Case Report

Orofacial granulomatosis as a manifestation of sarcoidosis: A rare case report

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Abstract

Orofacial granulomatosis (OFG) is an uncommon clinicopathological condition describing patients who have oral lesions characterized by persistent labial enlargement, oral ulcers and a variety of other orofacial features. Sarcoidosis is a systemic non-caseating granulomatous disorder of unknown etiology that may affect multiple organ systems. As a systemic disease, it is well documented in existing literature. However, the rarity and its often non-specific oral manifestation emphasize the need to reinforce the literature with more clinical case examples and draw additional attention to clinical features that general practitioners may encounter. In this report, a rare case is presented with definite association of sarcoidosis and orofacial granulomatosis with two years follow up after treatment. This report aims to expand the literature surrounding orofacial sarcoidosis and support oral and general health practitioners in recognizing its orofacial manifestations along with the clinical presentations that all led to a diagnosis of sarcoidosis in this case.

Keywords: Gingival enlargement, orofacial granulomatosis, sarcoidosis

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INTRODUCTION

Orofacial granulomatosis (OFG) is described as a granulomatous inflammation of unknown origin that can occur on all parts of oral mucosa.[1] OFG may be the oral manifestation of systemic conditions such as Crohn's disease, sarcoidosis, granulomatosis with polyangitis and Melkersson–Rosenthal syndrome (MRS).[2] Local and systemic conditions characterized by granulomatous inflammation should be excluded by appropriate clinical and laboratory investigations.[3] This article reports a case of sarcoidosis with generalized gingival overgrowth and persistent upper lip swelling treated with gingivectomy and systemic steroids with two years follow-up without recurrence.

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CASE REPORT

A 38-year-old woman reported to our Dental clinic with chief complaint of enlargement of gums and persistent upper lip swelling [Figures 1 and 2]. She gave a significant dental history of similar gingival overgrowth one year back which was treated by a dental surgeon. Eight months after the treatment, patient noticed a recurrence of gum enlargement that had gradually increased to the present state. Patient further described a recurrent swelling of her lips that had eventually become permanent. The swelling had begun a few months earlier and was not associated with any change in oral hygiene products or cosmetics. She had difficulty while eating and brushing teeth, with

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occasional bleeding. Extra oral examination showed diffuse, nontender, soft, upper and lower lip swelling, which had a normal temperature on palpation. Mild fissures were present on the vermilion border of upper and lower lips while the upper lip showed an enlarged and swollen tubercle.

Intraoral examination revealed diffuse generalized enlargement of the gingiva. The gingiva was reddish pink in color, soft, spongy and shiny. Stippling was absent. Bleeding on probing was present. She had missing teeth 18, 35, 45, 48. Grade I mobility was found in 21, 31 and 41. Her orthopantomogram revealed alveolar bone loss in the maxillary and mandibular anterior region [Figure 3]. There were no appreciable changes on the dorsal surface of the tongue. However, her floor of the mouth was edematous and palatal rugae were hyperplastic. The rest of the intraoral examination was unremarkable. The patient's medical history included fibrous dysplasia of tibia bone which was operated upon two years back. Patient was not taking any medicines or drugs. Gingival enlargement was initially treated by doing phase I therapy including scaling and root planing to remove the local deposits. After one month, on reevaluation, it was observed that the enlargement had not reduced in severity, size, clinical appearance and consistency was the same as it was before phase I therapy. Hence, the inflammatory cause of gingival enlargement was ruled out and she was given appointment for surgical excision. After taking informed consent, external bevel gingivectomy under local anesthesia was performed on mandibular and maxillary anterior regions one week apart. Histopathological examination of the excised tissue using H and E staining showed chronic inflammatory cell infiltrate, peri and paravascular aggregation of lymphocytes, plasma cells, non-caseating granuloma formation with epitheloid cells and Langhans type of giant cells [Figure 4a-c]. Based on the histopathological findings, a number of special investigations were undertaken to rule out various granulomatous diseases. The suggested differential diagnosis was the tubercular gingival enlargement, sarcoidosis, Crohn's disease, angioedema and Melkersson–Rosenthal syndrome. She reported no intestinal problems that would suggest Crohn's disease, nor did she complain of chronic fatigue. An in-depth gastrointestinal investigation did not appear justified in this case, since there were no signs of anemia or symptoms suggestive of Crohn's

