

Skin diseases that affect the mouth

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Much of the oral mucosa is derived embryologically from an invagination of the ectoderm and perhaps not surprisingly, this, like other similar orifices, may become involved in disorders that are primarily associated with the skin. Sometimes the oral lesions occur along with skin lesions although the lesions in the mouth may be the only manifestation. Careful examination of the mouth can be difficult, particularly in circumstances where there is inadequate illumination.

Lichen planus

Lichen planus is a common skin disorder that may affect the oral mucosa in up to 0.2% of the adult population. There is approximately a 25% crossover between skin and mucosal lesions but often there are only oral manifestations. These also tend to be longer lasting than skin lesions, sometimes persisting for many years.

The aetiology of lichen planus in the mouth is commonly idiopathic but it may be associated with the ingestion of a multitude of medications (Table 1). It is also a feature of graft-versus-host reactions and there is some evidence to associate lichen with a form of reaction to the mercury content of amalgam dental restorations as, possibly, to other restorative materials.

Lichen planus can involve any part of the lips and oral mucosa but is most common on the buccal mucosa and the lateral margins of the tongue. There are several clinical forms in which it appears.

White striae or a lace-like, reticular appearance is common. These are often symptomless, only being detected on routine examination. Interspersed between the striations, there may be epithelial atrophy, with the mucosa assuming a red appearance. This tends to be uncomfortable, causing a burning sensation either spontaneously or especially with eating spicy, sour or hot foods. A feature of lichen is that it tends to exhibit a degree of symmetry in its oral involvement. This can be useful in differentiating it from other white lesion in the mouth, such as leukoplakia.



Figure 1. Dark red, raised lesions of lichen planus on the forearm. Some scratching has occurred.

Table 1

Medications commonly associated with lichen planus

Beta blockers
ACE inhibitors
Calcium channel antagonists
Thiazide diuretics
NSAIDs
Oral hypoglycaemics
Carbamazepine
Phenothiazines



Figure 2. Lichen planus. White striae present on the lower lip. Can be easily missed if the patient is wearing lipstick.

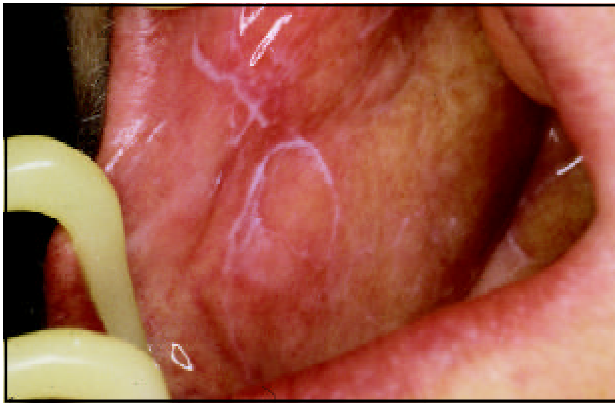


Figure 3. Lichen planus. White striae on the buccal mucosa. May be symptomless or else associated with sensitivity and a burning sensation.



Figure 4. Lichen planus. White striae and plaques alongside an erythematous area on the buccal mucosa.



Figure 5. Lichen planus. Chronic ulceration on the buccal mucosa. The posterior, lower quadrant is a site commonly affected.



Figure 6. Lichen planus. White striae and atrophy of the papillae on the lateral aspects of the dorsum of tongue. Lichen often tends to be symmetrical in oral involvement.



Figure 7. Lichen planus presenting as desquamative gingivitis. The epithelium is partially shed, leaving an erythematous, bleeding base.

Rather than striae, lichen may present as white plaques. These may be extensive or as smaller papules. Locating these on the dorsum of the tongue requires careful observation.

The erythematous, atrophic mucosa may break down, resulting in ulceration. The term 'erosive' lichen planus has been used for this although, more accurately, it is ulcerative. These ulcers can cause considerable discomfort and persist for months or even years.

In dentate individuals, a form of gingivitis may develop. This has been termed desquamative gingivitis and can also be a clinical feature of pemphigoid, dermatomyositis and orofacial granulomatosis. There is erythema of the attached gingivae, sloughing of the epithelium and bleeding on minimal provocation.

Recently, it has been recognised that individuals with oral lichen planus have a greater chance of developing squamous carcinoma, possibly about fiftyfold the normal population. It appears that this occurs more frequently in red, atrophic or ulcerated areas rather than with striae.

