Digital Journal of Clinical Medicine

Volume 5 | Issue 2 Article 3

Summer 6-8-2023

Eosinophilic granulomatosis with polyangiitis (Churg Strauss Syndrome) – a case report

Rudrani Mukherjee
JSS Medical College and Hospital, JSSAHER, rudranimukherjee1@gmail.com

Bhanukumar M
JSS Medical College and Hospital, JSSAHER, mbhanukumar@jssuni.edu.in

Recommended Citation

Mukherjee R, M B. Eosinophilic granulomatosis with polyangiitis (Churg Strauss Syndrome) – a case report. *Digital Journal of Clinical Medicine*. 2023; 5(2): 50-55. doi: https://doi.org/10.55691/2582-3868.1132

Eosinophilic granulomatosis with polyangiitis (Churg Strauss Syndrome) – a case report

Abstract

Eosinophilic granulomatosis with polyangiitis (EGPA), previously known as Churg Strauss syndrome, is a rare small vessel vasculitis characterized by asthma, eosinophilia, and systemic involvement. This case report presents a 51-year-old man with a history of asthma, who presented with neurological symptoms including blurring of vision, bilateral ptosis, and altered sensorium. The patient had a previous history of cervical and lumbar spine surgeries for symptoms attributed to spondylosis. Further examination revealed hyperpigmented lesions on the limbs, wasting of small muscles, and foot drop.

Investigations showed eosinophilia and thrombocytosis on blood analysis, with bone marrow aspiration revealing eosinophilic and megakaryocytic hyperplasia. The cerebrospinal fluid analysis indicated eosinophilia and lymphocytic predominance. Imaging studies revealed hypodense areas in the midbrain and pons, as well as age-related cerebral atrophy.

Based on the diagnostic criteria for EGPA, the patient fulfilled the requirements of asthma, blood eosinophilia, peripheral neuropathy, transient pulmonary infiltrates, paranasal sinus abnormalities, and the presence of extravascular eosinophils on biopsy. The diagnosis was further supported by the presence of positive antineutrophilic cytoplasmic autoantibodies (ANCA-MPO). Treatment was initiated with intravenous corticosteroids and azathioprine due to the severity of the disease.

EGPA is a heterogeneous disease with varying clinical presentations and organ involvement. Early and accurate diagnosis is crucial for appropriate management and improved outcomes. Glucocorticoids, such as prednisolone, are the mainstay of treatment. While the overall mortality rate is relatively low compared to other systemic vasculitides, cardiac involvement can contribute to significant morbidity and mortality.

In conclusion, this case report highlights the importance of considering EGPA in patients with asthma, eosinophilia, and multi-system involvement. Prompt recognition and initiation of appropriate therapy can help mitigate complications and improve patient outcomes. Further research is needed to better understand the pathogenesis and optimal management strategies for this rare vasculitic disorder.

Keywords

Churg Strauss, Hypereosinophilic syndromes, Vasculitis, ANCA-MPO

Eosinophilic granulomatosis with polyangiitis (Churg Strauss Syndrome) – a case report

Clinical History

Chief complaints:

- History of blurring of vision since 5 days, sudden onset

- History of drooping of both eyelids since 2 days

- Altered sensorium in the form of decreased responsiveness since 1 day

A 51-year-old man, with a 5-year history of asthma, was brought to our emergency department with complaints of blurring of vision in both eyes for 5 days, sudden onset drooping of both side eyelids for 2 days, and altered sensorium for the past few hours. There was no history of fever, trauma, headache, double vision, nausea or vomiting, or convulsions.

He had presented with complaints of difficulty in holding objects in his left hand together with numbness in the left index and ring fingers 6 years ago; he was investigated by an orthopedic surgeon in another hospital, and his symptoms were then attributed to cervical spondylosis, and he underwent cervical spine surgery.

Four years ago, he had presented with insidious onset, gradually progressive weakness and numbness of the right lower limb, initially resulting in slippage of slippers and dragging of the right foot while walking that progressed to difficulty in getting up from sitting position and later requiring support for walking. He was once again assessed by an orthopedic surgeon in another hospital. Following investigations, his right lower limb weakness was attributed to lumbar spondylosis, and he underwent lumbar laminectomy 6 months before his current presentation.

