

PEMPHIGUS VEGETANS

By

(A case report with a brief review of literature)

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INTRODUCTION

Pemphigus signifies a group of relapsing usually fatal bullous diseases (Andrews 1957). It is an important dermatological entity inasmuch as its aetiology is as yet not understood and its treatment is a difficult problem. The name Pemphigus is derived from the Greek word Pemphix meaning pustule.

Credit goes to Civatte (1943) who pointed out that this condition is clearly distinguishable histologically from other bullous lesions such as erythema multiforme and dermatitis herpetiformis even when clinical differentiation is difficult. Other authors (Dupont 1951 and Director 1952) also subscribe to this view.

The purpose of this paper is to report a case of Pemphigus Vegetans which on follow-up for over 6 months terminated as a classical case of Pemphigus Vulgaris. In view of the fact that such reports are extremely rare in Indian literature and none to our knowledge from Kerala, we thought it worthwhile placing this on record.

CLINICAL ASPECTS

Pemphigus Vegetans is a rare type of pemphigus, characterised by fungoid vegetations, involving particularly the axillary and crural folds. The onset of this condition may solely be manifested by bullae in the mouth or about the genitalia or the umbilicus. Its course is protracted with intervals of long remissions. Even though pemphigus is usually associated with systemic symptoms such as pyrexia, anorexia and adrenal exhaustion, the subjective symptoms in vegetans are slight if these vegetative lesions involve the anogenital region. Conditions like syphilitic condylomata, granuloma inguinale and amoebic granuloma merit consideration in the differential diagnosis.

Histologically certain diagnostic features are observable and most elementary change is the disappearance of intercellular bridges in the epidermis—a process Civatte called 'acantholysis'. This usually occurs in the supra basilar layers. Director (1952) states that this change may also be seen in the basal layer and may even extend to pilosebaceous follicle and at times to the sebaceous glands themselves.

As regards the aetiology, this is a subject of much speculation. Hence only numerous theories have been propounded which state that viruses and bacteria may have a aetiological role. There may even exist some hitherto undetected metabolic and/or genetic factors that may have a role in its causation. The beneficial effect of Vitamin B₁₂ in certain intractable skin diseases like psoriasis and



Fig. 1—Showing on first admission the vegetative growths in the axilla—Pemphigus Vegetans



Fig. 2—Lesions of the lip. (on second admission)



Fig. 3—Showing on second admission evidence of typical eruptions of Pemphigus Vulgaris in the region of the scrotum.



Fig. 4—Showing on second admission the change of the vegetations to eruptive lesions of Pemphigus Vulgaris in the axilla.

seborrhic states has to be reckoned with. The exact role of these factors and the extent to which they act remain to be studied in future.

CASE REPORT

First admission :—7-1-1964. A patient (N), Hindu, aged 57 years, presented himself in the Skin Department of the Medical College, Calicut with multiple vegetation in both the axillae, groin and neck (Figure 1). He said that the disease started in the right axilla and had received treatment with some antibiotics for this. However it recurred and with recurrence he noticed the lesion had also appeared in the left axilla, groin and neck. The duration of this disease is about a year. General examination showed that the patient was a wellbuilt individual who was otherwise fairly well but for this skin condition. There were many vegetative growths in both the axillae, groin and neck. The mucous membrane of the mouth was also involved. His blood pressure was 110/70. Urine examination showed no abnormality. Biopsy was done on one of the vegetative growths. The histopathological report was as follows: The epidermis appears hypertrophied and hyperplastic with numerous downward projections. There were many abscesses composed mainly of eosinophils and a few neutrophilic polymorphs in varying stages of degeneration. These abscesses were mainly seen in the elongated projections of the epidermis. There were in addition collections of chronic inflammatory cells in the sub epidermal zone.

Diagnosis :—Pemphigus Vegetans (Microphotograph 1).



Microphotograph 1—Hyperplastic epidermal downgrowths c̄ evidence of intraepidermal micro abscesses. A large abscess is shown consisting predominantly of eosinophils.

Treatment :—(1) Prednisolone 20 mgm, daily for the first 15 days.
(2) Prednisolone 15 mgm, daily for the next 30 days.

- (3) Prednisolone 10 mgm. daily for the next 30 days.
- (4) Demethylchlor tetracycline 1 gm. daily for 5 days.
- (5) Chloramphenicol 2 gm. daily for 5 days.
- (6) Supportive therapy with liver extract and vitamin B complex given throughout the period of treatment.

The patient showed fairly good response to this line of treatment. The patient then left the hospital.

Second admission :—13-8-1964. He gave the history of stopping Prednisolone 2 weeks prior to seeking admission for a second time. This time the lesions were confined to the scalp, groin and mucous membrane of the mouth. There was one ulcer on the tongue. The cutaneous lesions were all infected and the patient looked very ill. (Figure II and III). Treatment given was Prednisolone 40 mgm. daily. Biopsy was done on one of these lesions and the report stated that it was Pemphigus Vulgaris.

COMMENTS

This case is a typical report clearly showing that Pemphigus Vegetans finally ends as Pemphigus Vulgaris. Lever (1961) has stated that Pemphigus Vegetans commences and terminates as vulgaris. On the other hand, Director (1952) is of the opinion that Pemphigus Vegetans is merely a reactive phase in the course of Pemphigus Vulgaris associated with increased resistance manifests itself with regression of dry papules and formation of vegetations.

True Pemphigus includes Pemphigus Vulgaris and Vegetans (Sneddon, 1961). The histology of this with its classical features is very important and has always remained true to type in contrast to conditions simulating Pemphigoid. The histology of Pemphigus Vulgaris and of Vegetans is more or less identical but for the villi formation and downward extension of epithelial strands both of which are very pronounced in the latter. With the formation of vegetations the typical acantholysis may not be detected, as it was so in the case reported here. However, the intra epidermal abscesses composed predominantly of eosinophils are highly pathognomonic of this condition.

Pemphigus like eruptions sometimes are apparently associated with malignant neoplasms of the internal organs (Goldsmith and Hellier, 1954). Whether true Pemphigus is similarly observed with such visceral involvement is a matter for study.

SUMMARY

A rare case of Pemphigus Vegetans is reported with a follow-up for a period of over 6 months. The termination of the condition agrees with the view expressed by others. A brief review of literature with discussion on certain salient features as regards aetiology is appended.

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