In the first instance, the management of oral lichen planus should be directed towards a definitive diagnosis, with biopsy if need be, and identifying potential aetiological factors. However, because a patient is taking particular medication does not necessarily mean that the drug is at fault. The question of removing dental restorations is problematic. There are no good predictors for this and the implications of removing a mouthful of fillings in the hope that it may help are significant. Patch testing has been of limited value.

Corticosteroids are widely used in the management of lichen planus. This may be in the form of a mouthrinse or depot injection. Surprisingly, there are no commercial steroid mouthrinses available and these have to be formulated according to local availability. In New Zealand, it is convenient to prescribe soluble 0.5mg betamethasone phosphate tablets (Betnesol. GlaxoWellcome) with the patient dissolving four of these in 10 mL water and then rinsing four times daily. It must be stressed that they do not swallow this solution. Nonetheless, here will still be a degree of oral retention, in the order of 5% to 10%. In Australia, an alternative solution can be prepared from dexamethasone injection vials (Decadron - MSD) with a final concentration of 2mg in 10 mL.

Alternatively, depot steroids, such as 40mg methylprednisone (DepoMedrol. - Pharmacia & Upjohn) can be injected under the ulcer.

Should steroid be unsuccessful, hydroxychloroquine (Plaquenil. - Sanofi-Synthelabo) can be used in a dose of 200mg to 400mg daily. Long term use requires supervision from ophthalmology, as retinal damage may develop.

Desquamative gingivitis is best managed in conjunction with a dental surgeon. Professional cleaning together with twice daily rinses with 0.2% aqueous chlorhexidine will reduce plaque accumulation and hence non-specific gingivitis. Thereafter topical steroids are used.

If a lesion should look in the least suspicious then biopsy is necessary. Long term observation is also important given the increased risk of transformation into a squamous carcinoma.

Lupus erythematosus

Lupus erythematosus may affect the oral mucosa as part of more widespread discoid lupus or systemic lupus. The lesions are similar to those of lichen planus with central areas of erythema or ulceration surrounded by white striae or plaques. There does not appear to be the symmetrical distribution, which is often seen with lichen, and striae alone are not a feature.

The management of mucosal lesions is the same as for lichen, with topical corticosteroids.

In cases of SLE, the possibility of co-existent inflammatory exocrinopathy (Sjögren's syndrome) should be considered with the problems of salivary gland hypofunction.



Figure 8. Lupus erythematosus. Central area of ulceration with white striated margins.

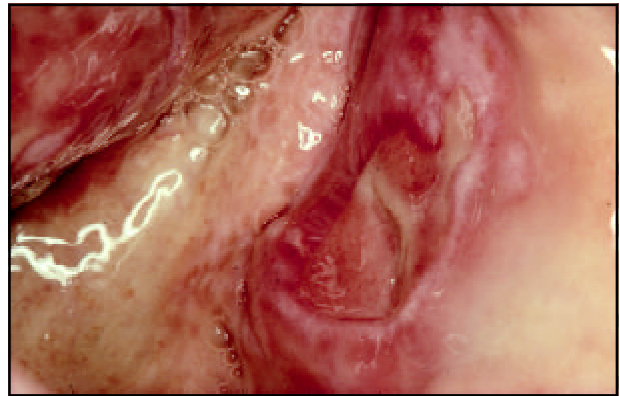


Figure 9. Lupus erythematosus. Solitary, chronic, deep ulcer on the buccal mucosa.

Erythema Multiforme

Erythema multiforme is a relatively common disorder, ranging in presentation from the milder erythema multiforme minor form to the more severe but rarer erythema multiforme major form (Stevens-Johnson syndrome). It occurs acutely, usually in young to middle age adults and apparently with a greater incidence in males. In many instances the aetiological factor remains unknown but ingestion of a range of medications (e.g. sulphonamides, NSAIDs) and herpes simplex infection may trigger an episode.

The oral lesions are non-specific areas of irregular ulceration but the diagnosis can often be established from the history of previous episodes as well as the characteristic target/iris skin lesions. It may be of a minor, fairly asymptomatic nature, with few skin lesions, or as severe and extensive ulcerative condition.

For the oral lesions, the mainstay of treatment has been corticosteroids; either topically as a mouthrinse or systemically using prednisone. Clearly, if a particular medication is implicated then it is appropriate that this not be repeated. More recently, antiviral drugs have been found to be successful both for the acute presentation and also for longer-term prophylaxis. Oral valaciclovir (trade name) or famciclovir (Famvir - SKB) are the drugs of choice.



Figure 10. Erythema multiforme. Characteristic target or bull's-eye lesions on the palm.



