

Case Report

Melkersson-Rosenthal Syndrome: a Case Report of the Classic Triad

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KEY WORDS

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ABSTRACT

Melkersson-Rosenthal syndrome (MRS) is a rare neurological condition that includes a triad of symptoms: recurring facial paralysis, orofacial swelling and fissured tongue. The diagnosis and treatment of this syndrome is difficult, because the classic triad is rarely possible to see in its complete form. The etiology of MRS is unknown, but it is thought to be caused by various factors such as infections, genetic predisposition, immune deficiency, food intolerance and stress. This case report presents a 22 years old male patient with classical triad of MRS.

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Introduction

MRS is a rare disorder that manifests by the clinical triad of recurring facial nerve paralysis, swelling of one or both lips (cheilitis granulomatosa) and fissured tongue [1-3]. MRS begins in childhood or adolescence [4]. This syndrome may appear alternately after the first time. After frequent attacks (from days to years), edema may remain and increase and finally become permanent [5]. The etiology of MRS is unknown, but there may be a genetic condition [1, 5].

The classic triad is rarely seen completely, therefore the diagnosis of this syndrome is difficult [1, 6]. The most common and first symptom of MRS is orofacial swelling. The fissured tongue is seen in about 20-40% of the affected people and may have been present since birth, and facial paralysis occurs in about 30% of patients [7]. In histopathologic features of a classic form of cheilitis granulomatosa edema and dilation of lymphatic vessels in superficial lamina propria is present, and Scattered aggregates of noncaseating granulomatous inflammation, consisting of lymphocytes and histiocytes, are also seen [8]. In this case report a patient with all aspects of the classical triad is presented.

Case Report

In this study, we presented a 22 years-old man applied to Prosthodontics department, Dental School, Tehran Medical Sciences University for prosthetic treatments. He had swelling of the right side of the lower lip, and paralysis of face (Figure 1). He has reported that the edema of lip had repeated with certain intervals. Also patient's history showed that he had suffered of facial paralysis and facial edema in right side of his face for many years ago. On intraoral examination it was observed that he had macroglossia, and deep fissures on the dorsal and lateral surfaces of the tongue (Figure 2).



Figure 1: Swelling and asymmetry in the right side of the lower lip and face of the patient



Figure 2: Fissured tongue

There was no systemic diseases, regular drug use and adenopathy. Patient was consulted with the Oral medicine Department, School of Dentistry, Tehran Medical Sciences University and he was diagnosed with MRS based on the clinical findings (complete triad of signs).

Discussion

MRS is a rare neuro-mucocutaneous disorder with a recurrent and progressive course, and characterized by the triad of lip swelling, fissured tongue and facial paralysis. The most common clinical presentation are oligosymptomatic or monosymptomatic forms [9].

The classic triad of symptoms of MRS is seen in only 8% to 18% of patients and the mono-symptomatic or oligo-symptomatic form is more common. This makes it difficult to diagnose this disease [9-10]. In our patient, all three clinical findings were observed at the time of referral to the clinic. Therefore, he was diagnosed with MRS based on clinical symptoms.

The first manifestation of MRS is usually edema of upper lip, lower lip, one or both species, eyelids or rarely one side of the scalp. Lips especially upper lip are involved more frequently [1]. In our patient, swelling was seen on the right side of the lower lip. Also, fissures was seen on the right side of the dorsal of tongue. The third finding in our patient was facial paralysis for many years ago.

In patients affected, swelling has a tendency for recurrence and usually completely subsides in the early stages [12-13] but it can to last longer and become a fixed enlargement of orofacial tissues. Therefore early

diagnosis and appropriate treatment can prevent of undesired complications [5].

This patient was 22 years old. Elias and *et al.* [9] in a study on the 75 cases with MRS showed the age range were 8-9 years. Feng and *et al.* [14] in a study on 44 cases showed the age onset was 14 years. However diagnosis may be delayed for several decades [9].

Etiology MRS is not clear. It seems that MRS is due to genetic factors in some cases, because it involves several members of the their families. Other factors as dietary, other allergens, infections (especially herpes simplex virus), immune deficiency, and stress may also be involved [4,15]. Therefore treatment of this rare disease is difficult [4].

However, treatment of this syndrome is symptomatic, and may include treatment with nonsteroidal anti-inflammatory agents, corticosteroids, antibiotics, and anti-depressant drugs [5,16-17]. Surgery may be prescribed to reduce the pressure on the facial nerves and reduce swelling, but its effectiveness has not been proven [18-19]. Regardless of the type of treatment, patients should be regularly examined, even if they have no clinical symptoms. Because over years the course of MRS is chronically progressive [5].

Patient referred to oral medicine department .For our patient oral corticosteroid therapy was administered. Dental impressions were taken for the diagnosis and treatment plan of prosthetic treatment and now he is completing his pre prosthetic treatments.

Conclusion

The Orofacial swelling of the MRS can be prolonged and become permanent. So early detection and proper treatment can prevent unwanted side effects. Dentists should consider MRS for patients with recurrent swelling in the lips, and they should refer them to the relevant specialists.

Conflict of Interests

Authors have no conflict of interest.

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