Cheilitis Granulomatosa—An Uncommon Clinical Presentation with its Non-Surgical Management
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Abstract

Cheilitis granulomatosa is characterized by recurrent or persistent swelling of one or both lips. The exact etiology is not certain. It may represent oral manifestations of Crohn's disease and sarcoidosis, so the complete diagnostic work up of the patient with suspected Cheilitis granulomatosa is a must. Microscopically, a non-necrotizing granulomatous inflammation is the typical feature. Management depends upon the severity of the condition and the patient's esthetic concerns. Here a case of successfully treated Cheilitis Granulomatosa along with gingival involvement in an 18 year old female is presented along with its clinical features.

Keywords: cheilitis; female; granuloma; Intralesional steroid

Introduction

Cheilitis Granulomatosa (CG) is a rare granulomatous disease of uncertain etiology, described by German dermatologist Miescher in 1945 as a distinct clinicopathological entity, characterized by diffuse, not tender, soft to firm recurrent or persistent swelling of one or both lips. [1] CG has often been associated with other orofacial Granulomatous disorders e.g. atypical Tuberculosis, Anderson-Fabry disease, Crohn's disease, sarcoidosis, and allergic reaction. It is also considered as an oligo-symptomatic or mono-symptomatic form of Melkersson Rosenthal syndrome (MRS). CG usually affects young adults, mostly in the 2nd decade of life with a female predominance with incidence of 0.08% [1]. The diagnosis of cheilitis granulomatosa is made by correlation of the patient's history and clinical features, supported by the histopathological findings. Management of cheilitis granulomatosa is difficult, as described in the literature. Different treatment modalities have been reported, from various conservative treatments to surgical interventions, with variable outcome. [2]
Case Report

An 18 year old female patient reported with a chief complaint of swelling in upper lip and anterior gums since last 5 years, which was painless and persistent. History of the presenting illness revealed that initially there was a swelling of the upper lip followed by the anterior gum and slowly progressed to the present extent, with exacerbation and remission periods since 3 years. There was no apparent history of trauma, allergy to any substance, insect bite, pain in the teeth, pus discharge, fever, facial paralysis or any other history of systemic ailment. Patient consulted a physician 2 months back for the same problem, and was prescribed symptomatic systemic medication, but got no relief. Patient was unaware about the medication prescribed as the records were not available. The family history and past dental history of the patient were non-contributory. The swelling was consistent in size without any aggravating or relieving factors. On general physical examination, the patient appeared moderately built and nourished with the other vital signs were within the normal limits.

Extra oral examination revealed a diffuse swelling of upper lip with smooth and intact overlying skin, with dry and scaling of upper lip. On palpation, the lip swelling was soft in consistency, non-tender, non-fluctuant and there was no localized increased in the temperature. Regional lymph nodes were not palpable. Intraoral examination revealed midline crack and bleeding spot in the upper lip. In upper anterior region, the gingiva was lobulated and enlarged, extending mesio-distally from upper right canine to left canine regions and superior-inferiorly from labial vestibule to partially covering the crowns with blunting and enlargement of interdental papillae. In the lower anterior region, gingiva was enlarged from left canine to right canine region, with apparent inflammation. Enlarged gingiva was reddish pink in color, soft to firm, with loss of stippling & bleeding on probing. [Fig.1]

Fig 1: Extra orally, diffusely enlarged upper lip which was soft, non-tender with smooth and intact overlying skin. Intra-orally, there was midline crack and bleeding spot visible. Also, the anterior gingiva inflamed, red, soft to firm and with bleeding on probing.
On the basis of history and clinical examination, a provisional diagnosis of Chronic Idiopathic Granulomatous lesion of the upper lip and gingiva was made. The differential diagnosis included cheilitis granulomatosa, angioedema, cheilitis glandularis, neurofibroma, exfoliative cheilitis, plasma cell cheilitis, sarcoidosis, Crohn's disease, and tuberculosis. Vitality test in relation to involved teeth region was done and teeth were found to be vital. Routine Blood investigations (Hb%, BT, CT, TLC, DLC), RBS, HbSAg and HIV were in the normal range except there was increased ESR. On endoscopic examination to rule out Crohn's disease, the intestinal mucosa appeared normal. The chest radiograph appeared normal. Patch testing was done for commonly used food products and cosmetics and the results were negative. An incisional biopsy of the lip and gingiva was performed under local anesthesia. Biopsy specimen revealed stratified squamous parakeratinized epithelium overlying an edematous connective tissue stroma with areas of noncaseating granulomatous infiltrate consisting of lymphocytes, foamy histiocytes, epitheloid cells and multinucleated giant cells, which were suggestive of chronic granulomatous lesion of the lip [Fig.2].

Fig 2: Photomicrograph of the lesion showing infiltrates of inflammatory cells in sub epithelial connective tissue. (H & E, x40).

