Case Report
Cheilitis Glandularis: A Case Report
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Abstract
Cheilitis Glandularis is an uncommon and rare inflammatory salivary gland disorder affecting the lower lip of adults with various etiological and predisposing factors. Based on clinical and histopathological findings three types of cheilitis glandularis have been described in the literature (i.e. simple, superficial suppurative and Deep Suppurative). The simple type being the most common but malignant changes have been observed in Deep Suppurative type. We report a case of cheilitis glandularis affecting lower lip in 20 years old female which is rare in that age and sex.

Key Words: Cheilitis; Glandularis; Suppurative; Autosomal Dominant; Mucopurulent; Minor Salivary Glands.

Introduction
Cheilitis glandularis (CG) was first described by Richard Von Volkman in 1870 as a chronic, suppurative inflammation of the lower lip characterized by swelling of the mucus glands and the mucopurulent discharge through the dilated ductal openings.\(^1\) It is an uncommon condition most commonly seen in the lower lip of adult male with little predilection to female, children and upper lip.\(^{1,2}\)

Historically three forms of Cheilitis Glandularis have been described, the simple, superficial suppurative, and deep suppurative with CG simplex being the most common.\(^{3,4}\) Initially it presents as simple type, if untreated, infection may progress to the superficial and to the deep suppurative type with malignant changes.\(^5\) Histologically CG is a nonspecific chronic inflammatory lesion characterized by localized dense accumulation of inflammatory cells and inflammation of mucus glands associated with glandular distension and ductal dilatation with notable loss of acinar architecture.\(^4\)

A variety of treatment modalities have been reported in the literature including topical steroid application, intralosional steroid injection and surgical excision.\(^6\) We hereby report an unusual case of CG of lower lip in a young female patient.

Case report
A 20 year old female reported with a swelling of lower lip since 2.5 year duration. She noticed swelling on right side which is gradually involving the left side of lower lip with frequent alteration in size (Fig 1). The lesion started with symptoms of dry and red lip which later was associated with bleeding from the dry areas upon stretching. History of topical application of Efudux (fluorouracil) for 2 - 4 weeks BID upon consultation with dermatologist relieved her symptoms with persistent swelling.

![Figure 1: Extra Oral Photograph of lips with areas of dryness and scars.](image)

Recurrence of the symptoms after 6 months of treatment made her consult a dentist. Upon examination there was diffuse swelling of lower lip and is everted with loss of normal contour and lip line. The surface was dry with bleeding from the cracks and scars in the previous healed areas (Fig 2). On Palpation, findings elicited soft to firm, tender swelling with bleeding upon stretching. With all the above findings the provisional diagnosis of cheilitis glandularis,
actinic cheilitis, atopic dermatitis, angioedema and cheilitis granulomatosa was made. Routine blood investigations performed were in normal limits except for decreased hemoglobin percent and elevated erythrocyte sedimentation rate. Biopsy was performed to confirm the diagnosis.

Figure 2: Prominent crusted areas with dry scaly appearance and small areas of bleeding points on the Lip when stretched.

Histopathology revealed hyperorthokeratinized and edematous stratified squamous epithelium with underlying dense admixture of acute and chronic inflammatory cells within the connective tissue, predominantly consisting of lymphocytes, plasma cells, and neutrophils surrounding the minor salivary glands and ducts (Fig 3). Lesion also revealed glandular distension and ductal dilatation with notable loss of acinar architecture (Fig 4). The above features were suggestive of Cheilitis Glandularis.

Figure 3: Photomicrograph showing hyperorthokeratinized stratified squamous epithelium with inflamed connective tissue and mucous salivary glands.

Figure 4: Photomicrograph showing loss of acinar architecture and inflammatory cells surrounding the ductal and acinar cells.

Discussion
Cheilitis Glandularis is a chronically descriptive diagnosis that refers to an uncommon poorly understood inflammatory disorder of the lower lip.7 In contrary to Volkmann (1870), Sutton (1914) stated that the characteristic lip swelling was attributable to a congenital adenomatous enlargement of the labial salivary glands.7,8 In 1984, Swerlick and Cooper analyzed retrospective cases along with 5 new cases stating that there was no evidence to support the assertion that submucosal salivary gland acinar hyperplasia is either responsible for or a consistent feature of established CG.9

Although the precise etiology of CG is still unknown but some have speculated that CG represents a hereditary autosomal dominant condition. Composite findings in most cases appear to indicate cheilitis glandularis represents a clinical reaction pattern to chronic irritation of the lip from a spectrum of highly diverse external causes like use of tobacco products, poor oral hygiene, and chronic exposure to environmental elements (sunlight and wind), compromised immune system and double lip associated with syndrome.1,10

CG appears to favor adult males however cases have been rarely reported in women.

