Churg-Strauss Syndrome in an elderly man

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ABSTRACT

Churg-Strauss Syndrome (CSS) is an uncommon systemic vasculitis affecting the medium and small sized arteries. CSS was earlier considered fatal, but prognosis has significantly improved with the use of steroids and other immunosuppressive therapy. CSS has been reported in patients as young as seven years old to as old as 74 but generally occurs in the third to fifth decade of life. Here we describe an 86-year-old Indian man who was affected by this disease with clinical presentations as described in younger individuals and excellent response to therapy. To our best knowledge, presentation at so late an age has not been documented in the literature.

Keywords: Churg-Strauss Syndrome, elderly, vasculitis, ANCA, hypereosinophilia syndrome

INTRODUCTION

Churg-Strauss Syndrome (CSS) was originally described in 1951 as 'allergic angiitis and allergic granulomatosis' based on autopsy studies of patients who exhibited similar characteristic lesions. Currently classified among the various antineutrophil cytoplasmic antibodies (ANCA)-associated vasculitides, its pathophysiological mechanisms are incompletely understood. The international annual incidence in the general population is reported to be approximately 0.11 to 2.66 per million (in 2007) with a prevalence of 10.7 to 14 per million adults. [1] There is a slight female preponderance and the mean age at diagnosis ranges from 38 to 53 years. [1] To the best of our knowledge, the oldest patient reported so far was 74 years old [1], thus highlighting the importance of physician awareness of this disease in the elderly. Since CSS shows a remarkable response to corticosteroid therapy, an early diagnosis will greatly improve the prognosis.

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CASE REPORT

An 86-year-old south Indian man who had never travelled outside his constituency was treated by multiple general practitioners for suspected chronic obstructive pulmonary disease (COPD). His medical history was also remarkable for recurrent allergic rhinitis without epistaxis or haemoptysis. He was referred to our hospital with dyspnoea, high grade fever, sudden onset productive cough, and polyarthralgia of five days duration. Prior medical records were unavailable and no laboratory

tests were performed previously. He was admitted to the intermediate care unit and did not require ventilator support as oxygen saturation was maintained at 96% on room air. On physical examination the patient was febrile and tachypnoeic, and had extensive wheeze that was audible bilaterally.

Haematological investigations revealed leucocytosis of 37,700/mm^3 with an absolute eosinophil count of 21,700/mm^3 (57.4%) and an absolute neutrophil count of 13,500/mm^3 (35.7%). Peripheral blood smear showed eosinophilia. The haemoglobin was 14.3 gm/dl, the platelet count was 258, 000mm^3, and the erythrocyte sedimentation rate (ESR) was 9mm at one hour. No parasites were demonstrable on stool examination. Chest radiograph (Figure 1) showed a few heterogeneous shadows in the lower zones. Renal function was slightly impaired (Table 1) and urine analysis detected albuminuria (1+) and haematuria (3+). Procalcitonin level was within reference range.

A provisional diagnosis of an acute exacerbation of COPD was made, and the patient was treated with Prednisolone which controlled the symptoms. Discontinuation of the steroid therapy saw a reappearance of the fever and deterioration of renal function (Table). The patient then developed bilateral weakness of the lower limbs with markedly

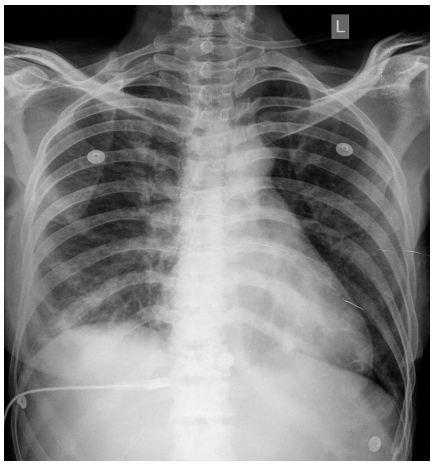


Fig. 1: Chest radiograph (PA view) of the patient showing heterogeneous shadows in lower zones.

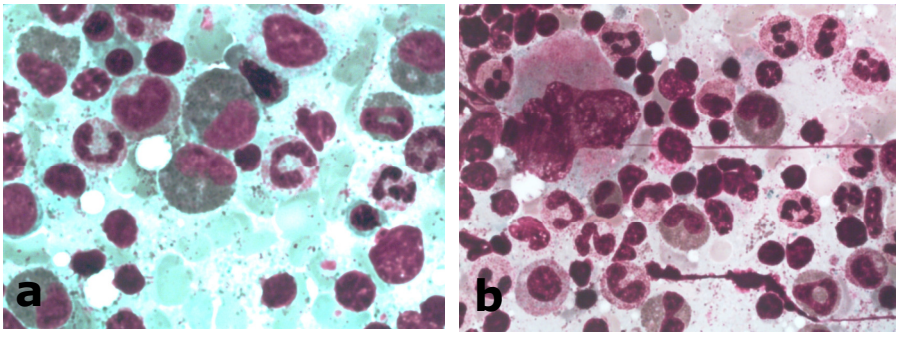
reduced deep tendon reflexes and slight numbness over the upper limbs. There were no specific sensory deficits. Soon afterwards, a few erythematous vesicles formed bilaterally over the lower limbs.

