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CASE REPORT

RARE CLINICAL ENTITY - IDIOPATHIC OROFACIAL GRANULOMATOSIS

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ABSTRACT

Orofacial granulomatosis (OFG), a noncaseating granulomatous inflammation is a rare disorder affecting the orofacial region which is clinically characterized by diffuse, nontender, soft to firm enlargement involving lip, tongue, gingiva, buccal mucosa, floor of the mouth and other intra-oral sites. The term Idiopathic Orofacial granulomatosis refers to conditions restricted to the oral region without any identifiable systemic granulomatous diseases. Though certain food and food additives, dental materials and various microbiological agents may be possible etiological agents, the definite cause has yet to be identified. The exact antigen inducing the immunological reaction differs in individuals. Evidence for the role of genetic predisposition to the disease is sporadic. Delayed type of hypersensitivity reaction appears to play a significant role. Early diagnosis and management prevent the systemic complications. Multidisciplinary approaches are evolved to achieve periodontal esthetics. This case report entails the overview of enlargement of lip and gingiva in a 28year old female of 2 years duration without any identifiable etiology and was treated by gingivectomy.

KEYWORDS: Orofacial granulomatosis [OFG], Gingival enlargement, Corticosteroid therapy

INTRODUCTION :

Melkersson first described a case of occurrence of granulomas in the orofacial region with facial palsy and orofacial oedema without a recognized underlying systemic condition in 1928 [1]. Wiesenfield proposed the term orofacial granulomatosis in 1985. Orofacial granulomatosis is a rare, idiopathic clinical entity causing swelling of soft tissues of the oral and maxillofacial region, with the histological evidence of noncaseating granulomatous inflammation in the absence of diagnosable systemic Crohn's disease or sarcoidosis. It also includes Melkersson Rosenthal syndrome and cheilitis granulomatosa. It is clinically characterized by diffuse, nontender, soft to firm, painless extra-oral swelling restricted to one or both lips and intraoral sites involving tongue, gingiva and buccal mucosa [2, 3]. Though several theories including infection, genetic, and allergy have been proposed, the exact etiology has still not been established [4 – 8]. Systemic and local causes must be excluded as the clinical and histological features of OFG which has close similarities with the manifestations of few systemic diseases such as tuberculosis, sarcoidosis, Crohn's disease, angioedema and local causes such as foreign body reaction and allergy[9]. Early diagnosis, treatment and monitoring of cases prevent the development of fatal systemic diseases.

CASE REPORT:

A 26-year-old female patient reported to the Department of Periodontics with the chief complaint of intermittent, painless swelling of lips and gums for the past 2 years. Patient's past dental, medical, drug history and history of allergy were non-contributory.

Extra oral examination and palpation showed diffuse, soft, nontender, upper and lower lip swelling of normal temperature (Fig. 1). Mild fissures on the vermilion border of upper and lower lips was present. On intraoral examination, diffuse gingival enlargement extending from 17 to 27 up to the mucogingival junction in the maxilla and involving 43 to 33 in mandible up to attached gingiva was seen. The enlargement extended coronally almost up to the incisal margins in the maxilla and up to the middle third in the mandible. The gingiva was pale pink in color, smooth and shiny with absence of stippling. Interdental spacing was seen where none existed before and plaque and calculus formation was seen. Bleeding on probing was elicited. There were no considerable changes on the dorsal surface of the tongue. Orthopantomogram did not show alveolar bone loss in the maxillary and mandibular region. (Fig:2) Investigations were made, red blood cells count, hemoglobin, hematocrit, total leukocyte count, differential leukocyte count, platelets count, hematocrit/ packed cell volume, mean corpuscular volume, mean corpuscular hemoglobin, mean corpuscular hemoglobin concentration and serum angiotensin convertase enzyme, chest radiograph and mantoux tests were advised. All blood and serum investigations were within normal limits. Chest radiograph was normal, and the mantoux test was also negative. History and clinical examination, aided us to make the provisional diagnosis of orofacial granulomatosis of the upper and lower lip with maxillary and mandibular gingival enlargement.

