Research Article

An Insight to Churg Strauss Syndrome (CSS)

Karthika Unnikrishnan[1], Kattakkayam Sukanya Mathew[1], Denna Lukose[1], Dr. Sreeraj[2], Remya Reghu[1]*

1. Department of Pharmacy Practice, Amrita School of Pharmacy, Kochi, Amrita Vishwa Vidyapeetham, Amrita University, India.
2. P G Resident, Department of Pulmonary Medicine, Amrita Institute of Medical Sciences, Kochi, Amrita University, India.

***Corresponding author's E-mail:** remyareghu@aims.amrita.edu

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ABSTRACT

Churg and Strauss in 1951 first time described Churg - Strauss Syndrome (CSS). It is characterised by asthma, peripheral and tissue eosinophilia, vasculitis of multiple organ systems, and extravascular granuloma formation. This is a case of a 41yr old gentleman who developed signs and symptoms of this syndrome but responded well to the initial iv antibiotics , pulse dose corticosteroids and iv cyclophosphamide and was discharged with adequate improvement in his general condition.

Keywords: Churg strauss syndrome, allergic angitis, granulomatosis, eosinophilia, vasculitis.

INTRODUCTION

Synonym for CSS is Eosinophilic granulomatosis with polyangitis (EGPA), which is a rare condition of blood vessel walls inflammation affecting small to medium-sized vessels. It is often presented with severe asthma and blood and tissue eosinophilia.[1,2] It was first noted in 13 patients who presented with asthma, eosinophilia, granulomatous inflammation, necrotizing systemic vasculitis, and necrotizing glomerulonephritis. The disorder consists of namely three phases: prodromal, eosinophilic and vasculitic phases.[1-3] The diagnosis of the same will be made by analysing various factors like the patient's history, blood tests, chest X rays or CT scans and biopsy reports of the affected organs. Treatment is usually done with steroids or other immune suppressing agents. [4]

CASE REPORT

41yr old gentleman with obstructive airway disease was admitted in a local hospital with complaints of shortness of breath, cough with mucoid expectoration for three days and running nose. No h/o haemoptysis, loss of appetite, loss of weight, fever, chest pain.

CT PNS: right ethmoid, mastoid, sphenoethmoidal recess opacity

Chest X-ray: bilateral pneumonia

CT thorax: multiple irregular soft tissue density lesions in all lobes of lung, few lesions show cavitation and ill defined ground glass opacities at periphery of lesions.

Bronchoscopy: nodular lesion in all airways with induration.

Bronchial biopsy: suppurative inflammation with lymphoid cells infiltrates.

IHC: negative for lymphoma

ANCA and Aspergillus specific IgM were negative.

The patient was started on I.V antibiotics (Piperacillin-Tazobactam) and antifungals (Flucanazole, Voriconazole).

The patient was referred to our hospital for further evaluation of bilateral non-resolving pneumonia. At the time of admission the patient was febrile and had elevated serum total counts and other inflammatory markers. Treated initially with I.V Meropenem and Clindamycin, nebulized bronchodialators and other supportives. Rheumatology consultation was sought to rule out vasculitis in view of elevated serum eosinophil counts and total IGE with background of uncontrolled asthma and persisting allergic rhinitis. Vasculitis work up was done and a provisional diagnosis of Churg Strauss syndrome was made. The patient was initially treated with pulse dose corticosteroids and later on IV Cyclophosphamide was given. ENT consultation was sought for B/L ethmoidal sinusitis, otitis medial and mastoiditis. DNE done showed B/L mucopurulent discharge with crusting. Nasal swab culture grew *staphylococcus aureus*. Nasal discharge fungal culture grew *Aspergillus species*. Since he had persistent headache and two episodes of vomiting, MRI Brain done as per Neurology opinion, but did not show any evidence of meningitis/ parenchymal changes. Dermatological consultation was sought for leision in B/L shins, they opined as seborrhoic dermatitis and palmo plantar eczema and their orders were carried out. Cardiology opinion was obtained to rule out cardiac involvement associated with Churg strauss. 2D Echo showed normal chamber dimensions. No RWMA. Good LV systolic function, Trileaflet aortic valve. No AS/AR MVP-AML-A2 and PML-P2 prolapse and no evidence of PAH. They opined to manage MVP conservatively. His repeat chest x ray showed significant resolution of B/L consolidation with adequate improvement in his general condition as well. The patient was discharged in a hemodynamically stable condition with resting room air oxygen saturation of 97%.

DISCUSSION

CSS can occur at any age but often excludes infants. As per the literature the mean age of onset was found to be 48yrs, with 1.2:1 female-to-male ratio. The CSS etiology is still unknown. One of the causal factor is hypersensitivity to an inhaled agent. [5,6] In this case, the patient has a h/o carbon inhalation. Asthma is one of the main sign of this syndrome. Rarely a parasitic infection or antigenic drug for desensitization can trigger the syndrome. Most frequently involved organs are Lungs. In few cases a rare finding of the presence of a pulmonary nodule has been reported.[6-8]

There are six criteria for the diagnosis of CSS which includes asthma, higher than normal count of eosinophils called eosinophilia [normal: 1-3%]. Above 10% is considered abnormally high and as a strong indicator of CSS. Damage to one or more nerve groups (mononeuropathy or polyneuropathy), lesions on a chest x ray, sinus problems, extravascular eosinophils are the other criteria. If four out of six of these features are seen, then it has a high specificity and sensitivity for the diagnosis of CSS.[9,10] Here the patient had asthma, elevated serum eosinophil counts, allergic rhinitis, multiple irregular soft tissue density lesion in all lobes of lung and bilateral maxillary and ethmoidal sinusitis.

Frequently treatment includes for the first three to six months a combination of corticosteroids and cyclophosphamide are used, and thereafter the cyclophosphamide is switched to methotrexate or azathioprine for maintenance for several additional months. immunoglobin is used to those who haven't responded to above treatments.[10-12] Here the patient was treated initially with pulse dose corticosteroids and later on IV Cyclophosphamide.

CONCLUSION

Survival chance of CSS patients is high, although relapses are not uncommon. For diseases like asthma the widespread use of steroid has resulted in a confusing situation in which the steroid therapy accidently suppresses CSS and changes in steroid treatment uncover the disease. The scope for future research in this area is huge.

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