

## Pemphigus Vegetans Presenting with Cheilitis

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### Abstract

Pemphigus vegetans is a rare variant of pemphigus vulgaris which differs from it by the presence of vegetating erosions, primarily affecting intertriginous areas. There are 2 types of pemphigus vegetans. There is a Neumann type that begins like Pemphigus vulgaris, with flaccid bullae, followed by peripheral pustules that resolve with hyperkeratotic verrucous vegetations. The less common Hallopeau type starts with pustular lesions which evolve into verrucous vegetations. Oral presentations with cheilitis have rarely been described in the literature. We report the case of patient with pemphigus vegetans presenting with cheilitis.

### Case Report

An 81 years old female patient has a history of hypertension treated by calcium channel blockers. She complained of painful erosions around the lips for the last 9 months. She also developed umbilical and groin lesions treated with local and oral antibiotic without any improvement. There was no history of any fluid filled lesion or any genital ulcer in the past. Physical examination showed an erosive cheilitis. There was also a well-defined, irregular and hypertrophic plaque located on the umbilical and inguinal areas (**Figure 1**).

There were no other systemic findings. VDRL in serial dilutions was non-reactive and TPHA was negative. A biopsy taken from the umbilical region showed suprabasal epidermal acantholysis, clefting and blister formation (**Figure 2**). Direct

immunofluorescence showed intercellular immunoglobulin G throughout the epidermis and intercellular substance antibodies were detected by indirect immunofluorescence. The diagnosis of pemphigus vegetans of Hallopeau was established. The patient was put on oral corticosteroids (2mg/kg per day). She had complete healing of the cheilitis and intertriginous areas within 8 weeks (**Figure 2**).

### Discussion

Pemphigus vegetans is a rare verrucous variant of pemphigus vulgaris, affecting intertriginous area. It presents as heaped up vegetating and verrucous plaques mainly in the flexures. Other areas are less described such as foot, oral mucosa, lips or toes. Oral manifestations presenting with cheilitis have been rarely reported in the literature [1,2].



Figure 1: (a) Erosive cheilitis and hypertrophic lesions plaque before treatment and (b) Clinical appearance after treatment.

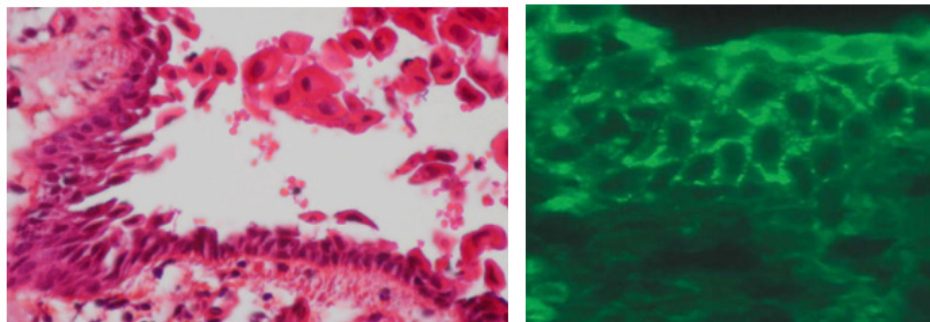


Figure 2: (a) Biopsy showing supra-basal acantholysis and (b) Intercellular immunoglobulin G throughout the epidermis

Condyloma lata form, paraneoplastic pemphigus and Pyodermitis-pyostomatitis vegetans should be taken into consideration as a differential diagnosis [3].

In the present case, histopathology and direct immunofluorescence findings confirm the diagnosis of pemphigus vegetans. The treatment is usually with corticosteroids with a good response. Our patient showed rapid response to oral corticosteroids.

### Conclusion

Pemphigus vegetans with cheilitis has very rarely been reported. In our case, histopathology, DFI and the excellent response to steroids confirm the diagnosis. This report describes the case of a patient presenting with a 6-month history of persistent cheilitis on the lower lip who was diagnosed as a pemphigus vegetans. Clinicians should be aware of this rare manifestation

and take into consideration the diagnosis of pemphigus vegetans in case of chronic cheilitis.

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