Desquamative Gingivitis: Clinical Tips

Joseph L. Jorizzo, MD
Professor, Founder and Former Chair
Department of Dermatology
Wake Forest School of Medicine
Winston-Salem, NC

Professor of Clinical Dermatology
Weill Cornell Medical College
New York, NY
Conflict of Interest

Advisory Boards/Honoraria
Amgen
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Maderal AD; Salisbury L; Jorizzo J

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Key Points

- Desquamative gingivitis describes mucosal inflammation with erythema and especially peeling
- Most common causes are oral lichen planus, cicatricial pemphigoid, pemphigus vulgaris
- Patients often have secondary reduced oral hygiene practices, leading to more severe gingival inflammation and possible bone loss
Mucosal skin

- No stratum corneum
- Any disruption of normal mucosa is potential site of yeast colonization or bacterial superinfection
Differential Diagnosis

Key Points

- Differential diagnosis of desquamative gingivitis can be divided into recurrent or persistent diseases.
- Screening for involvement of other mucosal sites should be performed in all patients with a thorough review of systems, as the prevalence especially of genital mucosa is high and is associated with significant morbidity.
Selected Diseases with Recurrent Desquamative Gingivitis

Erythema multimforme
Complex aphthosis
Behcet’s Disease
Selected Dermatologic Diseases with Persistent (Chronic) Desquamative Gingivitis

Oral Lichen Planus
Cicatricial Pemphigoid (Mucosal Pemphigoid)
Pemphigus Vulgaris
Epidermal Bullosa Acquisita
Paraneoplastic Pemphigus
Graft versus Host Disease
<table>
<thead>
<tr>
<th>Disease</th>
<th>Histology</th>
<th>DIF</th>
<th>IIF/ELISA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral Lichen Planus</td>
<td>Apoptosis, band-like lymphocytic infiltrate</td>
<td>Non specific</td>
<td>N/A</td>
</tr>
<tr>
<td>Pemphigus Vulgaris</td>
<td>Acantholysis, intra epidermal cleft formation, preserved basal keratinocytes</td>
<td>Intercellular IgG in the epidermis</td>
<td>Antibodies are intercellular, Desmoglein 3 +/- Desmoglein 1</td>
</tr>
<tr>
<td>Cicatricial (Mucosal) Pemphigoid</td>
<td>Sub epidermal blister, mixed infiltrate</td>
<td>Linear deposits of IgG and C3 along the BM2</td>
<td>Often negative 1) BPISD 2) Alfa-6 beta-4 integrin 3) Laminin 332</td>
</tr>
</tbody>
</table>
Mucosal lichen planus

- Can affect oral and vaginal mucosa
- Apoptosis is a hallmark
- Red erosions and plaques with white lines present (Wickham striae)
- Oral erosive lichen planus has increased risk of mucosal squamous cell carcinoma if not controlled
- Treat with strong topical corticosteroids, topical tacrolimus, or systemic immunosuppressive medications like methotrexate or mycophenolate mofetil
Lichen planus
Fig. 11.16 Oral lichen planus. (A) White lacy pattern and an erosion on the buccal mucosa, the most common location for the reticular form. Note the ring configuration with short radiating spines. (B) Erosions on the lateral aspect of the tongue in addition to lacy white plaques and scarring.

B, Courtesy, Louis A. Fragola, Jr, MD
TIPs for Oral Lichen Planus

- Water pick
- Manage Candida acutely with fluconazole and chronically with daily clotrimazole troche
- CREST whitening (dilute peroixde)
- 1mg tacrolimus capsule – open & dissolve in ½ liter water swish and spit for 2 minutes (Ortonne)
- Topical and/or intralesional corticosteroids
- Oral methotrexate or mycophenolate if needed
- Biopsy as indicated for exclusion of SCC

Torti DC, Jorizzo JL. Arch Dermatol 2007;143:511-515
Pemphigus vulgaris

- Autoimmune disease with antibodies to desmoglein I and III
- Affects individuals in their 5\textsuperscript{th} or 6\textsuperscript{th} decade of life
- Clinical features
  - Big flaccid bullae, easily ruptured
  - Erosions
  - Oral mucosa affected > skin
  - Positive Nikolsky sign
- Treatment
  - Prednisone, mycophenolate mofetil, topical steroids, rituximab
Fig. 29.5 Pemphigus vulgaris.

A, B.
Essentially all patients develop painful oral mucosal erosions, with
the most common sites being the Buccal and palatine mucosae.