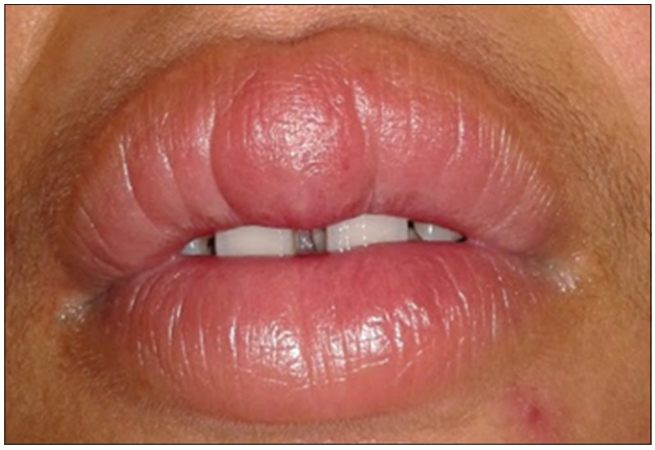

Figure 1: Pretreatment clinical photograph showing lip swelling

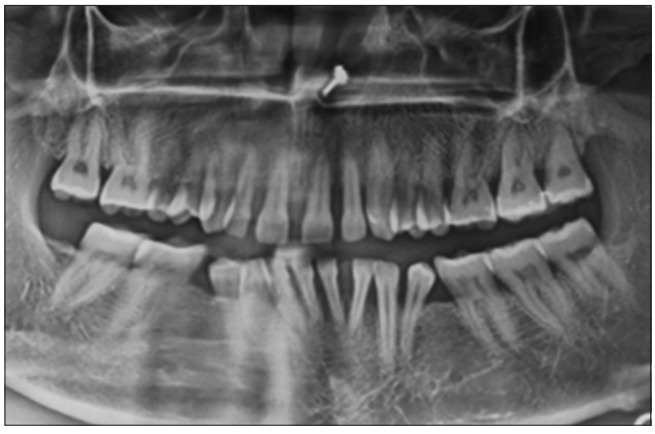

Figure 3: Orthopantomogram showing alveolar bone loss and missing teeth

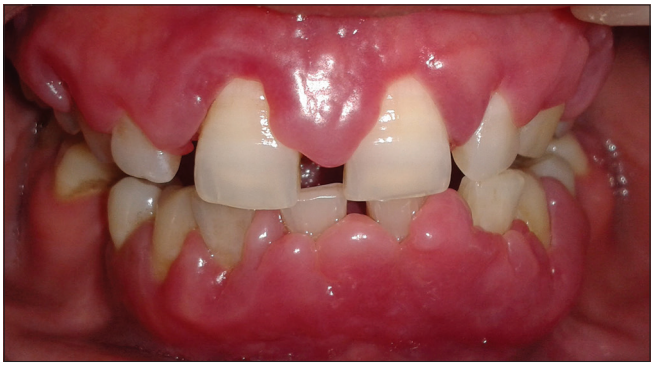

Figure 2: Pretreatment clinical photograph showing generalized gingival enlargement

