ISSN 2689-8268 | Volume 8

American Journal of Surgery and Clinical Case Reports

Case Report and Review Article Open Access

Oro Facial Granulomatosis: A Rare Case Report and Review

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Received: 28 Oct 2024
Accepted: 28 Nov 2024
Published: 03 Dec 2024
J Short Name: AJSCCR

Citation:

Shantala R Naik. Oro Facial Granulomatosis: A Rare Case Report and Review.
Ame J Surg Clin Case Rep. 2024; 8(4): 1-4

1. Introduction

Orofacial granulomatosis (OFG) is characterized by non-necrotizing granulomatous inflammation of the oral and maxillofacial area and presents clinically as labial enlargement, perioral and/or mucosal swelling, oral ulcerations, and gingivitis. Several etiological factors such as food substances, food additives, dental materials, and various microbiological agents have been associated with the disease process, but precise pathogenesis is yet to be elucidated. Delayed type of hypersensitivity reaction plays a significant role, although the exact antigen inducing the immunological reaction varies in individual patients.[1] We present a case of OFG in a female patient who presented to us with a history of long standing swelling of the face. No etiological factor was traced, however patient had come to us after long duration of numerous treatments at several places but was of no help. This case was managed very well and patient was extremely happy after treatment that got her confidence back. This case report highlights the use of intra lesional steroids in the management of OFG.

2. Case Presentation

A 61-year-old female patient came to the department of Oral Medicine and Radiology with a complaint of asymptomatic persistent swelling in the lips, right and left cheek since 2-3 years (Figure 1a, Figure 1b). There was no history of trauma, hypersensitivity reactions. No history of any topical application of drugs. Patient did not give any known history of allergy to any kind of agent. Patient was hypertensive since 15 years and was on medication (tab metaprolol) for the same. Patient had visited a local doctor for the same complaint and was medicated (unaware of the type of medicine) but was not fruitful. Extra orally there was diffusely swollen upper and lower lip, right and left cheek, prominent nasolabial fold, obliteration of the philtrum. Lower lip appeared everted due to heaviness and hence fall out phenomena noted. Pigmentation were present on the vermillion border of lower lip. Swelling was soft and non pitting and non tender. Intraorally bucccal mucosa and floor of mouth appeared bulbous, there was generalized attrition seen. A provisional diagnosis of orofacial granulomatosis (secondary to delayed hypersensitivity) and differential diagnosis like angioneurotic edema, lymphangioma, chron's disease, sarcoidosis, wegeners granulomatosis, tuberculosis, leprosy, systemic fungal infections, foreign body reaction were considered. There was no history of cough, weight loss, fever, gastro intestinal manifestations. Patient did not reveal any swelling in any other part of her body. Patient was advised ultrasonography of facial swelling, thyroid function test, liver function tests, renal function test and complete hemogram, anti nuclear antibody, blood sugar levels. Complete hemogram revealed 12.2gm% of haemoglobin, 81% of polymorphs (raised), and 7 eosinophils. Patient had raised SGPT (47 IU/L) and SGOT (46 IU/L) levels. Rest of the parameters were within normal range. USG revealed edematous tissue in cheek and lip region. Patient was advised endoscopy, chest radiograph and lip biopsy. Endoscopy, Chest radiograph was normal. Lip biopsy was suggestive of hyperplastic and hyperkeratotic stratified squamous epithelium with subepithelial mild chronic inflammation, suggestive of chronic inflammatory lesion. There was no evidence

of amyloidosis or malignancy. Hence this case was diagnosed as Idiopathic orofacial granulomatosis. Patient was advised systemic (oral) prednisolone 30mg in three divided doses for a period of 7 days. She was also prescribed pantoprazole (40 mg) orally. Patient was reviewed after a week and swelling had reduced by 25%. Although the swelling reduced in size but we decided to give her intralesional steroids with 4mg of dexamethasone since patient was hypertensive. This drug was infused slowly on right and left buccal mucosa, upper and lower labial mucosa. The systemic steroid (oral) was tapered thereafter. Patient was recalled every week once for buccal mucosal intralesional injections. Patient reported to us after a week and swelling had reduced by 75%. When patient reported to us after another week, by this time patient had received 4 doses of intralesional steroid and patient compliance achieved was 9/10. This day was the last dose of intralesional steroid. Following this patient was put on immunomodulator (Tab Levamisole, 150mg once daily 3 times a week for 3 weeks for 3 months) (Figure2). Patient was followed up regularly every month. When patient first visited us, she said that she had stopped attending social gatherings because of swelling and after treatment, since her swelling subsided, she was confident and happy attending her family events. The results are evident as displayed Figure 3. This case highlights the treatment modality of OFG. We managed this case successfully with intralesional steroids and maintenance therapy with immunomodulators that gave quicker and faster and long lasting results.