After the establishment of the provisional diagnosis, intra-lesional injection of triamcinolone (40 mg/mL) was injected weekly into each lateral edge of the upper and lower lip till 3 weeks. Upper and lower lip was anesthetized by infraorbital and mental nerve block to reduce the pain and discomfort arising after the intra-lesional injection. Thereafter, gingival enlargement was initially treated in phase I therapy including scaling and root planing. After phase I therapy, the gingival tissue become firmer due to the reduction of edematous component of the enlargement. Then, gingivectomy was planned for maxillary region. A written informed consent was obtained. After administration of local anesthesia to the affected area, the depths of the pockets are assessed using a periodontal probe and bleeding points are marked. The bleeding points are used as a guide for incision and to determine the depth of the tissue to be excised. Gingivectomy was done for maxillary region and periodontal dressing was placed for 1week (Fig:3) Histopathological examination of the excised tissue showed the presence of stratified squamous epithelial lining with acanthosis and scattered chronic granulomatous inflammation in the underlying fibro-collagenous stroma (Fig: 4) The case was followed for one year without any recurrence (Fig:5)

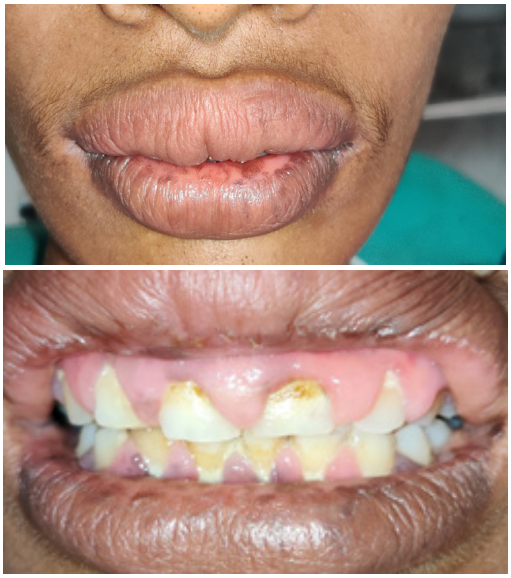

Fig 1: Preoperative photograph showing upper and lower lip swelling and maxillary and mandibular gingival enlargement

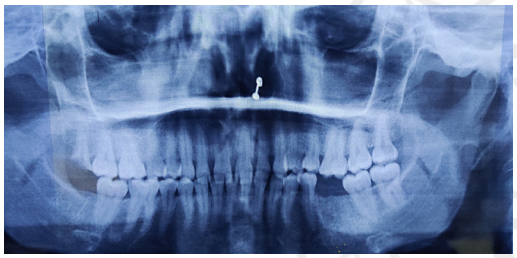

Fig 2: Orthopantomogram X-ray showing no alveolar bone loss

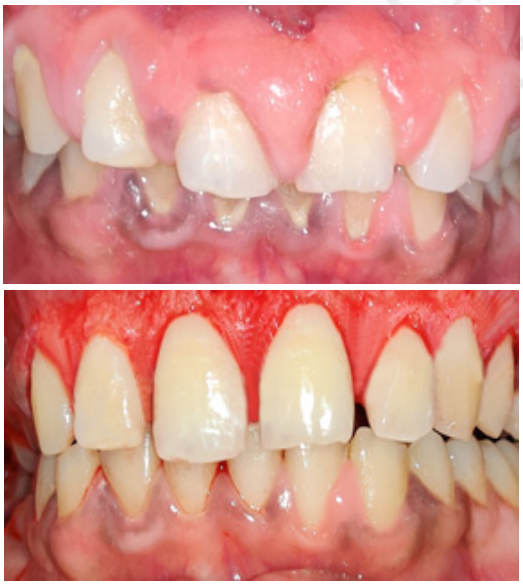

Fig 3: Gingivectomy performed in maxillary region

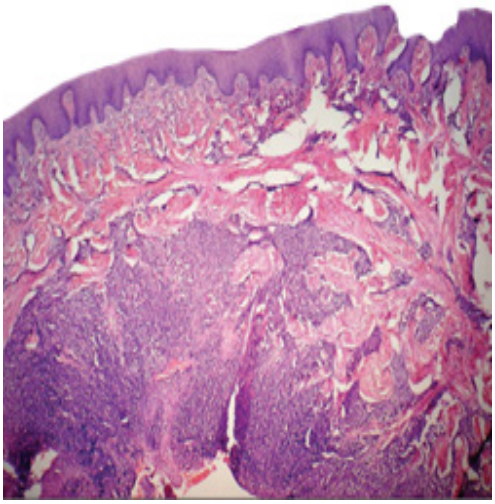

Fig 4: Stratified squamous epithelial lining with acanthosis and scattered chronic granulomatous inflammation in underlying fibro-collagenous stroma

