

Eosinophilic Granulomatosis with Polyangiitis (EGPA) - A Case Review

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Abstract- Eosinophilic granulomatosis with polyangiitis (EGPA) – is a specific variant of the group of diseases characterized by necrotizing vasculitis of small and medium-sized systemic blood vessels. It is highly variable in its presentation and course; the manifestations may range from mild symptoms (asthma, nasal polyps, cutaneous lesions) to life-threatening conditions (severe gastrointestinal involvement, heart disease, disabling multiplex mononeuropathy). The condition has a male predisposition. Here we discuss a 44 year-old male patient who presented with weakness and numbness of both upper and lower limb and EGPA was diagnosed following a comprehensive workup.

Keywords: EGPA, Necrotizing vasculitis, Multiplex Mononeuropathy, Asthma.

INTRODUCTION:

Eosinophilic Granulomatosis with Polyangiitis (EGPA,) formerly known as Churg Strauss Syndrome (CSS) or Churg Strauss disease (CSD) is a rare autoimmune disorder characterized by excess circulating tissue eosinophils and vasculitis, which affects the lung and skin. It is a primary vasculitis characterized by late onset of asthma, multisystem involvement, and eosinophilic vasculitis of small and medium sized vessels. Pulmonary infiltrates and pleural or pericardial effusions due to serositis may be present. It was first described by Churg and Strauss in 1951.

DEFINITION:

Eosinophilic Granulomatosis with Polyangiitis (EGPA) is characterized by necrotizing vasculitis of small and medium sized systemic blood vessels.

SYNONYMS:

- Eosinophilic Granulomatosis with Polyangiitis (EGPA)
- Allergic Granulomatous Angiitis
- CSS
- Churg-Strauss Vasculitis

INCIDENCE AND PREVALENCE:

EGPA is a rare condition with an estimated annual incidence of 1-3 per million. It can occur at any age (mean age of onset is 48 years). Usually, infants are exempted. Male to female ratio is 1.2:1

CAUSES:

- Genetics (Family history of autoimmune diseases)
- Environmental triggers (Chemicals, drugs- Leukotriene receptor antagonist, and infections)
- Abnormal immune response (immune system over react to certain triggers leading to inflammation and damage to blood vessels)
- Eosinophilia

PATHOPHYSIOLOGY:

The pathophysiology of EGPA involves three main stages. These stages may or may not occur sequentially. In some individuals it will not progress through all these three stages.

- Allergic stage: In the first stage, the body undergoes an allergic reaction to an unknown trigger, which causes an increase in the number of eosinophils in the blood.
- Eosinophilic stage: In the second stage, eosinophils accumulate in various organs of the body, such as the lungs, skin, and gastrointestinal tract, causing inflammation and tissue damage. This stage is characterized by the presence of eosinophilic granulomas in affected organs.

- **Vasculitic Stage:** In the third and final stage, inflammation of the blood vessels leads to damage and disruption of blood flow to various organs. This can result in a range of symptoms, such as asthma, skin rashes, peripheral neuropathy, and organ damage.

SIGNS AND SYMPTOMS:

- **Asthma:** Many people with EGPA have a history of asthma, which may be severe and difficult to control with standard treatments.
- **Eosinophilia:** Eosinophils are a type of white blood cell that play a role in the immune response to parasites and allergies. In people with EGPA, there is an abnormally high number of eosinophils in the blood and tissues.
- **Systemic symptoms:** Fever, weight loss and fatigue
- **Skin rash:** Some people with EGPA develop a rash on the skin, typically on the arms or legs.
- **Nerve damage:** EGPA can cause nerve damage, leading to symptom such as numbness, tingling, or weakness in the limbs.
- **Sinus and nasal symptoms:** EGPA can cause chronic sinusitis, nasal polyps, and other symptoms of upper respiratory tract inflammation.
- **Gastrointestinal symptoms:** EGPA can cause abdominal pain, diarrhoea and other GI symptoms.
- **Kidney damage:** in some cases, EGPA can cause damage to the kidneys, leading to the symptoms such as haematuria, proteinuria and decreased kidney function.

DIAGNOSTIC FINDINGS:

The diagnosis of EGPA, can be challenging because the symptoms of this condition can be similar to those of other autoimmune diseases. Common investigations to help diagnose EGPA includes,

- **Blood tests:** A complete blood count (CBC) may show an increase in eosinophils, which is a common feature of EGPA. Other blood tests may also be performed to find out signs of inflammation or autoimmunity.
- **Urine tests:** A urinalysis may be performed to check the signs of kidney damage, which can occur in some cases of EGPA
- **Biopsy:** A tissue biopsy may be performed to look for signs of inflammation and damage to blood vessels. The most common site for a biopsy is the skin, but other organs including nerves may also be biopsied if they are affected.
- **Imaging tests:** Imaging tests, such as X-rays, CT scans, and MRI scans, may be performed to look for signs of inflammation or damage to organs such as the lungs or sinuses.
- **Clinical criteria:** A combination of clinical criteria, that is a history of asthma or other respiratory manifestations eosinophilia and signs of systemic vasculitis, skin rash or nerve damage

MANAGEMENT:

