Cutaneous Granulomas in a Patient with Common Variable Immunodeficiency

Sir,

Common variable immunodeficiency (CVID), one of the most frequent of the primary specific immunodeficiencies, is a heterogeneous group of syndromes characterized by a deficiency in antibody synthesis associated with panhypogammaglobulinaemia and recurrent bacterial infections (1). The occurrence of non-caseating granulomas in different organs, including the skin, has been described previously. We report here a new case of cutaneous granulomas in a patient with CVID.

CASE REPORT

An 18-year-old woman had been diagnosed as suffering from CVID 16 months before and had since been treated successfully with monthly intravenous immunoglobulin therapy. Her past medical history included an abdominal tuberculosis at 5 years of age and a postprimary pulmonary tuberculosis at 11 years of age, which were correctly treated in both cases, as well as recurrent bacterial respiratory infections without any other complications.

The patient was referred to our department with a 10-day history of a non-pruritic papular eruption on both legs, coinciding with one of the intravenous immunoglobulin doses. Review of her systems did not reveal any other symptomatology.

Dermatological examination disclosed multiple reddish-violaceous papules on both legs. The lesions, 0.5 – 1 cm in diameter, were tender, firm, infiltrated and not well defined. The remaining physical examination was normal.

Routine laboratory tests revealed normal blood cell count, sedimentation rate, creatinine and liver function. The levels of serum immunoglobulins, once established replacement therapy, were as follows: IgG 501 mg/dl (normal 694 – 1860 mg/dl), IgA <7 mg/dl (normal 85 – 400 mg/dl) and IgM 264 mg/dl (normal 42 – 320 mg/dl). The lymphocyte subpopulations were normal except for a discrete increase in the activated CD3+ T-cells. All serological tests for viruses and bacteria were negative. Tuberculin testing was 3 mm and sputum and urine analysis did not show any acid-fast bacilli. Pulmonary function studies, chest X-ray, abdominal ultrasonography and thorax CT scan revealed no abnormalities. The histopathological picture of one of the papules on the legs showed multiple non-caseating perivascular granulomas throughout the deep dermis and the subcutaneous fat. These granulomas were compact, well-circumscribed and composed of epithelioid cells, occasional multinucleated giant cells and peripheral lymphocytes. The overlying epidermis was normal. Periodic acid-Schiff, methenamine silver, Zhiel-Nielsen and Gram's stains gave negative results. Examination under polarized light failed to demonstrate birefringence.

The skin lesions spontaneously regressed in 2 months without therapy. The patient is currently asymptomatic, and is receiving monthly intravenous immunoglobulin, which has not been interrupted at any time.

DISCUSSION

Cutaneous granulomas have been reported previously in patients with CVID (2 – 6) and with other primary immunodeficiencies (7) and have been described as erythematous scaling plaques (2), nodules (6) and, most frequently, as in our patient, as a papular eruption. In immunodeficient patients it is important to rule out other causes of granulomatous inflammation in the skin, principally mycobacterial and fungal infections, by using special stains and appropiate cultures. We must also exclude a non-infectious aetiology, such as sarcoidosis (3).

The pathogenesis of these granulomas is unknown; one hypothesis is that in a state of humoral deficiency the cell-mediated component overreacts and forms granulomas (5). Similar immunological responses could explain the occurrence of granulomas in patients with tumours (8), infections (9) or following the administration of several drugs (10).

These lesions usually regress spontaneously, as occurred in our patient, and only in cases of aggresive evolution with tissue destruction systemic corticosteroids may be useful (11), though they should always be used with caution.

Finally, it is neccessary to emphasize the importance of systemic study and exhaustive follow-up in all these patients because the appearance of cutaneous granulomas may be the first manifestation of multisystemic granulomas (4).

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