INTRODUCTION

Idiopathic Orofacial granulomatosis refers to conditions restricted to the oral region without any identifiable systemic granulomatous diseases. Wiesenfield et al. proposed the term Orofacial Granulomatosis (OFG) in 1985 as descriptive for non-infectious granulomatous disorders of the lips, face and oral cavity that are histologically associated with non-caseating epithelioid granulomas and multinucleated Langhans (foreign body) type giant cells within the oral mucosa. It may be a separate entity or as a component of localized or generalized Crohn’s disease, tuberculosis, actinomycosis and sarcoidosis. The main clinical features of OFG are facial or lip swelling, angular cheilitis, oral ulcerations, vertical fissures of the lips, gingival enlargement, mucosal tags and sometimes lymph node involvement will also be there.

Although the enlargement of the lips is the most common presenting complaint, on rare occasions oral signs may precede all the other local and systemic manifestations. Here we describe a case of idiopathic OFG occurring in a nine year old female patient with gingival enlargement as sole manifestation. Usually OFG presents as swelling of upper or lower lips with an associated swelling in the orofacial region. Unlike the reported manifestations in literature, an exclusive gingival enlargement alone was reported in a young female patient which is a rare entity. Since the early diagnosis, treatment and monitoring of such cases are essential in preventing the development of fatal diseases such as Crohn’s disease or sarcoidosis.

CASE REPORT

A nine year old female patient reported to the Department of Oral Medicine and Periodontics with the chief complaint of bleeding from the gums while brushing and gives history of enlarged gums for the past six months. History revealed that the enlargement of gingiva which was initially localized to the anterior two teeth in the upper and lower jaw gradually over a period of six months spread to other teeth. There was no pain associated with the enlargement and the only symptom was bleeding from the gums which occurred during brushing. On consultation with general dentist who did a thorough oral prophylaxis and put the patient on short course of systemic antihistaminics (diphenhydramine maleate). The clinical condition did not regress and the patient was referred to our hospital for further evaluation and management.

Clear history was recorded for any gastrointestinal disturbance, loss of weight, loss of appetite, cough, fever and any other systemic signs or symptoms. There was no history of any medication and allergic reactions to any food substances or drugs.
Intra oral examination revealed enlarged gingiva of the maxillary and mandibular anterior region, which is diffuse and curtain like enlargement as shown in Figure 1. There was slight bleeding on probing and minimal plaque and calculus were present. The enlargement involved the marginal gingival, interdental papilla and attached gingival extending till the mucogingival junction. Secondary changes like pus, ulcerations were not seen over the lesion. On palpation the enlarged gingiva was uniformly soft in consistency and non tender. The teeth in the region of the gingival enlargement were normal except for the fractured 21 which occurred two years before due to trauma. The fractured tooth was asymptomatic. The intra oral periapical radiographs of upper and lower anterior teeth didn’t show any abnormality. Based on the history and clinical features, clinical differential diagnosis of tuberculous gingivitis, plasma cell gingivitis, histoplasmosis, allergic gingival enlargement, gingival enlargement due to any hormonal changes or an idiopathic granulomatous enlargement.

Hematological investigation was performed for RBC. Count, WBC count, Differential Count, platelet count, ESR, Hb%, serum ACE, PCV, MCV, MCH and MCHC. All the levels were within the normal range and the ACE level was 40 U/L which is also within the normal range.

The mantoux test was also negative and the chest radiograph didn’t reveal any abnormal pathology. Polymerase Chain Reaction (PCR) was done to rule out Tuberculosis infection. PCR for detection of Mycobacterium tuberculosis genome by nested PCR which detects MPB64 and IS 6110 were negative, thus ruling out tuberculosis involvement.

An incisional biopsy of the enlarged gingiva was performed in relation to 13, 14 region and 32, 33 region and the tissue was sent for histological examination. The histological report showed the presence of parakeratinised stratified squamous epithelium of variable thickness and multinucleated giant cells with peripherally arranged nucleus suggestive of Langhans giant cell surrounded by modified macrophages suggestive of epitheloid cells as shown in Figure 2. There was also a rim of inflammatory cells primarily lymphocytes. The connective tissue showed areas of moderate vascularity and areas of hemorrhage as shown in Figure 3. Multiple granulomas containing lot of giant cells suggestive of a granulomatous lesion.

Thus correlating the history, clinical findings, laboratory
investigations and histopathological examination, a final diagnosis of idiopathic orofacial granulomatosis was made.

Since in our case there was only the gingival enlargement without any systemic involvement, we planned for surgical excision. Gingivectomy of both upper and lower arches was carried out under local anesthesia. The case was followed for one year without any recurrence as shown in Figure 4.

DISCUSSION

The patient fulfills the clinical and histological criteria for a diagnosis of OFG. The conditions with granulomatous lesion histologically includes Melkersson Rosenthal syndrome, Crohn’s disease, sarcoidosis, tuberculosis, hypersensitivity reactions, angioneurotic edema, thus complete red blood cell investigation, white blood cell investigation, platelet count, ESR was done.

Chest radiograph was taken and the possibility of tuberculosis, sarcoidosis was ruled out. Sarcoidosis was further ruled out by serum angiotensin converting levels. Tuberculosis has been ruled out by chest radiograph, mantoux test and Polymerase Chain Reaction of gingival tissue as well as serum. Leprosy also been ruled out. Endoscopy and colon vital biopsy are required only if the patient has a history (or) signs and symptoms of gastrointestinal discomfort that could be features of Crohn’s disease.

Recently hypersensitivity reactions to various food, preservatives (or) components of oral hygiene products have been implicated in inducing tissue changes consistent with OFG. Hence, the potential allergic factors should be evaluated in the early diagnostic process, querying any history of contact allergies to oral hygiene products, food additives, flavorings, cosmetics.

Similar clinical presentation of OFG with exclusive gingival hyperplasia was reported in two females aged 28 and 18 by Mignogna et al. This finding corresponds to our case of an exclusive gingival enlargement in a female patient of much younger group and hence it is a rarity.

As regards the treatment, the condition resolves spontaneously, but generally the therapy is necessary. The various treatment modalities include intraleisional steroid injections, topical and systemic steroids, and surgical excision. Since in our case there was only the gingival involvement, we planned for surgical excision.

Since there is an increase in the incidence of OFG and the gingival lesions are frequently associated with the disease the periodontist has to be the first health care professional to suspect and recognize the disease. There are many conditions which may cause gingival enlargement including drug induced gingival enlargement, idiopathic gingival fibromatosis, inflammatory enlargement, conditioned gingival enlargement, leukemic enlargement or neoplastic etc. All the above mentioned conditions should be ruled out by careful clinical as well as histological examination. Thus, the early diagnosis of OFG is a crucial step in the prevention of a possible Crohn’s disease or a coeliac disease and its associated complications developing later in the life inspite of the patient not presenting with the characteristic gastrointestinal findings of an inflammatory bowel disease at the time of presentation.

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