

**CHURG STRAUSS SYNDROME ASSOCIATED WITH CNS  
INFARCTION: A RARE MANIFESTATION OF A RARE DISEASE-  
CASE REPORT**

<sup>1</sup>Siddharth Jain, <sup>2</sup>N. K. Agrawal, <sup>3</sup>\*Lalit P. Meena, <sup>4</sup>Jaya Chakravarty, <sup>5</sup>Madhukar Rai,  
<sup>6</sup>Shyam Sundar

<sup>1</sup>Junior Resident, Department of General Medicine, Institute of Medical Sciences,  
Banaras Hindu University.

<sup>2</sup>Junior Resident, Department of General Medicine, Institute of Medical Sciences,  
Banaras Hindu University.

<sup>3</sup>Associate Professor, Department of General Medicine, Institute of Medical Sciences,  
Banaras Hindu University.

<sup>4,5,6</sup>Professor, Department of General Medicine, Institute of Medical Sciences, Banaras  
Hindu University.

Article Received on  
20 Jan. 2018,

Revised on 09 Feb 2018,  
Accepted on 02 March 2018,

DOI: 10.20959/wjpr20185-10726

**\*Corresponding Author**

**Dr. Lalit P. Meena**

Associate Professor,  
Department of General  
Medicine, Institute of  
Medical Sciences, Banaras  
Hindu University.

**ABSTRACT**

Churg strauss syndrome is the least common among the ANCA positive other vasculitis with an incidence around 1-3 per million and is associated with ANCA in only 40% of cases. Clinically patient has varied presentation with background history of asthma in more than 90% of cases. Common features include asthma with sinusitis, skin rash, peripheral neuropathy, renal involvement (glomerulonephritis). CNS involvement occurs in only 5-7% of cases. Here, We report a case of Churg Strauss which itself is a rare disease with its rare presentation in form of CNS involvement.

**KEYWORDS:** Churg strauss syndrome, Granulomatosis with polyangitis, Anti-neutrophil cytoplasmic antibodies.

**INTRODUCTION**

Anti-neutrophil cytoplasmic antibodies (ANCA) associated vasculitis are rare group of disorders and includes Granulomatosis with polyangitis, Microscopic polyangitis and

Eosinophilic Granulomatosis with Polyangiitis (EGPA) which is also known as Churg Strauss syndrome. Eosinophilic Granulomatosis with Polyangiitis is the least common among the ANCA positive other vasculitis with an incidence around 1-3 per million<sup>[1]</sup> and is associated with ANCA in only 40% of cases<sup>[2]</sup> (directed against MPO). Clinically patient has varied presentation with background history of asthma in more than 90% of cases. Common features include asthma with sinusitis, skin rash, peripheral neuropathy, renal involvement (glomerulonephritis). CNS involvement occurs in only 5-7% of cases.<sup>[4]</sup>

We report a case of Churg Strauss which itself is a rare disease with its rare presentation in form of CNS involvement.

### CASE SUMMARY

35 years, male working in a medical shop with a background history of asthma since childhood, presented to us with chief complains of headache for 15 days and altered sensorium 1 day prior to admission. There was no history of fever, vomiting, seizure, vision changes, cough or fall. No history of ATT intake or tubercular contact. He is on inhaled corticosteroids and LABA for asthma for last 10 years. On clinical examination- Patient was in altered sensorium with Glasgow coma score (GCS) -E2V1M1 with Pulse Rate- 88/min, rhythmic, normovolemic. All peripheral pulses were palpable. Blood pressure was 124/76 mmHg in both arms. Patient was febrile with temperature-101.5<sup>0</sup>F. Pallor was present. Icterus, cyanosis, clubbing and lymphadenopathy were absent. He had palpable rashes in both lower limbs suggesting some vasculitis phenomenon (Image-1). Systemic Examination of Cardiovascular system and abdomen was within normal limit. Respiratory system examination showed some abnormalities in form of reduced chest expansion and diffuse rhonci and coarse crepts in both lungs. In Central nervous system examination, meningeal signs were absent without any signs of cranial nerve involvement. Motor examination could not be done with only positive finding being atrophied thenar and hypothenar eminences. Sensory examination could not be done due to poor GCS. Based on clinical suspicion investigations were done. Two CBC reports showed eosinophilia with eosinophil percentage of 48.7 and 36.6 with AEC of 13100/mm<sup>3</sup> and 7,700/mm<sup>3</sup>. NCCT head showed multiple hypodense lesion in bilateral cerebral hemisphere suggestive of acute/subacute infarct. CSF examination was normal with cells-3, protein-34.7 and sugar-78 with corresponding RBS-140mg/dl. Urine routine showed 15-20 dysmorphic RBCs with 1+ protein and no pus cells. Based on above finding with background history of asthma we suspected him to be a case of

