CASE REPORT

Orofacial Granulomatosis: A Case Report with Review of Literature

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ABSTRACT:

Orofacial granulomatosis is a granulomatous disease affecting orofacial region. It is characterized by persistent and/or recurrent labial swelling, ulcers which on biopsy shows lymphoedema & noncaseating granulomas. It is important to establish the diagnosis accurately because this condition is sometimes amanifestation of other granulomatous diseases like Crohn's disease or sarcoidosis. Here we report a case of orofacial granulomatosis presenting as a persistent swelling of lip. The diagnostic approch & treatment of orofacial granulomatosis are reviewed.

Key words: Orofacial granulomatosis, Crohn's disease, cobble stone appearance, mucosal tag.

INTRODUCTION

Orofacial granulomatosis(OFG) unrelated to a certain systemic disease was first reported and described by Melkersson in 1928 as an orofacial swelling accompanied with facial nerve palsy.[1] But the term OFG was first introduced by Wiesenfeld in 1985.[2, 3] OFG is an uncommon disorder characterized bypersistent and/or recurrent labial enlargement, oralulcers and a variety of other orofacial features, in theabsence of identifiable Crohn's disease of the intestineor sarcoidosis. There remains no consensus on whether OFG is a distinct clinical disorder or an initial presentation of Crohn's disease, as it can be difficult reliably to exclude Crohn's disease, and also to predict if gastrointestinal Crohn's will develop at some stage in the future. Nevertheless, it is distinct fromclassic Crohn's disease, or it may represent a variant,since patients with OFG may have different humanleucocyte antigen (HLA) profiles to those with Crohn'sdisease.[3]

The exact cause is unknown but several theories have been suggested, including infection, genetic predisposition and allergy.[3, 4] However, an immunologic origin (cell-mediated hypersensitivity reaction) is favoured because of the presence of activated helper T lymphocytes expressing interleukin-2 receptors in

these lesions.[3] Patients with OFG may have ahistory of atopy, and there are occasional associationswith food intolerance, e.g. monosodium glutamate andfood preservativesand chocolate. Delayed hypersensitivityto dental materials has occasionally beenimplicated, and the removal of amalgam has causedreduction of swelling of buccal mucosa and lips of OFGin isolated cases.[3]

The clinical presentationof OFG is highly variable, with persistent, painless swelling of the orofacial tissues as consistent finding.Lips are the most frequent site of involvement andother intraoral sites commonly affected are the tongue,gingiva and buccal mucosa.[5]

CASE REPORT

A 17 year old male patient came to the department of oral medicine & radiology with a chief complaint of swelling of lower lip since 2 months. Swelling was sudden in onset, started 3 months back in right & left buccal mucosa; extended to lower lip & persisting in the samemanner. Patient had similar swelling in the lip 6 months back & has been treated with fexofanadine by a dermatologist. Swelling subsided but recurred again after 3months. Past medical history & family history were not significant. There was no history of allergy, tuberculosis, Crohn's disease. Vital signs were within normal limits.Extraoral examination revealed no lymphadenopathy. Lower lip showed a diffuse swelling (fig 1) which is firm & non tender on palpation. On intraoral examination, buccal mucosa was swollen with folds (fig 2) giving it a cobble stone appearance. Upper labial vestibule showed red mucosal tag (fig 3). Based on the history & clinical features a provisional diagnosis of OFG was made & a differential diagnosis of Crohn's disease, sarcoidosis & angioedema was given.

Incisional biopsy was done on labial mucosa & submitted for histopathological examination. Histopathology revealed stratified squamous epithelium & underlying connective tissue which revealed perivascular & paravascular collection of lymphocytes, histiocytes & focal noncaseating granulomas (fig 4). Correlating the clinical & histopatholgy, a final diagnosis of Orofacial granulomatosis has been etablished.

Once the diagnosis has been established, systemic steroid therapy (prednisone 40 mg thrice daily for 1 month) & doxycycline 200 mg once a day for 2 months was started. Swelling decreased in size. Steroid therapy was tapered with a maintainence dose of 20 mg once a day.

DISCUSSION

OFG is normally seen in the second decade of life and has a female predilection. [6]The differential diagnosis of a persistent labial swelling includes angioedema (idiopathic or hereditary), sarcoidosis, Crohn's disease, OFG, Chelitis granulomatisum (CG), Melkerson Rosenthal Syndrome (MRS) and some specific infections like tuberculosis, leprosy and deep fungal infections. Amyloidosis&Ascher's syndrome may also be included in the differential diagnosis. All of these conditions must be taken into account during the investigation of a patient with persistent lip swelling. The medical history and clinical examination help to direct the investigation. The biopsy helps in establishing the correct diagnosis.[3,5,7] Confusingly, it has beensuggested that 10–37% of the patients with OFG havebeen found to have Crohn's disease, and oral lesionsmay precede the development of intestinal involvement.[3]

Chest radiography andassessment of serum levels of angiotensin-convertingenzyme for sarcoidosis; complete blood count, erythrocytesedimentation rate and serum levels of folic acid, iron andvitamin B12 for Crohn's disease; and tuberculin skin testand chest radiography for tuberculosis. Gastrointestinalassessment is essential, especially if symptomssuggestive of Crohn's disease are present. The diagnosis ofOFG is therefore a diagnosis of exclusion and is based onappropriate clinical and pathologic correlation.[3,5]