He was taking intermittent inhalers for the management of asthma; he was never admitted for the management of exacerbation of asthma or chest infection. There was no history suggestive of sinusitis.

He was detected to have type 2 diabetes mellitus 20 years ago, which was controlled by oral hypoglycemic agents. He was found to have hypertension 5 years ago and was on regular anti-hypertensive medications with good control of blood pressure. There was no other past medical history of significance. There was no significant family history. He is a non-smoker. He has been consuming alcohol regularly for the past 20 years.

Clinical Examination

General Physical Examination:

Patient is moderately built and nourished, is alert, cooperative and well oriented.

Multiple hyperpigmented lesions present on both upper and lower limbs

Vitals:

Pulse – 80 beats per minute

BP – 110/70 mm of Hg, measured in the right arm in supine position

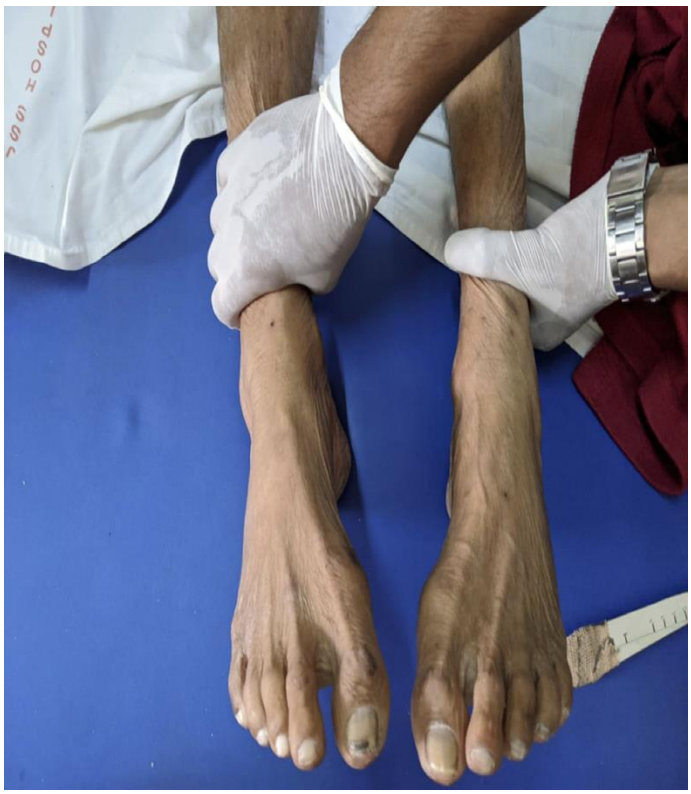
RR – 16 breaths per minute

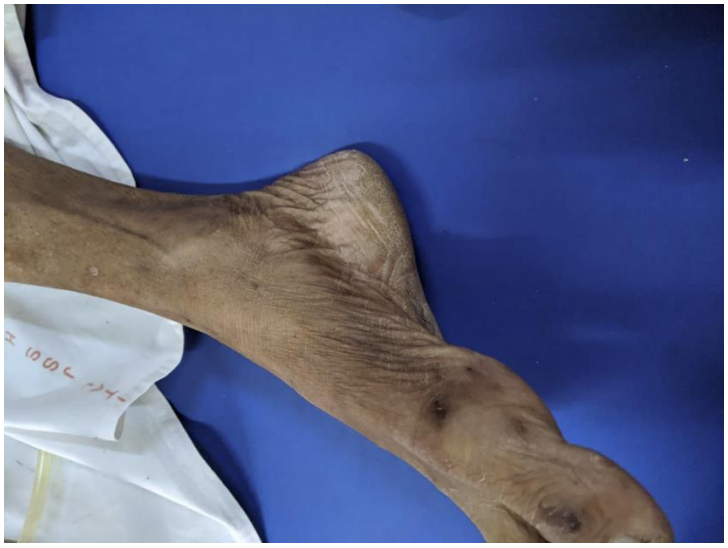
Temp - afebrile

No pallor, icterus, clubbing, cyanosis, lymphadenopathy, pedal oedema.

Wasting of small muscles of right hand and right foot noted.

Foot drop present on the left side.