No active treatment was under taken because of lack of symptoms and infection. But to relieve symptoms topical application of Tacrolimus 0.1% twice daily for 6 weeks was advised. Patient is under follow up.
and in children with wide age range between 4th to 7th decades of life. According to English literature only one case in a child, few cases (12 cases) in teenagers and Young adults 1,11,12 have been reported with possible predisposing factors like poor oral hygiene, chronic sun exposure, and a compromised immune status. However the present case is unusual because of its occurrence in a young aged female with possible etiological role of exposure to sunlight and wind.

Patients with CG typically present for diagnostic consultation within 3 - 12 months of onset. Complaints vary according to the nature and degree of pain, the enlargement, loss of elasticity of the lip and the extent of evident surface change. It manifests as progressive, often multinodular enlargement, with erosion and induration. Prolonged exposure to the external environment results in desiccation and disruption of the labial mucous membrane, predisposing it to inflammatory, infectious and actinic influences. In more supplicative cases application of gentle pressure can elicit mucopurulent exudates. With advancing lip prominence and mucosal eversion the mucosal vermillion junction is obfuscated.1,4 In contrary to above findings the present case also revealed same findings but due to the early treatment with fluorouracil the symptoms was mild with obfuscated vermilion zone.

CG historically been subdivided into 3 types believed to represent evolving stages in severity of a single progressive disorder.3 In simple type, multiple, painless, papular surface lesions with central depressions and dilated canals are seen. The superficial supplicative type (Baelz disease) consists of painless, indurated swelling of the lip with shallow ulceration and crust. Cheilitis glandularis of deep supplicative type comprises a deep seated infection with formation of abscesses, sinus tracts, and fistulas and potential for scarring. The latter two types have the highest association with dysplasia and carcinoma.3

The histological features of CG outlined in the literature have been characterized by dilated and tortuous minor salivary gland ducts, many of which are lined by oncocytic cells showing foci of hyperplasia and mucus metaplasia.13 Other well established features include acinar atrophy with infiltration of the glandular parenchyma by chronic inflammatory cells and the presence of extravasated mucin.14 In our case there was Inflammation of mucus gland lobules with glandular distension and ductal dilatation along with notable loss of acinar architecture favoring the diagnosis of simple superficial type of CG. Unfortunately the microscopic features are not specific enough and there is much overlap between the different clinical categories.

The approach to treatment for CG is based on diagnostic information obtained from histopathological analysis, the identification of likely etiological factors responsible for the CG, and attempts to alleviate or eradicate those causes. Emollients, topical corticosteroids and chemotherapeutic agents have been used in the treatment of the simple subgroup of CG successfully.6 Lederman proposed the avoidance of systemic corticosteroids due to ineffectiveness, however when injected intralesionally, superficial CG was treated successfully.15,16 Antibiotic therapy may be required concurrently if the lesions are infected, but selected after sensitivity testing of the supplicative exudates. According to Bovenschen, combined oral Minocycline (100 mg OID) along with Tacrolimus 0.1% twice daily for 6 weeks was successful in treating CG with deep infection.16 In Deep Suppurative group the recommended treatment of choice is vermillionectomy or labial stripping along with antibiotic therapy.14 The association of squamous cell carcinoma with deep supplicative type has been proposed in 18-35% of cases.13

**Conclusion**

Cheilitis Glandularis represents as an unusual disease that may be encountered in clinical practice and commonly seen in the lower lip. Patients with documented actinic cheilitis need to be monitored once or twice a year for an indefinite period. This is because, some patients may be at risk for the development of supplicative episodes if trauma to the lip surface is continuous leading to chronic ulceration or erosion and form portals of entry for bacterial invasion and inflammatory sequelae, few cases also has the potential for the development of lip carcinoma. In cheilitis glandularis, cases in which a history of chronic sun exposure exists, biopsy is strongly recommended to rule out actinic cheilitis or carcinoma. The present case is under follow up since 2 years with all local measures in controlling the recurrence by palliative treatment. We
also propose that this entity should be considered as a differential diagnosis of minor salivary gland swelling and focal ulceration.

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References

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