CASE REPORT

Melkersson Rosenthal Syndrome: A Case Report.

Mohammed Azamulla[1], Suman Sen[2], Sufia Khan[1], Subhashini Singh[1]

Abstract

Introduction: Melkersson-Rosenthal Syndrome (MRS) is a rare neuro-mucocutaneous condition. It mainly includes three triads as orofacial swelling, facial palsy and fissured tongue. All of the classical triad may not be seen at the same time, as symptoms may appear in different interval of time and has a recurrence. MRS is diagnosed based on oligosymptomatic when two or more clinical features are present or monosymptomatic cases when there evidence of a non-necrotising, granulomatous cheilitis in patients with isolated facial or lip edema can be noted in biopsy.

Case Presentation: In this case report a 18 year old male patient all the three classic clinical characteristic features as granulomatous cheilitis, unilateral facial muscle palsy and a fissured tongue are seen. The clinical features of this rare condition along with treatment procedure are presented.

Conclusion: Oral corticosteroids are the mainstay in the treatment of MRS. Intralesional triamcinolone acetonide used in oro-facial edema. In recurrent cases of MRS need second-line immunosuppressants like methotrexate and thalidomide. In refractory recurrent facial nerve palsies Surgical decompression of the facial nerve may be considered.

Keywords: Granulomatous cheilitis, fissured tongue, facial palsy, corticosteriod

Oral and Maxillofacial Pathology Journal (2023): ***https://www.ompj.org/archives***

Introduction

Melkerrson-Rosenthal syndrome is a rare disorder of unknown aetiology and characterized by the triad of oro-facial edema, facial nerve palsy, and furrowing of the tongue. Two or more of the above are essential for making a clinical diagnosis. However, cases in which the patient experiences all the three symptoms are relatively rare. It was initially described by Melkersson in 1928 and followed by Rosenthal in 1931. In 1931 it was named after Ernst Melkersson and Curt Rosenthal. Rosenthal emphasized and included its association with fissured tongue also called as "scrotal tongue". Around only 8–25% of the cases had all the three triad of this syndrome.[1] There is no racial or gender predilection of the syndrome. The onset of the condition initially noted in late childhood or young adult life. It has no definite etiology, though genetic, infectious and immunologic factors may be considered.[2] Genetic inheritance with autosomal dominant gene on chromosome 9 may be present.[3] Risk factors include some food allergy, lymphogranulomatosis, dental infections, tonsillitis, adenoid hypertrophy, viral infections including herpes simplex, and other bacterial or viral infections have been considered as causative factors.[3] The first episode may resolve in few hours or days, but swelling may become more intense that may last longer in next few episodes and can become permanent in due course of time. Classical triad of MRS can only be observed in 8% to 18% of the patients that makes it difficult to diagnose this rare condition.[4,5]

Case Presentation

A 18 year old male came to the department of oral medicine with a complain of swelling of upper lip and inability to close right eye with reduced taste sensation. He had a history of facial paralysis on the right side of his face 1 month back. During extra-oral inspection there was loss of facial expression were observed over the right side of face (Figure 1) and swelling of the lips (Figure 2). Patient had an acute onset of weakness over right side of face with deviation of nasolabial fold to left along with inability to close right eye completely (Figure 3) and loss wrinkling over forehead on frowning (Figure 4). He did not have any fever, vesicles on the ear, trauma, otalgia, vertigo, tinnitus. On examination showed right facial paralysis associated with loss taste sensation in anterior part of tongue. He had inability to blow

[1]Department of Periodontology, Awadh Dental College, Jamshedpur, India [2]Department of Oral Medicine and Radiology, Haldia Institute of Dental Sciences and Research, Haldia, West Bengal, India

Corresponding author: Suman Sen, Department of Oral Medicine and Radiology, Haldia Institute of Dental Sciences and Research, Haldia, West Bengal, India. Email- sumansen20@yahoo.co.in

How to cite this article: Azamulla M, Sen S, Khan S, Singh S Melkersson Rosenthal Syndrome: A Case Report. Oral Maxillofacial Pathol J 2023;14(1): page no. 113-115

Source of Support: Nil

Conflict of Interest: None

Acknowledgement: To the patient for his consent

the right cheeks. During intraoral examination it was observed that the patient had multiple fissured dorsum of tongue (Figure 5). The rest of the neurologic examination, fundoscopy and external and middle ear examination were normal.

Treatment was advised based on symptoms of the cases. Oral corticosteroid therapy was prescribed to him for a total 4 weeks with tapered doses. Initially it is given of 60 g in the first seven days, 40 g in the following seven days, 20 g for the second seven days and 10 g in the last seven days. Patient was given eye pad to be used at night during sleeping and eye lubricant was advised. Physical therapies were performed as exercise, bio feedback and massage are used for fasten recovery of facial palsy. Patient advised to oral hygiene instruction.

Discussion

The reported incidence of Melkersson-rosenthal syndrome (MRS) is between 0.2 and 80 in 100,000 per year.[6] This incidence may be underestimated, since this syndrome is both mis- and under-diagnosed, and a mean diagnostic delay of nine years has been reported.[1,2] MRS is a rare neurological entity of unknown etiology. Classical triad of these syndrome includes recurrent facial palsy, swelling of face but most commonly lips, and deep fissured over dorsum surface of tongue.[7] This entity is seen equally in both gender and has racial predilection. Due to inconsistency of signs and symptoms and appearance most of patients go undiagnosed for prolonged period.[8] The unilateral facial palsy is of lower motor neuron type may mistake clinically as Bell's palsy that can be unilateral or bilateral and of varying severity. Involvement of other cranial nerves such as olfactory, auditory, glossopharyngeal and hypoglossal nerves is also noted in certain cases.[9] It is commonly seen in second decade of life.