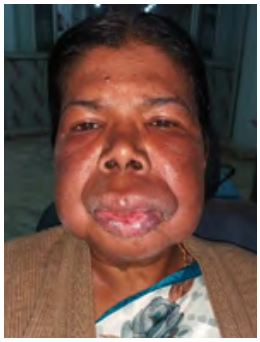

Figure 1 and b: baseline photograph shows swelling of lips and cheeks, exaggerated nasolabial fold, pouted lips.

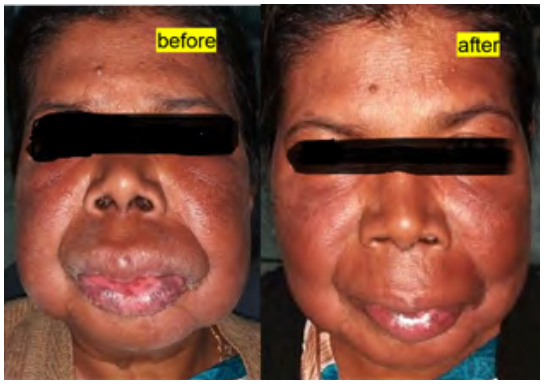

Figure 2: Recall check up photograph shows comparison between baseline and recall check up and shows improved aesthetics of patient and a smiling face.

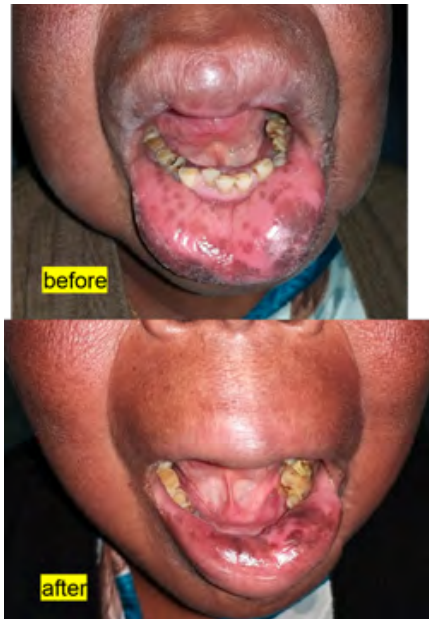

Figure 3: picture showing the comparison between baseline and post treatment effects of lip swelling.

3. Discussion

Oro facial granulomatosis is a multifactorial disease and its treatment might be challenging [1], was the first person who introduced the term Orofacial granulomatosis (OFG). He described it as a uncommon chronic granulomatous condition involving lips, face and oral cavity and histologically associated with non caseating granulomas and multinucleated Langans type giant cells [1]. OFG is considered as nonspecific granulomatous inflammation presenting as facial lip swelling, cheilitis, ulceration, gingival enlargement, mucosal tags and lymphadenopathy.[2] Persistent or recurrent lip swelling is the most common presentation and hence the term cheilitis granulomatosa. It is also considered as a monosymptomatic form of Melkerson Rosenthal syndrome [2]. OFG is also considered as a delayed hypersensitivity (type IV hypersensitivity reaction. Local immune and host response promotes accumulation of macrophages and monocytes. It can occur due to local and systemic factors. Primary factors for OFG are foreign material like amalgam, cosmetic fillers and suture material. Delayed hypersensitivity secondary to flavouring agents, ingredients in oral hygiene products and dental restorations are associated with granuloma. In case of OFG with no definite etiology and systemic causes, it is called as idiopathic OFG [3]. Idiopathic orofacial granulomatosis was described by Tilakaratne et al. [4], in the year 2008, as the lesions restricted to the oral region without identifiable granulomatous disease. He also suggested that diagnosis be not changed until patient develops systemic manifestation of a specific granulomatous condition [4]. Hence idiopathic OFG needs to be followed up to evaluate for secondary chron's disease at a later stage [2]. The unifying term "OFG" has been introduced to integrate the spectrum of various disorders, including Melkersson-Rosenthal syndrome and granulomatous cheilitis (which is sometimes con-

