

PYODERMA GANGRENOSUM ASSOCIATED WITH IgA PARAPROTEINEMIA. TREATMENT WITH CYCLOSPORIN A AND PREDNISONE

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SUMMARY

A 58-year-old woman with a 20-year history of recalcitrant pyoderma gangrenosum associated with IgA paraproteinemia was treated with cyclosporin A 5 mg/kg/day combined with prednisone 30 mg/day without significant amelioration. After raising the dose of cyclosporin A to 10 mg/kg a rapid improvement was observed. Subsequently the cyclosporin A and prednisone doses were reduced progressively and a complete cure was achieved after 7 months of treatment. Three years later no relapse has occurred on a maintenance therapy consisting of 3 mg/kg/day of cyclosporin A and 6 mg/day of prednisone.

KEY WORDS

pyoderma gangrenosum, treatment, cyclosporin A, prednisone

INTRODUCTION

Pyoderma gangrenosum (PG) is a rare disease of unknown etiology and slowly progressive course. It is characterized by sterile inflammatory lesions, sometimes granulomatous and sometimes pustular or ulcerative, which show a tendency to scar at the center and to extend centrifugally. The course is chronic with alternating exacerbations and remissions, and vast ulcerations several decimeters in diameter may develop over months or years, without apparent precipitating factors. In 50 to 75% of cases PG is associated with a systemic disease, most often ulcerative colitis, Crohn's disease, polyarthritis or a gammopathy.

Immunologic mechanisms have been shown to be

involved in the pathogenesis of PG: this explains the successful use of cyclosporine A (CsA), which is characterized by a potent activity mainly on cell-mediated immunity but also on antibody-dependent immunity and chronic inflammatory reactions. CsA in the treatment of PG has been well documented (1-3).

The first case of PG treated with CsA was described by Curley et al. in 1985 (4); many other reports have followed (5-7). The literature sometimes describes therapeutic successes with low dosages of CsA (3-4 mg/kg/day (18,9)), while other authors suggest the need for a dose of 7-10 mg/kg/day (10-12), also in association with systemic steroids (4,6,9,10,13).

The following description is a case report of PG