In this case also the patient is teenager and had all the clinical features of Bell's palsy along with swelling of lips and fissured tongue. Differential diagnosis of MRS can be thyroid orbitopathy, allergy, angioedema, bacterial, viral or filarial infections, systemic lupus erythematosus, dermatomyositis, bell's palsy and Ramsay Hunt syndrome.[10] In this case biopsy is not essential as all classic clinical condition is seen to confirm the MRS. Inflammatory bowel disease like Crohn's disease may present with oral manifestations such as lip swelling with fissures on lips, mucositis, gingivitis, glossitis, and cobblestone appearance of the oral mucosa.[11]

There is a significant role of abnormal immune function, immune deregulation, allergic tendencies in patients with MRS. So, short courses of immunosuppressants are often used in the

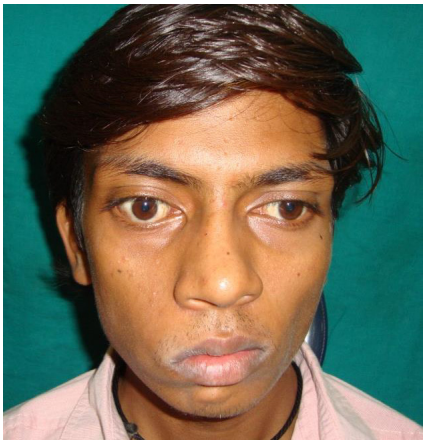

Fig.1: Front facial profile showing loss of facial expression were observed right side

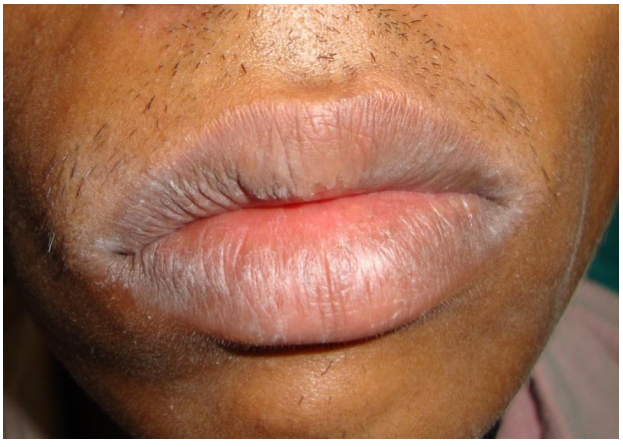

Fig. 2: Swelling of upper and lower lips

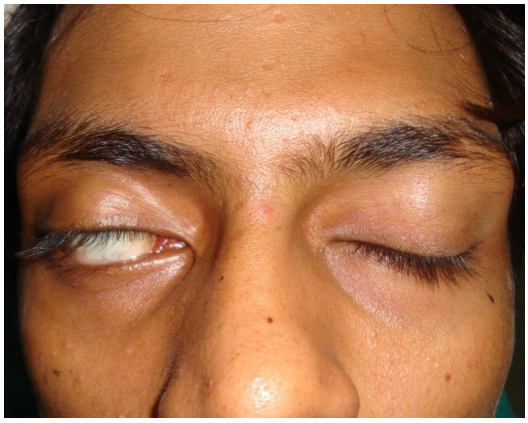

Fig. 3: Inability to close right eye completely of affected side

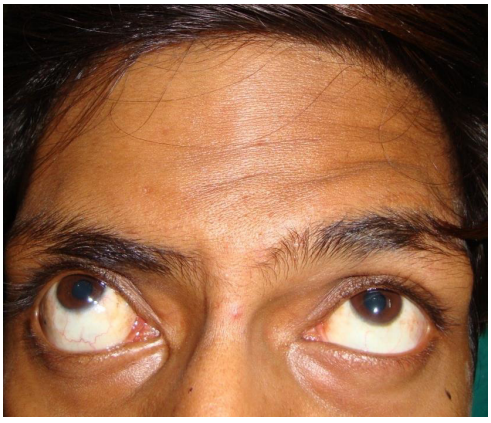

Fig. 4: Loss of wrinkling on right side forehead on frowning

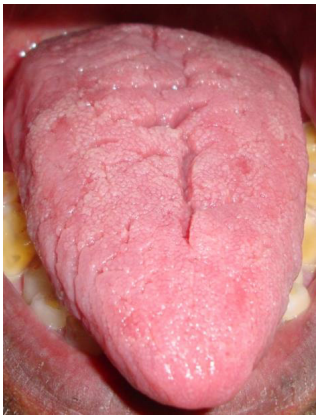

Fig. 5: Multiple fissured dorsum of tongue

treatment of MRS. There is no specific treatment for MRS.[11] The initial therapeutic approach is based on intralesional or systemic corticosteroids administration tapered over 3–6 weeks, depending on the severity of the condition, improves symptoms in 50–80% of the patients, with a recurrence rate decrease of 60–75%.[12] In this case methylprednisolone is given along with eye care of affected side and physical therapy for facial palsy recovery. Vitamins like thiamine, niacin, riboflavin, pyridoxine, ascorbic acid and vitamin E have can be given along with the symptomatic treatment for eye care that includes eye patching and lubrication, lubricating drops should be applied frequently during the day and a eye protective pad should be used at night to protect as eye mostly do not closed completely. Physical therapies as exercise, biofeedback, electrotherapy, massage and thermotherapy are used for facial palsy.

Conclusion

MRS is diagnosed based on physical findings and history. It may be misdiagnosed since facial palsy is not present in all the cases. Condition can be treated with corticosteroid and facial palsy heals within 2 weeks but it has chances of recurrences. As oral diagnostician we should keep the MRS in mind for the patients who have recurrent or permanent swelling of lips, unilateral facial palsy and fissured tongue.

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