Cheilitis glandularis: a rare occurrence involving the upper lip

ABSTRACT
A variety of pathologic conditions of diverse etiologies may involve the lips. Cheilitis glandularis is a rare disorder of unknown etiology characterized by inflammation of the minor salivary glands, predominantly those of the lower lip. With externalization and chronic exposure, the delicate labial mucous membrane lining the minor salivary glands are altered by environmental influences, leading to erosion, ulcerations, crusting and, occasionally, infection. Importantly, susceptibility to actinic damage is increased. Therefore, cheilitis glandularis can be considered a potential predisposing factor for the development of actinic cheilitis and squamous cell carcinoma. This report is of a 12-year-old boy with cheilitis glandularis in an uncommon site, the upper lip. An overview of the etiology, clinical presentation, histopathologic features, and treatment are also discussed.

Key words: Child; Cheilitis; Salivary glands

Introduction
Cheilitis glandularis (CG) is a clinical diagnosis that refers to an uncommon, poorly understood, benign inflammatory disorder of the minor salivary glands in the lower lip. Although some researchers have speculated that CG represents a hereditary autosomal dominant condition, composite findings in most patients indicate that the condition represents a clinical reaction to chronic irritation of the lip from diverse external causes, such as actinic damage, factitial injury, atopy, infection, and tobacco irritation. Therefore, CG can be considered a potential predisposing factor for the development of actinic cheilitis and squamous cell carcinoma. This report is of an unusual occurrence of CG in an uncommon site, the upper lip of a 12-year-old boy.

Case report
A 12-year-old boy presented to the M. R. Ambedkar Dental College and Hospital, Bangalore, India, in 2005 with diffuse swelling of the upper lip for the previous 1.5 years. The patient was conscious of the lip swelling, although it was asymptomatic, and desired orthodontic treatment for his proclined upper front teeth. He had a habit of licking and biting the lip for the previous 3 years. He also complained of excessive drying of the lip from mouth breathing and constant exposure to wind.
At examination, his lips were incompetent and the swelling measured approximately 1.5 x 2.5 cm, and was oval-shaped and pale pink in color (Fig 1). There was associated crusting and scaling of the lip. On palpation, the swelling on the upper lip was soft in consistency, mobile, and fluctuant. On application of pressure, a discharge of mucinous fluid was seen. Intraoral examination revealed increased overjet and deep bite with two supernumerary teeth present between the upper central incisors, with no evidence of dental caries. Based on these findings, a provisional clinical diagnosis of CG was reached and differential diagnoses of double lip, angioedema, cheilitis granulomatosa, orofacial granulomatosis, chronic factitial injury, and actinic cheilitis were considered.

The patient underwent routine blood and radiological investigations. Based on the patient’s desire for orthodontic treatment, an orthopantomograph was taken, which revealed an impacted permanent left upper canine (23) and a retained primary left upper canine (63). A lateral cephalograph was taken to see the soft tissue profile where the increase in the size of the upper lip was evident (Fig 2). Routine blood investigation showed normal parameters for all tests except for erythrocyte sedimentation rate, which was slightly increased to 26 mm/h (reference range, 0-20 mm/h). A differential diagnosis of double lip was considered as the condition could be associated with Ascher’s syndrome, but this was ruled out by a complete physical examination and the clinical opinion of a general medicine consultant. The patient was referred to the Department of Oral and Maxillofacial Surgery for cosmetic management, and a transverse elliptical incision with debulking by surgical cheiloplasty was done under local anesthesia to restore normal lip architecture and function.

Hematoxylin and eosin–stained section revealed parakeratinized stratified squamous epithelium with a few areas of spongiosis (Fig 3). The underlying connective tissue showed numerous dilated blood vessels with perivascular inflammatory infiltrate and inflamed minor salivary acinar units and ducts (Fig 4). The histology findings confirmed the diagnosis of CG.

Discussion

Von Volkmann introduced CG for a rare disorder of unknown etiology characterized by inflammation of the minor salivary glands, predominantly those of the lower lip. Von Volkmann also described a clinically distinct, deeply supplicative, chronic inflammatory condition of the lower lip characterized by mucopurulent exudates.
from the ductal orifices of the labial minor salivary glands. Sutton 6 proposed that the characteristic lip swelling was attributable to a congenital adenomatous enlargement of the labial salivary glands. The incidence of CG is rare. In many patients, dysplastic (premalignant) surface epithelial change is evident histopathologically and frank carcinomas have been reported in 18 to 35% of patients 7. The condition often occurs in middle-aged or elderly men, but it also occurs in children, teenagers, and young adults. The risk for dysplasia and carcinoma increases with age, especially in fair-skinned individuals with sun-damaged skin, because the characteristic eversion of the lower lip results in long-term chronic exposure of the thin vulnerable labial mucosa to actinic influence.

Patients typically present for diagnostic consultation within 3 to 12 months of onset 8. The presentation varies according to the degree of pain, enlargement, loss of elasticity, and surface changes. Asymptomatic lip swelling initially occurs with clear viscous secretion expressed from dilated ductal openings on the mucosal surface. Some patients report periods of relative quiescence interrupted by transient or persistent painful episodes associated with suppurative discharge 9. Cheilitis glandularis affects the lower lip almost exclusively, and CG of the upper lip is a rare oral anomaly 10. Cheilitis glandularis manifests as progressive, often multinodular enlargement, eversion, and induration. Salivary gland duct orifices may be dilated, with spontaneous discharge of viscous clear secretions. In more suppurative disease, application of gentle pressure can elicit mucopurulent exudates. With advancing lip prominence and mucosal eversion, the mucosal-vermilion junction is obscured. Prolonged exposure to the external environment results in desiccation and disruption of the labial mucous membrane, predisposing the underlying minor salivary glands to inflammation, infection, and actinic influences 11. In this patient, CG could have been caused by a combination of factors such as actinic exposure, mouth breathing, and habitual lip biting.

Cheilitis glandularis has been subclassified into three types: simple, superficial suppurative, and deep suppurative. The superficial suppurative type has been termed Baelz’s disease, and the deep suppurative type has been referred to as myxadenitis labialis or cheilitis apostematosa 5,6. It is thought that these subtypes represent a continuum of disease wherein the simple type, if not treated, could become secondarily infected and progress to become Baelz’s disease and, eventually, the deep suppurative type. Chronic abscess and fistula formation are commonly seen in immunosuppressed patients. The latter two types of CG have the highest association with dysplasia and carcinoma. Lip enlargement is attributable to inflammation, hyperemia, edema, and fibrosis. Surface keratosis, erosion, and crusting develop consequent to long-standing actinic exposure,
unusual repeated manipulations that include self-biting or other factitial trauma, excessive wetting from compulsive licking, drying (sometimes associated with mouth breathing, atopy, eczema, and asthma), and other repeated stimuli that serve as a chronic aggravating factor. The histopathologic findings for this patient were consistent with those of CG.

Differential diagnosis should include double lip, angioedema, lymphangioma, chronic factitial injury, cheilitis granulomatosa, and orofacial granulomatosis. Treatment of CG is by vermilionectomy and antibiotic therapy for suppurative disease. Medical management of CG includes antibiotics, steroids and antihistamines. Cryosurgery and labial mucosal stripping are other surgical techniques for the treatment of CG.

Cheilitis glandularis of the upper lip is a rare oral anomaly. Cheilitis glandularis of the superficial suppurative type (Baelz’s disease), which this patient had, has a predisposition for dysplastic changes if left untreated. Given the fact that CG is known to coexist with immune suppression and other malignancies, careful evaluation and follow-up are critical for the management of these patients.

References