Case Report

Miescher’s cheilitis: A case report with literature review

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ABSTRACT

Miescher’s cheilitis is an inflammatory disorder characterized by chronic lip swelling due to granulomatous inflammation. It is a rare disorder first described by Miescher in 1945. It is a monosymptomatic form of Melkersson-Rosenthal syndrome. We report a case of Miescher’s cheilitis in a 59-year-old lady presented with recurrent swelling and erythema of upper lip and submental area with no features of facial palsy and fissuring of tongue. Laboratory tests including serum complement C3, C4 and C1 esterase inhibitor functional were within normal limits. The biopsy from the upper lip revealed dermal edema, dilated lymphatic channels and multiple granulomas. After exclusion of other causes of orofacial granulomatosis, the diagnosis of Miescher’s cheilitis was made. Patient was treated with oral hydroxychloroquine and topical tacrolimus and mometasone cream with gradual improvement.

INTRODUCTION

Miescher’s cheilitis (MC) is a rare condition, also known as cheilitis granulomatosa (CG), characterized by recurrent lip swelling. This inflammatory disorder was first described by Miescher in 1945.¹ It is a monosymptomatic form of Melkersson-Rosenthal syndrome (MRS), which is characterized by edema of the lips, facial palsy and fissuring of the tongue. MC is a granulomatous inflammatory disorder and an entity included under the umbrella of orofacial granulomatosis (OFG), which was coined by Wiesenfeld in 1985.¹ Here, we report a case of Miescher’s cheilitis due to its rarity.
CASE REPORT

A 59-year old lady presented with recurrent non-painful swelling and erythema of upper lip (fig.1A) and submental area (fig.1B) since 2 years, previously treated with intralesional and topical steroid. There was no history of cutaneous or systemic allergic manifestation or any other systemic disease. Features of facial palsy and fissuring of tongue were not present. On examination, the upper lip showed erythema and swelling with focal crusting. Submental area also showed erythema and mild swelling. Such lesions were not seen intraorally.

Ultrasonography revealed tortuous labial vessels and soft tissue thickening in upper lip and edematous thickening of subcutaneous tissue in submental region.

Laboratory examination revealed complete blood count within normal limits. No eosinophilia noted. Serological investigations for HIV, HBsAg, Anti-HCV, CRP and anti dsDNA were negative. Serum complement C3, C4 and C1 esterase inhibitor functional were within normal limits. Fine needle aspiration cytology of the lip was reported as inflammatory lesion. Provisional diagnoses of angioedema, cheilitis granulomatosa, leprosy and tuberculosis were made and punch biopsy from upper lip was taken.

The biopsy from the upper lip revealed dermal edema, dilated lymphatic channels (fig.2) and multiple granulomas with Langhans’ type multinucleated giant cells (fig.3).

Ziehl-Neelsen stain and modified Fite stain for acid fast bacilli were negative. PAS stain did not reveal any fungal elements. Congo red was negative for amyloid. PCR for mycobacterial DNA and immunohistochemistry for mycobacteria were negative. Immunohistochemistry for Leishmania was also negative. With all of these findings the diagnosis of Miescher’s cheilitis was made.

Patient was treated with oral hydroxychloroquine sulfate 200 mg once daily and topical tacrolimus and mometasone cream. After 2 months of treatment, submental swelling and erythema disappeared completely and lip swelling was still persisting but with significant improvement.

DISCUSSION

Miescher’s cheilitis is a rare disease with unknown etiopathogenesis and the swelling of lip due to granulomatous inflammation. It is a monosymptomatic form of Melkersson-Rosenthal syndrome, which is characterized by the triad of orofacial edema, facial palsy and fissured tongue. The complete triad is present in 8% to 25% of patients diagnosed with MRS, while oligosymptomatic forms of the condition account for 47% of cases. Miescher’s cheilitis is the most common form of MRS constituting 80% of cases. Our patient had monosymptomatic form of the syndrome without facial palsy and fissured tongue. No neurological signs were present in our patient.

In 1985, Wiesenfeld et al coined the concept of orofacial granulomatosis to describe a series of non-infectious, non-necrotizing granulomatous disorders of the lips, face and oral cavity that included CG, Crohn’s disease and sarcoidosis. OFG is an umbrella term that encompasses a number of entities of diverse etiology, pathogenesis and clinical features.

The etiology of MC is poorly understood, but has been associated with genetic, allergic, and infectious factors. Some cases may demonstrate an autosomal dominant inheritance pattern, with the responsible gene mapping to chromosome 9p11. There are reports that associate the pathogenesis of MC with odontogenic infection and autoimmune mechanism. Authors even mentioned that the episode of MC persists even after the infection has resolved in some patients. Various food items including chocolate, food additives and cinnamon compounds have been implicated as possible etiologic agents; the elimination of these items from food have led to improvement in
Miescher’s cheilitis occurs most frequently in the second or third decade of life, but can occur at any age with no racial or sexual predilection. In a study of Martinez et al, the age of patients with MC ranged between 29 and 74 years. Our patient was 59-year-old female. Histopathological findings of MC include dermal non-caseating granuloma, dermal edema and dilatation of lymphatic channels. The overlying epidermis is usually hyperplastic. There is no acanthosis of epidermis in our case and all other histological features were present.

Differential diagnoses of MC include mycobacterial infection, foreign body reaction, sarcoidosis, Crohn’s disease, Wegener’s granulomatosis, histoplasmosis, amyloidosis, rosacea, medications such as ACE inhibitors and calcium channel blockers; toxic reactions to a wide variety of allergens and hereditary diseases such as C1 esterase deficiency. In our case, we have excluded mycobacterial infection, amyloidosis, fungal infection and C1 esterase deficiency by special stains, EIA, PCR or immunohistochemistry. Features of vasculitis were not present excluding the possibility of Wegener’s granulomatosis. History of intake of ACE inhibitors or calcium channel blockers was not found in our case. Crohn’s disease is difficult to rule out based on morphology, however Crohn’s disease is more likely to involve the oral cavity and in our patient, no lesions were noted in oral cavity. In addition, there has been an association noted between MC and Crohn’s disease.

In patients presenting with lip swelling, it is important to perform an appropriate evaluation, which includes history of atopy or current therapy with an ACE inhibitor or calcium channel blocker, chest X-ray and mantoux test to exclude angioedema, sarcoidosis or mycobacterial infection. Gastrointestinal tract endoscopy may exclude Crohn’s disease. A family history of orofacial swelling should prompt an investigation into the autosomal dominant C1 esterase inhibitor deficiency (hereditary angioedema). Patch test or serum allergy test shall be used to rule out food and food additives sensitivity.

There is no definite treatment for MC as the pathogenesis is obscure. White A et al recommended a cinnamon- and benzoate-free diet as first line therapy for OFG as lip inflammatory score goes down at 8 weeks. Various other treatment options for MC have been reported including antibiotics like tetracycline, roxithromycin and clofazimine, metronidazole, hydroxychloroquine, oral and intralesional steroids, adalimumab (TNF alpha inhibitor) and surgical resection.

CONCLUSION

Miescher’s cheilitis is an independent entity within the group of orofacial granulomatosis. It is rare and difficult to diagnose and treat. In this case, the diagnosis of MC was made after exclusion of other causes of OFG and importance of thorough investigation is highlighted.

Conflict of Interest: None

REFERENCES


