Abstract

Pemphigus vegetans is a rare vesiculobullous autoimmune disease which is a rare variant of pemphigus vulgaris characterized by vegetating plaques in the flexures and lesions in the oral cavity. It is a less common disease and involves the mucosa and skin due to disintegration of cellular adherence (acantholysis) resulting in intradermal split. The lesions are usually painful and if untreated it may be fatal. A 68 years old male patient was admitted with complaints of fluid filled skin lesions over trunk since 3 months. The lesions later ruptured spontaneously and healed with crusting. The lesions gradually progressed to involve axilla, scalp and extremities with no itching. Successful implementation of dexamethasone –cyclophosphamide –pulse (DCP) therapy along with other adjuvant drugs has induced disease remission.

Key words: Pemphigus vegetans, Autoimmune disease, Acantholysis.

Introduction

Pemphigus vegetans (Pveg) is a rare vesiculobullous autoimmune disease which is a less common variant of pemphigus vulgaris characterized by heaped up, cauliflower like vegetating plaques in the flexures and rarely in oral cavity. This disorder affects chiefly middle aged adults(1,2).It is caused by auto antibodies directed against desmogleins 1 and 3, which are transmembrane glycoprotein’s and results in loss of intercellular adhesion between intact keratinocytes which results in acantholysis. The incidence of the disease is 0.09 to 1.8%. In a study conducted in India majority of pemphigus patients have been diagnosed to have pemphigus vulgaris followed by pemphigus foliaceus, pemphigus erythematosus and pemphigus vegetans. This disease occurs mainly on interiginous areas, scalp and face but may occur anywhere on the skin surface. The oropharynx, oesophagus, stomach, duodenum, anus, nasal vulvovaginal, laryngeal and conjunctival mucosa can also be affected (3). Oral involvement is reported in 60-80% of pemphigus cases and cerebriform tongue, characterized by a pattern of sulci and gyri of the dorsum of tongue has been reported in 50% cases of Neumann type vegetans (4). In Pemphigus vegetans blisters are fragile that usually rupture and leave an area of erosion and have a tendency to develop excessive granulation tissue and crusting in some patients. A positive Nikolsky sign (positive when gentle shearing pressure to pink or normal looking skin results in the formation of an erosion or extension of bulla) often can be elicited. Possible complications of pemphigus include infection of skin, sepsis, rarely death. Pemphigus isn’t contagious and there is no way to predict who will get it.

Case report

A 68 years old male patient was admitted in dermatology ward with complaints of fluid filled skin lesions over trunk since 3 months. The lesions later ruptured spontaneously to leave

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behind raw areas, which got covered with crusts. The lesions gradually progressed to involve axilla, scalp and extremities. The lesions were painful and not associated with itching.

On Cutaneous examination, multiple crusted plaques well noted over anterior and posterior aspect of trunk and lower extremities, vegetating plaques, vesicles and bullae on bilateral axilla, umbilicus and hyper pigmented patches on scalp and upper extremities were observed (Fig. 1 and Fig. 2). Hyper pigmented ridges of the nails and buccal mucosa was observed with few hyper pigmented patches, some of the erosions were secondarily infected with pus formation.

On investigation complete haemoglobin and liver function tests were within normal limits except for mild hypoproteinemia. Culture from pus produced grants of gram positive cocci and gram negative bacilli.

**Diagnosis:** A provisional diagnosis of Pemphigus vegetans was made based on the clinical features. A Tzanck smear was prepared from a fresh vesicle and acantholytes cells were noted. Confirmation by skin biopsy is done after observing suprabasal split in histopathological examination and immunoglobulin G deposits within the epidermis in direct immunofluorescence.

**Medication:** After confirmation of diagnosis, patient was treated with DCP (140 mg of dexamethasone dissolved in 500 ml of 5% dextrose for 3 consecutive days, 500 mg of cyclophosphamide was also given on the second day) After 3 days patient was started on prednisolone 30 mg/day. Patient was also given appropriate antibiotics, calcium supplements, antacids and anti- histamines.

**Discussion**

Pemphigus vegetans is a rare autoimmune disease and affects people irrespective of gender. This disease occurs mainly on interiginous areas, scalp and face, Similar to that in the present case patient developed fluid filled lesions initially over trunk which later ruptured and got covered with crusts. The lesions gradually progressed to axilla, scalp and extremities. The aetiology in the present case is unknown. As it is auto immune disease in most cases it is unknown what triggers the disease but the mechanism underlying the pathology is IgG auto antibodies directed against desmogleins, a keratinocytes protein which helps in intercellular adhesion and results in acantholysis histologically and bulla formation clinically. Sometimes, pemphigus develops as a side effect of medications, such as antihypertensive drugs and chelating agents, this
type of disease disappears when medication is stopped. A preliminary diagnosis of Pemphigus vegetans was established in the present case on the basis of presenting clinical features and a Tzanck smear test, which reveals multiple acantholytic cells (Tzanck cells). The smear test is quite significant in the early diagnosis of Pemphigus vegetans and can be performed with ease and rapidity (5).

The goal of treatment is to induce complete remission while minimizing treatment related adverse effects. The first priority for patient management is to attain rapid disease control. This is typically achieved through the administration of systemic glucocorticoids. Although systemic glucocorticoids therapy is highly effective, the high doses and long treatment periods that are needed to maintain the clinical response may lead to serious or life-threatening side effects. Introduction of dexamethasone-cyclophosphamide pulse (DCP) therapy for pemphigus group of disorders by Pasricha and Gupta in 1984 has revolutionized the therapy of pemphigus. Administration of suprapharmacologic doses of drugs in an intermittent manner is known as “pulse therapy”, which refers to intravenous (IV) infusion of high doses of steroids for one or more days for quicker, better efficacy and to decrease the side effects of long term steroids. The most common side effects of pulse therapy are mood and behavior alteration, hypokalemia, diarrhea, arrhythmias and shock. If administered properly, dexamethasone - cyclophosphamide pulse (DCP) therapy has the potential to affect lifelong recovery from pemphigus (6). In the present case, the patient was managed with dexamethasone - cyclophosphamide pulse (DCP) therapy to control and prevent the development of new lesions. The DCP regimen was followed by oral prednisolone therapy 30mg/day and the patient was managed symptomatically.

Conclusion
The treatment of Pemphigus diseases is a challenge. However the mortality rate of pemphigus vegetans has reduced with the advent of new therapies and treatment modalities. Appropriate treatment and personal care helped patient to improve health condition. Untreated Pemphigus vegetans is often fatal because of many possible complications hence importance should be given for pemphigus vegetans treatment.

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REFERENCES