The treatment of OFG is difficult, particularly in the absence of an etiologic factor. Objective of treatment is to improve the patient's clinical appearance and comfort.[5] Spontaneous remission of OFG is rare.[3,5] Variety of drugs have been tried in the treatment of OFG, including corticosteroids.[6] Systemic corticosteroids are consideredthe best treatment. Glucocorticoids effectively suppress the activated T-helper-induced cell process occurring at the site of disease.[3,5-10] The usual therapy is prednisolone 1mg/kg for 4-6 weeks followed by slow tapering over 2-3 months.[6,9] Intralesional corticosteroids (e.g. triamcinolone acetonide 40 mg/

mL) may cause some improvement in some but not all patients.[3] Recently, higher concentrations of the drug (40 mg/mL) have been suggested. The higher concentration offers the advantages of reducing the volume of fluid injected, the administration of a higher dose and the maintenance of remission.[5]

Drugs like salazosulfapyridine, hydroxychloroquine, azathioprine, dapsone, methotrexate and thalidomide have been tried with partial success.[3, 9]

The literature on OFG, MRS and CG shows an important problem in the classification of these entities. This is probably related to a lack of understanding of etiologic and pathogenic mechanisms. The term "OFG" in cases of noncaseating granulomatous inflammation has the advantage of describing a clinicopathologic situation without linking it to a specific disease entity. Terms like "OFG in thecontext of sarcoidosis or in the context of Crohn's disease"and "OFG secondary to a chronic dental infection or tocontact hypersensitivity" are recommended. A diagnosis ofidiopathic OFG is made on the basis of negative results ofa thorough investigation.[5, 9, 10]

Our case showed clinical presentions like lip swelling, cobble stone appearance of buccal mucosa & mucosal tag; histopathologic features of noncaseating granulomas which are characteristic of OFG. By excluding involvement of gastrointestinal system & other granulomatous diseases we came to diagnosis of idiopathic OFG. In our case the patient responded well to the systemic steroid therapy.

CONCLUSION

OFG is a disorder with variable clinical presentations & can be associated with other granulomatous diseases. Dentist is the first person to diagnose & manage OFG & also recognize the systemic diseases associated with it at the earliest.

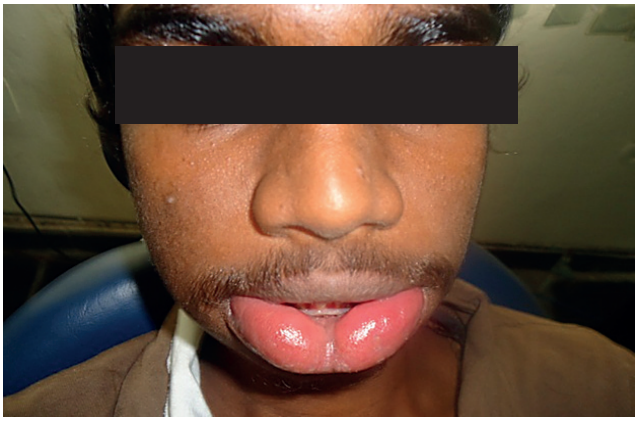

Fig 1: showing lip swelling

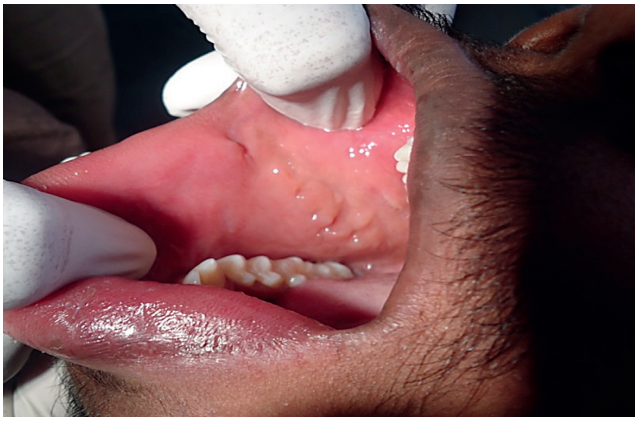

Fig 2: showing cobble stone appearance of buccal mucosa

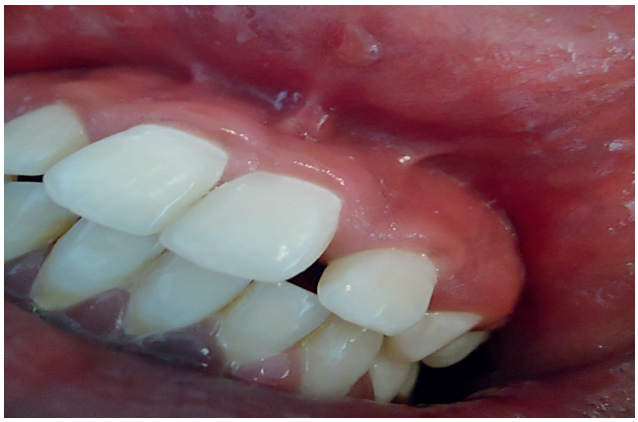

Fig 3: showing mucosal tag

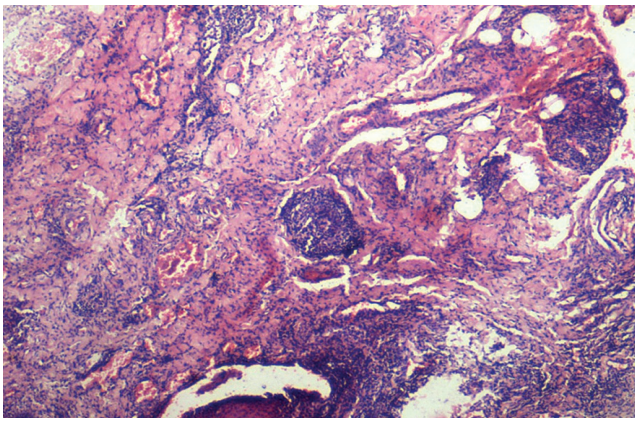

Fig 4: showing noncaseating granuloma

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