www.jmscr.igmpublication.org
Impact Factor 5.84
Index Copernicus Value: 71.58
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v5i9.120

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Pemphigus Vegetans of Hallopeau Type

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Abstract
Pemphigus vegetans is a rare variant of pemphigus vulgaris that is characterized by vegetating lesions primarily in the flexures. A 49 year old male having chronic kidney disease had a vegetating growth of groin, perianal area and over scalp for the last 6 months and was clinically diagnosed as multicentric squamous cell carcinoma and after skin biopsy it was diagnosed as pemphigus vegetans of Hallopeau as histology showed intraepidermal eosinophilic abscess and acanthosis and responded dramatically by systemic steroids.

Keywords: *Pemphigus Vegetans, Pemphigus Vulgaris, Hallopeau.*

Introduction

Pemphigus vegetans is a rare variant of pemphigus vulgaris, first described by Neumann in 1876.[1] Two subtypes –Neumann and Hallopeau.[2,3] Pemphigus vulgaris is an autoimmune disorder characterized by production of IgG autoantibody against intercellular adhesion protein desmoglobin leading to vescicobullous lesion and acantholysis and easy to diagnose most of the times. Unlike pemphigus vulgaris which produces lesions on skin and mucosa, pemphigus vegetans, especially Hallopeau variant produces extensive vegetation which mimics squamous cell carcinoma.

The cutaneous lesions in the Hallopeau type begin as pustules and heal as vegetating plaques while those in Neumann type, with a worse prognosis, are characterized by vegetations developing during the course of pemphigus vulgaris.[3] Vegetating lesions seem to be a reactive pattern of the skin to the auto-immune insult , with certain areas of the skin showing more of a tendency to form vegetations. Local moisture, heat and friction are important factors in their development. The sites involved are the intertriginous areas (the axillae, groins and inframammary folds), scalp and the face.[3,4,5]

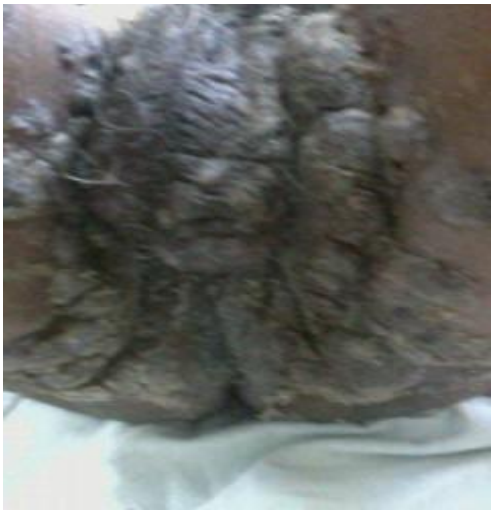
Case Report

A 49 year old male, farmer came to the dermatology department with cauliflower like proliferative growth in the groins and perianal area for the past 6 months. He also developed similar lesion over the scalp with bleeding and pus formation. The scalp lesions was well-defined, approximately 6 × 5 cm. Cutaneous examination revealed multiple, gray colored, irregular hypertrophic, vegetating, oozing plaques present in the both groin, perianal region and on scalp [Figure 1]. No definite vescicobullous lesions are noted neither over the skin nor on the oral mucosa. Patient had inguinal lymphadenopathy 1x1 cm, discrete, non-tender, mobile and firm with normal overlying skin. The patient was a

heavy smoker and a known patient of chronic kidney diseases. The case was suspected as multicentric squamous cell carcinoma.

Investigations

His routine investigations revealed eosinophilia (absolute eosinophil count- 750/cu mm)and raised serum creatinine (2.4 mg/dl). Other findings like hemoglobin concentration, liver function tests, and X-ray chest were normal. Rest of the muco-cutaneous and systemic examinations were within normal limits. VDRL in serial dilutions was non-reactive and TPHA was negative. KOH preparation from the lesion was negative for any fungus. Gram stain from the lesion demonstrated numerous neutrophils, eosinophils along with gram positive and gram negative bacilli. Repeated culture showed contaminants only. A skin biopsy was done from peri-anal papule and received for histopathological examination. Histopathology revealed intraepidermal eosinophilic abscesses, few acantholytic cells and a supra-basal cleft. Dermis showed intense inflammatory infiltrate comprising of plasma cells and eosinophils [Figure 2]. Immuno-fluorescence studies could not be performed due to lack of facility. A diagnosis of pemphigus vegetans of Hallopeau was established on the basis of classical clinical and histopathological picture. The patient was treated with systemic steroids (1 mg/kg body wt/day) and showed significant improvement with drying up of lesions near about in 8 weeks. [Figure3].



