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# A CASE OF NECROTIZING VASCULITIS PRESENTING AS EOSINOPHILIA- CASE STUDY

General Medicine

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## ABSTRACT

A woman with history of bronchial asthma presented with weight loss, malaise, cough with mucopurulent sputum, fever and blood blisters on hand and feet. Chest X-Ray revealed pulmonary infiltrates. There was no definite evidence of necrosis of vessel walls but the appearances were suggestive of allergic vasculitis. On treatment with prednisolone the response was remarkably good with normal Chest X-Ray.

## KEYWORDS

Eosinophilic pneumonia, Allergic vasculitis, Wegner's granulomatosis, churg's allergic granulomatosis, pulmonary infiltrates, Steroids

## INTRODUCTION

An unusual case of necrotizing vasculitis is reported with bronchitis which preceded the onset of allergic vasculitis of the skin, pulmonary infiltrates, marked blood eosinophilia and extra- vascular proliferation. The relationship between benign pulmonary eosinophilia, Churg's allergic granulomatosis and Wegner's granulomatosis is discussed.

Pulmonary eosinophilia is characterized by coughing, which is persistent in nature and gets aggravated at night, asthmatic attacks, weight loss, low grade fever and an enlarged spleen. Investigations includeperipheral eosinophilic count, serum IgE levels. Diethylcarbamazine is the main therapeutic drug used.

Churg's allergic granulomatosis is an extremely rare autoimmune condition which leads to the inflammation of small and medium sized blood vessels. Some of the symptoms are joint pain, muscle pain, bleeding inti tissues under the skin a rash with hives, small bumps, cough, night sweats, fever, fatigue and abdominal pain.

Wegner's granulomatosis is an extremely rare long-term systemic disorder that involves the formation of granulomas and inflammation of blood vessels. It mainly affects the upper respiratory tract, lungs and kidneys. Typical signs and symptoms include nosebleeds, stuffy nose and crustiness of nasal secretions and inflammation of the uveal layer of the eye. Treatment is usually with immunosuppressants like rituximab or cyclophosphamide in combination with high dose corticosteroids.

## CASE HISTORY:

A female aged 27 years presented with history of weight loss, malaise, cough with muco-purulent sputum, fever without chills and rigor, stinging and burning sensation and blood blisters on face, hand and feet (figure1) for 4 weeks. She also developed weakness in left hand which slowly progressed, she found difficulty in holding glass of water and in wearing clothes. She had past history of bronchial asthma since 4 years and was on treatment with β agonist.

On examination pulse was 90 per minute regular, blood pressure 110/70 mm/Hg, heart sounds were normal. There were occasional rhonchi in basal region bilaterally. Touch, pain and temperature was diminished on two medial fingers of left hand. Power of different muscles of left hand were as follows (lumbricals medial two 2/5, interossei medial three 2/5, abductor digitis minimum 2/5, flexor digiti minimum 2/5. There were hemorrhagic blisters on extremities suggestive of vasculitis.

Investigations revealed hypochromic anemia (Hb 8.9 g/100ml) with WBC count of 8100/mm of which 22% (1780/cumm) were eosinophils and an ESR of 96 mm/hour. Platelets were normal. There was no proteinuria or microscopic hematuria. Sputum for AFB by concentration method was negative in 3 consecutive samples. X-ray chest PA view showed multiple pulmonary infiltrates in both lower zone of lung field (Figure 2). ECG, blood glucose, urea, creatinine clearance, liver function tests fecal occult blood, anti-nuclear factor, USG abdomen and pelvis were normal. Skin biopsy showed only small focus of inflammatory cells in the upper epidermis which were mainly nuclear fragments. There was no definite evidence of necrosis of vessel walls, but the appearance suggested allergic vasculitis.

The patient was given I. V. cloxacillin and ampicillin (1 gm IV 6 hourly) along with oral prednisolone 60 mg daily for 1 week. The repeat X-ray chest PA view showed clearance of the patchy shadows. The skin biopsy revealed small sub-epidermal vesicle with collection of mononuclear cells in the underlying epidermis, composed largely of histiocytes with occasional langhans type giant cells and a few plasma cells along with mild perivascular lymphatic infiltration in the dermis. Acid and alcohol fast bacilli were not seen in appropriately stained sections.

The clinical response was remarkably good and within 1 week the patient recovered with normal chest x-ray, ESR 24 mm/1 hour and WBC count of 7100/mm$^3$ with 2% eosinophilia. The patient had good progress on decreasing dosage of prednisolone and was maintained on dose of 5 mg/day.

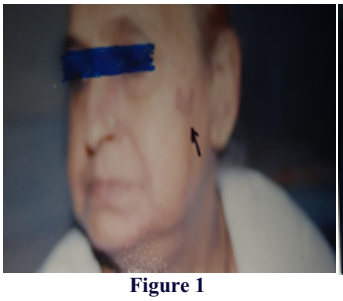

Figure 1

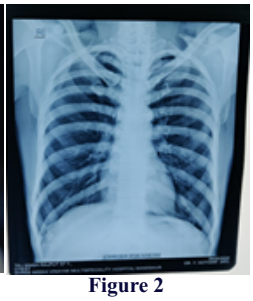

Figure 2

## DISCUSSION

Allergic granulomatosis has been recognized as a clinico-pathological syndrome by Churg and Strauss' characterized clinically by severe asthma, fever, eosinophilia and histologically by widespread necrotizing vasculitis with characteristic changes in the vascular and extra vascular connective tissue. Braverman[1] pointed out that asthma precedes the systemic illness by months or years and characterically remits before the onset of the vasculitic phase Abdominal pain and bloody diarrhea is present invariably along with arthralgia and peripheral neuropathy.

Rose and Spencer[2] in a retrospective study found that in the majority of cases the lungs are completely spared, even when there is severe polyarteritis in almost every other organ but some cases with pulmonary involvement form a distinctive subgroup characterized by marked blood eosinophilia, granulomatous lesions in the lungs and

other viscera. The clinical and radiological findings in this case are compatible with an eosinophilic pneumonia. The rapid resolution of these changes following steroid therapy suggests the changes were due to eosinophilic infiltration rather than pulmonary arteritis.

Sokolov[3] suggested a possible relationship between widespread necrotizing vasculitis with pulmonary involvement and benign transient eosinophilic infiltration of the lungs which they called Loeffler's syndrome. Thus there may be a range of syndromes from benign transient pulmonary eosinophilia through chronic eosinophilic pneumonia and cases with higher incidence of vasculitis and extra-vascular granuloma formation to the full blown Churg's syndrome.

**CONCLUSIONS:**

SYNDROMES CHARECTERIZED BY VARYING DEGREES OF PULMONARY EOSINOPHILIA, NECROTIZING VASCULITIS AND GRAULAMATOUS PROLIFERATION.

Loffler syndrome - Benign transient pulmonary eosinophilic infiltration, usually due to ascaris infestation

Churg's syndrome - Severe asthma, fever and eosinophilia preceding the onset of disseminated vasculitis and granulomata.

Wegner's granulomatosis - Granulomatous ulcers of upper and lower respiratory tract, progressing rapidly to disseminated vasculitis and granulomata, with renal involvement in the late stages.

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