Nervous System Examination:

Deficits:

- Bilateral ptosis
- External and internal ophthalmoplegia
- Asymmetrical weakness of both lower limbs and left upper limb (distal more than proximal) with sensory involvement
- Deep Tendon Reflexes – generalised areflexia, except both the knee reflexes which were normal

Respiratory exam revealed bilateral rhonchi

Other systems examination were unremarkable.

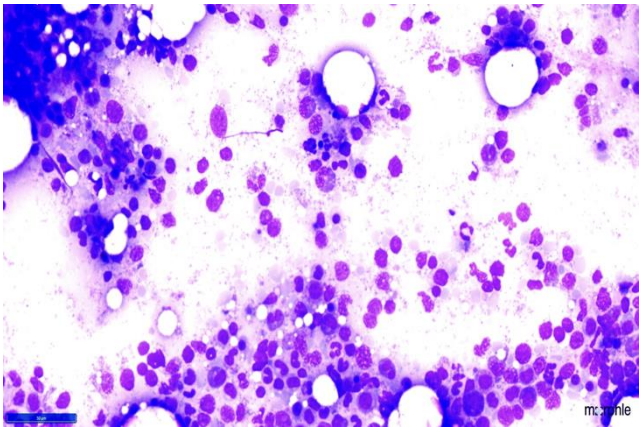
https://rescon.jssuni.edu.in/djcm/vol5/iss2/3
DOI: 10.55691/2582-3868.1132

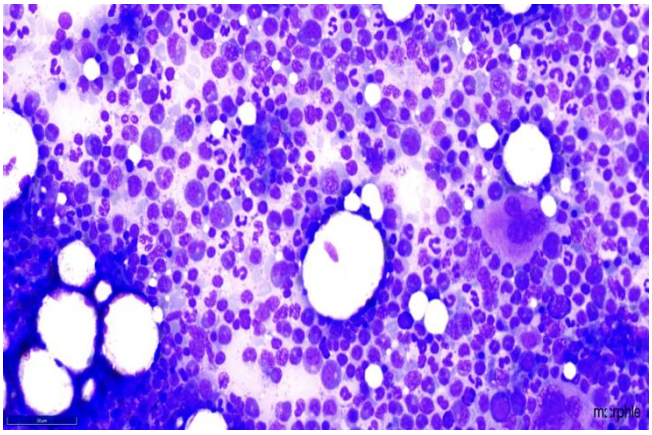
Investigations:

Blood Investigations -

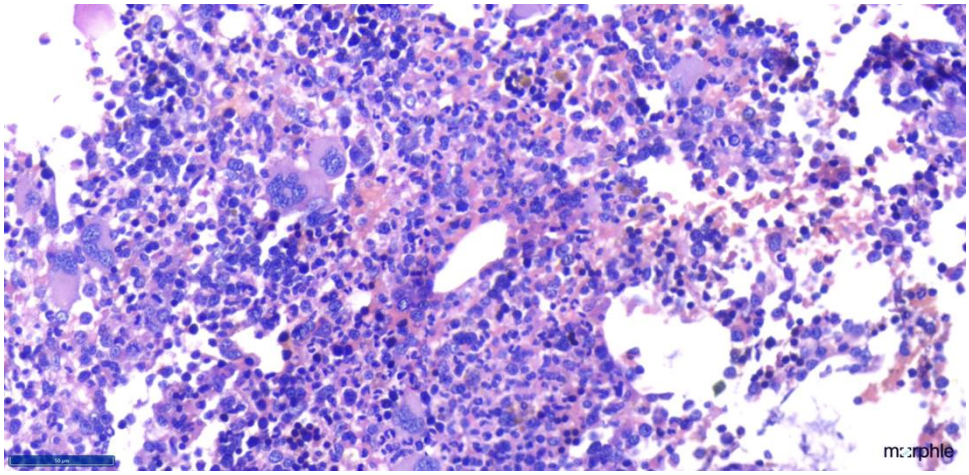
Ix	31/7/20	1/8/20	2/8/20
Hb	12.5 gm/dl		
PCV	36.3		
RBC Count	4.54		
MCV	80		
MCHC	34.4		
RDW	13.9		
Reticulocyte	1.56		
TLC	**23340**		**21740**
DLC (N/ L/ E)	37/ 8/ **51.2**	**AEC - 11954**	35/ 5.8/ **55.8**
Platelets	**6.1**		
ESR	80		
Urea/ Creatinine	19/ 0.61		
PBS	**Eosinophilic leukocytosis with thrombocytosis.**		
Na/ K/ Cl	**125/** 4.5/ 86 mEq/L	**128/** 4.2/ 90 mEq/L	130/ 4.2/ 92 mEq/l

Bone Marrow Aspiration - revealed eosinophilic and megakaryocytic hyperplasia.





