Eosinophilic Granulomatosis with Polyangitis – Case Report

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Introduction
EGPA is an uncommon small-medium vessel vasculitis. The presentation varies from asymptomatic to severe clinical manifestations including cardiovascular, gastrointestinal, renal and CNS involvement.

Case Report
52/M patient with history of asthma from childhood, on regular medication, now presented with short febrile illness and acute onset right wrist drop, followed by development of left foot drop 3 days later.
Patient also gives history of positive sensory symptoms involving distribution of right radial nerve and B/L common peroneal nerve.
No history of proximal muscle weakness, no history suggestive of higher function or cranial nerve involvement. No history of back pain or autonomic symptoms.
On examination PR 78/’ regular, BP 140/80, no postural hypotension, Temperature 101F.
There was b/l pitting pedal oedema. Palpable purpura was noted on right lower limb above medial malleolus

On central nervous system examination, there was weakness of extension of right wrist and fingers and weakness of dorsiflexion of left ankle and decreased sensation along the distribution of right radial nerve and bilateral common peroneal nerve.
On auscultation, Bilateral rhonchi was present.
All other systemic examinations were normal.
Clinically we suspect the possibility of a confluent type of mononeuropathy multiplex in view of involvement of more than two peripheral nerves and was investigated for the cause.
Investigations showed Hb 14.5, ESR 53, Total count 15800, Platelet count 2.72L Absolute
eosinophil count 7740, IgE 1593, CRP positive, C-ANCA, P-ANCA negative, Peripheral smear showed severe eosinophilia. N37E50L3
Eosinophils 9000/mm3
ECG: Normal sinus rhythm, 2D ECHO: Normal
CXR: B/L reticulonodular calcifications were present.
USG Abdomen: Hepatomegaly
VDRL, HIV, HBSAg, Anti HCV were negative,
Tumour markers, URE, RFT, LFT, RBS, TFT,
FBS, FLP, RBS, PT, INR, APTT, BT, CT, ANA
Profile were within normal limits.
Video bronchoscopy, BAL, split skin smear, MRI
brain and whole spine screening showed no
significant findings.
Skin biopsy from purpura showed Eosinophilic
vasculitis with fibrinoid necrosis and tissue eosinophilia.

We arrived at the diagnosis of Eosinophilic
granulomatosis with polyangitis according to 2022
American college of Rheumatology / European
alliance of association for rheumatology
classification criteria for EGPA.
Patient was started on pulse steroid therapy.

**Discussion**

EGPA is an uncommon disease with an estimated
annual incidence of 1-3 per million
The mean age of onset is 48 years, with female to
male ratio of 1.2:1. It involves small – medium
sized muscular arteries, capillaries, veins and
venules. A characteristic histopathological feature
is granuloma that may be present in tissues or
within the walls of vessels themselves., associated
with infiltration of the tissue with eosinophils.
Precise pathogenesis of disease is uncertain and it
points to aberrant immunologic phenomena.
Patients with EGPA often exhibit non-specific
manifestations like fever, malaise, anorexia and
weight loss. Severe asthmatic attacks and
pulmonary infiltrates dominate the clinical picture.
Mononeuritis multiplex is the second most
common manifestation and occurs in 72%
patients. Skin manifestations occur in 51%
patients and include purpura in addition to
cutaneous and subcutaneous nodules. Myocarditis,
pericarditis, endocarditis or coronary vasculitis
can occur in 14% of patients and is an important
cause of mortality.
The characteristic laboratory findings include
striking eosinophilia >1000 cells/ ul. in >80%
people. Elevated ESR, CRP, about 48% have
circulating ANCA. Although diagnosis of EGPA
is optimally made by biopsy in a patient with
evidence of asthma, peripheral blood eosinophilia,
and clinical features consistent with vasculitis.
Treatment include glucocorticoids, dosetapering is
often limited by asthma and many patients require
low dose prednisolone for persistent asthma, even
after clinical recovery from vasculitis. In patient
who present with fulminant multi system disease,
particularly cardiac involvement the treatment of
choice is a combined regimen of daily
cyclophosphamide and prednisolone followed by
azathioprine or methotrexate.
Our patient had confluent type of presentation
with involvement of more than one nerve, within a
short time. It is an extremely rare presentation of
mononeuropathy multiplex due to EGPA.
The prognosis of untreated EGPA is poor with
reported 5-year survival of 25%. With treatment,
prognosis is favourable, hence timely diagnosis and proper management is essential for a favourable prognosis.