# PEMPHIGUS VEGETANS

**( A case report with a brief review of literature )**

By

RATAN SINGH* & KHURANA J. C.**

The word "Pemphigus" is derived from the Greek word 'Pemphix' meaning pustule. The term is applied to a group of relapsing usually fatal bullous disease characterised histologically by the phenomenon of acantholysis. Its commonest variety, pemphigus vulgaris is not uncommonly met with by workers in the field all over the world.

Pemphigus vegetans is the rarest variety and was first described by Neumann in 1886. It is a rare variant of Pemphigus vulgaris and occurs in those who have increased resistance to the disease. Two types have been described, the so called Neumann type in which the patient's resistance is slightly increased and begins and ends as Pemphigus vulgaris. The second variety known as Hallopeau type (Pyodermite Vegetantes) occurs in those with considerable resistance to disease, is more benign and may terminate as Neuman type.

The intertrignious areas eg. axilla, groins, inframammary regions, genitalia & perineum and scalp are the sites which show fungoid vegetative lesions. Papillomatous and hyperkeratotic vegetative granulation tissue slowly develops on erosive areas left after the bullae have ruptured. These lesions become verrucous masses covered with moist, malodorous, papillomatous, crusted excrescences which coalesce to form large areas. Subjective symptoms are slight and often there are long remissions in the course of the disease. Occasionally high fever and other constitutional symptoms develop due to sepsis.

The disease has to be differentiated from vegetative syphilides, Iodo-or Bromodermas, and in the anogenital region, from granuloma Inguinale, condyloma accuminata, and lymphogranuloma venereum. Mycotic and amoebic granulomas may also resemble pemphigus vegetans.

Histology : By the time the verrucous vegetations have formed, acantholysis and formation of clefts often are no longer apparent. The verrucous vegetation are characterised by papillomatosis, acanthosis and irregular downward proliferation of the strands of epidermis, not infrequently containing intraepidermal abscesses composed almost entirely of eosinophils (Lever, 1967). However, erosions and bullae show essentially the same histological picture as seen in pemphigus vulgaris.

Desai and Rao (1960), Kandhari & Pasricha (1965) & Ambady et al (1965) in their series of 21, 34 and 21 cases respectively, did not come across any case of pemphigus vegetans. A few case reports of pemphigus vegetans, all from the southern peninsula

*Professor and Head of the Department, **Postgraduate; Department of Dermatology & Venereology, Maulana Azad Medical College & associated Irwin & G. B. Pant Hospitals, New Delhi-1, India.

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of the Indian subcontinent have been reported (Kutty & Joy–1965, & Amin et al 1968). Of 141 cases of different varieties of pemphigus diagnosed and treated in Skin & V. D. Department of Irwin Hospital, New Delhi from 1964 to 1968, not a single case of pemphigus vegetans is on record. In view of the facts that such reports are rare in Indian literature and none to our knowledge from Northern India, it was thought worthwhile, placing on record, a case of pemphigus vegetans, diagnosed and treated in the Skin & V. D. Department of Irwin Hospital, New Delhi.

## CASE REPORT

A married Hindu male aged 25 years, resident of Punjab (North India) was referred from a sister institution to the V. D., O. P. D. of Irwin Hospital on 25–1–69 as a case of lympho granuloma venereum. The patient had a foul smelling vegetative mass in the inguino-perinial regions of 2½ years duration. The disease first started on the coronal sulcus of the penis and received treatment with some antibiotics and was relieved. After a short while, it relapsed; this time involving the scalp, and right axillae also. A year later vesico-bullous lesions appeared on the abdomen and this graduaily spread to all other parts of the body. The lesions healed with antibiotics and corticosteroids leaving behind hyperpigmented macular lesions. Muccus membrane was never involved and itching was mild. No other subjective symptoms were present.

Examination revealed foul smelling, cauliflower like, reddish-brown vegetative mass studded with pustules, crusts and ulceration involving both groins more on right side and adjoining areas. Similar lesions, but smaller in size and less offensive were seen on the scalp and right axilla. Trunk and proximal parts of extremities showed numerous hyperpigmented macular lesions of different sizes. Oral mucosa was free and there was no adenopathy. Some of the finger nails showed beaus lines; otherwise the patient was well nourished and well built.

## LABORATORY INVESTIGATIONS

Hb. 10 gm%. Total Leucocytes count 9000/ cmm. with Polymorphs–64%, Lymphocyte 30%, Eosinophils 4%. and Monocyte 2%. E. S. R. 10 mm/first hour.

Urine & Stool N. A. D. Serum Electrolytes Na 138 mEq/l and K. 4.6 mEq/l. Tzanck Test & S. T. S. were negative.

X–ray chest was normal.

Biopsy was taken from the vegetative Lesions. It showed intra-epidermal bullae containing a few acantholytic cells, and numerous eosinophils and polymorphs. Epidermis was acanthotic with marked proliferation of reteridges, some of them containing intraepidermal abseesses composed chiefly of eosinophils.

He was put on prednisolone 40 mg daily to begin with, antibiotics to control sepsis and supportive therapy. Topically he was given condy's washes and lotio gentian violet 1%. The lesions showed dramatic improvement but later firm papular lesions, which failed to regress were treated with superficial X–ray therapy and

responded well. The patient is on a maintenance dose of 15 mg. of Prednisolone daily and is being followed up.

## COMMENTS

The etiology of Pemphigus has now been sighted in the distant horizon. The auto-immune concept is fast gaining ground (Donold et al, 1966; Edwin et al, 1966; Tadeusz et al, 1966; Ernest et al, 1968 & Snedden, 1968). But why out of the four related variants of Pemphigus namely Pemphigus vulgaris, Pemphigus foliaceous, Pemphigus erythematosus and Pemphigus Vegetans, Pemphigus Vulgaris is the one seen most widely all over the world and Pemphigus Vegetans is the rarest one, is not precisely known. Director (1962) is of the opinion that Pemphigus Vegetans is merely a reactive phase in the course of Pemphigus Vulgaris associated with increased resistance of the host. This is further supported by the observation that patients of Pemphigus vulgaris, who have turned the corner and lesions start healing, often develop vegetative lesions very akin to pemphigus vegetans in the axilla and other body folds. But we feel that racial susceptibility and environmental factors have equally important bearing to the type of disease. This may explain the high incidence of Pemphigus foliaceous (Fogo-selvagem) in South America. Desai and Rao (1962) from Western India had 8 cases of Pemphigus folicaeus out of a total of 21 cases, whereas Kandhari and Pasricha (1961) from Northern India had only 3 cases, of Pemphigus foliaceus out of a total of 34 cases. Also the few case reports of Pemphigus Vegetans from India, are all from the South. Racial and environmental factors thus seem to influence the pattern of Pemphigus and need further study.

## SUMMARY

A case of Pemphigus which is a rare variant of Pemphigus vulgaris is reported. A brief review of literature with discussion on its etiology is presented.

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