Asian Journal of Research in Dermatological Science

4(3): 23-26, 2021; Article no.AJRDES.71153

Granulomatous Cheilits: A Rare Entity

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Authors' contributions

This work was carried out in collaboration between both authors. Both authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Giuseppe Murdaca, University of Genova, Italy.
Reviewers:
(1) Sundaramoorthy Srinivasan, Chettinad Hospital & RI Chettinad Academy of Research & Education, India.
(2) K.G.Padmanaban, Tamilnadu Dr.M.G.R.Medical University, India.
Complete Peer review History: https://www.sdiarticle4.com/review-history/71153

Case Study

Received 20 May 2021
Accepted 27 July 2021
Published 02 September 2021

ABSTRACT

Granulomatous cheilitis (GC) also known as Miescher cheilitis, is a rare chronic inflammatory idiopathic disorder of lips characterized by painless swelling of lips and was first described by Meischer in 1945. Wiensenfeld described it as non caseating granulomatous inflammation without any systemic cause and termed it as Oro facial granulomatosis (OFG). We hereby wish to present this rare entity, the mode of presentation and its various treatment modalities.

Keywords: Cheilitis; granulomatous cheilitis; melkerson rosenthal syndrome.

1. INTRODUCTION

Granulomatous cheilitis (GC) also known as Miescher cheilitis, is a rare chronic inflammatory idiopathic disorder of lips characterized by painless swelling of lips and was first described by Meischer in 1945 [1]. Wiensenfeld described it as non caseating granulomatous inflammation without any systemic cause and termed it as Oro facial granulomatosis (OFG) [2,3].

GC can occur at any age without any sexual predisposition and had an incidence of 0.08% [4]. Melkerson Rosenthal Syndrome (MRS) is the syndrome in patients with GC showing features of facial palsy and fissured tongue [5,6,7]. This

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rare entity should be considered in the differential diagnosis of persistent lip swelling [8].

2. CASE REPORT

A 38-year-old man presented to our Dermatology Department with left-sided swelling of and below lower lip since 2 months, which was gradually progressive. The size of the lesion was 3×2cm There were no complaints of itching or pain. There were no lesions elsewhere over the body (Fig. 1). On palpation, swelling of and below lower-lip was present, which was non-tender and soft with intact sensations, no rise of temperature. There was no gingival swelling. There was no history of any drug intake/trauma/insect bite/teeth pain or pus discharge. He had no history of angioedema, anaphylaxis, seasonal allergic rhinitis, or any malignancy. There was no family history of angioedema. All routine investigations were normal. Chest-X-ray, ACE levels were normal. Allergic patch testing was not done. Mantoux and SSS for AFB were negative. A punch biopsy was obtained from the lower lip, which showed mild acanthosis of the epidermis and scattered, noncaseating, poorly formed granuloma comprised of epithelioid cells and multinucleated giant cells with lymphoplasmacytic inflammatory infiltrate in the dermis. Perivascular lymphocytic infiltration was also present, and changes were consistent with diagnosis of GC. Special stains like ZN and PAS were negative for AFB and fungal-elements respectively.

The patient was given oral clofazimine 100 mg b.d for 3 months followed by administration of intralesional triamcinolone acetonide 10 mg/ml with an insulin syringe to the lower lip once weekly for a month along with combination of oral doxycycline 100 mg for a month showed marked reduction in the swelling (Fig. 2).

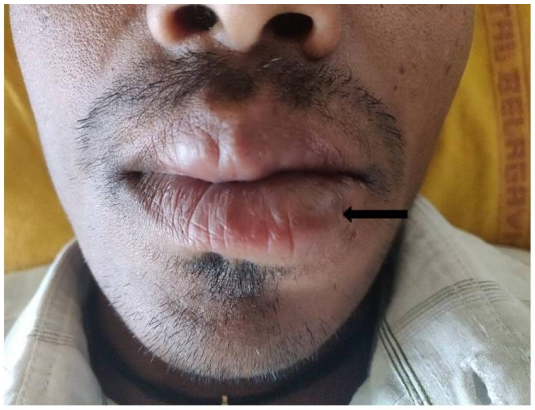

Fig. 1. On presentation: Non-tender, soft –firm swelling with intact sensations over left-side of and below the lower lip