Eosinophilic Granulomatosis with Polyangiitis (Churg Strauss Syndrome) or hypereosinophilic syndrome. Immunological profile was sent. MRI and NCV were done and treatment with methyl prednisone with dose of 1gm was started. On 2<sup>nd</sup> day after steroid treatment, he developed multiple episodes of hemoptysis and landed to shock and was shifted to acute care unit with continuation of pulse steroid therapy for 5 days. Results of other investigations showed positive p-ANCA-MPO with level of 83 U. ANA and anti ds-DNA tests were negative. NCV showed decreased amplitude of MNCV of right ulnar(4.81mV) and right median nerve(3.5mV) while no detectable tracing in MNCV and SNCV of common peroneal and posterior tibial nerve. MRI showed multifocal areas of acute and subacute infarct in b/l cerebral hemisphere and basal ganglia which were most probably secondary to thromboembolic phenomenon or vasculitis infarcts (Image-2). CECT thorax was done post stabilisation which had diffuse ground glass opacification with small areas of consolidation in both lung parenchyma suggestive of diffuse alveolar hemorrhage.(image 3).

**After the pulse steroid** Patient was started on oral cyclophosphamide 2mg/kg dose and oral prednisolone at 1mg/kg. He was stabilised with completely improved sensorium. X-ray at the time of discharge was normal. Currently patient is doing well on maintenance therapy using azathioprine 50 mg BD.

## DISCUSSION

Eosinophilic Granulomatosis with Polyangiitis (EGPA) is a multisystem disorder first described by Churg in 1951. It is characterised by asthma, prominent eosinophilia and necrotizing vasculitis predominantly affecting small and medium sized arteries.<sup>[3]</sup>

EGPA presenting with CNS manifestation is a rare presentation with prevalence of around 5-9%<sup>4</sup> EGPA related CNS lesions form four distinct neurological picture: ischemic lesions, intracerebral haemorrhage, cranial nerve lesions, loss of visual acuity. A review of 26 patients of EGPA with CNS manifestations 52% had ischemic lesions, 24% has ICH and/or SAH, loss of visual acuity in 32% and 21% had cranial nerve palsy with 25 patients having more than 1 of these manifestations.<sup>[5]</sup> Long-term neurological sequelae are common, and intracerebral hemorrhages had the worst prognostic impact. Although CNS manifestation of EGPA is not common but these type of manifestation should prompt physicians to consider Eosinophilic Granulomatosis with Polyangiitis as a differential diagnosis in such conditions.



Image 1: Palpable rashes in lower limb.