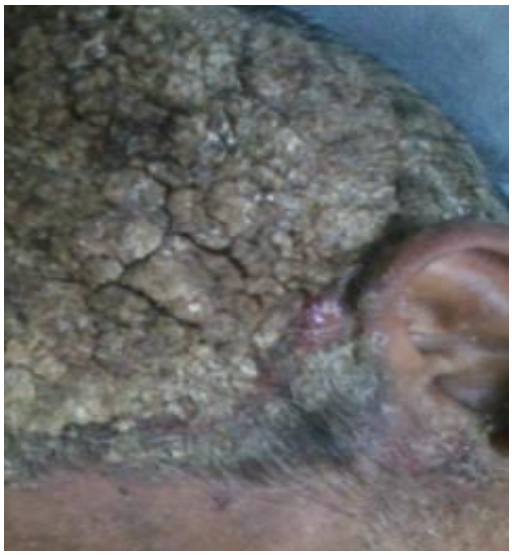
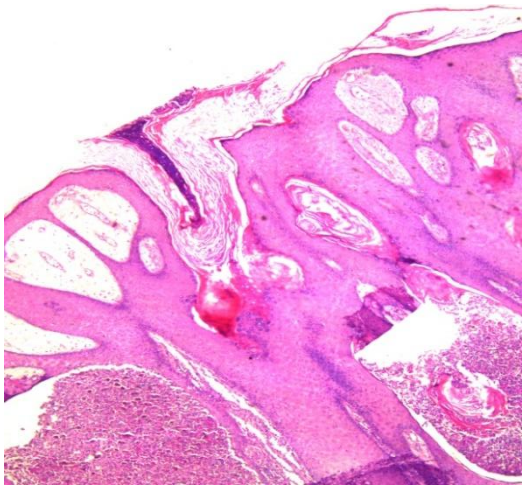
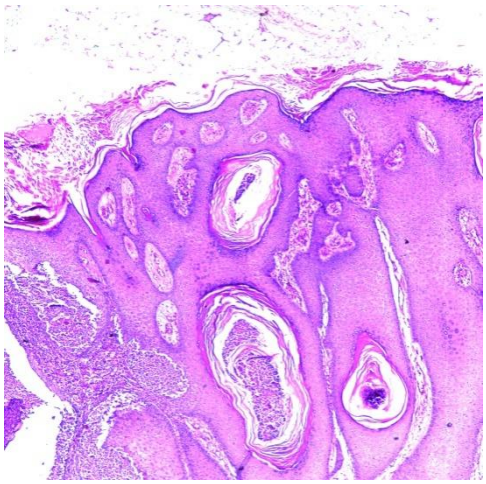
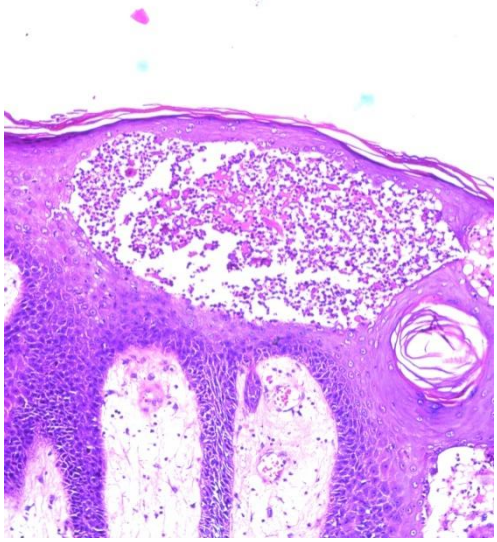


