

# Orofacial granulomatosis affecting lip and gingiva in a 15-year-old patient: A rare case report

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## Abstract

Orofacial granulomatosis (OFG) is a rare disorder affecting the orofacial region, and clinically characterized by diffuse, nontender, soft to firm, painless swelling restricted to one or both lips and intraoral sites such as tongue, gingiva and buccal mucosa. Histologically, OFG is characterized by noncaseating granulomatous inflammation. The early diagnosis of OFG is essential for the better prognosis of the lesion. Delay in diagnosis of OFG results into formation of indurated and permanent swelling of the lip that not only compromises esthetic appearance but also causes impairment in function such as speaking and eating. Early diagnosis of OFG is challenging to the health care professionals due to clinical and histological resemblance to other chronic granulomatous disorders. Thus, dentists may act as a first person to diagnose the lesion and play an important role in the multidisciplinary treatment of granulomatous disorders. Here, we present a case of OFG affecting lips and gingiva in a 15-year-old patient without any identifiable systemic or local causes.

**Keywords:** Corticosteroid therapy, gingival enlargement, noncaseating granulomatous inflammation, orofacial granulomatosis

## Introduction

Orofacial granulomatosis (OFG) is a rare, idiopathic, granulomatous disease involving the face and oral cavity, which is clinically characterized by diffuse, nontender, soft to firm, painless swelling restricted to one or both lips and intraoral sites such as tongue, gingiva and buccal mucosa and histologically characterized by noncaseating granulomatous inflammation.<sup>[1]</sup> OFG term was proposed by Wiesenfeld in 1985. Melkersson–Rosenthal syndrome and cheilitis granulomatosa of Miescher are subtypes of OFG.<sup>[2]</sup> Clinical and histological features of OFG may resemble with the manifestations of few systemic diseases viz., tuberculosis, sarcoidosis, Crohn's disease and angioedema as well as local causes, that is, foreign body reaction and allergy.<sup>[3]</sup> So the aforesaid systemic and local causes

should be ruled out before making a diagnosis of OFG. Here, we present a case of OFG affecting lips and gingiva in a 15-year-old patient without any identifiable systemic or local causes.

## Case Report

A 15-year-old girl reported to the dental outpatient department, Sir Sunderlal Hospital, BHU, Varanasi, with the chief complaints of swelling of painless and gradually increasing swelling of upper and lower lips along with swelling of gums from last 2 years. A detailed history was taken, and there was no contributory medical history, drug history, history of allergy to any food substance, dug or cosmetics, asthma or atopic eczema and family history.

Extra oral examination showed diffuse, nontender, soft, upper and lower lip swelling, which has a normal temperature on palpation. Mild fissures were present on the vermilion border of upper and lower lips while the upper lip showed the mild cobblestone appearance. On intraoral examination, the gingival enlargement was extended from 13 to 23 involving the attached, interdental and marginal gingiva and reached mucogingival junction in the maxilla and mandible. The enlargement covered the crown coronally up to the middle third and cervical third approximately in maxilla and mandible respectively. The gingiva was red in color, smooth and shiny. Stippling was absent [Figure 1]. Periodontal pocket probing revealed the bleeding on probing from the base of the pocket with mild local deposits and the presence of false pocket without clinical attachment loss. Orthopantomogram did not show alveolar bone loss in the maxillary and mandibular anterior region [Figure 2].

The suggested differential diagnosis was the tubercular gingival enlargement, sarcoidosis, Crohn's disease, angioedema, Melkersson–Rosenthal syndrome and OFG. Therefore, further

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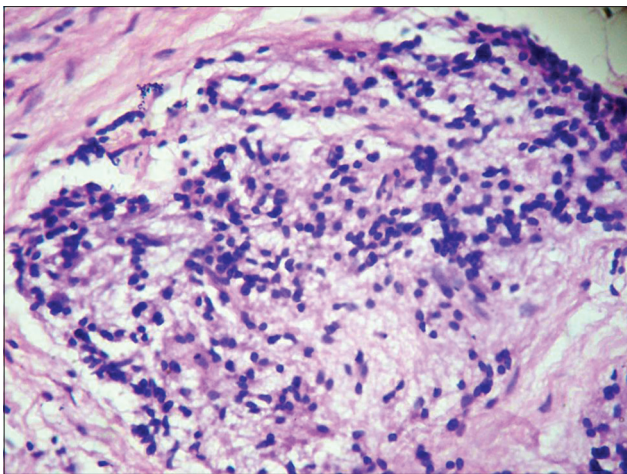
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**Figure 1:** Preoperative photograph showing upper and lower lip swelling and maxillary and mandibular anterior gingival enlargement



