# Eosinophilic granulomatosis with polyangiitis presenting with Mononeuritis Multiplex

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## Introduction:

- Eosinophilic granulomatosis with polyangiitis (EGPA), is an small vessel vasculitis associated with antineutrophil cytoplasmic antibody (ANCA).
- The hallmarks of the disease are asthma, eosinophilia, and systemic vasculitis with varying degrees of neurological, cutaneous, cardiac, gastrointestinal, and renal involvement.
- The pattern of neurological involvement may be mononeuritis multiplex and symmetrical polyneuropathy.

## Case Summary:

### Patient 1:

- A 64 year old male presented with 3 months history of progressive weakness and paresthesia in right hand followed by left hand and right foot.

- On examination he had ulcerations over bilateral upper and lower limbs ,weakness of hypothenar muscles in right hand, thenar muscles in left hand and right ankle dorsiflexion, and hypesthesia in distribution of right ulnar, left median and right peroneal nerves.
- He had history of Chronic Obstructive Airway Disease and also had decreased urine output and facial puffiness.
- **Patient 2:**
- A 33 year old male who was a known asthmatic presented with 1 month history of progressive weakness and paresthesia of right hand followed by left hand. Later he noticed drooping of left eye.
- On examination he had left $3^{rd}$ ,$4^{th}$ and right $7^{th}$ cranial nerve palsy ,weakness of hypothenar muscles in right hand, thenar muscles in left hand and hypesthesia in right ulnar and left median distribution.
- Additionally he had breathlessness on exertion, palpitations and had history of episodes of abdominal pain.

| Clinical criteria | Score |
|---|---|
| Obstructive airway disease | +3 |
| Nasal polyps | +3 |
| Mononeuritis multiplex | +1 |
| **Laboratory/biopsy criteria** | |
| Blood eosinophil count ≥1000 cells/μL | +5 |
| Extravascular eosinophilic-predominant inflammation on biopsy | +2 |
| Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies | −3 |
| Hematuria | −1 |

## Investigations and Treatment:

- Investigations shown moderate eosinophilia ,elevated IgE levels and elevated CRP in both the cases.
- In first case - RA factor and p ANCA was positive and elevated blood urea and creatinine was seen.
- In second case -CECT shown bilateral GGOs, EBUS with FNAC – Reactive lymphoid tissue. and 2d Echo shown global LV hypokinesia with EF 30 %. CEMRI Brain shown small vessel ischemic changes.
- NCS suggestive of Mononeuritis Multiplex pattern in both the patients. Nerve biopsy shown inflammatory changes.
- Other causes of MNM has been excluded.
- In view of asthma, eosinophilia and multiorgan involvement EGPA was considered and Steroids were given .Injection Rituximab was given in second case.
- Both the patients responded well to treatment.

## Discussion:

- In EGPA ,PNS manifestations like Mononeuritis multiplex are more commonly seen. Other include Plexitis and distal sensorimotor polyneuropathy.
- Meningitis is the most frequent central manifestation.
- Treatment includes high dose IV MPS followed by oral prednisolone taper with the addition of Rituximab or Cyclophosphamide.
- Mepolizumab ,a monoclonal antibody targeting IL-5 has been approved for EGPA.

## Conclusions:

- Diagnosis of EGPA is often difficult as the symptoms are diverse, history of asthma with blood eosinophilia and multiorgan involvement are the important clues to suspect EGPA.

## References:


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