Coexistence of Cheilitis Glandularis and Lichen Planus: Remarkable Response to Anti-inflammatory Treatments

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Sir,
Cheilitis glandularis (CG) is a rare disorder with an unknown etiology characterized with erythematous dilated ostia of the minor salivary glands, varying degrees of lip enlargement (macrocheilitis) due to the retention of saliva in dilated ducts, and discharge of thick saliva when palpated.\(^1,2\) Cheilitis glandularis with lichen planus has been reported in only one case.\(^3\)

A 36-year-old female patient presented with gradually enlarging and painful bleeding of her lower lip within 20 years. The patient did not define mechanical irritation, smoking, atopic state, chronic sun exposure, or photosensitivity. She was on oral antidiabetic treatment for Type 1 diabetes mellitus for 5 years. She did not have xerophthalmia, xerostomia, or arthritis. Organomegaly, lymphadenopathy, or palpable mass or any glandular involvement such as submandibular, sublingual, lacrimal, and parotid glands were not detected. Dermatological examination revealed fine desquamation, lacy white streaks, dilated and erythematous multiple milimetric ductal openings, and mild serous discharge by palpation [Figure 1]. There was no vermilion or adjacent skin involvement. A wedge resection biopsy of the lower lip showed epidermal keratinization, granular layer, apoptotic cells lined in the basal layer, and lichenoid inflammation. Chronic lymphocytic inflammation of the minor salivary glands, periductal dense lymphocytic inflammation, and mild ductal ectasia were detected in the dermis [Figure 2]. The inflammatory infiltrate in the tissue did not contain any plasma cells staining with CD138. Periodic acid Schiff/alcan blue (PAS/AB) staining did not show any dermal Schiff/alcan blue staining did not show any dermal mucin or thick basal membrane. Fibrosis and obliterative phlebitis within the tissue were not present.

Laboratory tests including total blood count with differential, renal, liver, and thyroid function tests, fasting glucose and lipids, erythrocyte sedimentation rate, serum levels of C3, C4, and urinalysis were all within normal limits. The antinuclear antibody test was negative. Serum IgG4 level was 5, 3 mg/dl (normal range: 3.92–86.4 mg/dl). Depending on clinicopathological correlation, the patient was diagnosed as having coexistence of CG and lichen planus. Two sessions of intralesional triamcinolone injection were performed and 100 mg/d oral doxycycline treatment and topical 5-fluorouracil (5-FU) ointment thrice a week were initiated. After 2 weeks, pain and swelling complaints markedly regressed. Topical 5-FU and steroid injections were ceased. Instead, 0.1% topical tacrolimus ointment was added to doxycycline. After 4 weeks, doxycycline was stopped. Topical tacrolimus was continued for extra 3 months; however, minimal erythema and discharge did not regress completely. The treatment was stopped since the patient had not any pain and edema and very satisfied for the outcome.

Cheilitis glandularis was considered to be a reaction pattern of salivary glands to chronic sun exposure,
smoking, atopy, or mechanical irritation; however, some researchers suggested the origin of CG was the lip epithelium.[4] On the contrary, thick saliva production associated with such mechanism is refused by some researchers. Nico et al. detected different expressions of aquaporins in CG and they suggested that CG may be associated with abnormalities of water transport mechanisms in saliva production.[5]

Clinical variants of CG include simple type, superficial suppurative, and deep suppurative types. Our case was the simple type without any secondary infection. Irritant or allergic contact or mechanic cheilitis, granulomatous cheilitis of Melkerson-Rosenthal syndrome, sialolithiasis, actinic cheilitis, and IgG4-related disease are the main differential diagnoses of CG.[1,6,7] The absence of dense lymphocytic infiltrate, fibrosis, and oblitative phlebitis, which are the major histopathological features of IgG4-related disease, lack of any other glandular involvement and plasma cell infiltration, and detection of normal serum IgG4 level provided us the exclusion of lip involvement of IgG4-related disease. The correct diagnose of CG needs taking a careful medical history, clinical examination, obtaining a deep biopsy including salivary glands and clinicopathological correlation.

The histopathological features of CG are not specific. Reiter et al. proposed diagnostic criteria for CG.[1] Involvement of more than 2 minor salivary glands and mucoid and/or purulent discharge from the ostia (clinical criteria) must be observed. Additional features are the 2 or more of the histopathological findings (sialectasis, chronic inflammation, mucin, and metaplasia in ducts). The two clinical and two histopathological criteria are present in our case. We did not detect an obvious ductal ectasia and mucus in the ducts which may be the result of easy serous discharge from the ostia without a remarkable retention.

Some epithelial alterations may be observed in CG including spongiotic dermatitis, variable epithelial atypia along the vermilion border associated with solar elastosis, in situ, or invasive squamous cell carcinoma.[1,8] Discoid lupus erythematosus (DLE) of the lips usually show up with round, scaly, atrophic or depigmented patches or plaques, involving the vermilion border with epidermal atrophy, hyperkeratosis, PAS-positive thick basal membrane, and dermal mucin accumulation.[9,10] The presence of hyperkeratosis, hypergranulosis, band-like lichenoid infiltrate, absence of atrophy, lack of thick membrane and mucin stained with PAS/AB, and no spread from vermilion to the adjacent skin provided us the exclusion of DLE and the diagnose of lichen planus over CG pathology. Coexistence of CG and lichen planus was only once reported in a 30-year-old male patient with diagnose of mucocutaneous lichen planus for 5 years.[1] He developed CG in the last 2 months. The authors suggested that CG emerged secondarily on the inflammatory oral cavity disease. In the present case, we could not find out when lichen planus had developed. The long period of lip enlargement since adolescence suggests that isolated mucosal lichen planus may have developed on CG secondarily.

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**Conflicts of interest**
There are no conflicts of interest.

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