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MELKERSSON ROSENTHAL SYNDROME: A CASE REPORT OF RARE DISEASE

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ABSTRACT Melkersson-Rosenthal syndrome (MRS) is a rare, neuro-mucocutaneous disorder of unknown etiology, classically defined by a triad of recurrent orofacial edema, relapsing facial nerve palsy, and a fissured tongue. While the complete triad is present in only a minority of cases, the presence of at least two of these features, particularly orofacial granulomatosis, is highly suggestive of the diagnosis. The clinical course is typically characterized by recurrent episodes of swelling, most commonly affecting the lips, which may eventually become permanent. The facial palsy is peripheral and can be unilateral or bilateral.The pathogenesis of MRS is not fully understood, though genetic predisposition, infectious triggers, and immune dysregulation have been implicated. Histopathological examination of affected tissue typically reveals non-caseating granulomatous inflammation, which is a key diagnostic feature.Diagnosis is primarily clinical, based on the characteristic signs and symptoms, and supported by biopsy findings. Management of Melkersson-Rosenthal syndrome is often challenging and primarily focused on symptomatic relief. Corticosteroids, both systemic and intralesional, are the mainstay of treatment, particularly for acute episodes of orofacial edema and facial palsy. Due to its rarity and variable presentation, increased awareness of MRS is crucial for timely diagnosis and appropriate management to improve patient outcomes.

KEYWORDS : Melkersson-Rosenthal Syndrome, Granulomatous cheilitis, fissured tongue, facial palsy, corticosteriod

INTRODUCTION

Melkersson Syndrome was first described in 1928 as peripheral facial paralysis and swelling of lips. In 1931, Rosenthal completed the triad by adding the presence of fissural tongue. The classical form of the Melkersson Rosenthal Syndrome (MRS) consists of the clinical triad of recurring facial nerve paralysis, swelling of one or both lips and fissural tongue (1).However, the majority of patients present with only one or two of these features, with research indicating that not all cases include the full triad. For instance, studies have shown that facial edema and fissured tongue are more commonly observed than facial paralysis, with prevalence rates varying.The onset of symptoms is most commonly between 25 and 40 years (range 1–69 years), with a female preponderance (sex ratio of 2:1).2,3.

The differential diagnosis of MRS includes a broad spectrum of heterogeneous conditions, mainly represented by other granulomatous disorders such as foreign body reaction, sarcoidosis, Crohn's disease, Wegener's vasculitis, amyloidosis and a wide variety of infections; Bell's palsy, orofacial herpes, rosacea, contact dermatitis and allergic reactions should also be considered(4).Diagnosis is mostly based on clinical findings (1) In this case report, a patient with MRS who had applied with complaints related to recurring swelling of the lower lip is presented.

Case Report

55 years-old male patient came to our hospital with complaint of swelling of upper lip and inability to close left eye with reduced taste sensation.Patient's history revealed that he had suffered facial paralysis on the left side of his face two years ago.During extra-oral examination, swelling of the lower lip, cheilitis and facial asymmetry were observed.Patient was unable to close his left eye, his angle of mouth deviates to right side on smiling and loss wrinkling over forehead on frowning (figure 1). During intraoral examination it was observed that the patient had multiple fissured dorsum of tongue(figure 2).Lip biopsy shows non-caseating granulomatous inflammation consisted of lymphocytes and epithelioid histiocytes, and few multinucleated giant cells, clustered around scattered vessels. Patient was diagnosed as Melkersson Rosthenthal Syndrome based on the clinical and histopathological findings. Oral corticosteroid therapy was administered to the patient for 26 days in total, with doses of 80 g in the first five days, 60 g in the following seven days, 40 g for the second seven days and 20 g in the last seven days. Swelling of the lip was reduced after the steroid therapy.Patient was given eye pad to be used at night during sleeping and eye lubricant was advised.

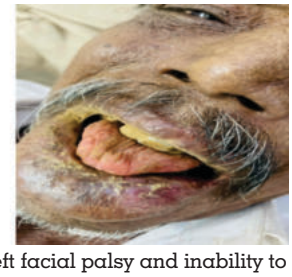

Fig 1: Lower lip swelling and left facial palsy and inability to close left eye

Fig 2: Fissured tongue

DISCUSSION

Melkersson–Rosenthal syndrome is a rare, neuro-mucocutaneous syndrome with an estimated incidence of 0.08% in the general population (5). Onset of this disease is more frequent in young adults, between the second and the third decades of life(6).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).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(8).Recurrent lip swelling, also termed Miescher's syndrome or Miescher's cheilitis granulomatosa (MCG), is the most common monosymptomatic presentation of MRS and histologic features include lymphomonocytic infiltration, non-caseating epithelioid cell granulomas, multinucleate Langerhans-type giant cells and fibrosis(9). The differential diagnosis of MRS includes a broad spectrum

of heterogeneous conditions, mainly represented by other granulomatous disorders such as foreign body reaction, sarcoidosis, Crohn's disease, Wegener's vasculitis, amyloidosis and a wide variety of infections; Bell's palsy, orofacial herpes, rosacea, contact dermatitis and allergic reactions should also be considered.(10)The third finding of MRS is the fissural tongue. Since fissural tongue is a widespread abnormality in the society, it is of lesser importance in the MRS diagnosis.Other findings of MRS can be listed as trigeminal neuralgia, ocular palsy, paresthesia, dysfunction of the lacrimal and saliva glands, migraine, keratitis, increases of secretions and psychological episodes(11).There is a significant role of abnormal immune function, immune deregulation, allergic tendencies in patients with MRS. So, short courses immunosuppressants are often used in the 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%.(13).Other therapeutic options include antibiotics, non-steroidal anti-inflammatory drugs (NSAIDs), and immunosuppressants. Surgical intervention may be considered for persistent, severe lip swelling or for decompression of the facial nerve in refractory cases. It has been reported that in most patients idiopathic facial paralysis can heal spontaneously within the first three weeks (14).

CONCLUSION

Diagnosis of MRS is difficult as it is a rare disease.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.Future research must be prioritized to unravel the complex etiology and pathophysiology of Melkersson-Rosenthal syndrome. A deeper understanding of the potential genetic, infectious, and immunological factors at play is essential for the development of targeted and more effective therapies.

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