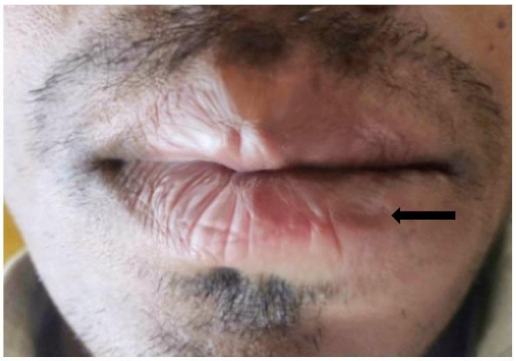

Fig. 2. The patient was given oral colfazimine 100 mg b.d for 3 months followed by administration of intralesional triamcinolone acetonide 10 mg/ ml with an insulin syringe to the lower lip once weekly for a month along with combination of oral doxycycline 100 mg for a month showed marked reduction in the swelling

3. DISCUSSION

The etiology of GC is complex. There are association with genetic, allergic, and infectious causes. A wide range of foods including wheat and dairy products, chocolate, eggs, peanuts, cocoa, cinnamaldehyde, monosodium glutamate, cormosine, food additives, toothpaste, and dental material such as amalgam and mercury has been implicated as the causative agents [9]. It may also be caused by an alteration in autonomic nervous system function localized to facial skin, resulting in increased vascular permeability and edema [10]. One recent hypothesis is that it can be due to random influx of inflammatory cells. Chronic antigen stimulation leading to monoclonal lymphocytic expression, cytokine production which leads to granuloma formation has been identified.

Clinically, the first episode typically subsides in hours or days. But both frequency and duration of the attacks increase until the swelling becomes persistent. The upper lip, lower lip, or both lips can be involved. The disease can also affect other oral and facial regions including the face, oral mucosa, gums, tongue, pharynx, and larynx. The oral mucosa is thickened and edematous with the buccal and labial (70%) mucosa assuming a corrugated or lobulated appearance. Cobblestone appearance of the buccal mucosa also may be observed.

The differential diagnosis of persistent lip swelling includes other granulomatous diseases such as a foreign body reaction, mycobacterial infection, sarcoidosis, Crohn's disease, Wegener's granulomatosis, and histoplasmosis; amyloidosis; rosacea; medications such as angiotensin-converting enzyme inhibitors and calcium channel blockers; atopic reaction to a wide variety of allergens; and hereditary diseases such as C1 esterase deficiency.

Diagnosis of GC is made excluding other conditions by history. History of atopy, drug history to rule out drug induced angioedema (angiotensin-converting enzyme inhibitors and calcium channel blockers should alert the physician to the possibility of angioedema) and family history of any hereditary angioedema. For patients with swelling lasting more than 2 weeks, causes for chronic cough (e.g. sarcoidosis and histoplasmosis, tuberculosis) should be asked.

Physical examination should also include a cranial nerve examination to determine facial nerve involvement, a full oropharyngeal examination to check poor dentition or lesions concerning malignancy, examination of the tongue for plication to diagnose MRS, inspection for aphthous ulcers that may suggest early Crohn's disease, and inspection for erythematous lesions suggestive of rosacea.

Histopathology shows noncaseating granulomatous inflammation in the dermis. Typically, the granulomas appear to cluster around scattered vessels and are not well formed or discrete. Fibrosis may be present in long-term lesions.

Corticosteroids (local or oral) have been used with temporary improvement. Combination therapy with intralesional triamcinolone 10-40mg once per week for 3 weeks and clofazimine 100-200 mg daily for 3-6 months or dapsone 100mg daily for 2 weeks followed by 50 mg daily for 25 weeks is one of the most commonly used treatment options.

Antibiotics such as sulfa drugs, tetracycline, macrolides, minocycline, hydroxychloroquine, sulfasalazine, antihistamines, thalidomide, isoniazid, roxithromycin 150 mg daily, metronidazole 1000 mg daily until clinical response is noted and immune modulators such as methotrexate, infliximab, adalimumab subcutaneous injection 80 mg week 1, 40 mg week 2 then 40 mg every other week until clinical response is noted, have been reported to provide good results. Elimination diets have been recommended, such as cinnamon-free and benzoate-free diets. Surgical treatment is an option for patients who are severely affected or have impaired function. Cheiloplasty, gingivectomy is reserved for treatment-resistant cases with major deformity [8].

4. CONCLUSION

Granulomatous cheilitis is a chronically recurrent disease. Treatment of the condition is unsatisfactory.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle4.com/review-history/71153