

# OROFACIAL GRANULOMATOSIS TREATED WITH SYSTEMIC STEROID: A CASE REPORT

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## ABSTRACT

Orofacial granulomatosis refers to conditions restricted to oral region mainly characterized by chronic swelling of orofacial tissues with histological evidence of non-caseating granuloma. It may be idiopathic where there is absence of identifiable granulomatous disease or may present as a component of localized or generalized Crohn's disease, tuberculosis and sarcoidosis. The clinical features are facial or lip swelling, angular cheilitis, oral ulcerations, vertical fissures of lips, gingival enlargement, mucosal tags and sometimes lymph node enlargement. A middle-aged female presented to our outpatient department with diffuse chronic swelling of lips which after treatment with systemic steroid showed visible improvement during follow up.

## KEYWORDS

Orofacial granulomatosis, orofacial swelling, steroid

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## INTRODUCTION

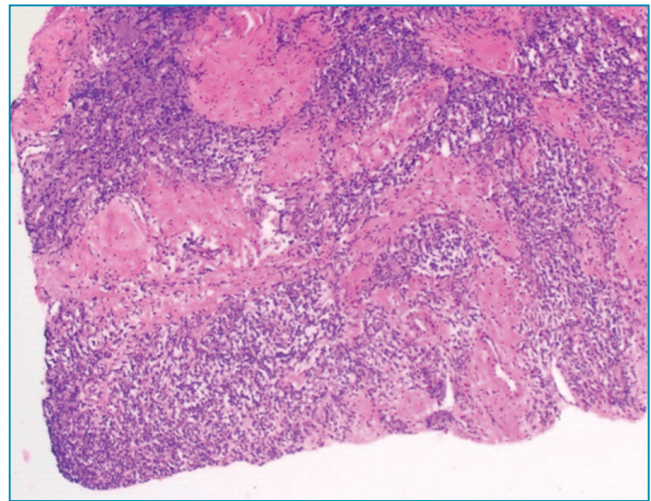
Orofacial granulomatosis (OFG) refers to chronic swelling of orofacial tissues with histological evidence of noncaseating granuloma.<sup>1</sup> Idiopathic orofacial granulomatosis refers to conditions restricted to oral region without any identifiable systemic granulomatous diseases. The term orofacial granulomatosis was proposed by Weisenfeld *et al* in 1985 as a description for non-infectious granulomatous disorders of the lips, face and oral cavity that are histologically associated with noncaseating epithelioid granulomas and multinucleated Langhans (foreign body) type giant cells within the oral mucosa.<sup>1</sup> It may be idiopathic or may present as a component of localized or generalized Crohn's disease, tuberculosis, actinomycosis and sarcoidosis.<sup>2</sup> The clinical features are facial or lip swelling, angular cheilitis, oral ulcerations, vertical fissures of lips, gingival enlargement, mucosal tags and sometimes lymph node enlargement. OFG may be the indication of certain conditions like sarcoidosis, Inflammatory bowel disease, granulomatosis with polyangiitis (GPA), Melkersson-Rosenthal syndrome (MRS).<sup>3,4</sup> Idiopathic orofacial granulomatosis has been linked to contact hypersensitivity with dietary factors and elements like cobalt.<sup>5-7</sup>

## CASE REPORT



**Fig. 1:** An erythematous, scaly plaque of size 5cm X 2cm was present over chin.

A 40-year-old female, presented in Banaras Hindu University, Varanasi Dermatology outpatient department with diffuse, soft, painless enlargement of lips for six months. Mild fissures were present on the upper and lower lips. The patient had gingival enlargement in



**Fig. 2:** Microphotograph of skin biopsy showing dense, diffuse, infiltrate of lymphocytes, histiocytes and plasma cells in a patchy pattern throughout the submucosa. There was no well-defined granuloma. Overlying epidermis showed mild focal spongiosis and slight hyperplasia. The histopathology was suggestive of orofacial granulomatosis (OFG). [H&E stain, 400x]



**Fig. 3:** After one month, the patient showed visible improvement.

the upper and lower arch. This enlargement involved attached, interdental gingiva and covered almost one-third to one-half of the crown length reaching mucogingival junction in the maxilla and mandible. An erythematous, scaly plaque of size 5cm X 2cm was present over chin (Fig. 1). We kept a differential diagnosis of cheilitis granulomatosa, sarcoidosis, cutaneous tuberculosis and oro-facial granulomatosis. Routine blood investigations, chest X-Ray, Mantoux test, stool occult blood, colonoscopy, serum angiotensin converting enzyme (ACE) levels revealed no abnormality. An incisional biopsy of the mandibular gingiva was performed which on histopathology showed dense, diffuse, infiltrate of lymphocytes, histiocytes and plasma cells in a patchy pattern throughout the submucosa. Non caseating granuloma was present but it was poorly defined. Overlying epidermis showed mild focal spongiosis and slight hyperplasia. The histopathology was suggestive of OFG (Fig. 2).

Our patient refused for intralesional triamcinolone injection, so we started her on Prednisolone 40 mg/day with tapering by 10 mg every ten days. After one month, the patient showed visible improvement (Fig. 3). After one month, the patient lost to follow up.

## DISCUSSION

Orofacial granulomatosis is a chronic swelling of orofacial tissues which can be a major cosmetic concern for the patient but also its association with systemic diseases which needs to be looked out and screened for during initial visit as well as follow up. The treatment of OFG is difficult, particularly in the absence of an etiologic factor during initial visit. Although rare, spontaneous remission is possible. Management includes excluding dietary allergens and materials containing cobalt in few case reports.<sup>5-7</sup> The various treatment modalities which have been tried include intralesional steroid injections, systemic steroids and surgical excision. Clofazimine has been reported to be effective in the management of OFG.<sup>8</sup> Low-dose thalidomide has been shown to be successful in treating OFG.<sup>9</sup> In recent years, infliximab, a chimeric monoclonal antibody against TNF- $\alpha$  has shown efficacy.<sup>10</sup>

Most of the cases of OFG has been linked to systemic involvement, which sounds the need for proper evaluation to rule out inflammatory bowel disease, tuberculosis or any other systemic illness, early diagnosis of which can lead to better prognosis.<sup>2</sup>

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