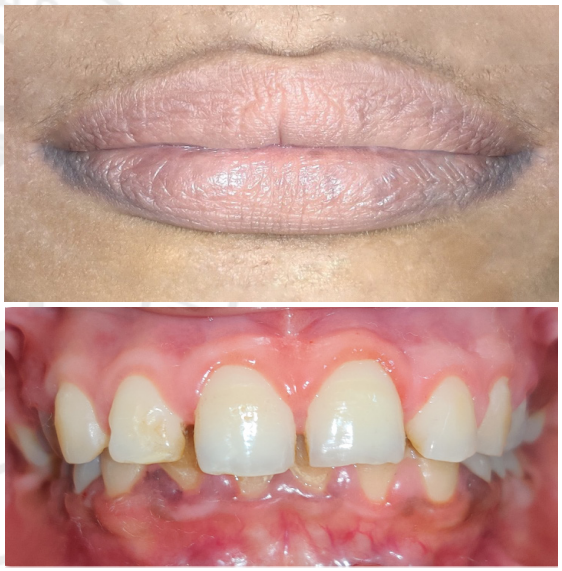

Fig 5: Postoperative photograph showing reduced upper and lower lip swelling and maxillary and mandibular gingival enlargement after 1 year

DISCUSSION:

The diagnosis of orofacial granulomatosis is made by histological presence of noncaseating granulomas with the clinical presence of orofacial swellings. Other conditions for granulomatous formation like sarcoidosis, Tuberculosis, Crohn's disease, angioedema, Melkersson – Rosenthal syndrome are excluded by appropriate clinical and laboratory investigations. The sarcoidosis was elicited on the basis of chest radiograph and serum angiotensin converting enzyme levels. Crohn's disease was ruled out on the absence of

signs and symptoms of gastrointestinal disorders. Tuberculosis was evaluated on the basis of history, chest examination, and Mantoux test. Angioedema was excluded by detection of allergic history to cosmetic, food, drug, atopic eczema or asthma [10]. The etiology of Orofacial granulomatosis has not been clearly evaluated. Studies have exhibited increased prevalence of oral allergic reactions indicated by history of allergic reactions in 80% of patients in contrast to 15–20% of the general population [11]. Exclusion of cinnamon and benzoate preservatives E211, E212, E213 in diet showed a distinct improvement in symptomatology and recurrence rate **12.** The treatment of Orofacial granulomatosis involves enhancing esthetics and preventing the recurrence of enlargement. Many treatment modalities executed for Orofacial granulomatosis are intra-lesional steroid injection, topical or systemic steroids, azathioprine, clofazimine, Ofazimine, methotrexate, metronidazole, minocycline, hydroxychloroquine, low-dose thalidomide and cyclosporine. Surgical therapy may be provided if needed [13, 14, 15]. The most effective drug therapy in reducing the facial swelling and recurrences is corticosteroid therapy. The synthetic glucocorticoid, triamcinolone has a potent anti-inflammatory effect which is eight times more potent than prednisone. Due to its metabolic effect, it has less sodium retention than that of hydrocortisone [16]. Due to the chronic nature of the disease and side effects associated with long-term use of corticosteroids, the use of systemic corticosteroid therapy is limited. Intra-lesional steroid injections have been evidenced as better therapy over the systemic corticosteroid therapy [17]. Monoclonal antibodies and TNF-α inhibitors are effective in some cases refractory to topical and systemic anti-inflammatory treatments [18]. A study with low-energy laser therapy has some efficacy and this method has not shown side effects and could be combined with the other therapies [19]. On the other hand, the chances of recurrences are often associated with the OFG. If orofacial granulomatosis is associated with gingival enlargement, it should be individualized from the other similar conditions of gingival enlargement - inflammatory gingival enlargement, hereditary gingival enlargement, drug-induced gingival enlargement and conditioned gingival enlargement [20]. Since our case presented with lip swelling and gingival enlargement without any systemic involvement, we planned for intralesional steroid therapy with surgical excision.