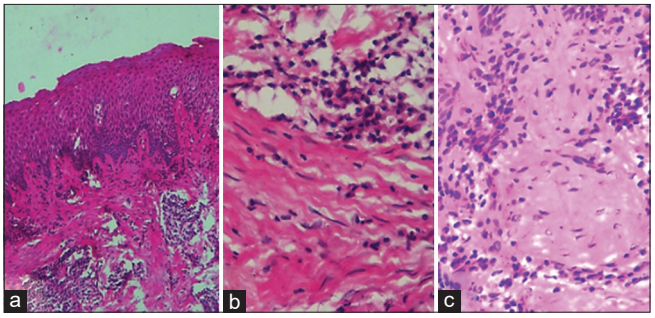

Figure 4: (a-c) Histopathological examination with H&E staining shows chronic inflammatory cell infiltrate, peri and paravascular aggregation of lymphocytes, plasma cells, non-caseating granuloma formation with epitheloid cells and Langhans type of giant cells

disease. There was no history of tuberculosis. Mantoux test and Sputum test were negative for tuberculosis. Melkersson–Rosenthal syndrome was ruled out because of no reported facial paralysis and the tongue appeared clinically normal. Patch testing was done for commonly used food products and cosmetics and the results were negative. This ruled out angioedema. Chest radiography and a series of blood tests, including assessment of serum levels of angiotensin-converting enzyme (sACE), were requested. Complete blood picture was normal, Angiotensin-converting enzyme was raised; 61 U/L (Normal range: 8-53 U/L). The chest x-ray showed hilar lymphadenopathy [Figure 5]. Based on the histopathological examination, elevated sACE levels and chest X-ray findings, diagnosis of sarcoidosis was made. Gingival enlargement was treated by gingivectomy [Figure 6] and for labial swelling systemic corticosteroid therapy was started which was well tolerated by the patient. Patient was administered Prednisolone 5 mg TDS for one week, then 5 mg BD for another week, and 5 mg OD for one more week. Long-term use of systemic steroids was avoided because of their potential side effects. The lip swelling had remarkably reduced after treatment [Figure 7]. The patient was referred to the medical specialist for further investigations and treatment. Since she did not have symptomatic pulmonary involvement, she was not given any medications for sarcoidosis and was kept under regular follow-up by the medical specialist. Patient was given strict oral hygiene instructions and was followed- up by us for two years and showed good healing with no signs of recurrence of gingival enlargement [Figure 8].

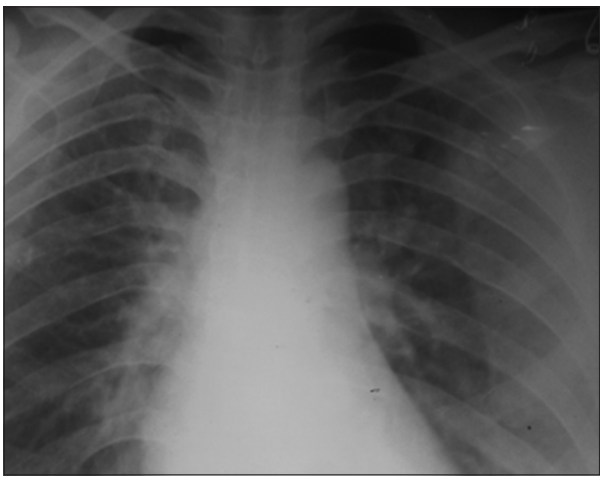

Figure 5: Posterior–anterior view of the chest X-ray shows bilateral hilar lymphadenopathy

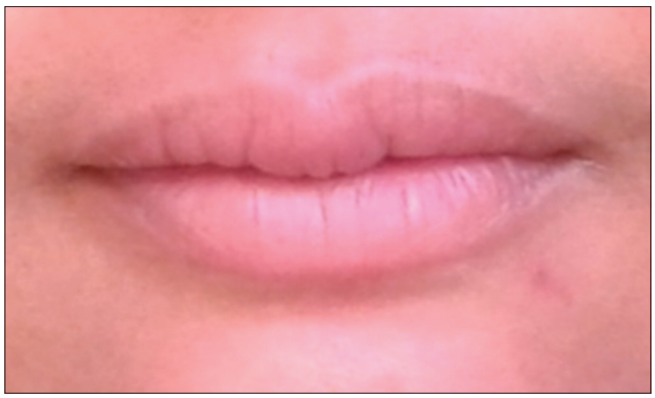

Figure 7: One-month Postoperative photograph showing reduced upper and lower lip swelling

