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A Case of Recurrent and Persistent Lip Swelling- A Case Report

Malathi[1], G. Vimala[2], M. Ramya[1] & G. Sneha[3]

[1]Associate Professor, Department of General Medicine,
[2]Professor and Principal, [3]Junior Resident,
Tamilnadu Government Dental College and Hospital

Abstract :

Orofacial granulomatosis is a chronic inflammatory disorder characterized by persistent or recurrent soft tissue swelling. Differential diagnosis includes a wide spectrum of diseases, like hereditary angioneurotic edema, Crohn's disease, tuberculosis, sarcoidosis, allergic reactions, C1 esterase inhibitor deficiency.

Keywords: *Orofacial granulomatosis, chronic idiopathic angioedema, gingivectomy, Melkersson - Rosenthal syndrome, Crohn's disease*

Introduction:

Orofacial granulomatosis (OFG) is characterized by the persistent enlargement of the soft tissues of the mouth, lips and face. It may be associated with other conditions, such as Crohn's disease, Tuberculosis and Sarcoidosis. Orofacial granulomatosis that affects the lips is known as Cheilitis granulomatosa. It is persistent, relapsing-remitting, idiopathic, non-tender swelling of lips. Although it can occur at any age, it's more prevalent in the age group between 20 to 40 years old. It is also often associated with Melkersson - Rosenthal syndrome. The exact cause of the disease is unknown. Several theories have been proposed, including infections, genetics and allergies. It may represent a delayed hypersensitivity reaction, but the causative agent is unidentified and varies from person to person. Suspected sources of allergens include metals, preservatives, additives in food, cinnamon, eggs, chocolate etc. There may be a genetic predisposition to the condition.

Case Presentation:

A 24-Year-old male presented to our hospital with the complaint of persistent lip enlargement of 6-year duration and gingival enlargement of 8-months duration. Initially, the lip enlargement was present and reduced by taking anti-histamines and other supportive measures. Later, it gradually enlarged and became persistent. The lip swelling was not associated with pain or itching. No history is suggestive of cough, loss of appetite, difficulty in swallowing, or sneezing. No history of drug allergy or food allergy was evident. No history of insect bite allergy or dental materials used. No history of family members being affected. No history of recurrent facial palsy, tongue and facial swelling.

On clinical examination, there was gingival and lip enlargement. This enlargement involved an attached gingiva and covered almost one–half of the crown length. Periodontal probing showed the presence of a false pocket with no signs of clinical attachment loss and with good oral hygiene. Painless diffuse swelling of lips was present, which was firm and nodular in consistency.

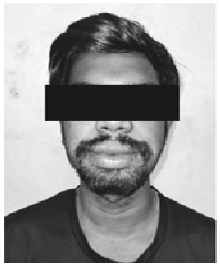

Fig1:*Patient presenting with lip swelling*

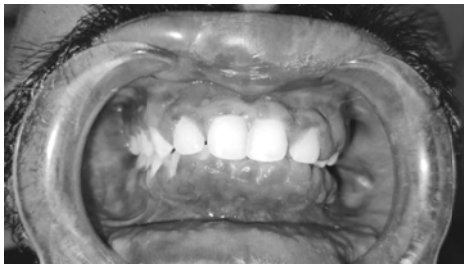

Fig 2: *Pre-Operative photograph showing generalized gingival enlargement*

Based on these clinical findings, a provisional diagnosis of Chronic idiopathic angioedema was given. The differential diagnosis includes angioneurotic edema, Crohn's disease, tuberculosis, sarcoidosis, cheilitis granulomatosa, foreign-body reaction, fungal infections and contact allergy.

Histopathology Findings:

A biopsy of the upper lip revealed circumscribed aggregates of non-caseating granuloma consisting of lymphocytes and epithelioid histiocytes with multi-nucleated giant cells suggestive of the granulomatous lesion. Following this, the patient was thoroughly investigated to rule out the list of granulomatous diseases.

Investigations:

The haematological investigation was performed for RBC count, differential count, platelet count, Hb%, ESR, serum folate, and iron. Serum ACE (Angiotensin converting enzyme) levels were within normal limits; hence sarcoidosis was ruled out. The Mantoux test was negative and the chest radiograph did not reveal any pathology and the acid-fast bacilli stain for mycobacteria was negative, thus ruling out tuberculosis. ELISA Test for HIV 1 and 2 was performed and the results were negative. The c-inhibitor negative was found to be within normal limits, hence Hereditary Angioneurotic edema was ruled out. There was no history of Gastrointestinal symptoms and a colonoscopy revealed no evidence of granulomatous caecal mucosa, Crohn's disease was ruled out. We ruled out Melkersson–Rosenthal Syndrome as there was no history of facial paralysis and the tongue was clinically normal.

