International Journal of Current Advanced Research
ISSN: O: 2319-6475, ISSN: P: 2319-6505, Impact Factor: 6.614
Available Online at www.journalijcar.org
Volume 7; Issue 7(D); July 2018; Page No. 14097-14099
DOI: http://dx.doi.org/10.24327/ijcar.2018.14099.2544

CASE REPORT

PEMPHIGUS VULGARIS A RARE AUTOIMMUNE DISEASE: A CASE REPORT

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A R T I C L E I N F O

Article History:

Received 5th April, 2018
Received in revised form 24th May, 2018 Accepted 20th June, 2018
Published online 28th July, 2018

Key words:

Pemphigus vulgaris; Rare Autoimmune disease; Dexamethasone-Cyclophosphamide-Pulse (DCP) therapy.

A B S T R A C T

Pemphigus vulgaris (PV) is a potentially life threatening disease that causes erosions and blisters of the skin and mucous membranes. It is a rare disease involve the mucosa and skin through the disintegration of cellular adherence (acantholysis), resulting in intradermal bullous disease. The lesions are usually painful and untreated generalized Pemphigus vulgaris may be a fatal one. A 39 year old male patient with known history of Pemphigus vulgaris completed DCP therapy I phase and is on interviewing oral Cyclophosphamide 50mg/day was admitted with chief complaints of new lesions since 2 days over scalp and trunk. Successful implementation of Dexamethasone-Cyclophosphamide-Pulse (DCP) therapy has reduced the mortality rate dramatically along with other adjuvant drugs. Untreated, Pemphigus vulgaris is often fatal because of the susceptibility to infection and fluid-electrolyte disturbances hence, utmost importance to be given for PV treatment.

INTRODUCTION

The term "Pemphigus", is derived from the Greek word Pemphix (bubble or blister) [1] and vulgaris is derived from Latin word (common).[2] Pemphigus is a potentially life threatening disease that causes erosions and blisters of the skin and mucous membranes.[3] Pemphigus vulgaris (PV) is a chronic, rare and intra-epidermal bullous disease with a potentially fatal outcome. It was originally named by Wickman in 1791. Pemphigus is an uncommon disease an incidence rate ranging from 0.5 to 3.2 per 100,000 per year.[4] Men and women are equally affected. The mean age of onset is 50–60 years. Pemphigus vulgaris has been observed in children and in the elderly too.[5] Pemphigus encompasses four related diseases with autoimmune etiopathogenesis: Pemphigus vulgaris, vegetans, erythematosus and foliaceus. Only the vulgaris and vegetans types attack the oral mucosa.[6] Pemphigus vulgaris is characterized by alterations in the intracellular links of the Malpighian stratum spinosum, where some structures become antigenic and stimulate the production of autoantibodies, mainly immunoglobulin G (IgG) The etiology of these modifications is not currently known, but the target of the autoantibodies is desmoglein-3 (Dsg3), a glycoprotein that interacts with components of the dense plaque of the desmosomes. The consequence of this interaction is damage to the links among epidermal cells, which creates suprabasal slots that appear clinically as bullous disease.[6]

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There are no population based surveys from India available to study the prevalence of this disease.[7] These rare diseases involve the mucosa and skin through the disintegration of cellular adherence (acantholysis), resulting in intradermal bullous disease.[8] The nasal mucosa, palpebral area, genitalia, pharynx, esophagus, and larynx can also be affected.[9] Pemphigus vulgaris presents as flaccid, thin walled vesicles and/or bullae, that usually rupture to leave an area of erosion and ulceration.[10] 80-90% of patients with pemphigus vulgaris develop oral lesions sometime during the course of disease, and, in 60% of cases, the oral lesions are the first sign.[3] The oral lesions begin as bleb like blisters or as diffuse gelatinous plaques. Rupture of the bullae occurs in an early stage and may be caused by slight rubbing or minimal mucosal trauma. The lesions are usually painful. Untreated generalized Pemphigus Vulgaris may be fatal. Therefore, by recognizing the oral lesions of Pemphigus vulgaris, the clinician has a responsibility in the early diagnosis of the disease, which is of the utmost prognostic importance.[11]

Dexamethasone- Cyclophosphamide Pulse Therapy

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.[11] The entire treatment was divided into four phases as per Pasricha *et al.* schedule.[12]

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.

Case study

A 39 year old male patient with known history of Pemphigus vulgaris completed DCP therapy I phase and is on interviewing oral Cyclophosphamide 50mg/day was admitted with chief complaints of new lesions since 2 days over scalp and trunk. History of moderate grade itching and burning sensation over the lesion. No History of associated fever and stoppage of medications. Not a known case of DM/HTN/BA/TB. Patient is a tobacco chewer since 15 years.