CONCLUSION:

Orofacial granulomatosis is arduous to diagnose clinically because of varied clinical manifestations and rare occurrence. Histopathology remains the most definitive way to diagnose this condition. Due to lack of evidence in the literature, it is difficult to relate a reference treatment of orofacial granulomatosis. For evidence-based diagnosis with detection of specific cellular markers remains as beneficial which results in targeted therapy for underlying pathology.

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CONFLICTS OF INTEREST:

There are no conflicts of interest.

REFERENCES:

1. Melkersson E. Case of recurrent facial paralysis with angioneurotic oedema. Hugiea 1928; 90: 737
2. Rana AP. Orofacial granulomatosis: A case report with review of literature. J Indian Soc Periodontol 2012; 16:469-74
3. Wiesenfeld D, Ferguson MM, Mitchell DN, MacDonald DG, Scully C, Cochran K, et al. Oro-facial granulomatosis - A clinical and pathological analysis. Q J Med 1985; 54:101-13.
4. Patton DW, Ferguson MM, Forsyth A, James J. Oro-facial granulomatosis: A possible allergic basis. Br J Oral Maxillofac Surg 1985; 23:235-42.
5. Pachor ML, Urbani G, Cortina P, Lunardi C, Nicolis F, Peroli P, et al. Is the Melkersson-Rosenthal syndrome related to the exposure to food additives? A case reports. Oral Surg Oral Med Oral Pathol 1989; 67:393-5.
6. Carr RD. Is the Melkersson-Rosenthal syndrome hereditary? Arch Dermatol 1966;

93:426-7.

7. Meisel-Stosiek M, Hornstein OP, Stosiek N. Family study on Melkersson-Rosenthal syndrome. Some hereditary aspects of the disease and review of literature. Acta Derm Venereol 1990; 70:221- 6.
8. Muellegger RR, Weger W, Zoechling N, Kaddu S, Soyer HP, El Shabrawi-Caelen L, et al. Granulomatous cheilitis and Borrelia burgdorferi: Polymerase chain reaction and serologic studies in a retrospective case series of 12 patients. Arch Dermatol 2000;136:1502-6.
9. Neville BW, Damm DD, Allen CM, Bouquot JE, editors. Allergies and immunologic diseases. In: Oral and Maxillofacial Pathology. 2nd ed. New Delhi: Saunders; Elsevier Publishing; 2005. p. 294-7
10. Tilakaratne WM, cooreysdottir J, Fortune F. Orofacial granulomatosis: Review on aetiology and pathogenesis. JOral Pathol Med 2008;37:191-5.
11. Armstrong DK, Biagioni P, Lamey PJ, Burrows D. Contact hypersensitivity in patients with orofacial granulomatosis. Am J Contact Dermat 1997;8:35–38.
12. Kauzman A, Quesnel-Mercier A, Lalonde B. Orofacial granulomatosis: 2 case reports and literature review. J Can Dent Assoc 2006; 72:325-9.
13. Sussman GL, Yang WH, Steinberg S. Melkersson-Rosenthal syndrome: clinical, pathologic, and therapeutic considerations. Ann Allergy 1992; 69:187-94.
14. Hegarty A, Hodgson T, Porter S. Thalidomide for the treatment of recalcitrant oral Crohn's diease and orofacial granulomatosis. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2003; 95:576-85.
15. Guillaume Feugueur, Maria Polina Konstantinou, Jocelyn Croze, Sara Laurencin, Sarah Custy Management of orofacial granulomatosis: a case report J Oral Med Oral Surg 2018;24:40-43
16. Singhal P, Chandan GD, Das UM, Singhal A. A rare case report of orofacial granulomatosis in a pediatric patient. J Indian Soc Pedod Prev Dent 2012; 30:262-6.
17. Hegarty A, Hodgson T, Porter S. Thalidomide for the treatment of recalcitrant oral Crohn's disease and orofacial granulomatosis. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2003; 95:576– 585
18. Merigo E, Fornaini C, Manfredi M, Meleti M, Alberici F, Corcione L, Buzio C, Rocca JP, Ferri T, Vescovi P. Orofacial granulomatosis treated with low-level laser therapy: a case report. Oral Surg Oral Med Oral Pathol Oral Radiol 2012;113: e25–e29
19. Kaarthikeyan G, Arvind M, Jayakumar N, Khakar M. Idiopathic orofacial granulomatosis in a young patient: A rare entity. J Oral Maxillofac Pathol 2012; 16:432-4.