

# Pemphigus Vulgaris with Tense Bullae

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## ABSTRACT

We report a case of a 51-year-old woman with a history of type II diabetes mellitus and dyslipidemia presenting with pain, swelling, and crusting of the lips. One year after onset of mucosal lesions, she developed an abdominal eruption with several tense vesicles and bullae on an erythematous base. The hematoxylin and eosin stain sample was consistent with a diagnosis of pemphigus vulgaris. The tense bullae of our patient highlight a rare phenotype of pemphigus vulgaris, which fits the mucocutaneous type because of involvement of the oral mucosa, with the exception of the findings of tense bullae.

## CASE REPORT

A 51-year-old woman with a past medical history of type II diabetes mellitus and dyslipidemia presented to the Dermatology Department with pain, swelling, and crusting of the lips. This was accompanied by anal pain, odynophagia, and weight loss of 20 kg since the onset of symptoms. Previous evaluations by Head and Neck Surgery, Allergy, and Infectious Diseases Departments included lip biopsy, viral and bacterial

cultures, and treatment with topical antifungals, an oral antiviral, topical and oral antibiotics, and topical steroids. The biopsy and culture studies did not reveal the etiology of her condition, and the treatments did not improve her symptoms.

One year after onset of her mucosal lesions, the patient developed an abdominal eruption consisting of several tense vesicles and bullae on an erythematous base (Figure 1). She also had lip edema and crusting, oral erosions and ulcerations, and an anal fissure. Histologic examination revealed suprabasal acantholysis (Figure 2), the histologic hallmark of Pemphigus vulgaris (PV) that is essential for diagnosis.<sup>1</sup> Furthermore, direct immunofluorescence showed intercellular staining with immunoglobulin G (IgG) and complement test C3, which was also consistent with PV.

After biopsy, the patient was empirically started on methylprednisolone, 48 mg daily. After confirmation of her diagnosis, the patient was also started on mycophenolate mofetil, 500 mg twice daily. The methylprednisolone was later switched to prednisone, 80 mg daily, because of gastrointestinal intolerance. Owing to minimal response after 2 months of therapy, the patient received 2 rituximab infusions of 1000 mg, 2 weeks apart. There was noticeable improvement within 2 weeks of her first dose of rituximab.



Figure 1. Physical examination findings of tense bullae on the patient's abdomen.

## DISCUSSION

PV is an autoimmune disease with the average age of onset between 40 and 60 years of age. It is characterized by involvement of the oral mucosa, such as painful oral erosions that often precede painful, cutaneous flaccid vesicles and bullae.<sup>2</sup> Involvement of the oral mucosa is usually accompanied by severe pain that can lead to weight loss and malnutrition. Although it is a rare disease, with incidence rates between 0.1 and 0.5 per 100,000 people per year, PV has the potential for life-threatening complications, and treatment is always indicated at the time of disease onset.<sup>3</sup> PV can be further subdivided into mucosal dominant type, presenting with mucosal lesions and minimal skin involvement; and mucocutaneous type, which presents with skin blisters and erosions along with mucous membrane lesions. IgG antibodies against desmoglein (Dsg) 1 and Dsg 3, cadherin-type cell-to-cell adhesion molecules in desmosomes, are believed to play a role in inducing vesicle and bullae formation in PV.<sup>4</sup> The dysfunction in Dsg 1 and Dsg 3 results in the loss of cell-to-cell adhesion in skin membranes and mucous membranes, respectively.

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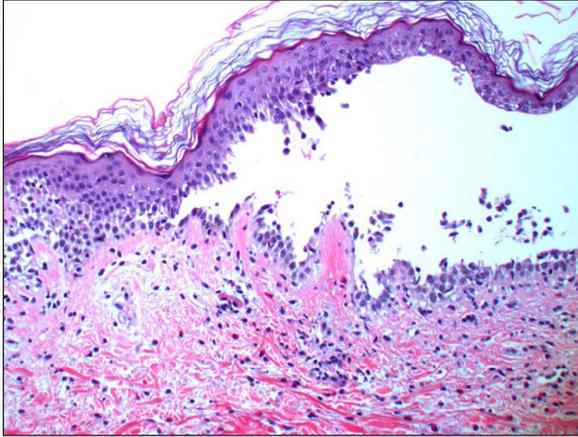


Figure 2. Biopsy specimen shows characteristic tombstone appearance of suprabasal acantholysis (hematoxylin and eosin stain sample; original magnification, x40).

## CONCLUSIONS

This case is of special interest because of the findings of tense, rather than flaccid, bullae (Figure 1). To our knowledge, there has been only one other case reported of PV with tense bullae with suprabasal acantholysis but no evident mucosal involvement, which Yoshida et al<sup>5</sup> described as cutaneous-type PV. Cutaneous-type PV, which has yet to be categorized as

a third type of PV, is thought to be a transient phenotype evolving from the previously described subdivisions of PV.<sup>4</sup> To date, there have been seven reported cases of putative cutaneous-type PV.<sup>6</sup> The tense bullae of our patient highlight a rare phenotype of PV that fits the mucocutaneous type caused by involvement of the oral mucosa, with the exception of the findings of tense bullae. ❖

## Disclosure Statement

The author(s) have no conflicts of interest to disclose.

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## The Whole Man

Medicine alone takes as its province the whole man ... It is concerned with ... man in all the complexity of his body and mind from his conception to his last breath; and its concerns extend increasingly beyond his sicknesses, to the conditions which make it possible for him to lead a healthy and happy life.

— Walter Russell Brain, 1st Baron Brain, 1895-1966, British neurologist and author of *Brain's Diseases of the Nervous System*