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Melkersson-Rosenthal syndrome : A new case

Case Report

Kaoutar Laamari*, Hanane Baybay, Samia Mrabat, Zakia Douhi, Sara Elloudi, Mernissi Fatima Zahra
Departement of Dermatology, University Hospital Hassan II, Fes, Morocco

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***Corresponding author:** Kaoutar Laamari, Departement of Dermatology, UniversityHospital Hassan II, Fes, Morocco

Abstract

Melkersson-Rosenthal Syndrome (MRS) is a rare diseasecharacterized by the triad of recurring facial paralysis, a fissuredtongue, and recurringswelling of the lips and/or face. The etiologyisunknown; itmaybecaused by hereditaryfactors or conditions occurringlater in life.

Keywords

Melkerssonrosanthal; Macrocheilitis; Facial Paralysis

Introduction

Melkersson-Rosenthal syndrome (MRS) is a rare orofacial granulomatosis, having an incompletely understood pathogenesis. It associates a recurrent palsy of the facial nerve, an edema of the superior lip and fissure grooves on the dorsal surface of the tongue.

Cases displaying the entire triad are very rare. The mono-symptomatic or oligo-symptomatic formis more common. Presence of granulomatous cheilitis in the biopsy together with one or two of these clinical findings could be sufficient for MRS diagnosis .

The differential diagnosis of MRS includes also chronic inflammatory and infectious diseases characterized by granulomatous infiltration, as well as rosacea, contact dermatitis, allergic reactions.

We report a case of a young adult suffering from macro cheilitis and we made the diagnosis of melkersson rosenthal syndrome.

Case report

35 years-old male patient who had no systemic diseases and regular drug use applied to our dermatology department, with painless and recurring swelling of the right side of is upper lip. patient's history revealed that he suffered from facial paralysis on the right side of his face. During extra-oral examination, swelling of the right side of the upper lip, macro cheilitis and asymmetry were observed. (Figure 1). During intraoral examination it was observed that the patient had no fissured tongue (Figure 2).

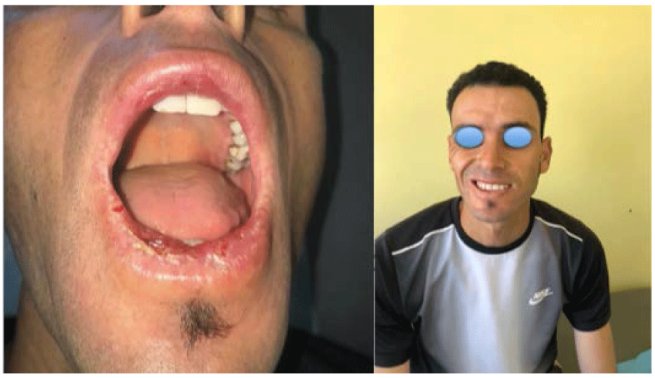

Figure 1: Macro cheilitis and asymmetry

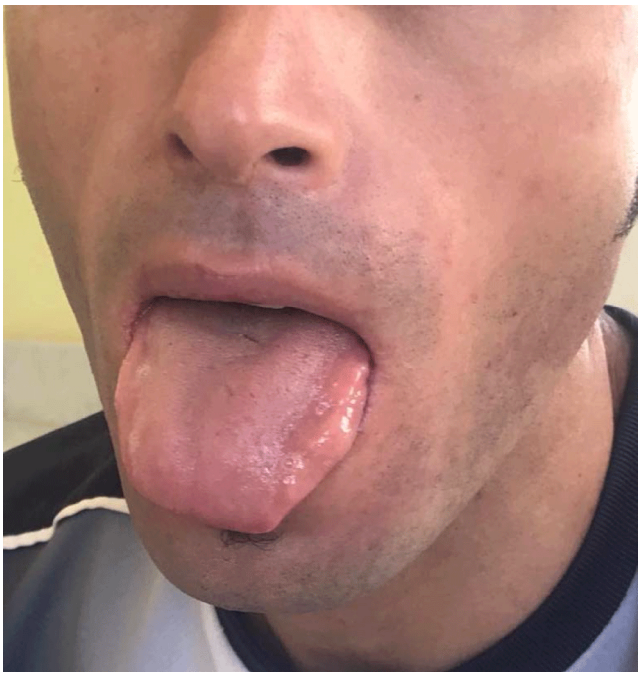

Figure 2: No fissural tongue

He was diagnosed with melkersson rosenthal Syndrome based on the clinical findings.

Oral corticosteroid therapy was administered to the patient with good evolution.

Discussion

The Melkersson-Rosenthal syndrome is a rare disorder of unknown etiology characterized by a triad of recurrent orofacial swelling, relapsing facial paralysis, and fissured tongue (1,2). Onset of this diseaseis more frequent in young adults, between the second and the third decades of life (3).

Exacerbations and recurrences are common. The orofacial swelling is characterized by fissured, reddish-brown, swollen, nonpruritic lips or firmedema of the face. The facial palsy is indistinguishable from Bell's palsy. The fissured tongue is seen in one third to one half of patients and, although the least common manifestation, its presence assists in diagnosis. The classic triadis not seen frequently in its complete form; therefore, diagnosis is difficult. This is particularly true because monosymptomatic and oligosymptomatic variants are seen more commonly. cheilitis granulomatosa treatment of Miescher is an example of a monosymptomatic variant of the Melkersson-Rosenthal syndrome. (4,5,6)

The differential diagnosis of MRS includes a broadspectrum 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' spalsy, orofacial herpes, rosacea, contact dermatitis and allergic reactions should also be considered (7,8,9,10).

Although not always necessary, a biopsy may be required to differentiate the syndrome from other conditions, especially Crohn's disease. The histopathology shows intra- and extravascular clusters of histiocytic-epithelioidcells and noncaseating sarcoidal granulomas (11). A multidisciplinary team approach involving otolaryngologists, dermatologists, and neurologist is required to manage these patients. Most patients are managed with steroids and nonsteroidal anti-inflammatory drugs, and other drugs that have been tried are danazol and sulfasalazine. Facial nerve decompression may become necessary for patients with recurrent facial paralysis. (1)

Conclusion

Melkersson–Rosenthal syndrome is a rare disorders which may present as a classic triad of orofacial swelling, facial palsy and fissured tongue or, more frequently, witholigo/mono-symptomatic features.

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