

DEFLAZACORT - CYCLOSPORINE A COMBINATION THERAPY IN THE TREATMENT OF PEMPHIGUS

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ABSTRACT

Two cases of pemphigus vulgaris and one of foliaceous pemphigus, treated with deflazacort combined with low doses of cyclosporine A, are reported.

Response and tolerance to treatment were markedly improved over the previous therapy.

KEY WORDS

pemphigus, cyclosporine A, immunosuppressants

INTRODUCTION

Presently treatment of pemphigus is mainly based on steroids alone or in combination with other immunosuppressants such as methotrexate, cyclophosphamide and azathioprine (1, 2, 3).

Over the last several years, various authors proposed the use of cyclosporine A in the treatment of this immunological skin disease (4, 5, 6).

This report describes our experience with cyclosporine A in combination with lowered steroid-dosage in three patients affected by different forms of pemphigus.

CASE REPORTS

The clinical diagnosis of pemphigus in these three patients was confirmed histo- and immunohistochemically.

Case 1.

M.B., a 72-year-old man, was affected by foliaceous pemphigus for 14 years. He also suffered from Basedow's syndrome, diabetes mellitus, and essential hypertension. He has been treated with doses of prednisone (40-60 mg/daily) over a long period of time. To reduce the side effects of this treatment, he has been receiving Deflazacort (mean dosage of 18 mg/daily) combined with cyclosporine A (from 2,5 to 3,2 mg/kg/daily) for the past nine months. Routine

hematochemical tests have been normal in the periodical checks: glycemia (range from 151-177 mg/dl) and blood pressure have not shown any remarkable increase.

Case 2.

D.L., a 35-year-old woman, has been affected with pemphigus vulgaris for the last 2 years. We treated her initially with Deflazacort (30 mg/daily). After 15 days no improvement was observed and we added cyclosporine A (3 mg/kg/day); this combination resulted in clinical improvement, allowing us to reduce the dosage to a maintenance level of 12 mg/daily Deflazacort and 2,2 mg/kg/daily cyclosporine A.

Case 3.

B.G., a 60-year-old woman, is affected with pemphigus vulgaris of the oral mucosa. The disease has been present for 3 years. At the outset she was treated with a combination of methylprednisolone (40mg/daily) - azathioprine (50mg/daily); the clinical picture showed no major changes.

Subsequently, she received Deflazacort (24 mg/daily) in combination with cyclosporine A (3 mg/kg/daily). This therapy resulted in a complete remission, and the treatment

was reduced to a maintenance dosage of 18 mg/daily Deflazacort and 2,25 mg/kg/daily of cyclosporine A.

DISCUSSION

Steroids, initially given at high dosage (100-200 mg/daily), and then decreased to a maintenance dose, are presently considered the standard therapeutic approach for pemphigus (1, 2, 3, 7).

Side effects related to steroid treatment are frequent and serious: infections, diabetes, osteoporosis, myopathy, cataracts, nervous and gastrointestinal disorders (1, 8).

Alternative therapies have been tried for the severe forms of pemphigus: methotrexate, cyclophosphamide, azathioprine, and in the past years cyclosporine A in monotherapy or in combination with steroids (1, 2, 3, 4, 5, 6, 8, 9).

The latter is considered useful in order for reducing steroid dosage with consequent reduction of side effects (4, 5, 6, 9). However, other authors stress that cyclosporine A in monotherapy displays little activity in the acute stage of pemphigus vulgaris (4).

Our data confirm that low dosages of cyclosporine A (less than 3 mg/kg/daily) permit a substantial decrease in the corticosteroid dosage with a concomitant reduction of the side effects.

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