**Figure 3:** Postoperative photograph showing reduced upper and lower lip swelling and maxillary and mandibular gingival enlargement after 3 months



**Figure 5:** Chronic granulomatous inflammation composed of epithelioid histiocytes admixed with lymphocytes (H and E,  $\times 400$ )

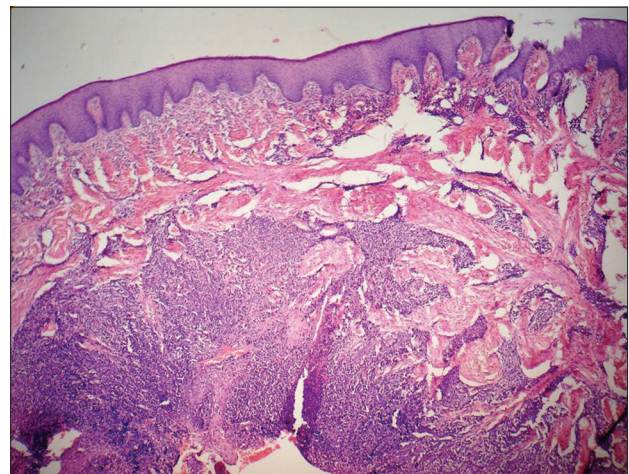
investigations, that is, red blood cells count, total leukocyte count, differential leukocyte count, platelets count, hemoglobin, pack cell volume, mean corpuscular volume, mean corpuscular hemoglobin, mean corpuscular hemoglobin concentration and serum angiotensin convertase enzyme, chest radiograph 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, clinical examination and investigations including the blood and serum investigations, chest X-ray, and mantoux test helped us to make the diagnosis of OFG of the upper and lower lip along with maxillary and mandibular gingival enlargement.

After the establishment of the diagnosis, intra-lesional injection in the upper and lower lip was planned. Upper and



**Figure 2:** Orthopantomogram X-ray showing no alveolar bone loss in the anterior region



**Figure 4:** Stratified squamous epithelial lining with variable acanthosis and scattered chronic granulomatous inflammation in underlying fibrocollagenous stroma (H and E,  $\times 40$ )

lower lip was anesthetized by infraorbital and mental nerve block to reduce the pain and discomfort arised after the intra-lesional injection. Intra-lesional injection of 3–10 ml of triamcinolone (10 mg/ml) was injected weekly into each lateral edge of the lower and upper lip up to 3 months. Gingival enlargement was initially treated by doing phase I therapy including scaling and root planing to remove the local deposits. After 1-month of phase I therapy, the gingival tissue was become more firm due to the reduction of edematous component of the enlargement. Thereafter, gingivectomy and gingivoplasty were done in the anterior region of maxilla and mandible under local anesthesia, and the periodontal dressing was placed for 1-week followed by another 1-week. After 3 months, the patient showed good response and is in regular follow-up from 6 months without any recurrence yet [Figure 3]. The patient was happy with the results of the treatment. Excised gingival tissue was sent for histopathologic examination. H and E staining at  $\times 40$  showed the presence of stratified squamous epithelial lining with variable acanthosis and scattered chronic granulomatous inflammation in the underlying

fibrocollagenous stroma [Figure 4]. H and E staining at  $\times 400$  showed chronic granulomatous inflammation composed of epithelioid histiocytes admixed with lymphocytes [Figure 5].

