Churg-Strauss Syndrome with Cutaneous and Neurological Manifestations Preceding Asthma

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Sir,

Allergic granulomatosis and angiitis (Churg-Strauss syndrome – CSS) is a relatively uncommon systemic disease with characteristic clinical and pathologic features. The syndrome is characterized by systemic vasculitis, extravascular granuloma and hypereosinophilia, occurring almost exclusively in patients with asthma or an allergic history. The American College of Rheumatology (ACR) has proposed six classification criteria, with four being necessary for CSS to be diagnosed. The six criteria are asthma, eosinophilia (>10%), paranasal sinusitis, pulmonary infiltrate, histological proof of vasculitis and mononeuritis multiplex (1). However, the ACR criteria are not designed to serve as diagnostic criteria for individual patients, and are thought to function poorly in this capacity (2–5). Although extremely rare, there have been some reported cases of non-asthmatic patients who present with other symptoms of CSS (6–10). We report a patient with CSS with various characteristic skin lesions and paraesthesias as initial findings preceding asthmatic symptoms. These findings may lead to an early diagnosis of CSS and prevent irreversible tissue damage with treatment.

CASE REPORT

A 42-year-old man was referred in September 2003 with a 1-week history of painful purpuric and papular skin lesions of the lower limbs and soles. He had no history of asthma or allergic rhinitis. His medical history and family medical history were not significant. Two weeks after onset, purpuric skin lesions developed and an erythematous rash resembling erythema multiforme appeared, progressively involving the patient’s inner thighs, trunk, upper extremities, face and head (Fig. 1). Additionally, he began to experience paraesthesias in conjunction with his cutaneous lesions (mainly on the extremities) and unintentional weight loss and fever were recorded. Laboratory tests revealed a leukocyte level of 9000/µl (normal 4000–9000/µl), eosinophils at 1746/µl (19.4%; 0–8%) and raised IgE 2700 IU/ml (<170 IU/ml). Serum eosinophil cationic protein (ECP) level was 147 µg/l (<15.7 µg/l). No significant laboratory abnormalities were found, including rheumatoid factor, antinuclear antibodies, total haemolytic complement, anti-neutrophil cytoplasmic antibodies (ANCA), hepatitis A and B serological tests, cryoglobulins, cryofibrinogen, serum protein electrophoresis, serum immunoglobulins and echocardiogram. Chest and abdominal X-ray, ultrasound examination and computerized tomography did not show any abnormalities. Stool examinations for ova and parasites were negative. Myelogram and bone marrow biopsy showed only a mild medullary hypereosinophilia without any morphological or neoplastic changes.

Microscopic examination of skin biopsy specimens obtained from the purpura on the left lower extremity and the abdominal erythema revealed a dermal perivascularity with neutrophils, red blood cells and numerous eosinophils, consistent with leukocytoclastic vasculitis and extravascular granuloma (Fig. 2). There was a predominance of eosinophils infiltrating the dermis around the nerve fibre.

The patient was given 30 mg prednisone daily, which led to an improvement in his symptoms. His condition stabilized and the peripheral eosinophilia and IgE levels normalized. The corticosteroids were tapered without any recurrence of the skin lesions. Two months after start of treatment, the patient received 15 mg prednisone daily, and his clinical and laboratory findings were normalized. The corticosteroids were then slowly tapered off. The patient was re-admitted to the hospital in April 2004 with light dyspnoea and firm palpable and haemorrhagic purpuric eruptions on the extremities with paraesthesias. He reported some asthma-like symptoms that coincided with the onset of the rash. Laboratory studies revealed an elevated white blood cell count of 9000/µl and eosinophilia of 13.0%. The IgE level was elevated to 2000 IU/ml. Serological tests were negative for perinuclear and cytoplasmic pattern ANCA. An end-expiratory wheeze was noted on chest auscultation. Pulmonary function

Fig. 1. (A) Sharply marginated dull red plaques with centrifugal spreading and central clearing in abdomen. (B) Erythematous and oedematous plaques with ill-defined borders on hand, face and head.
constituents of eosinophil granules, are neurotoxic and may contribute to direct nerve injury. IL-5 stimulates a massive expansion of eosinophils, which in turn secrete ECP (15). In our patient, the increased serum ECP concentration returned to normal during periods of disease remission. Thus, high ECP levels may play a direct role in the development of paraesthesias. The findings in our patient strongly suggest that this disorder is triggered by an eosinophil and IgE inflammatory response.

**REFERENCES**