Orofacial Granulomatosis of the Upper Lip: A Case Report

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ABSTRACT:
Orofacial granulomatosis (OFG) is a granulomatous disease of the orofacial region which appears usually as a persistent and/or recurrent labial soft tissues swelling in the orofacial region and is characterized histologically by a granulomatous inflammation. OFG may cause significant cosmetic and functional problems but can be prevented if the disease is diagnosed early and promptly treated. The etiology of oral lesions with non-caseating granulomas includes oral Crohn’s tooth associated infections, sarcoidosis and food or contact allergies. Treatment of OFG is not always necessary, although most patients do require some medical intervention.

Key-words: Diagnosis, Orofacial Granulomatosis, Treatment.

INTRODUCTION
Orofacial granulomatosis (OFG) is an uncommon, but increasingly recognized disorder characterized by persistent and/or recurrent labial swelling, oral ulcers and a variety of other orofacial features, in the absence of identifiable Crohn’s disease of the intestine or sarcoidosis. Granulomatosis seems to cause lymphatic blockage, leading to a diffuse swelling of the lips and other sites because of lymph-oedema. (1)

The term OFG was introduced in 1985 by Wiesenfeld et al. referring to the presence of granulomatous inflammation in the orofacial region. (2, 3)

OFG usually presents as swelling of upper and/or lower lip. In some cases unilateral or bilateral orofacial region, including chin, cheeks, peri-orbital swelling zygomatic area, eyelids and forehead can be seen.

A number of other features can be developed such as oral ulcers, oral mucosal swelling and tags, gingival enlargement, fissuring of the tongue, facial nerve palsy, erythema of the face and cervical lymphadenopathy. (1, 4)

The etiopathogenesis of OFG remains elusive, although minor immunological changes have been reported in a number of patients. Patient with OFG may have a history of atopy occasionally associations with food intolerance, e.g. monosodium glutamate and also allergy to food preservatives and chocolate have been considered. As the removal of amalgam resulted in reducing of swelling of buccal mucosa and lips of OFG in isolated cases, delayed hypersensitivity to dental materials has occasionally been implicated. (5-10)

Microbiological agents and genetic predisposition factors are considered as etiological factors. Furthermore, attempts have been made to analyze the relationship between T cell and cytokines in the pathogenesis. (2)
CASE REPORT

A 19-year-old woman was referred to dental faculty with complaining of lip swelling with exacerbation and remission periods since 9 months ago. The patient had no systemic disease and there was no history of foreign body reaction or trauma. Clinically, upper lip had rubbery consistency and scaling of lip was present. There was no palpable lymph nodes in the face and neck regions. Routine laboratory tests (CBC diff, ESR, Ca, P, Platelet count, ACE, SI, TIBC, and Ferritin) or were within normal limits. Radiographic studies of the chest did not reveal any abnormalities. There was no hilar lymphadenopathy on the chest radiograph (CXR). No clinical signs or symptoms of gastrointestinal (GI) disease were present. The patient had taken a course of antibiotic and antihistamine drugs. A mild recovery had occurred just for a short time following treatment and then the swelling relapsed again. The patient underwent incisional biopsy.

Histopathological examination revealed a non-necrotizing granulomatous lesion composed of epithelioid histiocytes and giant cells with a peripheral rim of lymphocytes. A diagnosis of OFG was made based on correlation of the histopathologic findings with normal laboratory test results and normal CXR findings (Fig.1).

Histologically, the lesion was characterized by non-caseating granulomas consisting of lymphocytes and epithelioid histiocytes. In the superficial lamina propria and perivascular aggregation of dilated vessels

![Fig. 1: Diffuse swelling of the upper lip before treatment](image1)

![Fig. 2: Low-power magnifications demonstrating granulomatous inflammation beneath the mucosa (H&E; magnifications 10x).](image2)

![Fig. 3: High-power magnifications demonstrating granulomatous inflammation beneath the mucosa (H&E; magnifications 40x).](image3)

Histiocytes and plasma cells were seen. Stains for acid fast bacteria and fungal organisms were negative and non-polarizing foreign materials were present (Fig.2 &3). The patient was treated by intra-lesion injection of Triamcinolone 10% twice weekly for a period of two month (Fig.4). She was followed up for three months and recurrence did not occur.
Fig. 4. Resolving of upper lip swelling after treatment.

DISCUSSION
OFG is a better term for those cases restricted to oral region which are not associated with any identifiable other granulomatous disease.(10) Diagnosis of OFG is confirmed through taking of incisional biopsy. Blood test, radiography and endoscopy are needed to differentiate OFG from Crohn’s disease, sarcoidosis, tuberculosis and foreign body reaction.(10) In patients presenting with classical OFG, the early diagnosis of the disorder is generally on the basis of clinicopathological findings, haematological and radiographic findings. However, it is not so difficult to rule out sarcoidosis, tuberculosis, leprosy (Hansen’s disease), deep fungal infections, hypersensitivity reactions, acquired and hereditary C1INH-related angioedema, leukemic infiltrates, and dentoalveolar abscesses.(4)

The most common reasons for lip swelling are trauma, infection and angioedema but all cause transient swelling. A number of disorders especially Crohn’s disease can mimic the more persistent features of OFG. The differential diagnoses of OFG include orofacial granulomatosis (cheilitis granulomatosa, Misser syndrome), Crohn’s disease, Sarcoidosis, Foreign body reaction, mycobacterial infection (tuberculosis, leprosy, a typical mycobacterium), deep fungal infection, contact allergy, chronic granulomatous disease (of childhood). Confusingly, it has been suggested that 10-37% of the patients with OFG have Crohn’s disease, and oral lesions may precede the development of intestinal involvement. Rarely, other conditions such as amyloidosis, microcystic adnexal carcinoma, cysts and Sjögren’s syndrome can cause labial swelling.(1,2)

Sometimes the swelling resolves spontaneously without treatment, but most often persists for years. As single treatment is not always effective; a number of options are suggested such as: eliminated diet, anti inflammatory drugs, radiotherapy and plastic surgery.(1,3,4)

REFERENCES