C. Flaccid blisters and an erosion due to rupture of a bulla.

B-D, Courtesy, Louis A Fragola, Jr, MD
**THERAPEUTIC LADDER FOR PEMPHIGUS VULGARIS**

### STANDARD TREATMENT

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Dose Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral prednisone</td>
<td>1.0 mg/kg/day as an initial dose (usually 60 mg/day) (1)</td>
</tr>
</tbody>
</table>

### AGGRESSIVE TREATMENT

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Dose Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>Immunosuppressive agents in combination with oral prednisone:</td>
<td></td>
</tr>
<tr>
<td>Azathioprine</td>
<td>2–4 mg/kg/day (usually 100 to 300 mg/day) (1)</td>
</tr>
<tr>
<td>Mycophenolate mofetil</td>
<td>2–3 g/day (2)</td>
</tr>
<tr>
<td>Cyclophosphamide</td>
<td>1–3 mg/kg/day (usually 50 to 200 mg/day) (2)</td>
</tr>
<tr>
<td>Cyclosporine</td>
<td>5 mg/kg/day (2)</td>
</tr>
<tr>
<td>Pulse methylprednisolone</td>
<td>1 g/day over a period of 2–3 hours for 3–5 consecutive days (2)</td>
</tr>
<tr>
<td>Methotrexate</td>
<td>7.5–20 mg/week (3)</td>
</tr>
<tr>
<td>Pulse cyclophosphamide</td>
<td>50 mg/kg/day × 4 days (3)</td>
</tr>
<tr>
<td>Plasmapheresis</td>
<td>1–2 times per week, at the onset (2)</td>
</tr>
<tr>
<td>High-dose IVIg</td>
<td>400 mg/kg/day for 5 consecutive days (1); may need to be repeated</td>
</tr>
<tr>
<td>Rituximab</td>
<td>375 mg/m² once weekly for 4 weeks (2)</td>
</tr>
<tr>
<td>Extracorporeal photopheresis</td>
<td>2 days per month (3)</td>
</tr>
</tbody>
</table>

### TOPICAL TREATMENT

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>Topical corticosteroids</td>
<td>(1), especially Class I topical localized persistent sites</td>
</tr>
<tr>
<td>Topical antibiotics</td>
<td>(2)</td>
</tr>
<tr>
<td>Topical immunomodulators (e.g. cyclosporine, tacrolimus)</td>
<td>(3)</td>
</tr>
</tbody>
</table>

**Table 29.5 Therapeutic ladder for pemphigus vulgaris.** Key to evidence-based support: (1) prospective controlled trial; (2) retrospective study or large case series; (3) small case series or individual case reports.
Tips for Pemphigus Vulgaris

- Waterpick
- Manage Candida acutely with fluconazole and chronically with daily clotrimazole troches
- CREST whitening (dilute hydrogen peroxide)
- 1mg tacrolimus capsule (open & dissolve in ½ liter of water – swish and spit for 2 minutes (Ortonne)
- Topical and/or intralesional corticosteroids
- Choose: Rituximab versus Prednisone and Mycophenolate

Cicatricial (Mucosal) Pemphigoid

Key Points

- Chronic, autoimmune, group of subepidermal blistering disorders, characterized by predominant involvement of the external mucosal surfaces and a tendency for scarring

- Associated with tissue-bound and less often circulating autoantibodies directed against distinct structural components of basement membrane in stratified and some complex epithelia:
  - Bullous pemphigoid antigen 180 (BP180, BPAG\textsubscript{2} or type XVII collagen)
  - Laminin 332 (laminin5)
  - Integrin (Beta4 sub unit)
Cicatricial (Mucosal) Pemphigoid

Key Points (cont.)

- Not a clinical entity but a disease phenotype shared by a heterogeneous group of diseases with lesions that favor mucosal surfaces and, less frequently, the skin
- When scarring affects the conjunctivae it can lead to blindness
- Diagnosis is clinical, histologic and confirmed immunofluorescence microsurgery and immunochemical studies
Cicatricial (Mucosal) Pemphigoid

Treatment

- Local gingival care (waterpick, dental inserts, etc.)
- Control oral candidiasis intermittent fluconazole prophylaxis with mycelectrodes
Cicatricial (Mucosal) Pemphigoid

Treatment (cont.)

- Potent typical corticosteroids (candida risk)
- Tacrolimus (topical or swish and spit)
- Dapsone
- Azathioprine
- Methotrexate
- Mycophenolate
- Systemic corticosteroids
- IVIG
- Rituximab
- Remember larynx genitalia, esophagus, and, of course eyes