

# Oral Manifestations of Autoimmune Blistering Diseases

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## Background

Oral lesions are observed commonly in autoimmune blistering skin diseases. Oral lesions can be the predominant or minor clinical manifestation of a given disease. Pemphigus vulgaris (PV) and bullous pemphigoid (BP) are the earliest recognized autoimmune blistering diseases, and, together, they account for about one half of the autoimmune blistering diseases. While most patients with pemphigus vulgaris have oral lesions, which usually are the first manifestation of this disease, only a few patients with bullous pemphigoid have oral lesions. Over the last few decades, many other autoimmune blistering diseases have been delineated, and some of these newly identified diseases have oral manifestations.

This article discusses the oral manifestations of several well-characterized autoimmune blistering diseases, including pemphigus vulgaris, bullous pemphigoid, linear immunoglobulin A (IgA) bullous dermatosis, and paraneoplastic pemphigus (PNP). A group of autoimmune blistering diseases affecting primarily the mucous membranes is termed mucous membrane pemphigoid (MMP) (also termed *cicatricial pemphigoid*). Because this topic is discussed in a separate article, it is not described in great detail in this article.

## Animal models

Spontaneous animal homologues of human autoimmune blistering diseases have been identified in the last 2 decades.<sup>[1]</sup> Those diseases in which oral involvement occurs include pemphigus vulgaris (dogs, cats), paraneoplastic pemphigus (dog, cat),<sup>[2]</sup> bullous pemphigoid (dogs, cats, horses, pigs),<sup>[3, 4]</sup> mucous membrane pemphigoid (dogs, cats),<sup>[5]</sup> linear IgA bullous dermatosis (dogs), epidermolysis bullosa acquisita (dogs), and bullous systemic lupus erythematosus (1 dog). The histopathologic and immunopathologic findings usually are the same as that of human diseases and are not discussed here.

## Pemphigus group

Pemphigus vulgaris is a very rare acantholytic skin disease. In most cases, oral involvement is severe, and the mouth sometimes can be the first site to exhibit lesions. Flaccid vesicles on the gums, tongue, and palate evolve rapidly into erosions and ulcerations with indistinct margins and peripheral sloughing of mucosal epithelium (Nikolsky sign). Pemphigus foliaceus, the most common form of pemphigus observed in animals, affects dogs and cats. It usually does not affect oral and other mucosal membranes.

## Pemphigoid group

The pemphigoid group includes the following:

- Bullous pemphigoid
- Mucous membrane pemphigoid
- Epidermolysis bullosa acquisita
- Bullous systemic lupus erythematosus

## Pathophysiology

As a group, autoimmune blistering skin diseases are recognized as autoantibody-mediated diseases. This group of diseases can be divided into 2 major subsets, the pemphigus subset and the pemphigoid subset. Whereas the pemphigus subset of diseases is mediated by autoantibodies that target the extracellular skin components that link one epidermal cell to another, the pemphigoid subset is mediated by autoantibodies that target the extracellular skin components that link the skin basement membrane components either to the lowermost layer of epidermal cells or to the dermal components. Accordingly, the pemphigus subset of diseases is termed intraepidermal blistering disease, while the pemphigoid subset of diseases is named subepidermal blistering disease. Passive transfer experiments have demonstrated that purified autoantibodies from patients with the pemphigus group of diseases can induce blister formation when delivered to newborn mice.

Passive transfer experiments using autoantibodies from human patients with 2 major forms of the pemphigoid group of diseases (ie, bullous pemphigoid, epidermolysis bullosa acquisita) failed to induce clinically observable blisters in

newborn mice; however, rabbit antibodies raised against the recombinant proteins encoded by the gene of mouse bullous pemphigoid antigen 2 (BP180) are capable of inducing blisters in newborn mice in a complement-dependent manner. Furthermore, anti-BP180 autoantibodies from patients affected with BP are capable of inducing dermal-epidermal separation in cryosections of normal human skin, further supporting the pathogenic role of BP180.

In addition, rabbit antibodies raised against type VII collagen (epidermolysis bullosa acquisita antigen) are also capable of inducing blisters in mice. So far, no truly active experimental animal models (in which healthy mice are induced to autoimmune disease de novo) are known to facilitate the studies on the induction phase of autoimmune blistering diseases. Nevertheless, autoantibodies can be induced by immunized healthy BALB/c mice with synthetic peptides of the mouse bullous pemphigoid antigen 2 NC16A domain.

In certain patient subsets, the development of the autoimmune disease has been proposed to have been triggered by an immune phenomenon, "epitope spreading," a concept stating that tissue injuries from an inflammatory event may expose the previously hidden autoantigen to autoreactive lymphocytes, leading to autoimmune disease.<sup>[6, 7]</sup> Possible clinical examples include mucous membrane pemphigoid and paraneoplastic pemphigus. For example, patients who developed ocular mucosal injury secondary to an inflammatory disease termed Stevens-Johnson syndrome are noted to subsequently develop ocular mucous membrane pemphigoid.<sup>[8]</sup>

## Epidemiology

### Frequency

#### United States

The prevalence at which autoimmune mucocutaneous blistering diseases occur in the United States is not known. Because of the rarity of these diseases and because of the wide clinical heterogeneity, epidemiologic study is difficult.

#### International

Likewise, the true prevalence of autoimmune mucocutaneous blistering diseases internationally is unknown. Nevertheless, it is now well recognized that this group of diseases does occur throughout Europe, Asia, the Americas, and Arabic countries.

### Mortality/Morbidity

The pemphigus vulgaris group of diseases is generally more severe and has higher mortality than mucous membrane pemphigoid. Both pemphigus and pemphigoid are chronic inflammatory diseases and, therefore, carry significant morbidity from the diseases themselves and from the adverse effects of therapeutic medications.

#### Pemphigus group

Before the availability of corticosteroids, most patients with pemphigus vulgaris died. The extensively denuded skin surfaces from the broken blisters in these patients made them very susceptible for all kinds of infections, water loss, and electrolyte imbalance. Severe oral erosions interfered with patients' proper eating and drinking and significantly hindered their nutrient intake and the health of their immune functions, thus further reducing their ability to defend against infections. The long-term use of corticosteroids and immunosuppressive agents introduces significant adverse effects (eg, osteoporosis, diabetes, susceptibility to infections) after long-term use. Several cases of cutaneous squamous cell carcinomas and one case of primary brain lymphoma have been reported to develop in patients with pemphigus vulgaris who received long-term immunosuppressive treatments.<sup>[6]</sup>

#### Pemphigoid group

As a group, a much lower mortality exists for this group than for the pemphigus group of diseases; nevertheless, the chronicity of the diseases can bring significant morbidity to patients. Adverse effects from chronic use of corticosteroids and immunosuppressives also can contribute to morbidity.

#### Paraneoplastic pemphigus

This disease is the most resistant to conventional medical treatment. If the primary neoplasm associated with the pemphigus is found and removed completely, patients usually responded to the treatments relatively well and could recover completely; however, if the primary neoplasm is not found or cannot be eradicated completely, the disease will likely lead to a fatal outcome.

### Race

No significant racial predilection for autoimmune blistering skin diseases exists other than an increase in frequency of pemphigus vulgaris in some Jewish populations.

### Sex

No sexual predilection for autoimmune blistering skin diseases exists other than a slight predilection of females for mucous membrane pemphigoid.

## Age

Autoimmune blistering diseases primarily affect elderly patients, although occasional cases of childhood onset have been reported. The noted exception is linear IgA bullous dermatosis; about one half of patients with this disease have onset during childhood.

## Clinical Presentation

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