Cheilitis glandularis – Review of a rare inflammatory disorder

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INTRODUCTION
Cheilitis glandularis is characterized by hyperplasia of minor salivary glands along with varying degrees of inflammation.1,2 It is a rare disorder which can be characterized by swelling of the lip and hyperplasia of the labial salivary glands. It is a chronic inflammatory disease of salivary acini and ducts, usually involving the lower lip.3,4

In 1870, Richard Von Volkmann coined the term "Cheilitis glandularis" to describe a disorder that presented as a chronic, suppurrative inflammation of the lower lip, characterized by swelling of the mucous glands and associated with mucopurulent discharge through dilated ductal openings.3,4 This was later reported under various terms such as Cheilitis glandularis simplex, Cheilitis glandularis superficialis and Cheilitis glandularis apostematosa.5

In contrary to Volkmann (1870), Sutton (1914) stated that the characteristic swelling of lip was attributable to a congenital adenomatous enlargement of the labial salivary glands.6,7

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 chelitis glandularis.8

PREVALENCE AND ETOLOGY
It is a rare condition mainly seen in the lower lip of adult males with little predilection for females, children and the upper lip.9,10 Despite the male predilection, cases have been reported in women and children.11,12,13,14,15,16

It is most frequently associated with the lower lip, and in middle-aged to older Caucasian men.17,18 Although the lower lip is more commonly involved, lesions
involving the upper lip have also been reported. Familial cases have also been stated in literature. Syphilis, bacterial infections, actinic radiation, tobacco, poor oral hygiene and genetic transmission are the numerous causes implicated in the etiology though the exact cause is unknown. It can occur as a separate entity or in association with immunosuppression and malignancy.

CLINICAL FEATURES

The vermilion border is the most obvious element of the anatomy of the lips, which represents the junction between the skin and the mucosa. A French dermatologist, Jean Darier first conceptualized this as the semimucosa. The vermilion is the external representation of the pars marginalis of the orbicularis oris muscle. The lips continue their course beyond the vermilion border through the pars peripheralis of the orbicularis muscle. The lips exhibit openings and dilated ducts. It lacks mucosal eversion and induration of the lip with superficial inflammation forming abscesses and fistulous tracts. An analogous pattern of the disease of the oral mucosa has been described as stomatitis glandularis. In some cases, it may show progression to squamous cell carcinoma.

Inflammatory disorders of the lips are designated as 'Cheilitis' when the vermilion borders of the lips are affected. Whether the condition is acute and symptomatic, chronic or disabling, a thorough evaluation is mandatory to determine the cause and rule out malignancy. A good history along with clinical examination will often reveal the cause.

Schauermann classified Cheilitis glandularis into three types: 1) Simple, 2) Superficial suppurative (Baelz disease), and 3) Deep suppurative, and this classification is commonly used. The simple type is characterized by multiple painless lesions which exhibit openings and dilated ducts. It lacks inflammation, but mucinous material is extruded when the lip is squeezed. Numerous small nodules can also be palpated. On palpation, there is irregular induration of the lip and, with progression becomes enlarged, firm and everted. If these lesions are infected, the disease may further progress to the superficial or deep suppurative type.

Superficial suppurative type is thought to result from a secondary infection of the simple type. This type characteristically shows painless crusting, swelling along with induration of the lip with superficial ulceration. A color change is seen in the surface mucosa, which may produce clear to cloudy fluid at the sites of ductal openings. The lip is covered with seropurulent and hemorrhagic crusts with enlarged and tender minor salivary glands. Deep suppurative type is a deep-seated infection accompanied by abscess formation and fistulous tracts and is thought to be associated with chronic infection.

DIFFERENTIAL DIAGNOSIS

Angioedema, lymphangioma, vascular tumors, salivary gland tumors, mucocele, Meischer syndrome (cheilitis granulomatosa), sarcoidosis and plasma cell cheilitis should be included under the differential diagnoses of Cheilitis glandularis. A condition involving the lower lip termed "Acute ectropionizing bacterial cheilitis" which has been described in German literature also marks a differential diagnosis. According to Bork et al, this rare disease is caused by exacerbation of a folliculitis or furuncle affecting the adjacent skin. Acute ectropionizing bacterial cheilitis manifests with edema of the lower lip. Within a few hours of inflammatory infiltration, eversion and crusts also develop on the mucosal part of the lip. Abscess-forming or phlegmonous inflammation has not been
extravasated mucin. Chronic inflammatory cells and the presence of infiltration of the glandular parenchyma by other well established features include acinar atrophy along with infiltration of the glandular parenchyma by chronic inflammatory cells and the presence of extravasated mucin.

**HISTOPATHOLOGY**

The clinical evaluation and histopathology of the salivary glands is a complex and difficult area of diagnostic pathology. Histologically, it is a nonspecific chronic inflammatory lesion which is characterized by localized dense accumulation of inflammatory cells along with inflammation of mucus glands associated with glandular distension and ductal dilatation with notable loss of acinar architecture. The histological outline described in literature has been characterized by dilated and tortuous minor salivary gland ducts, many of which are lined by oncocytes cells showing foci of hyperplasia and mucus metaplasia. Other well established features include acinar atrophy along with infiltration of the glandular parenchyma by chronic inflammatory cells and the presence of extravasated mucin.

**TREATMENT**

The approach to treatment for Chelitis glandularis depends upon diagnostic information received from histopathological analysis, the identification of possible etiological factors responsible for the CG and the attempts to alleviate or eradicate those causes. The treatment ranges from conservative treatment with steroids - topical and systemic, to more extensive treatment such as resection. The first step in treatment is the reduction or elimination of predisposing factors. If predisposing factors are not identified or eliminated, conservative treatment should ensue, including the use of topical steroids, intralesional steroids, systemic anticholinergics, systemic antihistamines, and/or antibiotics. Emollients, topical corticosteroids and chemotherapeutic agents have been successfully used in the treatment of the simple subgroup of Chelitis glandularis.

Lederman has proposed the avoidance of systemic corticosteroids due to ineffectiveness. However, when injected intralesionally, superficial type was treated successfully. If the lesions are infected, antibiotic therapy may be needed concurrently, but selected after sensitivity testing of the supplicative exudates. According to Bovenschen, combined oral Minocycline (100 mg OD) along with Tacrolimus 0.1% prescribed twice daily for 6 weeks was successful in treating CG with deep infection. If conservative therapy fails, surgical intervention is often necessary to reverse lip erosion and to remove the sources of inflammation of the minor salivary glands.

Patients with deep supplicative type of the condition, should be considered for surgical excision and must be closely monitored due to the risk of developing squamous cell carcinoma. Surgical options are inclusive of cryosurgery, vermilionectomy, and/or labial mucosal stripping. With the Deep Suppurative type, vermilionectomy or labial stripping along with antibiotic therapy is the recommended treatment of choice. The patients should be scheduled for periodic follow up in order to rule out any suspicious clinical changes.

**REFERENCES**

17. Bender MM, Rubenstein M, Rosen T. Cheilitis...


