

New-onset oral lichen planus and granulomatous cheilitis in a 66-year-old woman



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Key words: Crohn's disease; inflammatory condition; Melkersson-Rosenthal syndrome; Miescher's granulomatous cheilitis; oral lichen planus; orofacial granulomatosis; oral ulcers.

INTRODUCTION

We report a case of a 66-year-old woman with the simultaneous onset of oral lichen planus (OLP) and granulomatous cheilitis (GC). Both diseases are relatively rare, and further investigation for systemic or environmental causes of disease is warranted in new patients with either OLP or GC. This case and reports of a so-called lichenoid and granulomatous dermatitis in the literature beg the question, is there some unifying etiologic explanation for OLP and GC?¹

CASE REPORT

A 66-year-old woman presented with 1 year of unremitting upper lip swelling and anterior gingival pain, made worse by spicy or citrus foods. Her history was negative for new oral hygiene products or foods in her diet, and she had no known allergies. She had undergone extensive dental work during the last 2 years. Physical examination found edema of the upper lip and reticulated plaques surrounded by erythema on the lower lip and the left lateral tongue (Figs 1 and 2). The upper anterior gingiva had a beefy red, well-demarcated erythematous appearance (Fig 3). A complete blood count, comprehensive metabolic panel, and chest radiograph were within normal limits. Her history was negative for gastrointestinal symptoms, and she had a normal screening colonoscopy 2 years prior. Microscopic examination of a 3- × 2- × 2-mm shave biopsy section of the lower lip found a superficial bandlike, inflammatory infiltrate focally obscuring the dermo-epidermal junction. Direct immunofluorescence showed irregular deposition of fibrinogen along the basement membrane and no specific deposition of IgG, IgA, IgM or C3. A 9- × 5- × 3-mm incisional biopsy of the upper vermilion border showed

Abbreviations used:

GC:	granulomatous cheilitis
OFG:	orofacial granulomatosis
OLCR:	oral lichenoid contact reaction
OLP:	oral lichen planus
OLTR:	oral lichenoid tissue reactions

hyperkeratosis overlying mild epithelial hyperplasia with spongiosis and a superficial perivascular to nodular infiltrate of lymphocytes, plasma cells, and histiocytes (Fig 4). Stains for fungal and acid-fast organisms were negative, and polarized microscopy did not show foreign material. The histopathology of the upper and lower lip was consistent with GC and OLP, respectively.

DISCUSSION

In 1945, the term *granulomatous cheilitis* was used to label granulomatous infiltration and lymphatic obstruction of the lips. Decades earlier, the neuromucocutaneous triad of orofacial edema, tongue fissuring, and unilateral facial paralysis, indistinguishable from Bell's palsy, was identified by Melkersson and Rosenthal.² In 1985, the term *orofacial granulomatosis* (OFG) was coined, and referred to noncaseating granulomas of the face and oral cavity in the absence of systemic granulomatous disease. OFG encompasses a spectrum of cutaneous granulomatous conditions from Miescher's GC to Melkersson-Rosenthal syndrome. OFG is a diagnosis of exclusion, as localized granulomatous swelling of the oral cavity may be caused by systemic disease. Therefore, perioral granulomas warrant further workup to rule out conditions including Crohn's disease, sarcoidosis and tuberculosis. Isolated

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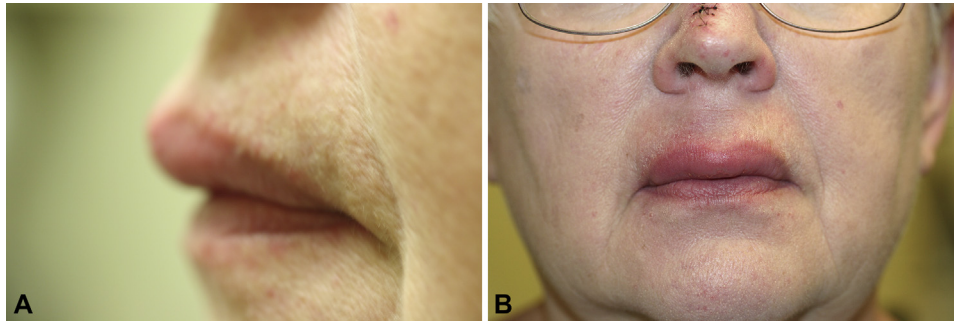


Fig 1. A and B, Upper lip swelling associated with granulomatous cheilitis.