Based on clinical history, laboratory investigations and histopathological findings, a confirmatory diagnosis of Idiopathic Orofacial granulomatosis was given. The patient was subsequently treated with biweekly intralesional injections of triamcinolone for 4 weeks. In our case, there was only gingival enlargement without any systemic involvement. Therefore, Gingivectomy surgery was performed under local anaesthesia. After a follow-up of 6 weeks, the patient showed uneventful healing of gingiva and moderate reduction in the lip swelling, however, it was persistent. The patient is currently under observation.

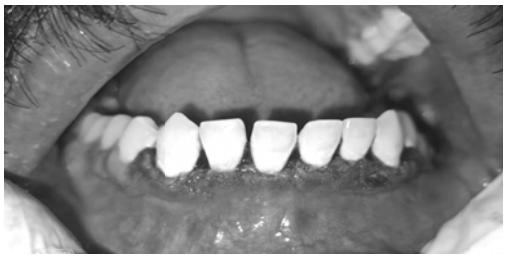

Fig 3: *Post Operative photograph: After Gingivectomy Surgery*

Discussion:

OFG is an increasingly yet uncommonly recognized entity affecting the oral and peri-oral structures. Orofacial granulomatosis can be a distinctive clinical problem or the primary sign of an underlying systemic illness like sarcoidosis or Crohn's disease. Orofacial granulomatosis (OFG) includes a group of diseases characterized by the presence of noncaseating granulomatous inflammation affecting the soft tissues of the oral and maxillofacial region.

The main clinical features of OFG are recurrent non-tender swelling involving the face/one or both lips, angular cheilitis, which may eventually become persistent, along with intraoral features which may be present like mucosal ulcerations, fissures on lips and tongue, mucosal tags, gingival enlargements, cobblestone appearance of the buccal mucosa, cervical lymphadenopathy. When it presents in a triad encompassing facial nerve palsy, lip swelling, and fissured or furrowed tongue it is called Melkersson–Rosenthal syndrome.

Routine blood investigations, serum ACE level, Mantoux test, and colonoscopy are performed to exclude diseases with similar clinical and histopathological features. Orofacial granulomatosis is a disease with a wide spectrum of clinical presentations. The causative agent in

the present case is unknown; hence it is categorized as Idiopathic Orofacial granulomatosis.

The treatment for OFG, given its unknown aetiology, is non-specific and subjective. Spontaneous remission is rare, hence symptomatic treatment is provided. Elimination diets are performed to identify the dietary allergens. Various treatments have been tried for OFG like Antihistamines, Systemic, topical and intralesional Corticosteroids, Antibiotics like dapsone, metronidazole, clofazimine and minocycline, thalidomide, intralesional injections of triamcinolone and Azithromycin,Tumour necrosis factor - alpha blocking agents such as infliximab an and combination therapy have been shown to reduce the facial swellings. Cheiloplasty has been advocated in certain cases where esthetics was the main concern for the patient.

Key Message:

Orofacial Granulomatosis is a distinct clinical entity or it can be an initial presentation of systemic illness like Tuberculosis, Crohn's disease, or sarcoidosis. Therefore, early diagnosis of Orofacial granulomatosis can lead to the identification of underlying systemic diseases. Patients with Idiopathic chronic angioedema those who are not responding to conventional therapy need to undergo further investigations to rule out granulomatous disease.

In the case of granulomatous disease, we have to always rule out other secondary causes like Sarcoidosis, Crohn's disease and Tuberculosis.

Conclusion:

Orofacial granulomatosis has become a topic of interest among professionals and it poses a challenge from the beginning of the presentation of the disease. It is a rare multifactorial disease with varying clinical manifestations. The growing incidence of Crohn's disease, leads to a detailed investigative analysis for suspected OFG, so as to deliver the appropriate treatment. A biopsy is the most definitive way to diagnose the condition along with the clinical symptoms. It is difficult to highlight a reference treatment for OFG, as there is a lack of evidence in the literature. The triamcinolone acetonide injection procedure appears to be the most suitable along with the change of diet to prevent recurrences.

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