

# WHAT IS . . .

## PEMPHIGUS VULGARIS

Pemphigus vulgaris (PV) is the most common form of pemphigus (~80%). It is an acquired autoimmune disease in which antibodies target desmosomal proteins, leading to intraepithelial, mucocutaneous blistering.

The oral mucosa is often the first site of involvement. Typically, patients will have had multiple oral ulcers that have persisted for weeks or months. PV affects the oral mucosa in nearly all cases. The oral mucosa is the first affected site in 80% of cases, and it is the only affected site in 24% of cases.

A minority of patients only experience skin lesions. Skin involved is typically the upper chest, back, scalp, and face, but lesions can occur on any part of the body. The condition progresses over weeks to months.

Note that the skin-dominant form manifests the Dsg1 autoantibody profile. The mucosal dominant form manifests the Dsg3 autoantibody profile. A mixed pattern of skin and mucosa manifests a mixed Dsg1 and Dsg3 autoantibody profile.



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**PV lesion**

## MUCOUS MEMBRANE PEMPHIGOID

Mucous membrane pemphigoid (MMP) is a chronic, autoimmune disorder characterized by blistering lesions primarily affecting various mucous membranes of the body, but also affects

the skin (MMP is now the preferred term for lesions only involving the mucosa). It is also known as cicatricial pemphigoid (CP) and often results in scarring.



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**MMP lesion**

The mucous membranes of the mouth and eyes are most often affected, but those of the nose, throat, genitalia, and anus may also be affected. The symptoms of MMP vary among individuals depending upon the specific site(s) involved and the progression of the disease. Disease onset is usually between 40 and 70 years, and oral lesions are seen as the initial manifestation of the disease in 90% of cases. Blistering lesions eventually heal, sometimes with scarring. Scarring from lesions in the eyes or throat may potentially lead to serious complications.

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## DENTAL MANAGEMENT TIPS

- Conduct a complete oral mucosal examination. Evaluate for any abnormalities, including oral complications of PV/MMP treatment, such as candidiasis.
- Be gentle during maintenance appointments. Schedule more frequent appointments to control plaque build-up.
- Expect that the patient may experience pain and bleeding during the procedure. Plan for it by scheduling extra time and using local anesthetics, suction, and gauze as needed.
- Use simple hand scaling instruments to increase control and disrupt the tissue as little as possible.
- Polish teeth with a nonabrasive toothpaste, avoiding harsh abrasives and air polishers.
- Provide a list of rinses that do not contain flavoring agents, alcohol, or other harsh ingredients that might irritate oral tissues.
- If patients are on a soft diet due to presence of oral ulceration and pain, suggest nutritious options such as vegetable soups, fruit smoothies, etc.
- If, after treatment and remission by a specialist, new lesions are observed, the general dentist should refer back to the specialist for re-evaluation and treatment.
- Oral hygiene instructions for home care may need to be tailored based on the level of mucosal involvement. When significant oral disease is present, gentle home care including extra-soft toothbrushes, mildly flavored toothpastes, and mild mouth rinses may be all that the patient can tolerate. Some patients may not be able to floss due to bleeding and pain. During this time, frequent dental cleanings become more important. Once the oral ulcerations and associated pain decrease, the standard home care routine, including regular brushing and flossing, can be recommended.

## BIOPSY TECHNIQUES

Whenever considering oral PV/MMP in a differential diagnosis, a biopsy must be obtained and the specimen must contain epithelium.

Here are some guidelines:

- Do not sample the bed of an ulcer because there is no epithelium there, resulting in a non-specific diagnosis.
- As a general rule, biopsies must contain intact epithelium and should be taken from perilesional (within 1 cm) or normal appearing tissue rather than the directly ulcerated tissue because an ulcer, by definition, is missing the epithelium. Without the epithelium, it is not possible to detect the destruction between epithelial cells (as in PV) or between epithelial cells and the underlying connective tissue (as in MMP).
- During the biopsy procedure, avoid any action that may cause separation of the epithelium from underlying connective tissue, e.g. rubbing the area with gauze to remove saliva or blood.
- Two specimens must be taken with samples submitted for BOTH routine hematoxylin and eosin (H&E) stain (storing specimen in 10% formalin) AND for direct immunofluorescence testing (MUST submit sample in Michel's transport medium and send to pathology laboratory as quickly as possible to preserve antibody detection).
- When performing DIF biopsies, have Michel's solution on hand. If planning to perform DIF biopsies, order and store in advance the Michel's transport medium. Identify IN ADVANCE a commercial laboratory equipped to perform DIF studies.

## CLINICAL PRESENTATION

Dental professionals should think of PV/MMP when there is a combination of:

- Multiple oral lesions
- Ulcerations preceded by bullae (however, in many cases, the bullae may not be evident to the patient or clinician)
- Chronic lesions (non-healing or very slow to heal)
- Primary lesions (lesion runs its course until resolved, as opposed to recurrent lesions, which come and go like cold or canker sores)
- Lesions may also occur outside of the mouth

A lesion can follow minor trauma. This is called a Nikolsky sign: cells pull apart and blister, and a lesion develops because the adhesion between cells is weakened.

The formation of a Nikolsky sign following minor lateral force on mucosa or gingiva can be a diagnostic aid. If a lesion develops, the patient almost certainly has PV or MMP. Patients can also experience "natural" Nikolsky phenomena: lesions following minor oral trauma (eating hard or sharp foods, etc.).