

## Oral cicatricial pemphigoid

Brenda L. Nelson, DDS, LCDR, DC, USNR  
Lester D.R. Thompson, MD

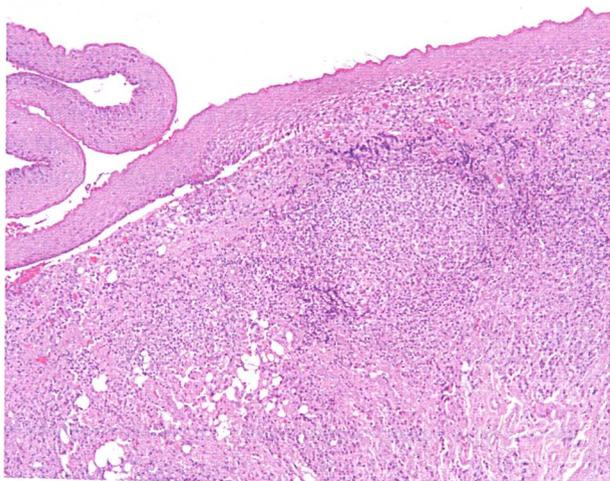


Figure 1. A low-power view demonstrates the characteristic smooth subepithelial clefting between the surface and the submucosal stroma.

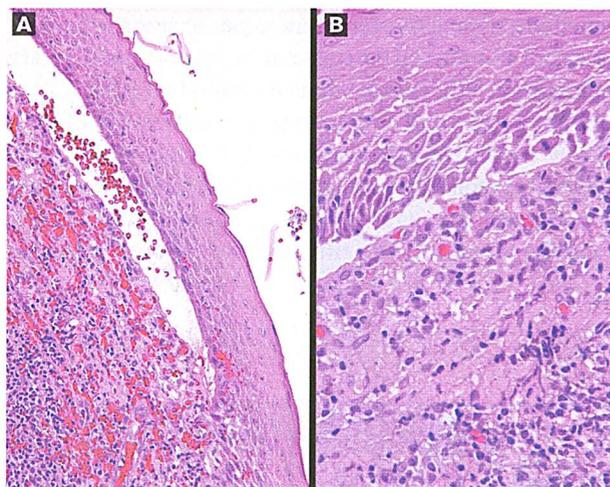


Figure 2. A high-power view shows the subepithelial cleft with hemorrhage (A), while a narrow subepithelial cleft is subtended by primarily chronic inflammatory cells (B).

Cicatricial pemphigoid is a vesiculobullous disease of the skin that may be found in the oral cavity. Previously designated "benign mucous membrane pemphigoid," cicatricial pemphigoid is a chronic, blistering, autoimmune disease that affects mucous membranes. Tissue-bound autoantibodies are directed against one or more components of the basement membrane in an affected individual. Cicatricial pemphigoid initially occurs in the fifth to seventh decades of life, and it is observed more frequently in women. Patients usually describe oral pain and/or ulceration, often of many years' duration. Clinically, the disease is characterized by the formation of bullae, which can be found anywhere in the oral cavity. The bullae rupture and produce ulceration, which may cause a scar (cicatrix) upon healing.

Microscopically, the affected tissue is characterized by a smooth, linear split between the surface epithelium and the underlying connective tissue at the level of the basement membrane rather than within the epithelium (figures 1 and 2). Surface keratinization may be seen. A mild infiltrate of chronic inflammatory cells, including lymphocytes and plasma cells, is noted subjacent to the

cleavage site. Direct immunofluorescence studies demonstrate a linear band of immunoglobulins (IgG) and complement (C3) along the basement membrane zone in 90% of affected patients. To be successful, immunofluorescence must be performed on fresh or frozen tissue (not on formalin-fixed, paraffin-embedded material).

The main differential diagnosis of cicatricial pemphigoid includes pemphigus, erosive lichen planus, and linear IgA disease, among other rare autoimmune diseases. Treatment, in most cases, involves the use of corticosteroids. Ophthalmic referral is essential once the diagnosis is confirmed, because nearly 25% of patients with oral lesions will have ocular lesions; ocular ulcerations that scar with healing may lead to blindness.

### Suggested reading

Lamey PJ, Rees TD, Binnie WH, Rankin KV. Mucous membrane pemphigoid. Treatment experience at two institutions. *Oral Surg Oral Med Oral Pathol* 1992;74:50-3.

Vincent SD, Lilly GE, Baker KA. Clinical, historic, and therapeutic features of cicatricial pemphigoid. A literature review and open therapeutic trial with corticosteroids. *Oral Surg Oral Med Oral Pathol* 1993; 76:453-9.

From the Department of Pathology, Woodland Hills Medical Center, Southern California Kaiser Permanente Group, Woodland Hills, Calif (Dr. Thompson).