

Management of Pemphigus Vulgaris Using Dexamethasone Cyclophosphamide Pulse Therapy: A Case Report

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Abstract

Pemphigus vulgaris (PV) is a life-threatening disease belonging to the pemphigus group of autoimmune intraepidermal bullous diseases of the skin and mucosae. The therapeutic management of PV remains challenging and, in some cases, conventional therapy is not adequate to induce clinical remission. The cornerstone of PV treatment remains systemic corticosteroids. Although very effective, long-term corticosteroid administration is characterized by substantial adverse effects. The introduction of dexamethasone-cyclophosphamide pulse (DCP) therapy for the pemphigus group of disorders has revolutionized the therapy of pemphigus. In this case report, standard DCP therapy has also been given to our patient.

Keywords: Corticosteroids, Dexamethasone, Pemphigus, Treatment.

INTRODUCTION

Pemphigus vulgaris (PV) is a life-threatening disease belonging to the pemphigus group of autoimmune intraepidermal bullous diseases of the skin and mucosa.^[1] Use of systemic corticosteroids has reduced the mortality rate from <10%.^[2] Corticosteroids are used in the dose of 1–2 mg/kg body weight or even higher doses in the initial phase of the disease with subsequent gradual tapering over 2–3 months to achieve remission followed by low maintenance dose for longer period of time.^[3]

Such long treatment are associated with various side effects such as diabetes, hypertension, secondary infection, and osteoporosis, leading to increase in comorbidities and treatment-related mortalities.^[1] To overcome these side effects and to achieve longer period of remission, Pasricha *et al.* in 1982 successfully started the use of dexamethasone cyclophosphamide pulse (DCP) therapy in Delhi. DCP therapy is given every 28 days in four phases. DCP therapy is a promising therapy providing long-term remission to patients with minimum side effects. It can be used as first-line therapy till the further advancement takes place in this field.^[4]

In this article, we will discuss about case report of PV treated using DCP therapy.

CASE REPORT

The patient reported to the department of oral medicine and radiology with a chief complaint of pain and fluid-filled ulcers in the lip, buccal mucosa region, and skin parts such as back region, hands, foot region, and legs for 1 month back, lesions first started in oral region exceeding toward hands further involving different parts of skin. The patient has visited a medical hospital and local application gel was given. He has habit of smoking from the past 12 years back 5 to 6 times a day. Family and dental oral history was non-relevant.

Skin examination

On inspection, it shows multiple symmetric, distributed, erythematous, ulcer with overlying hemorrhagic thick, crusting, and foul smelling present over scalp of varying size

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[Figure 1] also involving nail, trunk, upper limb, back, and lower limb region. A few erosions present without crusting over left, chest, and back region. On palpation, crusting presents over various parts of skin.

Intraoral examination

On inspection, multiple symmetric, distributed, erythematous, ulcer with overlying hemorrhagic thick, crusting over oral mucosa, and extending over left and right buccal mucosa were seen and on palpation, painful areas on the left and right buccal mucosa [Figure 2].

Oral examination shows superficial ulceration, history of fluid-filled bullae, and Nikolsky sign with sliding and loosening of adjacent mucosa. This sign is present when slight rubbing of the skin results in exfoliation of the outermost layer. There was reduced mouth opening due to painful erosive ulcers.

On the basis of various clinical findings, provisional diagnosis of pemphigus was made. Intraoral biopsy was done and immunofluorescence test was done, and after histopathological examination, final diagnosis of PV was confirmed.



Figure 1: Erosive areas on the head region



Figure 2: Erosive areas on the left buccal mucosa

Treatment

DCP therapy is divided into four phases^[4]

Phase I is considered from the start of DCP therapy till the remission is achieved usually taking 3–6 pulses on average. The treatment schedule is divided as follows:

Phase II (induction phase): Dexamethasone (100 mg dissolved in 500 ml 5% dextrose) by slow intravenous drip over 2 h (1 dose) on 3 consecutive days, along with cyclophosphamide (500 mg) on day 1. Three pulses DCP are repeated every 4 weeks. Between the pulses, the patients receive oral cyclophosphamide (50 mg daily). After that in phase II, another six pulses of DCP are continued.

In Phase III, only oral 50 mg of cyclophosphamide is given daily for another 9 months.

Phase IV is the phase of observation where all other medicines were withdrawn.

Antibiotics (amox-clav 1.2 g), iv tramadol, antibacterial creams (fusidic creams), and calamine lotions are also given along with therapy. For oral pemphigus, measures such as soft diets and soft toothbrushes help to minimize local trauma.

Topical analgesics or anesthetics are useful in alleviating oral pain, particularly before eating or tooth brushing.

Oral hygiene is crucial, tooth brushing should be encouraged and antiseptic mouthwashes used. Patients susceptible to oral candidiasis should be treated.

Follow-up after 15 days, 1 month [Figure 3a and b], and 3 months [Figure 4a and b] shows significant healing of the lesions.

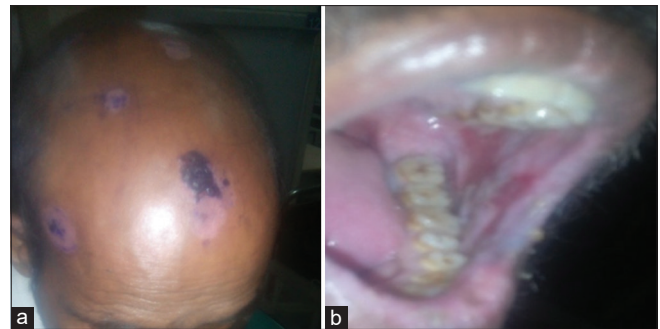


Figure 3: (a and b) Follow-up after 1 month with healing lesions



Figure 4: (a and b) Follow-up after 3 months with healing lesions

DISCUSSION

DCP therapy is primarily used for pemphigus. It was initially described as a treatment that produced quick control of the disease and reduced hospital stay in pemphigus.^[5] Pulse therapy has also been used for other conditions, chiefly autoimmune diseases and found to be an effective therapy. Patients were treated with DCP and no side effects were noticed and no recurrence was there. Thus, DCP has less long-term adverse events, has decreased both morbidity and mortality of pemphigus patients.^[6]

Systemic corticosteroids can be combined with an immunosuppressive agent, particularly when complications due to prolonged use (4 months), such as hypertension, diabetes mellitus, and osteoporosis, are expected. In cases where doses of steroid exceeding 100 mg/day are required, pulse treatment is considered.^[7] The aim of high-dose pulse therapy is to reduce the duration, cumulative dose, and iatrogenic effects of corticosteroids. However, the main disadvantage of DCP therapy is only longer follow-up (18 months). Another factor responsible for the effectiveness of pulse therapy is the patient compliance. On testing, pituitary-adrenal function was not found to be suppressed.^[8]

A study was done by Kothari *et al.* on 25 patients of PV treated with DCP therapy every 28 days where they also found that DCP therapy has shown promising results in terms of long-term remission and minimal side effects.^[9]

CONCLUSION

With the current clinical evidences, it can definitely be said that pulse therapy has changed the treatment and outcome of pemphigus patients. It is a promising therapy providing long-term remission to patients with minimum side effects. Further studies on single or long-term follow-up should be done to find out its true efficacy.

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