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Original Research Article

Granulomatous cheilitis: a rare case report

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ABSTRACT

Granulomatous cheilitis is a rare granulomatous disorder characterized by a recurrent firm swelling of one or both lips. This is called the cheilitis granulomatosa of Miescher (CGM) when it occurs in isolation. It is called Melkersson-Rosenthal syndrome (MRS) if the classical triad of recurrent or persistent orofacial edema, plicated or fissured tongue (lingua plicata), and relapsing peripheral facial nerve paralysis is present. A 40-year-old female patient came to the Dermatology Department with persistent painless swelling of both lips and perioral area for the last one year. On histopathological examination there was a nodular tuberculoid granulomatous inflammation in patchy pattern present throughout the submucosa. The granuloma consists of lymphocytes, histiocytes and occasional plasma cells. AFB stain and PAS stain were negative. Diagnosis of granulomatous cheilitis was made and patient was put on tab clofazimine and intralesional corticosteroids. The case is being reported due to its rarity and the role of dermatopathologist to make a diagnosis.

Keywords: Granulomatous cheilitis, Melkersson-Rosenthal syndrome, Granulomatous disorder

INTRODUCTION

Granulomatous cheilitis is an uncommon, persistent, painless condition characterized by chronic swelling of the lip with no known cause. It is regarded as a form of orofacial granulomatosis (OFG), a term used to describe swelling in the orofacial region due to non-caseating granulomatous inflammation without any associated systemic disease.[1] Granulomatous cheilitis can be considered as a monosymptomatic form of melkersson-rosenthal syndrome which is characterised by triad of peripheral facial nerve palsy, fissured tongue and granulomatous chelitis.[2]

CASE REPORT

A 40-year-old female presented in department of dermatology with persistent painless swelling of upper lip, lower lip and extending to perioral area for the last one year. Swelling started from upper lip and then gradually spread to involve lower lip and to perioral area. No history of pain or itching over swelling. On examination lips and perioral area showed induration with crusting and fissuring, touch sensation was intact over the swelling. There was no facial asymmetry and facial expression was intact and equal bilaterally. There was no tenderness over lip and perioral area. No such lesions were seen intraorally. chest radiograph was normal, Mantoux test, serology for hepatitis B and C viruses and human immunodeficiency virus, angiotensin-converting enzyme levels, erythrocyte sedimentation rate were normal. patients had no family history of labial swelling, a personal history of neurologic, digestive, or respiratory symptoms, or a previous diagnosis of a granulomatous disease. On histopathological examination there was a nodular tuberculoid granulomatous inflammation in patchy pattern present throughout the submucosa. The granuloma consists of lymphocytes, histiocytes and occasional plasma cells. Overlying epidermis showed mild spongiosis and slight hyperplasia. Ziehl-Neelsen, PAS stain was negative for acid fast and

fungal organism. Diagnosis of granulomatous cheilitis was made. For treatment patient was given tab clofazimine 100 mg thrice a day and was injected intralesional triamcinolone injection every two weekly. Patient was followed up and showed mild improvement after 6 weeks of treatment.

DISCUSSION

Granulomatous cheilitis is a rare condition that can develop at any age, even during childhood3, but it is most commonly seen in individuals in their twenties or thirties4. The upper lip is typically affected, though the lower lip can also be involved less often. Additionally, the disease can impact other areas of the oral and facial regions, including the face, oral mucosa, gums, tongue, pharynx, and larynx.[3] The most common presentation is acute, painless swelling of the lips that lasts from hours to days. Initially, these swelling episodes are self-limiting and resolve entirely; however, after several recurrences, the lips may retain a degree of firm, hardened edema. Patients might experience pain or a burning sensation, particularly if oral involvement includes erythema, fissures, erosions, or scaling of the lips. When other facial areas are affected, regional lymph nodes may become enlarged.[5] Granulomatous cheilitis is the most common presentation of Melkersson-Rosenthal syndrome, occurring in 80% of cases. Melkersson-Rosenthal syndrome is a systemic granulomatous disorder with an unknown cause, characterized by a triad of orofacial edema, facial palsy, and a fissured (or scrotal) tongue. The full triad appears in 8% to 25% of those diagnosed with the syndrome, while oligosymptomatic forms account for 47% of cases. In 28% of cases, the only symptom is orofacial swelling or granulomatous cheilitis.[6,7]

Multiple studies have connected granulomatous cheilitis with Crohn's disease. In a study of 14 patients with granulomatous cheilitis, 30% were found to have an association with Crohn's disease, while 43% exhibited minor gastrointestinal symptoms without any detectable signs of inflammatory bowel disease.8 Several substances, including chocolate, food additives, cinnamon compounds, and benzoates, have been suggested as potential causes of granulomatous cheilitis, with elimination diets leading to improvement in some patients.[9] Additionally, sensitization to metals like gold and mercury has been associated with the development of the disease.[10] The differential diagnosis for persistent upper lip swelling includes other granulomatous diseases like foreign body reaction, mycobacterial infection, sarcoidosis, Crohn’s disease, Wegener’s granulomatosis, and histoplasmosis; amyloidosis; rosacea; medications such as ACE inhibitors and calcium channel blockers; allergic reactions to various allergens; and hereditary conditions such as C1 esterase deficiency.[2] The diagnosis of granulomatous cheilitis is based on clinical observations confirmed by histological examination. Histopathology typically shows non-necrotizing granulomas, edema, lymphangiectasia, and perivascular lymphocytic infiltration. However, non-caseating granulomas may not always be present in the dermis, particularly in the early stages of the condition, So their absence does not exclude the diagnosis. Consequently, granulomatous cheilitis is not always associated with distinct histological changes.[11] The difficulty of managing granulomatous cheilitis is reflected in the variety of treatments that have been used and the lack of consensus regarding the preferred treatment. A frequently used treatment option is combination therapy with intralesional triamcinolone and either clofazimine or dapsone.[12] Other reported treatments include prednisone, hydroxychloroquine, sulfasalazine, antihistamines, tetracyclines, metronidazole, macrolides, thalidomide, and infliximab.[13]

Figure 1 (A and B): Erythematous swelling of upper lip, lower lip and perioral area.

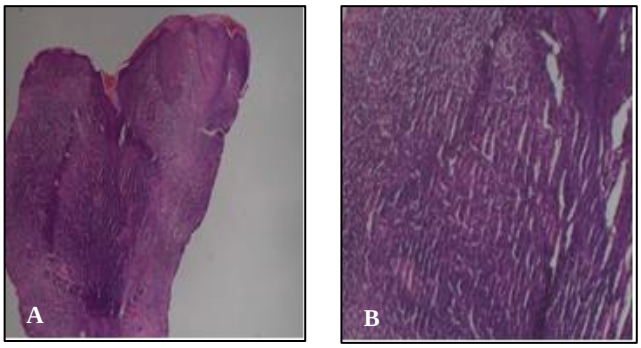

Figure 2: (A) histopathology 10x slide showing nodular tuberculoid granulomatous inflammation and (B) 40×slide showing granuloma consists of lymphocytes, histiocytes and plasma cells.

CONCLUSION

Above described is a case of granulomatous cheilitis, diagnosis was made based on clinical presentation and histopathology. No effective treatment which completely cures the disease is found till date.

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