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# Unusual clinical and pathological presentation of pemphigus vegetans

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**Abstract**
Pemphigus vegetans is a rare form of pemphigus vulgaris. It is an autoimmune disease characterised by flaccid bullae or pustules that erode to form hypertrophic papillated plaques involving predominantly skin fold, scalp, face and mucous membranes. Here, the authors reported a case of 59-year-old male presenting with chronic verrucous lesions involving the scalp, oral cavity and perianal area, which was initially mistaken as malignancy.

**Keywords:** Pemphigus vegetans, hallopeau type, unusal presentation, diagnosis, treatment

## Introduction
Pemphigus vegetans is an uncommon subtype of Pemphigus Vulgaris. This disorder usually classified as- Hallopeau Type, and Neumann Type.
The former starts with circumscribed pustules, runs a benign course and heals as vegetating plaques; while in contrast, the later type has vesicles and bullae as 1st degree lesions that are often refractory to therapy [1]. The diagnosis of Pemphigus vegetans is based on clinical manifestations and confirmed by histology.

## Case Report
A 59 yr Indian male presented with a large fungating lesion over the scalp for 4 months. The lesion was crusted, foul smelling with honey colored exudates. It started as small pustule, rapidly increased in size, was slightly itchy. It involved the perineal region and both inguinal regions subsequently. Oral cavity involvement included apthoid ulcers that waxed and waned with time.
The patient is non diabetic, normotensive with no family history of any major ailments. Initial diagnosis was pyoderma vegetance or pyostomatitis vegetance. His general physician started with a course of antibiotic (amoxicillin-clavulunate),topical antifungal (miconazole) + antibiotic (mupirocin).Discomfort worsened and was referred to surgery OPD where 56 maggots were removed and dressing was done along with blood sugar estimation and continuation of same antibiotic was suggested. Later biopsy was done ruled out any verucuous carcinoma.
Biopsy was done from scalp to rule out squamous cell (Verrucous) carcinoma. Histopathology shows hyperkeratosis, parakeratosis, prominent epidermal hyperplasia and intra corneal pustule formation. But it was not conclusive of malignancy. Pain and discomfort alleviated but lesion progressed and spread. A repeat biopsy was suggested from more representative area. Repeat deep biopsy from scalp and biopsy from inguinal region was done. The histopathology shows hyperkeratosis, parakeratosis, acanthosis, epidermal hyperplasia, papillomatosis, intraepidermal acantholysis extending into adnexal structures resulting to intraepidermal and suprabasal clefts formation. Prominent eosinophilic and neutrophilic celluiar infiltrate with formation of eosinophilic spongiosis, eosinophilic microabscesses and subcorneal pustules. There was no evidence of malignancy. Histopathological diagnosis done as Pemphigus vegetans, hallopeau type.
Immunoflourescense was suggested but the patient could not afford it. He was started on oral corticosteroid (prednisolone 1mg/kg/day) to which he responded dramatically. After a few weeks there is complete healing of the lesions and only hyperpigmentation on the perianal area was present.

## Discussion

The most common type of Pemphigus is Pemphigus vulgaris. [1, 15]. Pemphigus vegetans,a rare variant of pemphigus vulgaris, was 1st described by Neumann in 1876 [2].

Pemphigus vegetans represents 1-2% of all Pemphigus [3].

Two subtypes – Neumann and Hallopeau types are recognised, which are differentiated based on their clinical presentation, histopathology, course and response to treatment [3, 5].

Pemphigus vegetans, especially the hallopeau variant has no resemblance to a vesico-bullous disorder and it presents as pustules as primary lesions instead of bullae. Their development is followed by the formation of gradually enlarging verrucous vegetations, especially in intertriginous areas. Heaped up vegetating and verrucous plaques mainly occur on the flexures. It is often found on the groin, armpits, thighs, eyelids and perioral region [6]. Oral lesions are common [7].

The most important differential diagnosis is pyodermatitis, pyostomatitis vegetans, which has similar clinical and histological manifestation but negative IF results [8] and close association with underlying inflammatory bowel disease. Other diseases which may mimic presentation of Pemphigus vegetans include deep fungal infections, halogenodemas, Tuberculosis, Verrucosa cutis, atypical mycobacterial disease, vegetative pyoderma gangrenosum, giant keratocanthoma, botryomycosis, verrucous carcinoma, squamous cell carcinoma and SJS [9-12].