- **Corticosteroids therapy:** Corticosteroids are viewed first line therapy in EGPA. Corticosteroid induced complications are common as a part of treatment such as iatrogenic Cushing's syndrome, osteoporosis with vertebral fracture, GI hemorrhage and infectious complications. Preventive agents such as Bisphosphonates are found to be effective in some patients.
- **Adjuvant treatment with cyclophosphamide:** Cyclophosphamide can be added as adjuvant therapy in relapse cases, patients with substantial vasculitic and organ involvement and patients not responding to corticosteroids.
- **Rescue and alternative therapeutic options:** Rescue and alternative therapies are found to be effective in treatment of refractory EGPA (Plasma exchange Therapy). Intravenous immunoglobulin administration is used for the management of systemic vasculitis.
- **Prognostic category-based treatment regimen:**
 - Patients having sign of good prognosis:
 - ❖ Corticosteroids alone
 - ❖ Prednisone 1 mg kg⁻¹ day⁻¹ for one month. Gradually taper the dose for a year and if disease reoccurs the course should be restarted.
 - Patients having sign of poor prognosis (or with systemic involvement)
 - ❖ Prednisone 1 mg kg⁻¹ day⁻¹ for one month
 - ❖ Cyclophosphamide concurrent with steroids- Oral 2mg kg⁻¹ day⁻¹/ Intravenous pulse 0.6g/monthly

COMPLICATIONS

Even though the disease has a favourable prognosis with early detection and treatment, asthma often remains refractory and impacts the quality of life. Given the prolonged use of corticosteroids, there is a heightened risk of experiencing associated side effects, notably including diabetes mellitus, myopathy, osteoporosis leading to vertebral fractures, and potentially osteonecrosis of the femoral head. Furthermore, it is worth noting that nearly all patients with EGPA

(Eosinophilic Granulomatosis with Polyangiitis) may develop enduring neuralgia and myopathy that are resistant to steroid treatment.

PROGNOSIS:

Prognosis for EGPA is generally good. Early assessments of prognosis included this disorder with polyarteritis nodosa, thus attenuating projected expectations of remission and cure. With the introduction of corticosteroid treatment, remission and survival have improved greatly.

CASE REPORT:

The current report delves into the experience of the patient with EGPA, investigating the process of identification and management through conversations regarding medical history, examinations, and the current handling of clinical issues. This case report aims to build awareness among health care professionals to provide competent nursing care to those affected with EGPA as well as their family.

CASE PRESENTATION:

During a clinical posting, the patient and his wife were interviewed to compile a comprehensive medical history. The patient was admitted to a tertiary care center with primary complaints of weakness and numbness in both upper and lower limbs, along with severe pain and a burning sensation. The symptoms began acutely 20 days prior, with severe pain developing in both lower limbs, followed by weakness after 5 days, accompanied by a burning sensation in both feet. The weakness initially affected the right lower limb, progressing to involve the left within 48 hours. Additionally, he reported experiencing edema in both lower limbs. Initially, he had received intravenous corticosteroids, which led to slight improvement; however, his weakness continued to progress despite this intervention.

PAST HISTORY:

No relevant past history

FAMILY HISTORY:

No relevant family history of autoimmune disease.

INVESTIGATIONS:

Lab investigations of the patient revealed Anaemia, Hyponatremia, Leukocytosis and positive C reactive protein. X-ray shows normal findings. ECG was done for cardiac evaluation and it was normal. Neuro examination revealed decreased pain, touch and temperature sensation in both lower limbs. Both upper and lower limb hypotonia persists, accompanied by bilateral foot drop. In the wrists, there's notable weakness with a grading of G4/5 on the right side and G3/5 on the left side during flexion. Nerve Biopsy from Sural Nerve done under LA and revealed the presence of eosinophilic infiltration accompanied by a mononuclear infiltrate. The motor conduction test was performed on 12 nerves. There were no results within the specified normal range. The sensory conduction test was performed on 14 nerves. The result were normal in left Radial – anatomical snuff box.

CURRENT HEALTH STATUS:

Patient is conscious and oriented. Now patient complaints of worsening of numbness and weakness of both upper and lower limbs.

MANAGEMENT:

The patient received intravenous methylprednisolone 1 g/day, for 5 days, followed by oral prednisone (1 mg/kg per day). In addition, he was administered non-steroidal anti-inflammatory drugs (NSAIDs), opioids, gabapentin, and topical analgesics to alleviate her severe pain.

DISCUSSION:

As mentioned earlier, EGPA has been infrequently documented in Indian medical literature. The index patient is a striking case of EGPA because the patient shows of weakness and numbness in both upper and lower limbs, along with severe pain and a burning sensation and laboratory data revealed leucocytosis and positive C Reactive protein. Following a thorough evaluation, the patient was diagnosed with EGPA at the age of 44, based on careful analysis of clinical and laboratory findings. Pain was the striking feature and required high-dose analgesics and opioids. Now the patient is getting the steroid therapy as well as supportive treatments like topical analgesics.

CONCLUSION:

Patients face many challenges, even before acquiring the diagnosis. symptoms are complex, often non-specific and life altering Future research into the area is vital to ensure an improvement in the quality of life of those affected and to ultimately improve the outcome from those suffering with EGPA.

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