Bone Marrow Biopsy – revealed increase in eosinophilic series with predominance of mature forms with increase in megakaryopoiesis.



CSF Analysis -

CSF Glucose	62
CSF Protein	65.2
Cell Chloride	111
Cell Type	Predominantly lymphocytes with eosinophils (38%) and neutrophils with few RBCs in the background. **CSF Eosinophilia**

Other -

CSF Culture, Urine Culture, Blood Culture	No growth
ANA	Negative
ANCA – MPO	**Positive- 18.92 (>9 – positive)**
C – ANCA	Negative

CT Brain -

Ill defined non enhancing hypodense areas in midbrain and pons with age related cerebral atrophy.

HRCT Thorax -

Patchy areas of ground glass opacities in the left lower lobe – CORADS 2 (typical for other infections but not COVID 19)

Small pulmonary nodule in posterior segment of left lower lobe – too small to characterise.

CT Abdomen and Pelvis -

Minimal right perinephric free fluid with minimal perinephric fat stranding.

Differential Diagnosis:

Gross peripheral eosinophilia with Nervous System involvement differentials:

- Eosinophilic meningoencephalitis
- Vasculitis
- Peripheral neuropathy with sensory predominant symmetrical or asymmetrical involvement
- Cerebrovascular accident

Hypereosinophilic Syndromes -

- Idiopathic hypereosinophilic syndrome

https://rescon.jssuni.edu.in/djcm/vol5/iss2/3
DOI: 10.55691/2582-3868.1132

- Familial hypereosinophilia
- Churg Strauss syndrome
- Eosinophil associated gastrointestinal disease

Final Diagnosis:

Churg Strauss syndrome with vasculitic brainstem infarct with peripheral neuropathy and chronic airway disease.

This patient satisfied the diagnostic criteria of Churg Strauss syndrome with asthma, eosinophilia and a vasculitis involving the peripheral nerves with pulmonary involvement, supported by positive ANCA – MPO.

This diagnosis of CSS is made based on ACR 1990 guidelines.

Treatment:

Patient was considered to have severe disease and was thus treated immediately with combination therapy – intravenous corticosteroids and azathioprine.

Conclusion:

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg Strauss syndrome, is a rare systemic vasculitis characterized by asthma, eosinophilia, and multi-organ involvement. This case report presented a 51-year-old man with a history of asthma who developed neurological symptoms, including bilateral ptosis and peripheral neuropathy. The patient had a previous history of spine surgeries and comorbidities such as diabetes mellitus and hypertension.

The diagnosis of EGPA was established based on the 1990 American College of Rheumatology criteria, which require the presence of vasculitis on biopsy along with at least four of six clinical criteria. The patient fulfilled these criteria with asthma, blood eosinophilia, peripheral neuropathy, transient pulmonary infiltrates, paranasal sinus abnormalities, and extravascular eosinophils on biopsy. The presence of positive ANCA-MPO further supported the diagnosis.

Management of EGPA involves a multidisciplinary approach. Glucocorticoids, such as prednisolone, are the mainstay of treatment to control inflammation. In severe cases, immunosuppressive agents like azathioprine may be added. Prompt initiation of therapy is crucial to prevent further organ damage and improve outcomes. Cardiac involvement, which can occur in a significant proportion of patients, should be carefully monitored as it may contribute to disease-related mortality.

This case report highlights the challenges in diagnosing EGPA due to its variable clinical presentation and the potential for overlap with other conditions. Clinicians should maintain a high index of suspicion for EGPA in patients with asthma and eosinophilia who present with multi-organ involvement. Increased awareness and early recognition of the disease can lead to timely intervention and improved patient outcomes.

Further research is warranted to elucidate the underlying pathogenesis of EGPA and identify novel therapeutic targets. Long-term follow-up studies are needed to assess the efficacy and safety of current treatment strategies and to optimize the management of this rare vasculitic disorder.