DISCUSSION

The clinical and histological findings of the present case report are consistent with manifestations of OFG. Orofacial granulomatosis (OFG) is an uncommon disease characterized by persistent or recurrent soft tissue enlargement, oral ulceration and a variety of other orofacial features. It could be an oral manifestation of a systemic disease. OFG is a disease that has a wide spectrum of presentation. In the present case, it was an oral manifestation of Sarcoidosis. For a correct differential diagnosis, local and systemic conditions characterized by granulomatous inflammation should

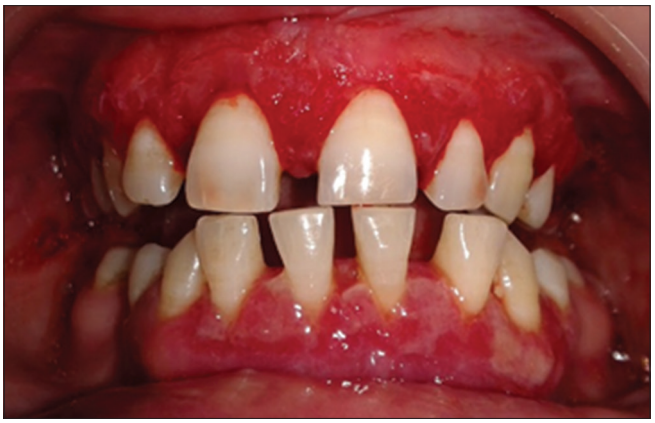

Figure 6: Clinical picture immediately after gingivectomy in maxilla

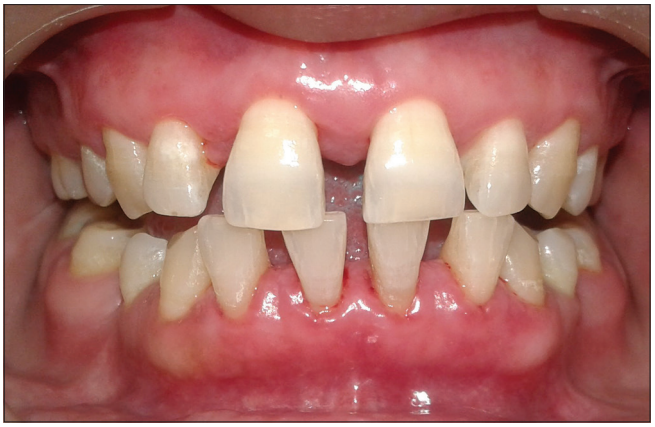

Figure 8: Post-treatment photograph on 2-year follow-up

be excluded using appropriate clinical and laboratory investigations.

Sarcoidosis is a systemic non-caseating granulomatous disease of unknown etiology. Although its etiology is unknown, genetic, infectious and environmental factors have been postulated as possible causes.[4] Sarcoidosis is a multiorgan disorder. One-third of the patients with sarcoidosis can present with non-specific constitutional symptoms such as fever, fatigue, malaise or weight loss.[5] The most common presentation of sarcoidosis consists of pulmonary infiltration and hilar lymphadenopathy, dermal, and ocular lesions.[6] Our patient had a history of fibrous dysplasia of tibia bone which was operated upon two years back with prophylactic tibia nailing [Figure 9]. Though there is no clear evidence in the literature of any association between sarcoidosis and fibrous dysplasia. Oral involvement in sarcoidosis is uncommon. Poe (1943) reported the first confirmed case of sarcoidosis affecting the oral cavity in the mandible.[7] Oral sarcoidosis usually appears in patients with chronic multisystemic sarcoidosis and presents as first manifestation of the disease in one third to almost two thirds of patients.[8]