sidered to be a monosymptomatic form of Melkersson-Rosenthal syndrome) and has been shown to be associated with diseases like chron's disease, sarcoidosis, wegeners granulomatosis, tuberculosis, leprosy, systemic fungal infections and foreign body reactions [5]. Several etiologic factors such as food substances, food additives, dental materials, and various microbiological agents have said to cause the disease process, but its precise pathogenesis is not known. However delayed type of hypersensitivity reaction appears to play a significant role, although the exact antigen inducing the immunological reaction varies in individual patients. There is no evidence of genetic predisposition of the disease [5]. Study has reported increased streptococcus in patients with OFG and chron's disease. Nisseriae Subflava is found to be important pathogen in causing OFG [7]. Studies have reported significant increase in interferon – c, interleukin-2 in tissues of OFG. Report suggests profound dysregulation of peripheral T-cell that suggests OFG as a systemic disorder with localized manifestation [8]. OFG predominantly occurs in 2nd decade of life and has female predilection (56%). It is said to be more commonly associated with chron's disease (10-37%). Oral lesions precede intestinal manifestations and hence OFG needs to be followed up for secondary chron's disease at a later stage of disease [2]. The classic presentation of OFG is a nontender, recurrent labial swelling that eventually becomes persistent. This swelling may affect one or both lips, causing lip hypertrophy. The swelling is initially soft, but becomes firmer with time as fibrosis occurs. However, the clinical presentation can be highly variable, making the diagnosis difficult. In case of idiopathic granulomatosis, no identifiable granuloma is seen histopathologically. Tissues present with inflammatory infiltrate. Idiopathic OFG can be non necrotising and nonspecific [9]. First line of treatment includes elimination of allergic factor, like allergic diet, dental material. Identifying the atopic patients by patch testing and avoiding the allergen. Topical and interlesional steroid therapy could be used. Intralesional corticosteroids with triamcinolone acetonide 40 mg/ml may reduce the swelling. Antileprotic drugs like clofazimine (50-100mg) has anti inflammatory properties that can be used in the dose of 100mg four times weekly for three to eleven months can be used. This has been useful in patients with severe granulomatosis.[10] Mild cases might resolve on own but lip swelling can be difficult to resolve. Cheiloscopy can be done in persistent cases where aesthetics is a concern to patients [9]. Treatment also includes use of nonsteroidal anti-inflammatory drugs, broad-spectrumantibiotics, antituberculous drugs, sulfa drugs (sulfasalazine), antimalarials (hydroxycholoroquine), TNF-alpha, infliximab. Surgically, cheiloplasty has shown some results, but is suggested only for resistant cases, particularly when long-lasting chronic inflammation has caused fibrosis and when it is too late to use medications. Hence, early diagnosis and treatment is mandatory for better outcome [2]. Thalidomide has recently been shown to be effective in recalcitrant cases [11]. Observed a significant improvement in lip swelling after 1 month of treatment with intralesional combination of triamcinolone, metronidazole, and minocycline [12]. Stein and Mancini successfully treated 2 children with a combination of oral prednisolone and minocycline [13]. Cheiloplasty is reserved for the most complicated cases with a major lip deformity or cases that are resistant to therapy [11]. Cases have been successfully managed with intra lesional steroids [14,15]. These data suggests use of intralesional steroid for better management of OFG. Our case was different from rest of the cases since it had involved not only lip, rather cheeks and both the lips. Our case was successfully managed with intralesional steroids. Since our patient was hypertensive, we put her on short term systemic steroids but we decided to give her intralesional steroids for speedy results. It reduced the swelling significantly in short period. So, intralesional steroid can be a best alternative treatment for effective management of OFG.

4. Conclusion

Orofacial granulomatosis is a rare occurrence in routine general practice. It poses a diagnostic challenge and treatment too. Most of the cases go unreported and undiagnosed and untreated. This case report highlights the treatment option for idiopathic OFG and diagnostic challenge since the histopathology may not always present in an ideal way. Idiopathic OFG is diagnosed after excluding other known causes for orofacial swelling, thus making it a diagnosis out of exclusion. It highlights the use of intralesional steroids in the treatment of OFG specially in patients who are diabetic and hypertensive. The systemic side effects are reduced while we use intralesional steroids.

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