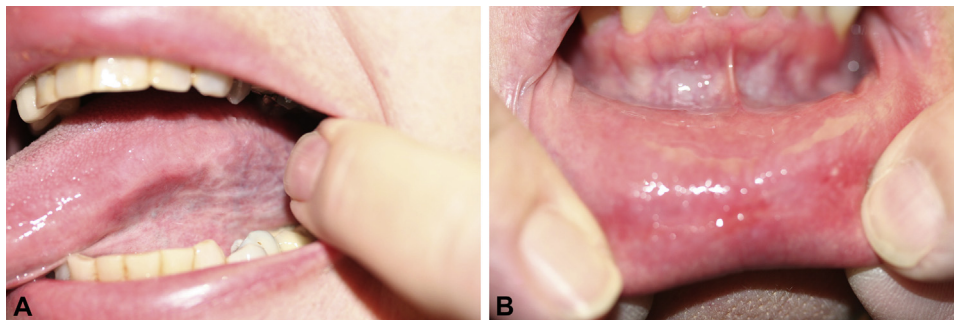


Fig 2. Oral lichen planus presenting as Wickham striae on the lateral tongue (A) and oral mucosa (B).



Fig 3. Edematous- and erythematous-appearing upper gingiva.

cutaneous granulomas may also occur as a foreign body reaction.

Macrocheilia is the most common sign of OFG, but oral ulcers, vertical fissures of the lips, mucosal tags, cobblestoned oral mucosa, gingivitis, and cervical lymph node involvement have been described.^{3,4} Melkersson-Rosenthal syndrome can also present with facial swelling, unilateral facial palsy, fissured tongue, and dysgeusia.

Clinically, GC presents with painless swelling of the lip, which may occur once and resolve but more commonly evolves into a chronic condition. Swelling is initially soft, but after several attacks becomes indurated. Fibrosis can cause permanent disfigurement, leading to problems with speech and eating,

requiring reconstructive plastic surgery in extreme cases. The etiopathogenesis is not definitively understood, although it is postulated to be a delayed-type hypersensitivity reaction.^{5,6} Foods, flavorings, additives, dental materials, and infectious agents have all been implicated. Case reports describe remission with dietary restriction. However, no rigorous *in vitro* or *in vivo* studies have been conducted to validate immunologic or inflammatory origins of OFG.^{5,6} A combination of systemic corticosteroids and minocycline is considered the best therapeutic regimen. There is considerable histopathologic mimicry among granulomatous skin conditions. OFG and oral Crohn's disease are histologically identical, and some investigators conjecture that OFG is a manifestation of Crohn's disease rather than an independent entity. Many OFG cases are rediagnosed as Crohn's disease because of the eventual onset of intestinal symptoms.⁷ Although not all patients with OFG go on to have full-blown Crohn's disease, this does not exclude the possibility that all cases of OFG are manifestations of Crohn's disease limited to the face and mouth. Debate continues, but patients with OFG require prolonged follow-up to evaluate for evolution into Crohn's disease.

