

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 dipropion 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 dipropion as 0.05% ointment and oral prednisolone 40mg /day as a single dose .

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

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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 vegetance 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 intralesional 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 intralesional 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 conferred by histopathological studies of skin biopsies , which were carried out on all patients . The patients were treated by topical clobetasol dipropionit 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 a 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

eosinophils , which produce eosinophilic spongiosis and eosinophilic pustules in the epidermis (Figures-1,2) .

The patients showed good response to topical clobetasol dipropionat 0.05% ointment , twice daily , with oral prednisolone 40mg /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 .

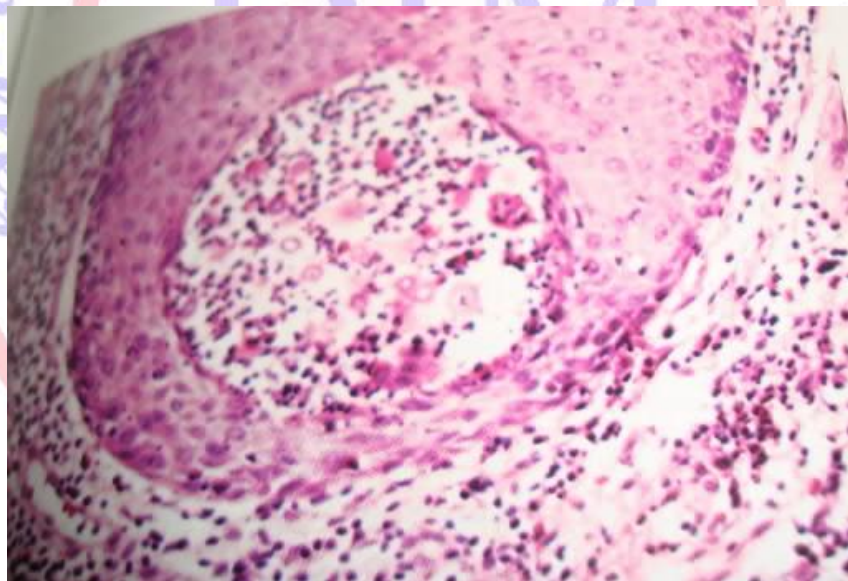


Figure (1)

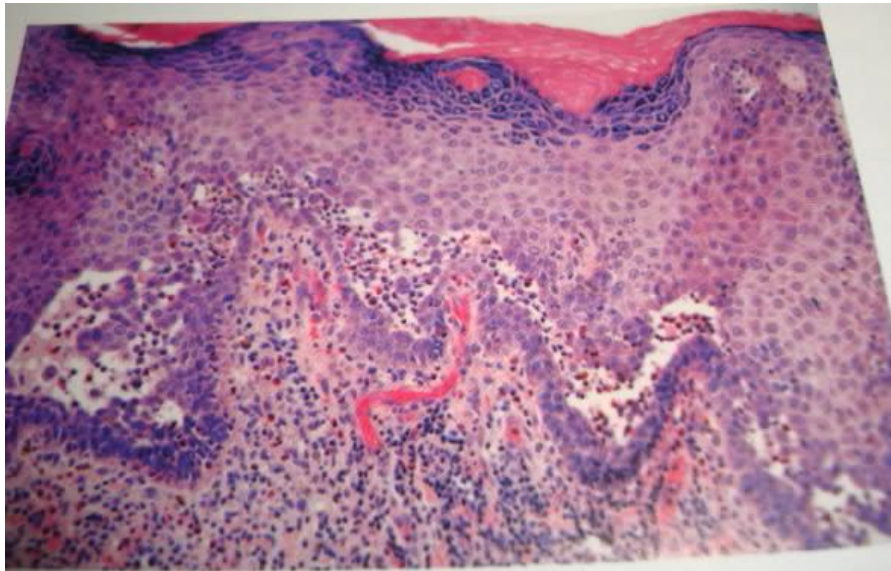


Figure (2)

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