Cheilitis Granulomatosis - A Case Discussion

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Abstract

Introduction: Cheilitis granulomatosa is an idiopathic, cosmetically unpleasant, non-specific, painless, persistent granulomatous inflammation which affects one or both the lips. It is considered under the entity orofacial granulomatosis with very high rate of recurrence inspite of good treatment.

Presentation of case: A 45 years old female patient reported with the chief complaint of swelling in the upper and lower lips for 2 years which had gradually increased in size since the onset. The swelling was asymptomatic, firm in consistency. On histopathological examination the condition was diagnosed as cheilitis granulomatosa

Discussion: Cheilitis granulomatosa is considered as an entity of orofacial granulomatosis. It is asymptomatic, persistent swelling of the lips. The primary goal of management should be elimination of the predisposing factor. Intralesional corticosteroids are considered as gold standard in the management of conditions. In severe cases surgical management is helpful.

Key words: *Cheilitis, Corticosteroids, Granulomatosis, Intralesional injection, Lip swelling*

Introduction

In 1985, the concept of orofacial granulomatosis was introduced by Wiesenfield and colleagues to include lesions that resemble Crohn's disease, clinically and histologically, but occur in patients who do not exhibit the gastrointestinal manifestations of Crohn's[1,2].

The exact etiology of cheilitis granulomatosa is undefined but the list of predisposing and aggrevating factors include exposure to sun, allergens, food allergies, and systemic illnesses like Crohn's disease, Sarcoidosis and Tuberculosis. On proper investigations we couldn't rule out the exact etiology, but we considered sunlight to be a predisposing factor as the patient was a farmer working for long durations in the sun[3].

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Case history:

A 45 year old female patient reported with a chief complaint of asymptomatic swellings of upper and lower lips. Patient gave a history of the swelling persisting for more than 2 years and with mild degree of regression since the onset (Figure 1). Patient was otherwise healthy and there was no history of similar complaints in the family.

On clinical examination, the lips were diffusely swollen, smooth, non-ulcerated with no evidence of any discharge on or around the swelling and a mild degree of surrounding erythema. The lips were chapped, normal in color and non-tender. The lips were firm in consistency and non-tender, non-reducible, non-compressible, non-fluctuant and non-pulsatile, with neither local rise in temperature nor positive transillumination.

On investigating the case, the vital signs were normal. Lab investigations for hematology showed normal values for CT,BT, ESR, DLC, RBS, HbsAg, HIV, serum folate, Iron, Vitamin B12 . Haemoglobin which was 9.5 grams/dl. The tuberculin skin test to rule out tuberculosis was negative.

There were no clinical or radiographic evidences of any odontogenic cause.

The patient was referred to an oral maxillofacial surgeon for biopsy of the upper lip that revealed stratified squamous parakeratinized epithelium, overlying edematous connective tissue stoma and delicate collagen fibres and chronic perivascular inflammatory cells infiltrating with some focal noncaseating granulomas in the submucosal connective tissue consisting of lymphocytes, histocytes, epitheloid cells and Langerhans type of multinucleated giant cells .The histopathological study was suggestive of a granulomatous lesion of the lip.

Based on the clinical and histopathological findings the case was diagnosed as Cheilitis granulomatosa. Thereafter treatment was planned with intralesional injection of triamcinolone 40mg -1ml mixed with 0.3 ml of normal saline which was deposited in both upper and lower lips at three different sites in between the junction of vermillion border and lining mucosa. After 2 intralesional injections there was around 40% reduction in swelling of the upper and lower lips (Figure 2).

A total of 6 intralesional injections were given to the patient every alternate week. After completion an 80% reduction in the swelling was acknowledged. Two more intra lesional injections were planned following which total remission was observed (Figure 3). The patient was followed-up for 6 months without any recurrence after which patient did not report.