Based upon the history, clinical examination and subsequent investigations, a final diagnosis of Cheilitis granulomatosa of the upper lip and anterior gingiva was made. Treatment included intralesional injections of 0.1ml of Triamcinolone Acetonide (TA) (40 mg/mL) into the upper lip vermillion and mucosa and anterior gingiva after mixing 0.05ml of adrenaline in the injection, on weekly bases for 2 months. There was a significant reduction in the lip and gingival swelling. The swelling regressed following our treatment and the patient was asymptomatic after treatment. [Fig.3]. The patient was re-evaluated each week for the next 6 weeks for any recurrence. Subsequently, the patient is under follow up since 18 months and has shown no signs of recurrence.
Fig 3: Showing reduction in the swelling of upper lip and anterior gingiva.

Discussion

The exact etiology & pathogenesis of CG is unknown. Hornstein suggested that the CG may be caused by an alteration in autonomic nervous system function localized to facial skin, resulting in increased vascular permeability and edema.[1] Other etiologic factor documented is cell mediated hypersensitivity reaction as there is presence of activated helper T lymphocytes expressing interleukin-2 receptors in these lesions.[3] It is also postulated that the cytokine production by lymphocytic clone could be responsible for the formation of granulomas in CG.[4] Chronic focal infections, sensitivity to food items and lymphatic vessels obstruction have been suggested as contributing factors.[1] According to Kano et al.,[5] some patients with CG are predisposed to Crohn's disease, and CG may precede intestinal Crohn's disease by few years. Worsaae and Pindborg reported that gingival swelling may precede the lip swelling and the gingival manifestations appeared mainly in the anterior part of the mouth.[6]. In the present case gingival enlargement preceded the lip swelling. In our case CG occurred on the upper lip which was in accordance to the available literature which stated that upper lip was the most common site of occurrence.

Histologically, there is dilation of lymphatic vessels, perivascular lymphocytic infiltration and to a varying degree non-necrotizing granulomas are seen in the lamina propria. Typically, the granulomas appear to cluster around scattered vessels and are not well formed or discrete. Fibrosis may be present in long term lesions.[1]

Diagnosis of CG is done by ruling out other conditions. The differential diagnosis for a swollen lip can be extensive but a good clinical history and thorough clinical examination will usually eliminate many diagnostic possibilities.[7] In approximately 42% of MRS patients, CG precedes other signs but reverse is not true as most of CG patients never develop MRS. Van der Waal et al., state that, although non-caseating granulomas are classically present, the histology may be nonspecific, with edema and perivascular lymphocytic infiltrate.[8] The diagnosis of CG is therefore a diagnosis of exclusion. It is necessary to exclude the possible local and systemic diseases that can

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have similar clinical manifestations, including hypersensitivity reactions. Further workup includes sarcoidosis, Crohn's disease, tuberculosis, deep fungal infections, leprosy, leukemic infiltrate, and foreign body reactions. Sarcoidosis is unlikely in the absence of respiratory problems, and if chest x-ray and ACE levels are normal. Biopsy of labial salivary gland can be of help in diagnosing sarcoidosis. Crohn's diseases can be ruled out if gastrointestinal complaints are absent.[9] IgE levels and patch test may be of diagnostic value when hypersensitivity reaction is suspected. Tuberculosis should be ruled out by acid fast bacillus staining and chest radiographs.[10]

Management of CG is dependent on correct diagnosis of the condition and identification of any precipitating factors. Patients without dental infections with CG should be enquired for the presence of systemic signs and symptoms of Crohn's disease, sarcoidosis or a history of angioedema. In the presence of positive findings, the patient should be referred for further systemic evaluation. Various documented treatment modalities includes surgery, oral and intralesional Corticosteroids and boiled water injections, drugs like antituberculous drugs, antibiotics, vitamins, phenylbutazone, ACTH.[1] Corticosteroids have been shown to be effective in reducing lip swelling and preventing recurrences and are considered the mainstay of therapy. Patient with mild swelling can be managed by using topical steroids. Severe cases of swelling of the lip can be treated with Intralesional Triamcinolone Acetonide (0.1%) injections. Patients with more extensive lip swelling can be initially treated with systemic medication.[11] Surgery is indicated only in severely disfiguring cheilitis and once the disease has been brought into a quiescent phase and should thereafter be treated with biweekly to monthly Triamcinolone 0.1% injections for 2-6 months to prevent relapse.[12] Recurrence, though minor, is not uncommon. Therefore, treatment is currently mainly empirical, and based largely on the severity of symptoms.

In our patient the intralesional Triamcinolone resulted in resolution of the lesion with no recurrence during the follow up period. Surgical re-contouring is not frequently used as there is an increased risk of recurrence. Prognosis is variable and is best in early diagnosed cases.[13]

References


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