Clinically, the disease can affect any site of the mouth. According to the literature reviewed by Bouaziz *et al.*[9] (2012) buccal mucosa has been described as the commonest site affected (30%), followed by gingiva (20%), lips (16%), tongue (16%) and palate (9%). Most cases of oral sarcoidosis present with mobility of the teeth due to rapid alveolar bone loss. Other oral manifestations include asymptomatic swelling of the involved mucosa, gingivitis and ulcers.[4] Sarcoidosis of the gingiva typically presents as a diffuse enlargement.[10] Involvement of the jawbones has also been reported,[11,12] with the most characteristic presentation being that of an ill-defined radiolucency involving a tooth-bearing region. In these cases, the patients may present with progressive bone loss and increasing mobility of the adjacent teeth, thus mimicking periodontal disease or Langerhans' cell disease (eosinophilic granuloma).[10,12] The oral manifestations in the present case were persistent lip swelling, recurrent diffuse gingival enlargement, alveolar bone loss with mobility of teeth, edematous floor of the mouth and hyperplastic palatal rugae.

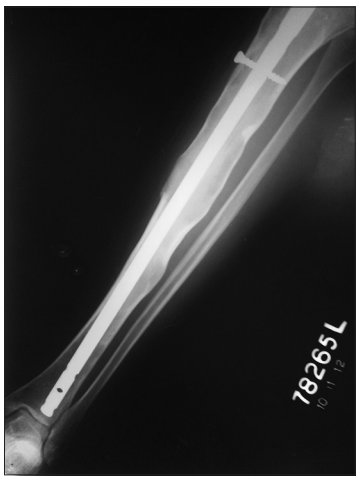

Figure 9: Radiograph showing prophylactic tibia nailing

Sarcoidosis is a diagnosis of exclusion. The diagnosis is based on history (occupational or environmental exposure), pulmonary function tests (forced expiratory volume, vital capacity), haematology (complete blood count, erythrocyte sedimentation rate), biochemical investigations (liver and renal function tests, serum calcium, and serum angiotensin-converting enzyme levels), chest radiograph, and histological studies.[4] Treatment is not required for all patients with sarcoidosis. The American Thoracic Society, European Respiratory Society and World Association of Sarcoidosis and Other Granulomatous Disorders (ATS/ERS/WASOG) have identified several specific conditions, which require treatment.[6] These include the sarcoidosis of the heart and nerves, hypercalcemia, and ocular involvement that do not respond to local therapy. Asymptomatic pulmonary involvement, as in the present case, does not require treatment, while treatment is indicated in symptomatic pulmonary sarcoidosis with worsening pulmonary function tests.[13] Corticosteroids have remained as the mainstay in the treatment of sarcoidosis.[14] The management of asymptomatic sarcoidosis with oral involvement appears to be based initially on close follow-up without any treatment, in consideration of the potential for spontaneous resolution. Corticosteroids or surgery should be indicated only for serious manifestations that obstruct oral functions.[15] In the present case report, a very loose dose (5 mg Prednisolone) of systemic corticosteroid therapy which was gradually tapered was prescribed for three weeks only to treat lip swelling as the patient had esthetic concerns. Steroids were stopped after 3 weeks. Gingival enlargement was managed by surgical excision. The patient started showing good response just after 2 weeks of the treatment and was under regular follow-up for 2 years without any recurrence.

CONCLUSION

OFG occurs as an independent entity but also in conjunction with systemic diseases such as sarcoidosis, tuberculosis and Crohn's disease. Diagnosis of sarcoidosis is by exclusion as no specific test is available. Radiographic, biochemical and histological findings are non-specific but helpful. Regular monitoring is important in such cases

to know the progress of the disease, as in some cases remissions may occur spontaneously or the disease may further progress. Oral lesions, although very rare, maybe the initial manifestation of the disease. Suspected cases of oral sarcoidosis should be biopsied and also be referred to a physician to rule out systemic involvement.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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