OLP is an inflammatory condition, ranging from asymptomatic lesions to severe, painful ulcers. OLP

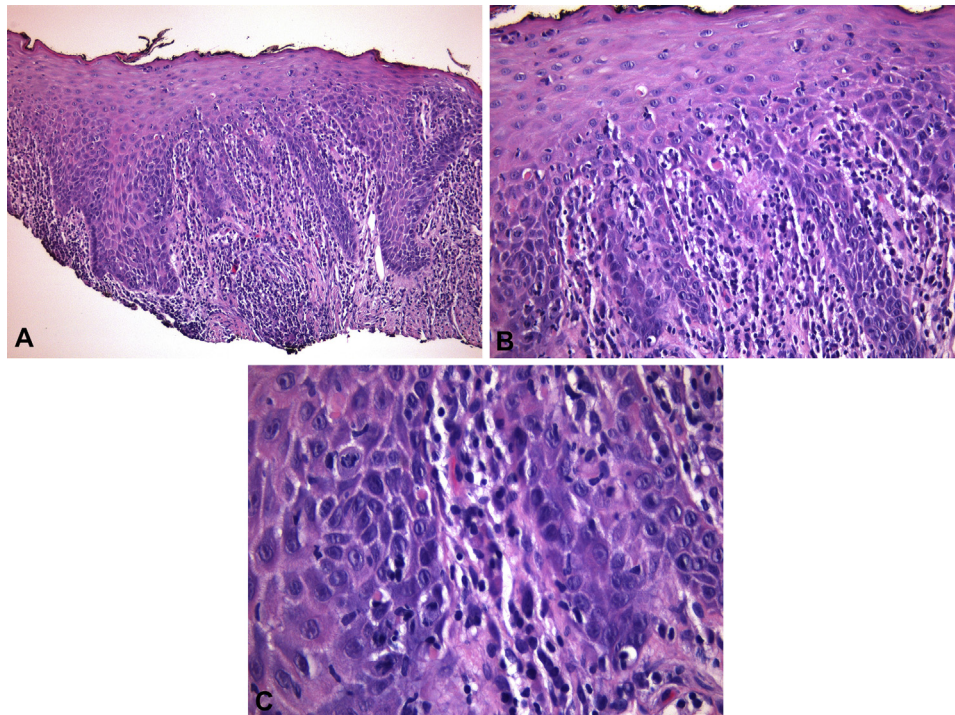


Fig 4. A-C, Biopsy section from the tongue shows irregular epithelial hyperplasia, basal layer vacuolar alteration, and lichenoid inflammation with scattered necrotic keratinocytes, typical of oral lichen planus. (Original magnifications: **A**, $\times 10$; **B**, $\times 20$; and **C**, $\times 40$.)

usually presents in middle age with slight female predominance and can affect the buccal mucosa, gingiva, tongue, and vermillion border of the lip. Idiopathic OLP is believed to be a T-cell-mediated response toward an unknown antigen and may represent an autoimmune reaction. Spontaneous remission is rare, and therapy is directed toward dampening the inflammatory response using corticosteroids or immune modulators. Patients may be refractory to treatment, and although reduction in symptoms can be achieved, no definitive cure has been identified. OLP carries a significant risk for transformation into squamous cell carcinoma, requiring surveillance for malignant changes.⁸ On examination, classic OLP is often indistinguishable from oral lichenoid tissue reactions (OLTR), highlighting the importance of thorough history taking. OLTR may be caused by systemic drug effects, localized contact reactions, or graft-versus-host disease.⁹ Adding to the diagnostic challenge, idiopathic OLP and OLTR have an identical histologic appearance that includes liquefactive changes of the basal keratinocytes with a bandlike array of activated T lymphocytes, macrophages, and dendritic cells. Histology isn't the diagnostic gold standard for OLP, and many infectious and immune-mediated diseases can induce oral lichenoid changes that mimic OLP. Oral lichenoid

contact reactions (OLCR) likely represent a delayed hypersensitivity reaction and removal of the allergen can lead to complete resolution.¹⁰ Given the similarities, oral lichenoid contact reactions can be misdiagnosed as OLP, leading to improper disease management. This patient presented with a rare and unique set of findings. The simultaneous onset of OLP and GC in an otherwise healthy woman may very well indicate a common immunologic or inflammatory etiology for 2 uncommon diseases of the oral cavity.

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