Systemic and Local Examination: CNS, CVS and RS were within normal limits.

Hair: Androgenetic Alopecia. (Fig.1).

Cutaneous: Few Flaccid vesicles over back of trunk. Crests over back of trunk, scalp, and beard area of face with underlying skinny layer. (Fig.2).

Hyper pigmented patches on base of the Nose and Labia. (Fig.3)

Blood Pressure: 140/90 **Pulse Rate:** 78
Patient Pathology report shows:
Total WBC Count: 6800 cu/mm
Neutrophils: 68.7%
Lymphocytes: 17.9%
Eosinophils: 1.8%
Basophils: 0.1%
Monocytes: 11.5%

Diagnosis

The diagnosis of Pemphigus vulgaris can be made based on the clinical and histopathological characteristics of the disease. However, histopathological examinations with conventional staining and indirect and direct immunofluorescence are essential to obtain a definitive diagnosis of Pemphigus. In this case, after histological confirmation of the disease, the patient was referred for treatment.

Medications

100 mg of dexamethasone dissolved in 500 mL of 5% dextrose, Tab Acuclav (Amoxicillin 500mg + Calvulinic acid 125mg) (BD), Tab Cycloxam 50mg (Cyclophosphamide) (OD), Calinta kit (Calcium 500mg + Calcitriol 0.00025mg + Zinc 20mg + Ibandronic acid 150mg)(OD), Tab Teczine (Levocetrizine 5mg) (OD)Tab Rantac (Ranitidine 150mg) (BD) Tab Omnacortil (Prednisolone 40mg), Candid mouth paint (Clotrimazole) (TD), Gentian Violet.

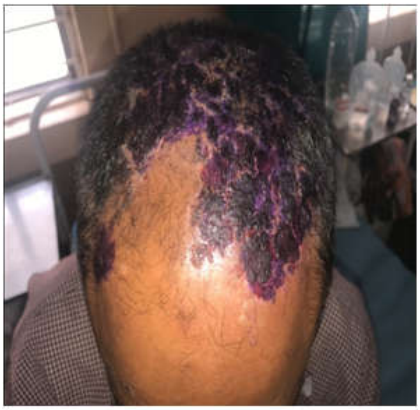

Fig. 01. Alopecia

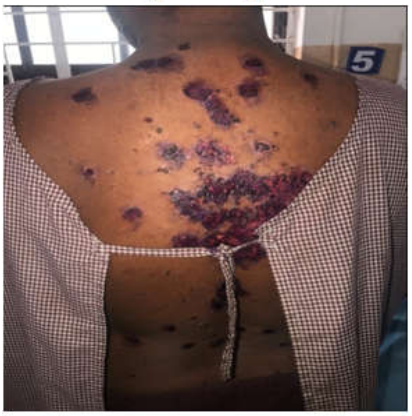

Fig. 02. Flaccid vesicles

Fig. 03. Hyper pigmented patches

DISCUSSION

Pemphigus vulgaris is a rare and autoimmune disorder if not treated well it ends up with fatality. In Pemphigus vulgaris, lesions at first comprise small asymptomatic blisters, although these are very thin-walled, they easily rupture giving rise to painful and hemorrhagic erosions.70-90% cases the first signs of disease appear on the oral mucosa. While lesions can be located anywhere within the oral cavity, they are most commonly found in areas like cheek, mucosa, tongue, palate and lower lip.[13] In the present case the lesions were seen on lower lips, nose, back of scalp and trunk and on head.

The etiology of this case is still unknown. These groups of diseases are characterized by the production of antibodies against intercellular substances so, therefore, classified as autoimmune diseases.[13] Other initiating factors reported included certain food (Garlic), infections, neoplasms and some drugs like Captopril, Pencillamine and Rifampicin.[14] In this

case multiple drugs like Amoxicillin calvulinic acid, Levocetrizine, Prednisolone were used to treat the Pemphigus vulgaris.

CONCLUSION

The treatment of Pemphigus vulgaris has been a challenge for decades. However, the mortality of patients with Pemphigus vulgaris has been significantly reduced with the advent of new therapies and treatment modalities. The Healthy food, time to time treatment with Zinc and Calcium and topical application of Clotrimazole helped patient to decrease lesions severity. Treatment with systemic steroids and Cyclophosphamide has reduced the mortality rate dramatically. Untreated, Pemphigus vulgaris is often fatal because of the susceptibility to infection and fluid-electrolyte disturbances hence, utmost importance to be given for PV treatment.

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How to cite this article:

Biradar S M *et al* (2018) 'Pemphigus Vulgaris A Rare Autoimmune Disease: A Case Report', *International Journal of Current Advanced Research,* 07(7), pp. 14097-14099. DOI: http://dx.doi.org/10.24327/ijcar.2018.14099.2544

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