The common aetiologies of eosinophilia were carefully excluded in the patient. Bone marrow aspiration and biopsy showed a predominance of myeloid precursors with increased eosinophil precursors but without any evidence of abnormal cells (Figures 2a and 2b). Computed tomography of the lungs and tissue biopsies were not performed owing to financial constraints that the patient faced.

Further investigations were directed towards CCS. Serology revealed elevated IgE levels (>2500 IU/ml) and positive perinuclear

Table 1: Correlations of laboratory investigation (serum creatinine) and temperature profiles with steroid therapy.

Day of admission	1	3	6	15	17	19	20	22	25
Serum creatinine (mg/dl)	1.19	1.01	0.91	1.35	3.06	5.28	5.33	3.93	1.28
Temperature (degree Fahrenheit)	99	98.6	98.6	98.6	101.8	98.6	101	98.6	98.6
Corticosteroid administration	Yes	Yes	Yes	No	No	No	Yes	Yes	Yes



Figs. 2: a) Bone marrow aspiration (H&E, x1000) showing a neutrophil-eosinophil ratio of 15:5 (normal 15-20:1) indicating increased eosinophil precursors, and b) Bone marrow aspiration (H&E, x1000) demonstrating the predominance of myeloid precursors with increased eosinophil precursors (almost 50%).

antineutrophil cytoplasmic antibodies (p-ANCA). Treatment with intravenous methylprednisolone resulted in rapid resolution of symptoms and normalisation of the eosinophil counts.

DISCUSSION

The American College of Rheumatology criteria established in 1990 were used to diagnose the disease in our patient. The four of the six criteria present in our patient were asthma, eosinophilia, neuropathy, and pulmonary infiltrates on chest radiograph. Tissue biopsies are not essential for diagnosis. The multiple organ system involvement which occurs in this disease is predominated by asthma (47-93%) and pulmonary symptoms (37-77%). [1] Dermatological manifestations occur in over half of the patients, the most common lesion being palpable purpurae. [1] Renal involvement is less common, and is often overlooked. It is an adverse prognostic indicator, and several lesions including obstructive uropathy have been described.

The primary aetiology of eosinophilia is parasitic infections and this had to be excluded in our patient given that the common occurrence in our setting. Medications such as leukotriene receptor antagonists have been implicated in the development of CSS. This has however been refuted in an extensive study by Weller *et al*, who thought the aetiology to be due to unmasking of underlying vasculitic process by these drugs. [2] Further studies are required before a definite relationship between the two is recognised. The discontinuation of corticosteroids may also be the cause, as seen in a series of case reports by Churg *et al*. [3]

Perinuclear ANCA usually specific to myeloperoxidase (MPO) are found in 40% of patients with CSS, though this is not distinctive of the disease. [4] ANCA positivity as seen in our patient has been associated with a significantly higher prevalence of renal involvement and mononeuritis multiplex, purpurae and pulmonary haemorrhage with lower prevalence of cardiac and other pulmonary involvement. [5] Studies have shown that ANCA may precede the clinical onset of CSS and may be thus used for early detection of the disease. [6]

Considered to be a rare disease, CSS may initially seem to be a hypersensitivity disease and may thus be underdiagnosed.

Determination of total leukocyte and eosinophil counts is thus warranted in all patients being treated for a diagnosis of COPD or bronchial asthma, so that possible early manifestations of this disease may be detected.

In conclusion, the clinical features in our elderly patient were typical of CCS described in younger patients. The response to corticosteroid therapy was noteworthy in that complete remission of symptoms was rapidly achieved. A high index of suspicion is required to diagnose this disease especially in the elderly, and often a correlation between the clinical and pathological findings may be necessary. In addition, careful regular monitoring of these patients for adverse reactions to medications is required.

REFERENCES:

1: Baldini C, Talarico R, Rossa AD, Bombardieri S. Clinical manifestations and treatment of Churg-Strauss Syndrome. Rheum Dis Clin N Am. 2010; 36:527–43.

2: Weller PF, Plaut M, Taggart V, Trontell A. The relationship of asthma therapy and Churg-Strauss syndrome: NIH workshop summary report. J Allergy Clin Immunol. 2001; 108:175-83.

3: Churg A, Brallas M, Cronin SR, Churg J. Formes Frustes of Churg-Strauss Syndrome. Chest. 1995; 108:320-3.

4: Sinico RA, Di Toma L, Maggiore U, et al. Prevalence and clinical significance of antineutrophil cytoplasmic antibodies in Churg-Strauss syndrome. Arthritis Rheumatism. 2005; 52:2926–35.

5: Pagnoux C. Churg-Strauss syndrome: evolving concepts. Discov Med. 2010; 9:243-52.

6: Zwerina J, Axmann R, Manger B, Schett G. The emergence of antineutrophil cytoplasmic antibodies may precede the clinical onset of Churg-Strauss syndrome. Arthritis Rheumatism. 2009; 60:626–67.