## Discussion

The clinical and histological findings of the present case report are consistent with manifestations of OFG, a rare disorder of the face and oral cavity that is characterized clinically by painless swelling of one or both lips and intraoral sites such as tongue, gingiva and buccal mucosa and histologically, by scattered aggregates of noncaseating granulomatous inflammation consisting of lymphocytes and epithelioid histiocytes are present with or without multinucleated giant cells.<sup>[3]</sup>

Other chronic granulomatous conditions like tuberculosis, sarcoidosis, Crohn's disease, angioedema and Melkersson–Rosenthal syndrome were ruled out before making a final diagnosis. Tuberculosis was ruled out on the basis of history, chest examination, and mantoux test. The sarcoidosis was excluded on the basis of chest radiograph and serum angiotensin converting enzyme levels. Crohn's disease was differentiated on the basis of blood investigations and absence of signs and symptoms of gastrointestinal disorders. Absence of allergic history to cosmetic, food, drug, atopic eczema or asthma was the criteria for the exclusion of angioedema. Melkersson–Rosenthal syndrome was ruled out on the basis of absence of signs of facial paralysis and fissured tongue.<sup>[3]</sup>

The etiology of OFG has not been clearly identified. Therefore, the treatment of OFG mainly aims at improving the esthetic appearance of the patient by reducing the swelling and preventing the recurrences because spontaneous remission is possible. Various treatment modalities for OFG are: Intra-lesional steroid injection, topical or systemic steroids, azathioprine, clofazimine, methotrexate, metronidazole, minocycline, hydroxychloroquine, thalidomide and cyclosporine. Surgical therapy may be provided if needed.<sup>[4]</sup> Choice of the treatment depends upon the site and extension of the lesion. Corticosteroid therapy is the most effective drug therapy in reducing the facial swelling and recurrences. Systemic corticosteroids therapy is of limited use due to the chronic nature of the disease and side effects associated with long-term use of corticosteroids. Intra-lesional injections have been proven a better therapy over the systemic corticosteroids therapy.<sup>[5]</sup> Higher volume intra-lesional 3–10 ml of triamcinolone (10 mg/ml) injection has been used in patients with OFG. Before injection, nerve block anesthesia enables adequate volume and repeated injections to be given painlessly.<sup>[6]</sup> Recently, small volume, higher concentration delayed release triamcinolone has been shown to be effective and offers the advantage of the reduction in the amount of volume injected and discomfort to the patient after injection. Nerve block anesthesia does not require in small volume

higher concentration injection therapy.<sup>[7]</sup> In the present case report, intra-lesional corticosteroid therapy for lip swelling and surgical excision of the gingival enlargement were provided to the patient. The patient showed good response after 3 months of treatment and is under regular follow up since 6 months without any recurrence yet. However, the chances of recurrence are often associated with the OFG.

If the gingival enlargement is the only affected site with OFG, it should be differentiated from the other similar conditions of gingival enlargement like chronic inflammatory gingival enlargement, hereditary gingival enlargement, drug-induced gingival enlargement and conditioned gingival enlargement.<sup>[8]</sup>

## Conclusion

The early diagnosis of OFG is essential for a better prognosis of the lesion. Delay in the diagnosis of OFG may result into formation of indurated and permanent swelling of the lip which not only compromises esthetic appearance but also causes impairment in function such as speaking and eating. Early diagnosis of OFG is challenging to the health care professionals due to clinical and histological resemblance with other chronic granulomatous disorders. Thus, dentists may act as a first person to diagnose the lesion and play an important role in the multidisciplinary treatment granulomatous disorders.

## References

1. Rana AP. Orofacial granulomatosis: A case report with review of literature. *J Indian Soc Periodontol* 2012;16:469-74.
2. 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.
3. Neville BW, Damm DD, Allen CM, Bouquot JE, editors. Allergies and immunologic diseases. In: *Oral and Maxillofacial Pathology*. 2<sup>nd</sup> ed. New Delhi: Saunders; Elsevier Publishing; 2005. p. 294-7.
4. Kauzman A, Quesnel-Mercier A, Lalonde B. Orofacial granulomatosis: 2 case reports and literature review. *J Can Dent Assoc* 2006;72:325-9.
5. 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.
6. Sakuntabhai A, MacLeod RI, Lawrence CM. Intralesional steroid injection after nerve block anesthesia in the treatment of orofacial granulomatosis. *Arch Dermatol* 1993;129:477-80.
7. Mignogna MD, Fedele S, Lo Russo L, Adamo D, Satriano RA. Effectiveness of small-volume, intralesional, delayed-release triamcinolone injections in orofacial granulomatosis: A pilot study. *J Am Acad Dermatol* 2004;51:265-8.
8. 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.

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