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Pemphigus Vegetans -Masquerading as malignancy of lip A Case Report

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Abstract

Pemphigus vegetans is a rare variant of pemphigus vulgaris characterized by pustules or papillomatous vegetations, preferentially affecting intertriginous and periorificial areas. Exceptional manifestations may be misdiagnosed resulting in delayed diagnosis and treatment. We herein report an unusual manifestation of Pemphigus Vegetans in a 58-year-old female presented with history of swelling and pain in the lower lip for 4 months. Suspecting malignancy of lip-edge wedge biopsy of the lesion was done. On examination it was a proliferative lesion of size 2 x 1 cm in the lower lip, irregular in shape with no discharge. On cut section a grey white to grey brown soft tissue fragment measuring 1.5 x 1 x 0.7 cm lesion was received. Microscopic examination of multiple sections showed a polypoidal ulcerative swelling with marked pseudoepitheliomatous hyperplasia, hyperkeratosis, marked irregular acanthosis and a focus showing suprabasal clefting. The base of the supra basal cleft showed villi formation covered by stratum basalis cells giving "tomb stone appearance". Dermis showed marked inflammatory infiltrates consisting of intraepidermal eosinophils, numerous plasma cells and lymphocytes, along with inflammatory infiltrates in the minor salivary glands. There is no evidence of malignancy. On further evaluation with immunofluorescence intercellular deposition of IgG and C3 in a fishnet pattern was seen confirming the diagnosis of Pemphigus vegetans.

Keywords: *Pemphigus Vegetans, Suprabasal Clefting*

1. Introduction

Pemphigus Vegetans is a rare variant of autoimmune vesiculo-bullous disorder with an incidence of 0.7/lakh. Most common cause is autoantibodies against Desmoglein 3 and Desmoglein 1 (DSG3 and DSG1). Pemphigus vegetans mainly affects the intertriginous and periorificial areas. Exceptional manifestations may be misdiagnosed resulting in delayed diagnosis and treatment (1).

Case History

A 58-year-old female presented with a history of swelling and pain in the lower lip for 4 months. Suspecting malignancy of lip, edge wedge biopsy of the lesion was done. Patient had nil significant past or personal history. Clinically there was a proliferative lesion of size 2 x 1 cm in the lower lip, irregular in shape with no discharge. On cut section grey white to grey brown soft tissue fragment measuring 1.5 x 1 x 0.7 cm lesion was received. Microscopic examination of multiple sections showed a polypoidal ulcerative swelling with marked pseudoepitheliomatous hyperplasia, hyperkeratosis, marked irregular acanthosis,acantholysis with a focus showing suprabasal clefting (Fig.1). The base of the supra basal cleft shows villi formation covered by stratum basalis cells giving "tomb stone appearance". Dermis shows marked inflammatory infiltrates consisting of intraepidermal eosinophils, numerous plasma cells and lymphocytes, along with inflammatory infiltrates in the minor salivary glands. There was no evidence of malignancy. On further evaluation with immunofluorescence intercellular deposition of IgG and C3 in a fishnet pattern was seen confirming the diagnosis of Pemphigus vegetans. Patient was started on corticosteroids and is on regular follow-up.

