Original Article

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Pulse Therapy - A Boon to Pemphigus: A Case Series from a Rural Tertiary Care Center

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Abstract

Background: Pemphigus is a group of autoimmune vesiculobullous disorders that carry high degree of mortality and morbidity. Dexamethasone-cyclophosphamide pulse (DCP) therapy has been proved to have promising outcome in the management of these diseases.

Aims: The objective of the study was to assess the outcome of DCP therapy in our pemphigus patients.

Methods: Pemphigus patients who were treated with DCP from 2005 to 2015 were prospectively and retrospectively analyzed.

Results: A total of 30 patients were enrolled. Male:female ratio was 1:1.3 with a mean age of 46.8 ± 15.8 with age range varying from 26 to 65 years. About 24 patients completed Phase I and were maintaining remission (80%). Out of 24, 5 were in Phase II, 6 were in Phase III, and 2 in Phase IV. Around 11 patients were declared cured. Only 3 patients relapsed (2 patients were in Phase II, 1 patient was in Phase III). Patients who relapsed were on irregular follow-up. No mortality was noted in our study. DCP therapy is not absolutely free from adverse effects. Most of the adverse effects were expected and tolerated and did not create any problem in continuing the pulse therapy. There was a slight increase in incidence of weight gain, cushingoid features, myalgia, pyogenic infections, oral candidiasis, headache, and hiccups. Laboratory parameters were within normal limits. Three patients showed an impaired glucose tolerance which reverted to normal with the continuation of pulse therapy.

Conclusion: DCP pulse therapy was effective in inducing, maintaining remission and offering cure for patients with pemphigus.

Key words: Cyclophosphamide, Dexamethasone, Pemphigus, Pulse therapy

INTRODUCTION

Pemphigus is a group of autoimmune disorders which present with blisters and erosions over skin, mucosa which is serious and life threatening. Dexamethasone-cyclophosphamide pulse (DCP) therapy designed by Pasricha *et al.* At All India Institute of Medical Sciences has revolutionized the management of pemphigus, since it was introduced, in 1982. If administered properly, DCP has the potential to offer cure for these diseases.[1-5]

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METHODS

Prospectively and retrospectively analyzed all pemphigus patients admitted in ward between 2005 and 2015. Diagnosis was based on clinical features, Tzanck smear, and skin biopsy. Immunofluorescence was not done due to non-availability. Baseline complete blood count, liver function test, HIV screening, renal function test, pus culture from erosions, serum electrolytes, electrocardiogram, chest X-ray, ultrasound abdomen was done for all patients before initiation of pulse therapy. Lab parameters were repeated on every subsequent pulse. Physician opinion was obtained before initiation of pulse therapy. Patients were maintained on daily dexamethasone to attain quick remission before initiation of pulse therapy. After evaluation, patients were started on dexamethasone 100 mg in 500 ml of 5% dextrose over 3 h for 3 consecutive days. Injection cyclophosphamide 500 mg was added to the same drip on the 2nd day. This

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constitutes one pulse. This was continued every 28 days until the patient achieved complete remission which was considered as Phase I. In between pulse patients were maintained on tablet cyclophosphamide 50 mg OD. Few patients who showed exacerbation of disease were maintained on oral steroids and few patients on interval pulse. After completion of Phase I, patient continued to receive the same dose of DCP for 9 more months which was considered as Phase II. In Phase III, patients were maintained only on tablet cyclophosphamide. In Phase IV, all drugs are stopped and patients are followed up for a period of 2-year. Patients that completed 2 years of disease free follow-up were declared cured. Data regarding age, sex, duration of disease, previous treatment details, comorbid diseases, phases of pulse, adverse effects were documented. Lab investigations such as urine analysis, complete hemogram, liver function tests, and renal function test were documented before each pulse. Pus culture was done from unhealthy erosions and pyogenic infections in needed patients.

RESULTS

Out of 30 patients followed up for DCP, 17 (56.6%) were females, 13 (43.3%) were males, male:female ratio was 1:1.3 with age range between 27 and 65 years. The highest incidence was noted in the age group of 41-50 (13 patients) followed by 8 patients in the age range of 31-40 years. The youngest range was in 4 patients (21-30 years) (Figure 1).

Disease Profile

Out of 30 patients, 28 was pemphigus vulgaris and 2 was pemphigus foliaceus. About 24 (80%) patients had severe skin and mucosal involvement (>30% body surface area), 6 (20%) patients had moderate skin and mucosal involvement (10-30% body surface area).

