Pemphigus Vegetance
a Clinical , Histopathological and Therapeutic Study
in Iraqi Patients

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Abstract

Background: Pemphigus vegetans is a very rare form of pemphigus vulgaris , with a similar etiology and early histological changes , it comprise only 1-2% of cases and of two variety (Neumann and Hallopeau) . It begins and ends as pemphigus vulgaris clinically and histologically , but many of the denuded areas heal with verrucous vegetations and epidermal hyperplasia , that may contain small pustules in the early stages .

Objectives: The study was done to identify that pemphigus vegetans may be manifested for the first time on the body of the nose , as a well defined, indurated, slightly elevated verrucous, hypertrophic and erythematous plaque .

Patients and methods: Eight patients were seen in the Department of Dermatology and Venereology in Baghdad and Baquba Teaching Hospitals , during the period from Nov. 2001 to Dec. 2004 . All of them were males with a mean age of 45 years (their ages ranged from 30-81years) . They complained of vegetating plaques on the body of the nose , which were diagnosed clinically and histologically as pemphigus vegetans and treated by topical clobetasol dipropionate 0.05% ointment twice daily and 40mg /day prednisolone as a single oral morning dose .

Results: The study shows that pemphigus vegetans may be manifested for the first time on the body of the nose , as a well defined slightly elevated erythematous, verrucous and hypertrophic plaques , with typical histological changes of pemphigus vegetans and good response to topical clobetasol dipropionate as 0.05% ointment and oral prednisolone 40mg /day as a single dose .

Keywords: Pemphigus vulgaris , pemphigus vegetans , clobetasol dipropionate , prednisolone .

Introduction

Pemphigus vegetans is a very rare form of pemphigus vulgaris , with similar etiology and early histological changes [1,2,3] . There are two variety of pemphigus vegetans (Neumann and Hallopeau) , which comprising only 1-2% of all cases of pemphigus , it begins and end as pemphigus vulgaris , but many of the denuded areas heal with verrucous vegetations that may contain small pustules in early stages [4,5,6,7] . Clinically the Neumann type starts at a slightly earlier age than pemphigus vulgaris but its presentation , with involvement of the oral mucosa is otherwise similar [1-3] . The early stages of the skin eruption are identical.
to pemphigus vulgaris, but the eroded areas soon develop hypertrophic granulation at their edges, these vegetations exude serum and pus and are studded with small pustules [1,3,8]. Frequently fresh blisters form at the edges of the vegetations and later give rise to new vegetations [1]. In time the vegetations become dry, heaped up, hyperkeratotic and fissured, flexures are more commonly involved, although vegetations may occur at any site [1,4]. The Hallopeau type is relatively benign, the early lesions consists of pustules arising on normal skin, instead of bullae, which is followed by the formation of gradually enlarged verrucous vegetations especially in intertriginous areas [3,4].

In pemphigus vegetans the early histological changes are similar to that of pemphigus vulgaris, but later downward growth of thick strands of epidermal hyperplasia into dermis gives rise to the appearance of papillomatosis and hyperkeratosis, the dermal infiltrate may consist largely of plasma cells and eosinophils. The infiltrate may produce both eosinophilic spongiosis and eosinophilic pustules in the epidermis and acantholysis may not be present in older lesions. The early lesions of Hallopeau type may reveal more eosinophilic abscesses than in the Neumann type [1,3,5,9,10,11].

Treatment of pemphigus vegetans includes topical and/or intralessional potent corticosteroid in mild cases and systemic corticosteroid, dapsone, etretinate and immunosuppressive drugs in sever and generalized forms. Prednisolone is the preferred treatment with a dose of 1-1.5 mg/kg in combination with topical or intralessional steroids is sufficient to control the disease [1-3].

The prognosis of pemphigus vegetans is there for similar to that of pemphigus vulgaris, however spontaneous remission has been reported and the course is generally more prolonged [1-3].

The aim of the present work is to identify that pemphigus vegetans may be presented for the first time on the body of the nose, as a well defined, indurated and verrucous plaque.

Methods

Eight patients were seen in the Department of Dermatology and venereology in Baghdad and Baquba Teaching Hospitals, during the period from the Nov. 2001 to Dec. 2004. All patients were males, with ages ranged from 30-81 years (mean age was 45 years). They complained of rash on the body of their noses, they were fully examined and interrogated regarding age, occupation, drugs intake, onset and duration of the rash, associated symptoms, other sites of involvement including oral cavity, previous similar attack and family history of the same condition, also the present and past medical and dermatological history was taken. The rash was diagnosed clinically as pemphigus vegetans and confirmed by histopathological studies of skin biopsies, which were carried out on all patients. The patients were treated by topical clobetasol dipropionate as 0.5% ointment, twice daily, with oral prednisolone of 40mg/day as single morning dose.

Results

Eight males patients were studied, with a mean age of 45 years (average 30-81 year), they were presented with plaques on the body of the nose, of two months to one year duration. The plaques are well defined, slightly elevated, erythematous, verrucous, hyperkeratotic, of different size, associated with itching and in 3 patients (37.5%) the mouth was involved later on. Skin biopsies showed changes similar to that of pemphigus vulgaris, with suprabasal cleft and there were a thick strands of downward growth of epidermal hyperplasia into the dermis gives rise to the appearance of papillomatosis, with dermal infiltrate of plasma cells and
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The patients showed good response to topical clobetasol dipropionate 0.05% ointment, twice daily, with oral prednisolone 40 mg/day as single morning dose. Complete clearance of the lesions were obtained after about one month of therapy, with a brownish post-inflammatory hyperpigmentation.

**Discussion**

The present study described a rare skin disease, which was a variety of pemphigus vulgaris, the pemphigus vegetans. In all studies and literatures, pemphigus vegetans like pemphigus vulgaris was usually manifested itself firstly in the oral cavity as bullae and erosions, latter on the skin become involved, usually as bullae, followed by crustation, than vegetation, verrucous and usually on the intertriginous areas [1-4], which was differ from this study, in which the disease was firstly appears on the body of the nose with out bullous eruption and involvement of intertriginous areas, the oral cavity may be involved later on in the course of the disease (37.5%). The lesions consisted of well defined erythematous verrucous and hypertrophic plaques of different size and duration with histological changes, which was similar to other studies and literatures, the age of onset like other studies was similar to that of pemphigus vulgaris [1]. Clinically the response to therapy was better than in pemphigus vulgaris and vegetans of intertriginous areas as complete clearance of the lesions was obtained after about one month of treatment.
References


