Dexamethasone-cyclophosphamide pulse therapy in pemphigus

Sir,

Pemphigus vulgaris is a potentially fatal disease in spite of a variety of treatment modalities available. The

introduction of dexamethasone–cyclophosphamide pulse (DCP) therapy for the pemphigus group of disorders by Pasricha *et al*. in 1981 has revolutionized the therapy of pemphigus.[1] The standard DCP therapy had also been given to our patients for the last 5 years and the results are as reported below.

Twenty patients of pemphigus admitted at the SKIMS Medical College Hospital, Srinagar were enrolled for the study prospectively from May 2001 to April 2006. Diagnosis of pemphigus was based on clinical features, Tzanck smear and skin biopsy. Confirmation was carried out by direct immunofluorescence examination. DCP therapy was given to those patients with positive Tzanck smear and histopathological features of pemphigus. Before starting the pulse therapy, investigations undertaken were – complete hemogram, erythrocyte sedimentation rate (ESR), urine analysis, blood sugar, kidney and liver function tests, chest radiography, electro cardiogram and stool examination for occult blood. Two patients had hypertension, one had diabetes and one had associated vitiligo. All investigations were repeated at monthly intervals and when necessary. The patients were monitored for side effects of DCP therapy.

The entire treatment was divided into four phases as per Pasricha *et al*. schedule. [1]

Phase I: DCP therapy was given in the presence of signs and symptoms. Patients received monthly doses of 100 mg of dexamethasone dissolved in 500 mL of 5% dextrose by slow intravenous infusion over 2 hour on three consecutive days along with 500 mg of cyclophosphamide in the infusion on day 2. In between, the patients received 50 mg of oral cyclophosphamide daily.

Phase II: Patients were in remission but monthly DCP therapy and daily oral cyclophosphamide were continued for 9 months.

Phase III: Only oral cyclophosphamide 50 mg was given to patients for an additional 9 months.

Phase IV: All treatments were withdrawn and patients were followed-up for relapse, if any.

Of the 20 patients of pemphigus vulgaris treated with this regimen, there were nine males and 11 females, the age ranged between 32 and 60 years. One unmarried male and one female who had not completed her family were given dexamethasone pulse (DP) without cyclophosphamide infusion but with daily oral cyclophosphamide. The duration of disease before treatment varied from a minimum of 1 month to a maximum of 7 years. Only six patients were treated with various other modalities before entering the study. Two patients were lost to follow-up. Of the remaining 18 patients, eight are in phase IV, six in phase III and four in phase II. The duration of phase I varied among patients, mostly being 3–4 months, with no correlation with age and sex of patients or the severity of the disease at presentation. Only two patients required daily oral corticosteroids in the first phase and none was given interval pulse. No case of death was seen amongst the cases studied. The duration of continuous remission in the patients is more than 2 years, the maximum being 5 years.

The side effects associated with prolonged treatment with corticosteroids and cyclophosphamide were virtually absent. The common side effects seen were generalized weakness and fatigue (7), gastrointestinal symptoms (5), menstrual irregularities (5), alopecia (4), candidiasis (2), dermatophytosis (1), hypertension (1) and urinary symptoms (1). No significant changes in laboratory parameters were seen.

Pemphigus is an autoimmune bullous dermatosis having a grave prognosis and is associated with high morbidity and mortality. Systemic steroids and other immunosuppressive therapies have remained the mainstay of treatment of pemphigus.[2] DCP therapy designed by Pasricha Gupta for pemphigus and was first used in 1981 with the aim of reducing the toxicity of corticosteroids and also to achieve better therapeutic results. Since then, the same pulse therapy has been used and complete remission of pemphigus has been reported.[3-6] In addition, the therapy also reportedly leads to a significant decrease in the mortality rate associated with the disease and there is a remarkable decrease in the side effects associated with long-term use of steroids and immunosuppressant drugs.[4-7] Our study included 20 patients of pemphigus vulgaris, of which eight are already in phase IV and others in different phases also showed remarkable response. The side effects profile was comparable with those from previous studies.[6,7]

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