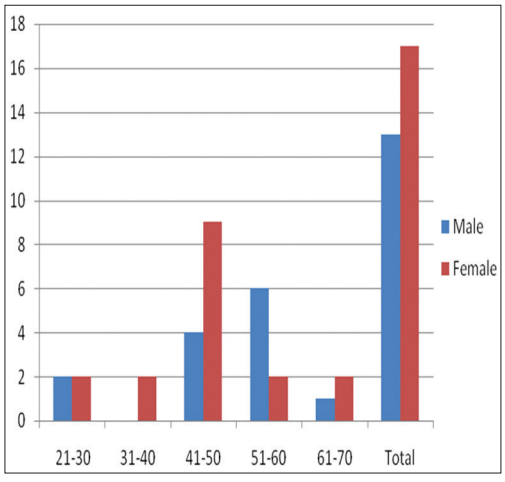

Figure 1: Age/sex distribution

Duration of Disease before Onset of DCP

Duration of disease before onset of DCP ranged from 5 days to 2 years. About 28 patients (95%) presented to us in the first episode.

Comorbidities

Out of 30 patients, 4 patients were diabetic, 2 patients had both diabetes and hypertension, one patient was a persons living with HIV/AIDS on antiretroviral therapy, one was treated Hansen with recurrent Type I Lepra reaction.

Phases of DCP (Table 1)

Phase I

Among 30 patients 6 patients (20%) are in Phase I out of which 3 are on irregular follow-up with recurrent exacerbations. Around 24 patients (80%) attained remission out of which 5 patients were in Phase II, 6 patients in Phase III, and 2 patients in Phase IV, 11 patients (36%) were declared cured. Relapse was observed in 2 patients while on Phase II, one patient in Phase III due to irregular follow-up. Good compliance was seen in 80% of our patients. No mortality was observed in our study.

Remission was achieved in 10 patients within 1-5 pulses (Table 2). The majority of patients (12) attained remission with 6-10 pulses. However, 2 patients needed 11-15 pulses to attain remission.

Adverse Effects Noted during Pulse Therapy (Table 3)

Adverse effects noted were myalgia, hiccups, dyspepsia, muscle cramps and headache in the immediate week following pulse therapy. Long-term adverse effects noted were weight gain, cushingoid habitus, cataract. Pyogenic infections - furuncles, abscess at intravenous cannulation sites, perianal abscess were also noted and was more in patients with comorbidities like diabetes, HIV disease.

Table 1: Number of patients in phases of DCP

Phases	Male	Female
Phase I	3	3
Phase II	3	2
Phase III	2	4
Phase IV	0	2
Cured	5	6
Total (*n*=30)	13	17

DCP: Dexamethasone cyclophosphamide pulse

Table 2: Number pulses required attaining remission

Number of pulse	Male	Female
1-5	2	8
6-10	7	5
11-15	1	1
Total (*n*=24)	10	14

Table 3: Adverse effects

Adverse effects	Number of patients
Immediate	
Dyspepsia	8
Hiccoughs	3
Tiredness	10
Muscle cramps	4
Delayed	
Weight gain	6
Cushingoid features	6
Furuncles	5
Abscess at IV cannulation site	1
Perianal abscess	2
Cataract	1
Amenorrhea	3
Oligomenorrhea	4
Oral candidiasis	2

IV: Intravenous

Amenorrhea and oligomenorrhea were noted in 5 patients. Laboratory parameters were within normal limits except for 3 patients who showed impaired glucose tolerance while on pulse who were non-diabetic before onset of pulse but reverted to normal values once pulse therapy was stopped.

DISCUSSION

Pulse therapy has been proved to offer quick remission and complete cure in these patients which previously was considered to carry a high degree of mortality.[1,5-7]

The results of our study show a positive outcome in terms of effectiveness. We were able to maintain remission in 80% (24 out of 30) patients.

Kandan and Thappa[7] reported a remission rate of 87.5% and Sachidanand *et al.*[3] reported remission rate of 82% which was comparable with our study. Patient compliance was good in 95% of our patients which was much higher than observations by Mahajan *et al.*[6] with 58% drop out. Mortality was not seen in our study whereas Kandan and Thappa[7] reported mortality in 5 out of 65 patients treated and Pasricha *et al.*[1,5] reported 19 deaths among 500 patients treated.

The adverse effects noted in our patients were tolerable, reversible, treatable and did not pose much problem in continuing the pulse therapy.

Even in patients with co-morbidities like diabetes and HIV disease adverse effects were comparable with the other patients except for slight increase in incidence of pyogenic infections. To our surprise, we noted that patient with recurrent Type 1 lepra reaction had no episode of lepra reaction while on treatment and follow-up.

CONCLUSION

The results of this study indicate a high degree of positive outcome in terms of effectiveness of pulse therapy in pemphigus. With this treatment modality, it is now possible to induce quick remission and offer cure in patients with these severe diseases.

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