Churg - Strauss Syndrome and Desensitization Therapy

A Case Report and Brief Review of Literature

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A 55 year old Saudi male with bronchial asthma and allergic rhinitis presented with a palpable purpuric skin rash, mononeuritis multiplex and marked eosinophilia developing during allergic desensitisation therapy. Based on the above finding a clinical diagnosis of Churg-Strauss Syndrome was made. The patient responded very well to steroids and cyclophosphamide. It has been postulated that repeated antigenic stimulation such as repeated injections of allergens during desensitisation therapy, drugs or parasitic infections may provoke systemic vasculitis in atopic individuals. In this article, we present the first reported case from Saudi Arabia with Churg-Strauss syndrome developing during desensitisation therapy. Until further proof, clinicians should be aware that allergic desensitisation therapy can rarely be complicated by Churg-Strauss Syndrome.

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The Churg-Strauss Syndrome (CSS) is a rare disease of unknown aetiology. In very few instances it has been postulated that repeated antigenic stimulation such as injections of allergens during desensitisation therapy, certain drugs or parasitic infections may have pathogenetic contribution to CSS in patients with allergic diseases such as allergic rhinitis and/or bronchial asthma[5-9]. In this connection, there have been a rare case report in the literature. Even though there have been three case reports of CSS from Saudi Arabia[2], our report is the first Saudi patient with CSS manifested during an allergic desensitisation process.

THE CASE

A 55 year old Saudi male with bronchial asthma for 5 years and allergic rhinitis for 10 years, presented with lower limb weakness, nocturnal fever, sweating, loss of weight, right pleuritic chest pain, dry cough, abdominal pain, and watery diarrhoea for 20 days. Three days prior to admission to Assir Central Hospital, he developed a sudden intense weakness and numbness of the right hand. While in the hospital and before initiation of treatment, he developed skin rash on the dorsum of the right hand. Before three months he had skin testing in a private clinic abroad and a 12 unlabelled but numbered bottles were prescribed for subcutaneous injection as desensitisation therapy. He took the injections for about 2 months on weekly basis and after the 6th dose he started to develop the above symptoms. He was on salbutamol inhaler, intermittent short courses of prednisolone and cromolyn sodium. There was no family history of bronchial asthma or allergic rhinitis.

Examination showed a temperature of 38° C, blood pressure of 110/70 mm Hg and expiratory rhonchi in both lungs. He had right facial asymmetry, wasting of right thenar muscles and distal weakness of the right upper and left lower limbs with absent ankle jerk. There was decreased skin sensation over the right thenar muscles and in both lower limbs in gloves and stocking distribution. The skin showed few palpable purpuric rash on the dorsum of the right hand. Cardiovascular system, and abdomen were normal. The initial impression was atypical Guillain - Barre' Syndrome. Investigation revealed a Hb 12.9 gm/dl, MCV 84.7, WBC 51.9 x 10^9/L with 73 % eosinophils and platelet count was 364 x 10^9/L. ESR was 75 mm during the first hour. Peripheral blood smear showed normochromic normocytic anaemia with markedly increased number of mature, normal looking eosinophils. Bone marrow smear was normal except for increased eosinophils infiltration. Urine analysis showed protein 1 +, RBC 10-12/HPF and WBC 6-8/HPF. 24 hours urine protein was 730 mg/dl and createnine clearance 46 ml/min. Stool analysis for ova, parasite and occult blood was repeatedly negative. Bilharzia, leishmania and hydatid serology were negative. HBsAg and anti hepatitis-C antibodies were negative. Rheumatoid factor and ANA were negative. ANCA was not done as facilities were not available. X-ray of sinuses showed thickened mucosa. Chest X-ray, ultrasound of abdomen, ECG and echocardiography were normal. Nerve conduction study showed diffuse low amplitude action potentials with normal velocities, consistant with axonal neuropathy. The patient refused biopsy from skin, sural nerve or kidney.

In view of the above clinical and laboratory findings, the patient was diagnosed and managed as CSS. He was treated initially with prednisolone 60 mg per day and showed remarkable clinical and laboratory improvement, but relapsed upon tapering the dose to 30 mg per day. He showed excellent response to cyclophosphamide (100 mg per day) while prednisolone dose was successfully reduced to 5 mg per day.

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DISCUSSION

CSS bears the name of two pathologists (Churg and Strauss) who first described the disease as a separate entity but related to polyarteritis nodosa. They describe three major histological criteria: eosinophilic tissue infiltration, extravascular granulomas and necrotizing vasculitis[1]. Subsequent reports showed that these histological findings were not specific for CSS, patchy in distribution, and rarely coexist in one tissue specimen[2,3]. When possible the diagnosis should be substantiated by biopsy of one of the involved organs.

Based on a series of 16 patients and the review of another 138 reported in the literature, Lanham et al, identified three clinical phases of the disease; the first phase (prodormal period) usually lasts several years and consists of allergic rhinitis and nasal polyposis. It is frequently followed by asthma. The second phase consists of blood eosinophilia in excess of 1.5 x 10^9/L and fleeting eosinophilic infiltrate. The third phase consists of systemic vasculitis. They considered that the clinical pattern of CSS is distinctive enough to justify its recognition on clinical ground.

The pathogenic mechanism is not yet clear. Experimental and clinical studies have shown a possible association between allergy as exemplified by asthma or its offending antigen and polyartetitis nodosa developing during its course[4]. On the other hand there are few reports in the literature about the possible aetiologic association between allergic desensitisation therapy and development of vasculitis. Phanuphak and Kohler described 20 patients with systemic vasculitis of whom 6 patients had features of CSS developed during allergic desensitisation therapy. This association was found in a subset of atopic individuals whose allergic diathesis was atypical with respect to late onset of symptoms, absence of family history, severity of upper respiratory tract disease and exaggerated eosinophilic response. A case report presented in the clinico-pathological exercise in the New England Journal of Medicine developed CSS during allergic desensitisation therapy[6]. CSS and polyarteritis nodosa were reported in patients with parasitic infections such as trichinosis and richinosis respecitvely[7,8]. Liver disease and vasculitis were reported in a patient taking cromolyn sodium[9]. It appears that hyperresponsiveness to repeated antigenic stimulation in a subset of individuals with atypical allergic diathesis, triggers a self-perpetuating vasculitic process[3,7].

The response to high dose steroids can be dramatic. The respiratory symptoms, asthma and eosinophilia may resolve quickly. In those who have more systemic involvement, such as cardiac (major cause of death) or neurologic involvement, the response may be delayed or limited. Cyclophosphamide has been shown to be extremely effective in the treatment of systemic vasculitis including CSS[10].

Our patient presented to Assir Central Hospital with bizarre neurological symptoms and signs consistent with mononeuritis multiplex, However, a history of allergic rhinitis and brionchial asthma with cromolyn use and immunotherapy with a battery of allergens heralded his symptoms. With this background and a finding of purpuric skin rash with a very high blood eosinophilia in the initial work up was suggestive of CSS. The dramatic improvement in his symptoms and resolution of eosinophilia to high dose steroid further supports the diagnosis of CSS. Since he could not tolerate tapering of steroid, the addition of cyclophosphamide resulted in his clinical and laboratory improvement.

CONCLUSION

We conclude by stating that as the practice of desensitization with unknown extracts of allergens for allergic rhinitis and asthma is performed erratically in some centres, clinicians should be aware that it can rarely be complicated by CSS.

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