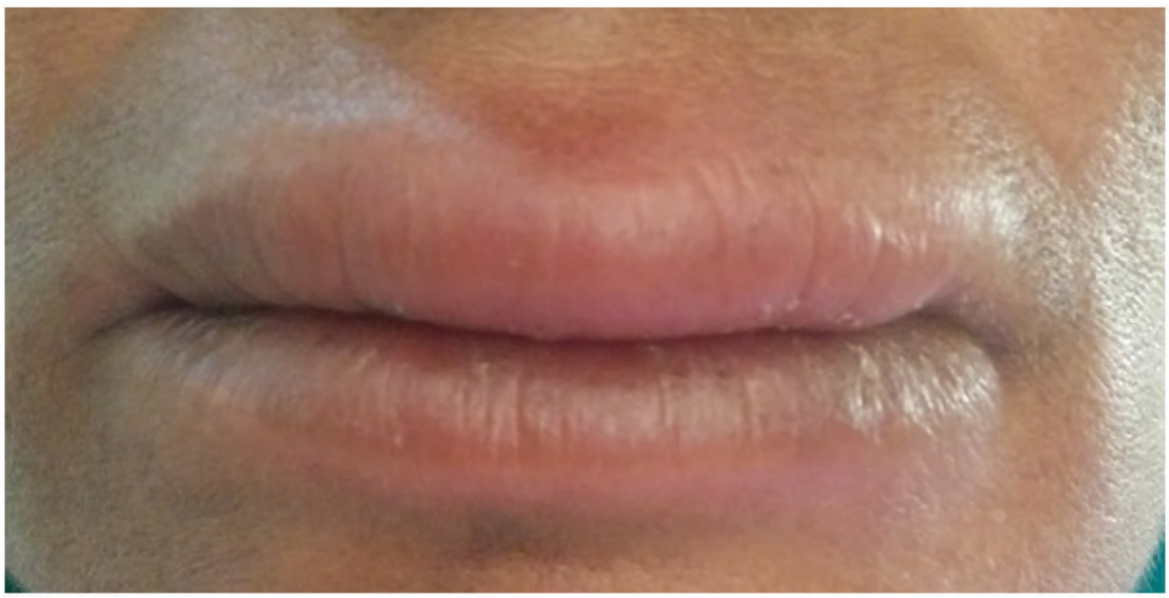

Figure 1: The appearance of upper and lower lips on the first visit to the hospital.

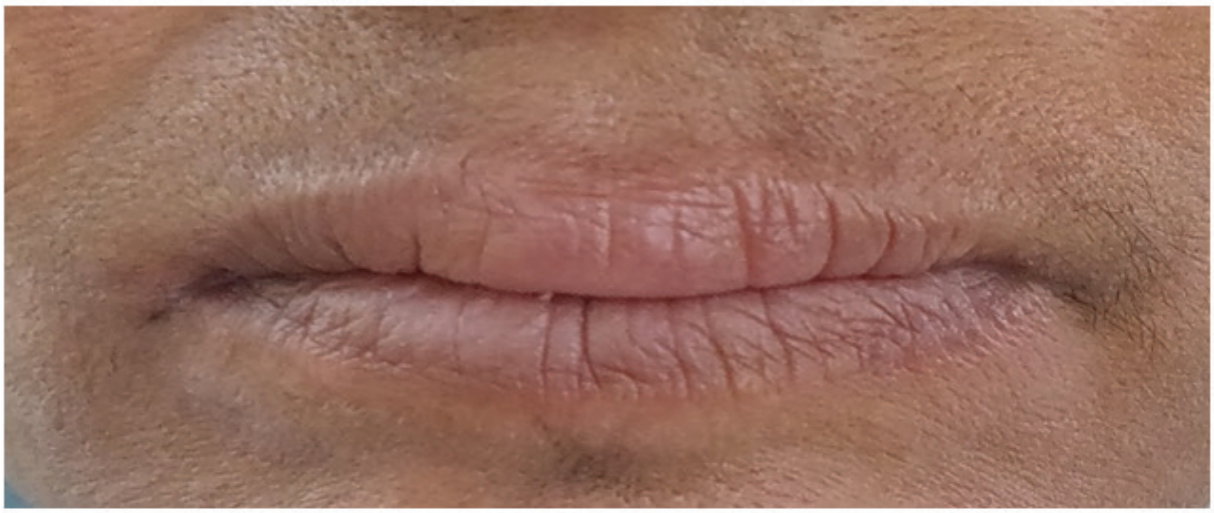

Figure 2 : The appearance of the lips after 2 injections , showing about 40% reduction in the swelling.