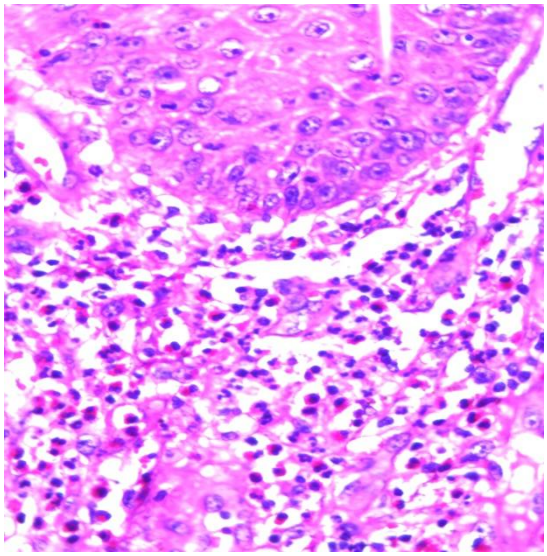
Figure 1: Clinical photograph showing well-defined, irregular, hypertrophic, fissured plaque with oozing and crusting at groin, peri anal region and scalp





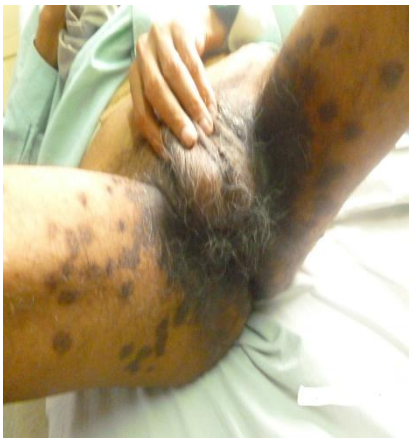


(Low power view)



(High power view)

Figure 2: Sections show verrucous or villi like hyperplasia of the epidermis with elongated rete ridges. Numerous eosinophils are present within multiple epidermal and dermal clefts producing eosinophilicspongiosis and eosinophilic pustules. No prominent acantholysis present in the sections. There is no evidence of atypia noted in the squamous epithelial cells. Mitotic figures are not noted above the basal layer of the epidermis.



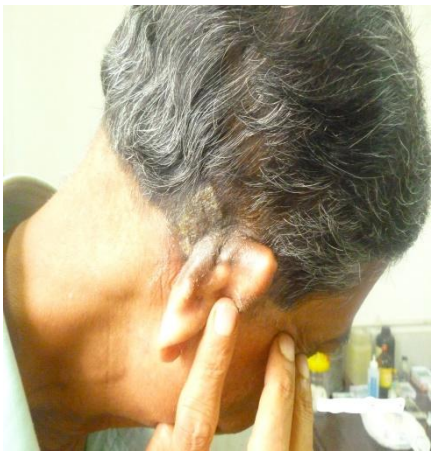


Figure 3: Clinical photograph showing dramatic response with significant clearing of the patient's lesions after treatment

Discussion

Diagnosis in the present case was established on the basis of characteristic clinical morphology in the form of vegetating papules to plaques in the groins and perianal region, involvement of the scalp, negative VDRL, eosinophilic micro-abscesses with acantholytic cells in the epidermis and dense inflammatory infiltrate in the dermis as seen in pemphigus vegetans.[6] The diagnosis was further confirmed by the excellent response to steroids.

Condylomalata form the most important differential diagnosis. These are the extremely infectious lesions of secondary syphilis which present as flesh colored moist oozing papules in the intertriginous areas. These lesions are often pruritic and might persist for months. These are often confused with condylomaacuminata and hemorrhoids.[7] The diagnosis of condylomalata

can be established on the basis of a positive DGI, a reactive VDRL and a characteristic histopathology in the form an intense plasma cell infiltrate with obliterative endarteritis and capillary proliferation in the dermis.

The Hallopeau type has a relatively benign course and responds to topical therapy, low dose steroids and occasionally to dapsone while the Neumann type has a poor prognosis and is resistant to treatment.

We would like to emphasize that, though rare, a differential diagnosis of pemphigus vegetans should be kept in mind in cases of condylomalata, especially in those not responding to therapy and also clinically looking squamous cell carcinoma cases.

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