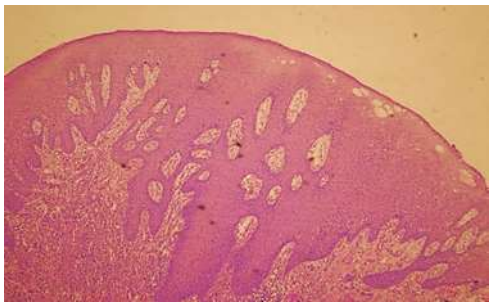

Figure 1: Pseudoepitheliomatous hyperplasia

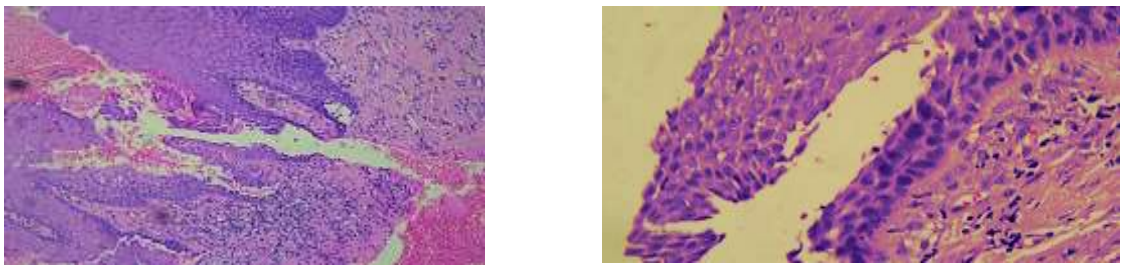

Figure 2: Suprabasal clefting

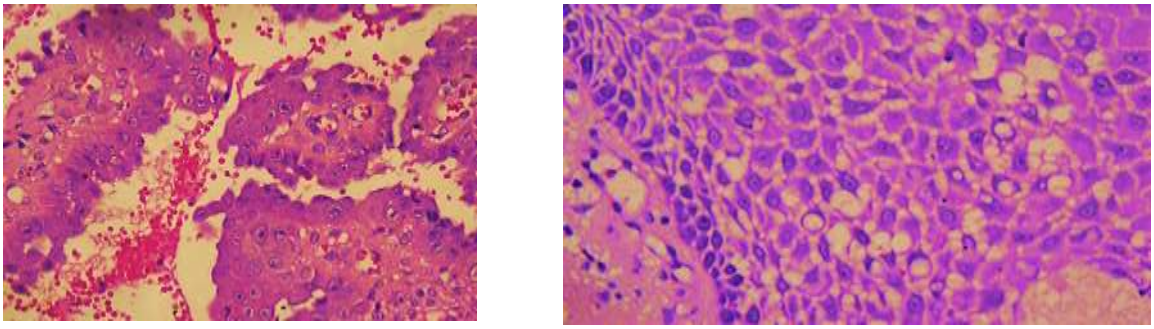

Figure 3: Tomb stone appearance Acantholysis

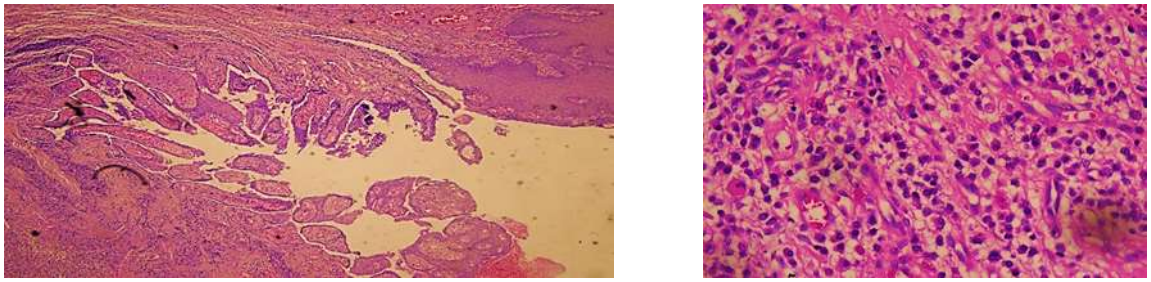

Figure 4: Villi Eosinophils and plasma cells

3. Results and Discussion

Pemphigus vegetans differs from pemphigus vulgaris by the designation "vegetans" presence of vegetating erosions, affecting flexural areas (2). Two variants have been recognized Neumann type, the lesions are vesicular and erosive, resembling pemphigus vulgaris, but the lesions evolve into vegetating plaques. Oral mucosa is involved. The less common Hallopeau type commences with pustular lesions, cobblestoned, verrucoid variant exhibiting localization to flexural areas. Oral mucosa is uninvolved. The tongue in pemphigus vegetans if involved show a cerebriform pattern. It exhibits spontaneous regression. Since the outbreak of the COVID-19 pandemic, oral ulcerative lesions have been described, associated with SARS-COV-2 infection. Bullous dermatoses can arise with similar lesions in the oral cavity, which is why the clinical picture must be very well known. So that when they mimic a malignancy picture it can be easily differentiated and patient can be relieved of a malignancy diagnosis. The final diagnosis can be arrived with immunofluorescence. Direct immunofluorescence studies will show intercellular deposition of immunoglobulin IgG and C3 in the intraepidermal space and surface keratinocytes (3). The presence of desmoglein 3 autoantibodies confirms the diagnosis of Pemphigus vegetans. On Electron microscopy Keratinocytes show a reduction of tonofilaments. Only rarely are the desmosomes identified. The basement membrane often appears damaged as a result of inflammatory infiltrates.

4. Conclusion

Spontaneous remission may occur in pemphigus vegetans, with complete recovery noted—a phenomenon not characteristic of pemphigus vulgaris. Autoimmune disease requires interprofessional approach for appropriate diagnosis and lifelong management (4). Pemphigus vegetans is often underdiagnosed or misdiagnosed. Although rarest it is fatal if untreated. Pemphigus vegetans has been reported to mimic malignancy on an 18F-fluorodeoxyglucose positron emission tomography scan with prominent involvement of the nose as well as multiple sites on the scalp, lips, and bilateral cervical lymph nodes. Underlying malignancy and a drug-based trigger, such as captopril and enalopril, are additional associations (5). Despite the dramatic appearance, the disease readily and rapidly responds to systemic steroids, and therefore, heightened awareness of a localized variant allows earlier therapeutic intervention, a critical cornerstone in optimal management.

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