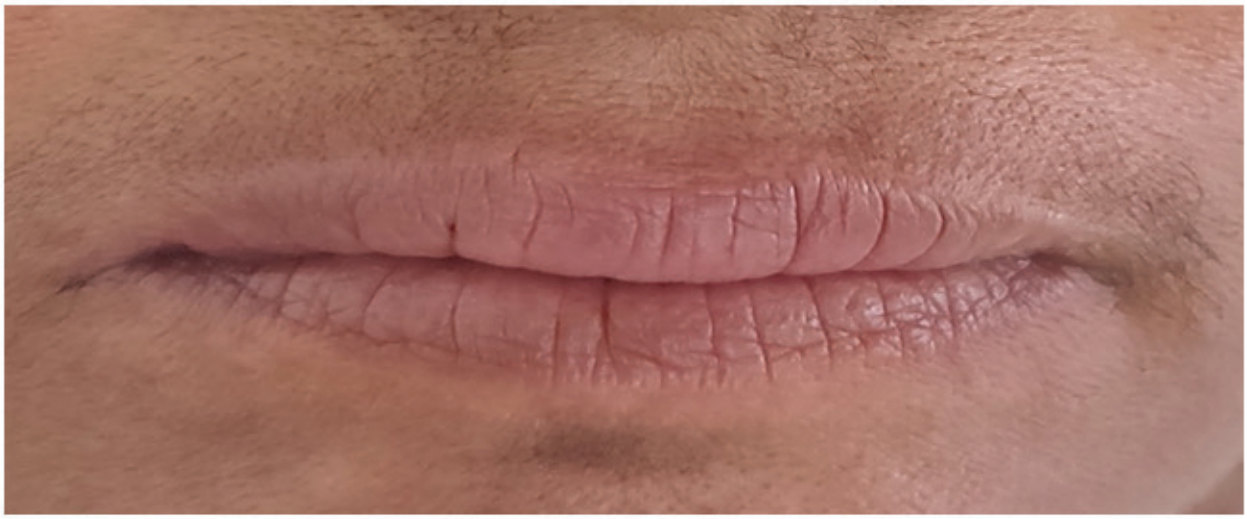

Figure 3: The appearance of lips after intralesional

Discussion

Cheilitis Granulomatosa (CG) presents as a persistent asymptomatic and recurrent lip swelling not associated with any particular age, sex or race. It is a presentation of orofacial granulomatosis which is a nonspecific granulomatous inflammation presenting as facial or lip swelling, cheilitis, ulcerations, gingival enlargement, mucosal tags and sometimes lymphadenopathy. The other differential diagnosis includes angioedema, Cheilitis glandularis, actinic cheilitis Melkersson-Rosenthal syndrome, Chron's disease, Sarcoidosis and tuberculosis[4].

Few of the differentiating features helped us to eliminate differential diagnosis and to arrive at final diagnosis. Presence of mucopurulent discharge as a result of pressure build- up most commonly in the lower lip due to the blockage of the salivary duct, surfacing the dilated salivary gland orifices as in cheilitis glandularis was missing in our patient. In this scenario patient had asymptomatic swelling of the lips for more than 2 years which helps in eliminating the condition angioedema where the symptoms regress with in 4 days. Actinic cheilitis usually affects lower lip as the upper lip is relative less exposed to sunlight. This condition usually results In loss of normal lip architecture which was not

seen in our patient. Even though patient presented with cheilitis the other components like facial paralysis and fissured tongue were missing to consider the condition as Melkersson-Rosenthal syndrome[5]. Endoscopy was performed to rule out gastrointestinal disturbances which helped in wiping off Chron's disease from our list. Oral manifestation of sarcoidosis are the presence of lesions on tongue, gingiva, palate and cheeks which were absolutely not found in our case thus excluding sarcoidosis. Tuberculosis was ruled out by the negative result of tuberculin skin test[6].

Non-specific granulomatous inflammation was seen on histopathology which confirmed cheilitis granulomatosa.

For the management a synthetic glucocorticoid triamcinolone which acts as anti-inflammatory and immunomodulator was chosen in our case because of its delayed release and high concentration[7]. It works by blocking the release of arachidonic acid from membrane phospholipids there by preventing the synthesis of prostaglandins and leukotriens, the mediators of inflammation. Similarly the condition was treated by Coskun et al. with a combination of intralesional steroid and metronidazole[8]. Likewise Stein and Mancini have used oral prednisolone and minocycline for treating cheilitis granulomatosis in children[9]. A combination of intralesional triamcinolone, metronidazole and minocycline was used by Dar et al. and they observed improvement in one month. Clofazimine was used by Gibson et al. for treating the condition. Miralles used 1000mg metronidazole to treat such cases[10].

The elimination of the differential diagnosis help in the proper final diagnosis. Elimination of all possible odontogenic causes is important in further management of this condition.

Conclusion

The complete resolution of the condition and prognosis may vary from patients to patients due to their immunological responses and adaptation to treatment. The primary treatment should be the elimination of the etiology of the condition. If the condition does not subside with a single drug usage, combination of drugs will be helpful. There is a need for extensive studies to find the etiology and the cure.

Conflicts of Interest - Nil

Financial support - Nil

Ethical Clearance – Patient's consent was obtained for publication of images and/or information about them.**injections showing complete remission of the swellings.**

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