Figure 11. Erythema multiforme in a young adult male. He has a history of similar recurrences, with ulceration on the lips, buccal mucosa and tongue.

Eczema (Endogenous)

While eczema is not a disorder that affects the oral mucosa, it may be associated with angular cheilitis, peri-oral scaling or vertical fissuring of the lips. These tend to become infected secondarily with *Staphylococcus aureus*.

Management measures are similar to elsewhere on the skin, applying an emollient or dimethicone barrier ointment to reduce crusting and so facilitate healing. Should there be a persistent infection, then application of mupirocin ointment (Bactroban. - SKB) may be helpful.



Figure 12. Eczema in a young girl. Crusted lesions on the lips and around the mouth.

Pemphigus vulgaris

Pemphigus occurs usually in adults over the age of 50 years. This bullous condition may manifest first in the mouth before progressing to the skin. The severity is variable from intermittent ulcers to severe and persistent ulceration. Bullae are not commonly observed in the mouth due to the fragile nature of the partial epithelial covering.

Patients present with irregular areas of ulceration affecting any part of the oral mucosa and these may remain for weeks or even months on end. The mucosa under a denture, particularly the upper, may slough off repeatedly in what might be considered as a form of Nikolski sign. The diagnosis is based on overall clinical presentation, serology for skin antibodies and both histology and immunofluorescence of mucosal (or skin) biopsies.



Figure 13. Pemphigus vulgaris in an elderly female. Irregular areas of ulceration.

Specialist management of pemphigus is appropriate where corticosteroids and other immunosuppressives may be required. Treatment of oral lesions can be augmented with topical steroids.

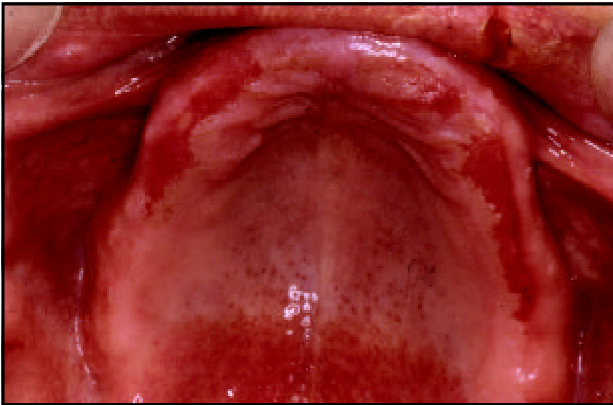


Figure 14. Pemphigus vulgaris affecting the hard palate. Normal loading of the mucosa by the adequate denture is resulting in ulceration.

Pemphigoid

For the present purposes, the oral lesions of Benign Mucous Membrane Pemphigoid and Bullous Pemphigoid will be considered together, although mucosal involvement with the latter is much less common. These bullous conditions are again more common over the age of 50 years.

As the bullae of pemphigoid arise along the epithelial/connective tissue interface, the roof is more robust

and the patient may be aware of the presence of blisters. They may also be noticed on examination and are often blood filled. Thereafter the bullae break down and present as irregular areas of ulceration, similar to those found in pemphigus.

Pemphigoid is another condition that can result in the clinical presentation of desquamative gingivitis, as described above under Lichen planus.

The diagnosis of pemphigoid is as for pemphigus.

Management of pemphigoid is along similar lines to pemphigus although less severe immunosuppression might be used due the more benign nature of pemphigoid. Steroid mouthrinses can be helpful and a regime for desquamative gingivitis, if present, has been outlined above.

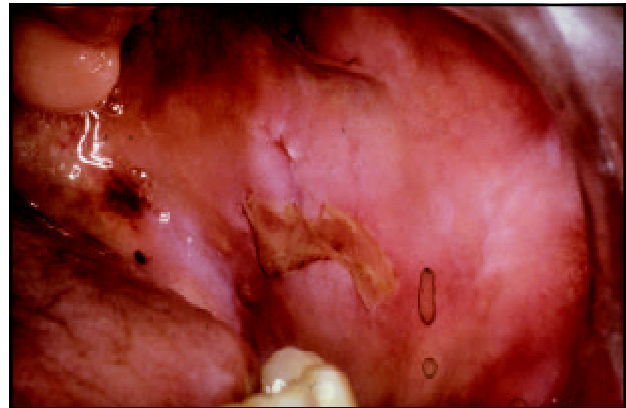


Figure 15. Benign mucous membrane pemphigoid. Irregular areas of ulceration that the patient may notice are preceded by bullae, which bleed on rupture.