-thorax few scattered pulmonary infiltrates were present. Pulmonary function test was suggestive of obstructive lung disease with reversibility with short acting beta-2 agonist.CT angio abdomen was unremarkable.

The skin biopsy from lesions came positive for eosinophilic and neutrophilic infiltration at vessel wall suggestive of leukocytoclasticvasculitis.

Nerve conduction velocity was done which suggested asymmetrical involvement of nerves of both upper and lower limb for axonal and demyelinating sensory and motor polyneuropathy suggestive of mononeuritis multiplex.

Thus patient was diagnosed as churg-strauss syndrome based on peripheral eosinophilia with skin biopsy suggestive of leukocytoclasticvasculitis, eosinophilic infiltrates around blood vessels; this was supported by long standing history of asthma as well as other organ system involvement.

Patient was treated with 3 doses of cyclophosphamide 500mg 15days apart and methylprednisolone 1gm for 3 days followed by oral steroids 40mg/day. Patient improved well as rash subsided and gastrointestinal bleeding was controlled and he was discharged on oral steroids at 20mg/day.

Other differential diagnosis such as Aspirin Exacerbated Respiratory Disease, Chronic Eosinophilic Pneumonia, and Allergic Bronchopulmonary Aspergillosis (ABPA) were ruled out in view of extrapulmonary organ system involvement.

Other vasculitis such as Granulomatous Polyangitis and Microscopic Polyangitis can involve lung but degree of eosinophilia and presence of asthma was characteristic for EGPA.

DISCUSSION

Churg-strauss syndrome was discovered by Churg and Strauss in 1951.

It is rare vasculitis with asthma and peripheral tissue eosinophilia with an incidence of 0.5-2.7 cases per 1 million, affecting both sex equally and commonly middle-aged patients.The incidence of EGPA in male is 0.7 per 100000 population compare to 0.1 per 100000 population in female. It mimics asthma so it is missed.Hence, patients with asthma and other organ involvement and skin lesion should prompt the diagnosis with vasculitis, so early treatment can be initiated.

Churg-strauss syndrome usually begins with respiratory manifestations (asthma or rhinitis) followed by manifestations of vasculitis. Cutaneous manifestations occurs in approximately 70% of cases as: palpable purpura, ecchymosis, hemorrhagic bulla, subcutaneous nodules frequently located on the scalp or distributed bilaterally over the extensor surfaces of the extremities, maculopapular eruptions, urticariform eruptions and livedoreticularis.

Peripheral neuropathy is expressed as a mononeuritis multiplex in 60% of patients. Central nervous system signs such as headache, convulsions, hemiplegia, and brainstem signs are rare.

Although most cases are idiopathic, inhaled antigens, vaccinations, desensitization, infections and suppression of oral corticoids have been considered as a triggering factors.
The probable pathogenesis involves immediate hyperse nsitivity and cytotoxic reactions with activation of Th2 lymphocytes, followed by degranulation of mastocytes and eosinophilia and activation of ANCA dependent neutrophils.

It develops in sequential phases:

1) **Prodromal Phase:**

Characterized by atopic disease, allergic rhinitis and asthma, which mostly occurs in 2nd and 3rd decade.

2) **Eosinophilic Phase:**

Peripheral blood eosinophilia with eosinophilic infiltration involving many arteries, lungs (as an eosinophilic pneumonia) and gastrointestinal tract (as gastroenteritis).

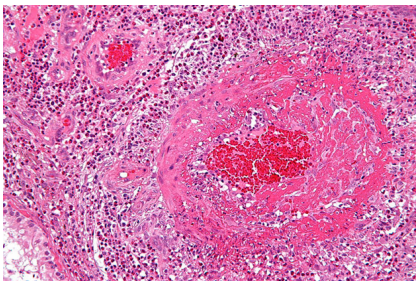
3) **Vasculitic Phase:**

Systemic vasculitis with granulomatous inflammation is characteristic of the disease. Patient can develop life threatening sequel which is due to vasculitis of the organs involved.

Criteria For Diagnosis:

In 1990 the ACR (American college of rheumatology) established six criteria for classification of EGPA in a patient with documented vasculitis[5] The presence of four or more of these criteria had a sensitivity of 85% and sensitivity of 99.7%:

1) Asthma(a history of wheezing or finding of diffuse high pitched wheeze on expiration)
2) Eosinophilia >10 % on differential leukocyte count
3) Mononeuropathy (including multiplex) or polyneuropathy
4) Migratory or transient pulmonary opacities detected radiographically
5) Paranasal sinus abnormality
6) Histological evidence of extravascular eosinophils[6]



Histological Findings:

1) Eosinophilic infiltration of tissue
2) Extensive areas of necrosis
3) Eosinophilic / giant cell vasculitis
4) Interstitial / perivascular necrotizing granulomas

Assessing Vasculitis Severity:

The FFS (five factor score) was initially devised in 1996 and was revised in 2011. IT includes:

1) Age > 65 years
2) Cardiac insufficiency
3) Renal insufficiency (creatinine > 1.7 mg/dl)
4) Gastrointestinal involvement
5) Absence of ENT manifestations (presence is associated with better prognosis)

FFS is scored 0-2(0=no factor, 1= 1 factor, 2= >1 factor)

TREATMENT:

Primary therapy of EGPA is systemic glucocorticoids.

In addition, immunosuppressant such as cyclophosphamide is required in patients with:

1) FFS 2
2) FFS 1 with cardiac or CNS involvement
3) FFS 0 with ANCA positivity (risk of renal complication in future)

Our patient had score 2 so, he was started on injectable steroids with cyclophosphamide.once remission is induced with cyclophosphamide and glucocorticoid therapy (which usually occurs in 6 to 12 months),patients are switched to maintainance therapy with less toxic immunosupressive drugs,usually azathioprine or methotrexate,in combination with tapering doses of glucocorticoids. Recent studies of mepolizumab (anti-IL-5 antibody) in EGPA have been encouraging, but require further investigation[7]

PROGNOSIS:

With current treatment options survival rate is 70-90% at 5 years.of the five factors, cardiac involvement, gastrointestinal disease, and age >=65 appears to be the strongest predictors of poor prognosis[8-9].

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