Pemphigus antigens are highly restricted in their distribution, and are normal constituents of suprabasillar cells and the apicolateral aspect of basal cells within stratified squamous epithelium The major antigen involved in Pemphigus vegetans is Desmoglein-3, which has a molecular weight of 130 Kd. This is the same antigen responsible for pemphigus vulgaris [9]. Mucosal dominant disease is more frequently associated desmoglein3 and mucocutaneous disease associated with autoantibodies against both desmoglein3 and desmoglein1 [3].

Early lesions of pemphigus vulgaris and Pemphigus vegetans, both show suprabasal acantholysis. The hallmarks for Pemphigus vegetans lesion are – epidermal hyperplasia,suprabasal cleft, papillomatosis, and prominent eiosinophilia [6, 13]. DIF studies of perilesional skin in Pemphigus vegetans are reported to be identical to there in pemphigus vulgaris [14].

In this case the clinical diagnosis was initially pyoderma vegetans or pyostomatitis vegetans. Later biopsy was done to rule out verrucus carcinoma. But initial biopsy was inconclusive of malignancy and a repeat biopsy was suggested to rule out verrucous carcinoma. The repeat biopsy was then diagnosed as Pemphigus vegetans, Hallopeau type.

There is excellent response to steroid of the patient, but not responding to Systemic and local antibiotic. The gold standard for treatment for treatment is systemic corticosteroids [3, 16].

Hence we can see that the clinical appearance of Pemphigus vegetans is difficult to differentiate from other non - neoplastic and neoplastic diseases. The main pitfall is clinical misdiagnosis or missing the diagnosis due to lack of familiarity or clinical suspicion. So, we would like to stress on the fact that even though Pemphigus vegetans may be a rare entity, it should be considered as a differential in verrucous lesions, while diagnosing, especially in cases not responding to standard treatment.

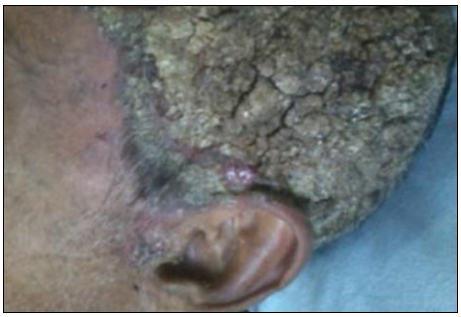

**Fig 1:** Fungating lesion over parieto-temporal region of scalp

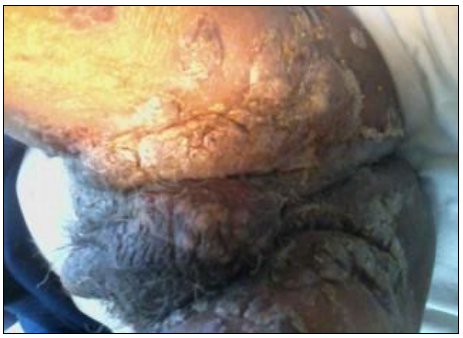

**Fig 2:** Honey crusted exudative lesion over perianal area

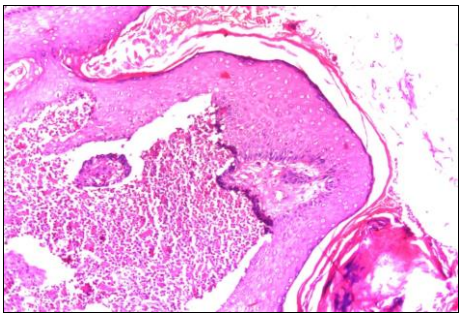

**Fig 3:** H & E stain (10x) showing intraepidermal acantholysis & Eosinophilic abscess in papillary dermis

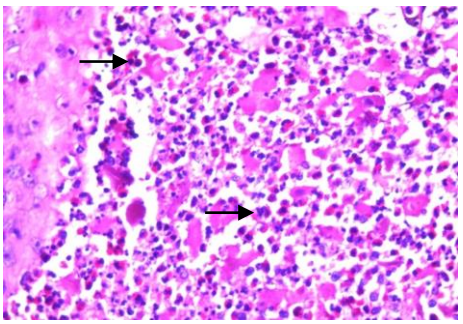

**Fig 4:** H & E stain (40x) showing details of eosinophilic abscess in papillary dermis

## Conclusion

Pemphigus vegetans is an uncommon subtype of Pemphigus Vulgaris. This disorder usually classified as- Hallopeau Type, and Neumann Type. Clinical appearance of Pemphigus vegetans is difficult to differentiate from other non-neoplastic and neoplastic verrucous lesion. Clinical

misdiagnosis is due to lack of familiarity or clinical suspicion of this entity. Pemphigus vegetans may be a rare entity but it should be considered as a differential in verrucous lesions, especially in cases not responding to standard treatment.

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