Discussion:

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg Strauss syndrome, is seen in people with asthma and eosinophilia, and is characterized by small vessel vasculitis, variably affecting many organs including nervous, cardiac, renal, pulmonary, and vascular systems; histologically there is necrotizing vasculitis, tissue eosinophilia, and eosinophil-rich granulomas. It is very rare, with a reported prevalence of approximately 1.0-1.4 cases per 100,000 people. It was first described in 1951 by **Jacob Churg** and **Lotte Strauss**, American physicians based in New York and is now classified along with Wegener's granulomatosis and microscopic polyangiitis as a small vessel vasculitis, associated with antineutrophilic cytoplasmic autoantibodies. (1)

The clinical phenotype is very heterogeneous, and hence, the diagnosis is often overlooked. Correct diagnosis is the key to initiating appropriate management for a successful outcome. Here we report a 51-year-old man, who presented acutely with bilateral ptosis, but following detailed assessment and investigations, was diagnosed to have, and appropriately managed for EGPA.

Diagnostic Criteria:

The 1990 American College of Rheumatology criteria requires a positive biopsy for vasculitis and at least four of the six criteria listed below (sensitivity 85% and specificity 99.7%):

- asthma: history of wheezing or diffuse high-pitched expiratory ronchi
- eosinophilia: >10% of the differential white blood cell count
- mono/polyneuropathy: development of mononeuropathy, multiple mononeuropathy, or polyneuropathy (glove/stocking distribution) attributable to systemic vasculitis
- pulmonary infiltrates, non-fixed: migratory or transitory pulmonary infiltrates (not including fixed infiltrates) attributable vasculitis
- paranasal sinus abnormalities: history of acute or chronic paranasal sinus pain or tenderness or radiographic opacification of the paranasal sinuses
- extravascular eosinophils: biopsy including artery, arteriole or venule showing accumulations of eosinophils in extravascular areas

The presence of 4 or more of these 6 criteria yielded a sensitivity of 85% and a specificity of 99.7%. (2)

CSS presents in three distinct phases: 1. allergic rhinitis, nasal polyposis, and asthma, persisting for years or decades; 2. eosinophilic pneumonia, gastroenteritis, and peripheral eosinophilia, with frequent recurrences; and 3. systemic vasculitis with granulomatous inflammation, occurring on average 3 years after the initial manifestations, although it is also possible for it to present up to 30 years later. (3)

Symptoms of patients vary with respect to severity of eosinophilic inflammation and necrotizing vasculitis, which can cause sequalae in any organ. It is a rare disease with poorly understood pathogenesis. Management consists of treatment with glucocorticoids (mainly prednisolone). The condition generally has a low mortality rate compared with other systemic vasculitides. Cardiac involvement may, however, be a significant contributor to disease-related death and may occur in up to 60% of cases. (4)

References:

1. Keogh KA, Specks U. Churg-Strauss syndrome. Semin Respir Crit Care Med. 2006 Apr;27(2):148-57. doi: 10.1055/s-2006-939518. PMID: 16612766.

2. Masi AT, Hunder GG, Lie JT, Michel BA, Bloch DA, Arend WP, Calabrese LH, Edworthy SM, Fauci AS, Leavitt RY, et al. The American College of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). Arthritis Rheum. 1990 Aug;33(8):1094-100. doi: 10.1002/art.1780330806. PMID: 2202307.

3. Mukamal LV, Sodré CT, Prata LB, Nakasato F, Cuzzi T, Ramos-e-Silva M. Churg-Strauss Syndrome or Eosinophilic Granulomatosis with Polyangiitis: Exuberant Classic Clinical Picture of a Rare Disease. Case Rep Dermatol. 2018 Jun 29;10(2):175–81.

4. Neumann T, Manger B, Schmid M, Kroegel C, Hansch A, Kaiser WA, Reinhardt D, Wolf G, Hein G, Mall G, Schett G, Zwerina J. Cardiac involvement in Churg-Strauss syndrome: impact of endomyocarditis. Medicine (Baltimore). 2009 Jul;88(4):236-243. doi: 10.1097/MD.0b013e3181af35a5. PMID: 19593229.

https://rescon.jssuni.edu.in/djcm/vol5/iss2/3
DOI: 10.55691/2582-3868.1132