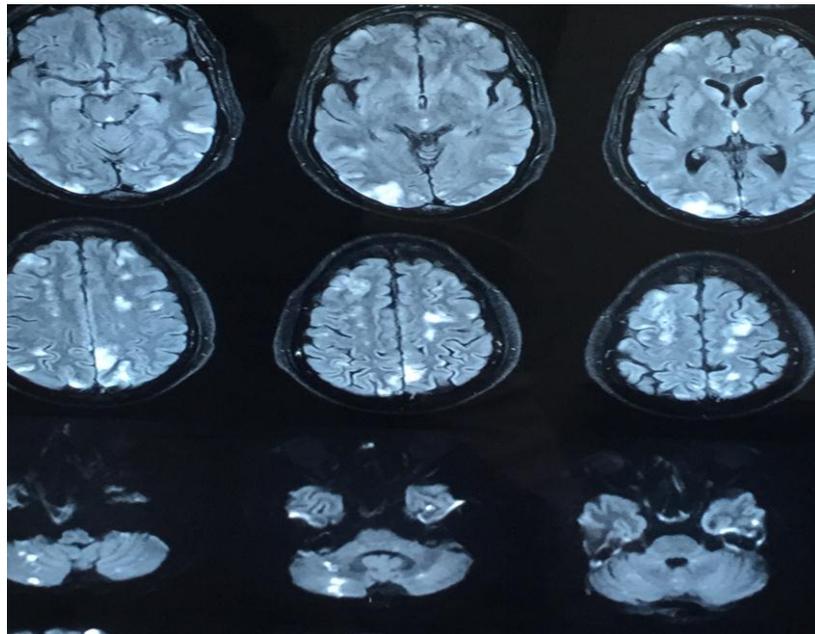


Image 2. MRI showed multifocal areas of acute and subacute infarct in b/l cerebral hemisphere.

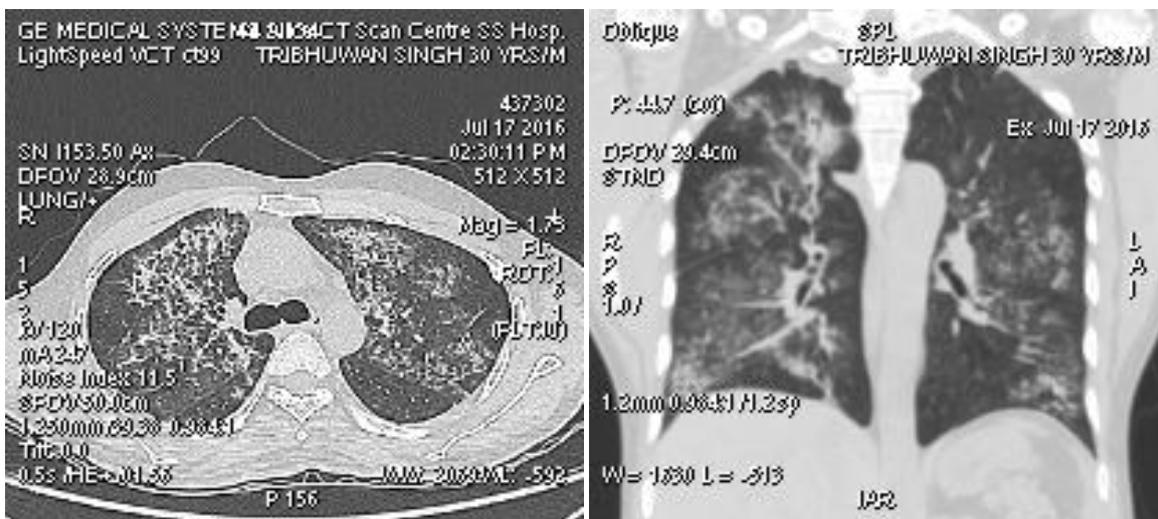


Image 3: Both lung parenchyma suggestive of diffuse alveolar hemorrhage.

**REFERENCES**

1. Harrold LR, Andrade SE, Go AS, et al: Incidence of Churg-Strauss syndrome in asthma drug users: a population based perspective, *J Rheumatol*, 2005; 32: 1076–1080.
2. Sablé-Fourtassou R, Cohen P, Mahr A, et al: Antineutrophil cytoplasmic antibodies and the Churg-Strauss syndrome, *Ann Intern Med.*, 2005; 43: 632–638.
3. Churg J, Strauss L: Allergic granulomatosis, allergic angiitis and periarteritis nodosa, *Am J Pathol*, 1951; 27: 277–301.
4. Comarmond C, Pagnoux C, Khellaf M, et al. Eosinophilic Granulomatosis with Polyangitis: clinical characteristics and long term followup of 383 patients enrolled in French Vasculitis Study Group cohort. *Arthritis Rheum*, 2013; 65(1): 270-81.
5. Andre R et al: Central nervous system involvement in eosinophilic granulomatosis with polyangitis: report of 26 patients and review of literature. *Autoimmune Rev.*, 2017; 16(9): 963-969.