Melkerson-rosenthal syndrome associated with hemifacial spasm

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INTRODUCTION

Melkerson-Nosenthal syndrome (MRS) is characterized by a triad of repeating unilateral or bilateral facial paralysis, permanent or temporary orofacial edema, especially in the lips, and fissured tongue.1,2 It may manifest as the result of one or more of these symptoms. MRS etiology is still unknown, however genetic, infectious and immunological factors are discussed in the literature.2 MRS is a syndrome in which the diagnosis is set with clinical symptoms, with no specific diagnostic test; it is rarely seen and difficult to treat.2 Herein, we report the case of an MRS patient who presented with lower motor facial paralysis repeated three times, fissured tongue, repetitive episodes of edema in the lips and hemifacial spasm. This is the first case of a patient with the MRS triad, accompanied by hemifacial spasm.

CASE REPORT

In September 2013, a 49-year-old female patient presented to our clinic due to suddenly developed edema of the lower lip and motor facial paralysis on the left side (Figure 1). It was learned from her medical history that she had experienced motor facial paralysis on the right side during pregnancy in 1998 and left lower side motor facial paralysis in 2003. Her right side facial paralysis had resolved with sequel and she had hemifacial spasm on the right side. The patient had experienced preeclampsia during the pregnancy period; Factor V Leiden mutation, activated protein C resistance, and low levels of protein S had also been determined. She was receiving antihypertensive and antiaggregan therapy. She was on paroxetine due to panic attacks. It was learned that she had eaten eggplant prior to the left lower motor facial paralysis and labial edema attack in her face. The patient has four siblings; the fraternal twin sibling of the patient had experienced facial paralysis and all her siblings had cold allergy. In the patient's neurological examination, left lower motor facial paralysis, edema in the lower lip, right hemifacial spasm and very pronounced fissured tongue were identified (Figure 2).

Cerebral magnetic resonance imaging (MRI), cerebral magnetic resonance angiography and cerebral magnetic resonance venography were normal. The patient did not have clinical symptoms of Crohn's disease, sarcoidosis or tuberculosis. Her chest radiograph was normal. Serum ACE and calcium levels were within the normal range.
Figure 1: Marked edema of the lower lip and lower motor facial paralysis on the left side.

Figure 2: Remarkable fissured tongue in the patient.

Borrelia Burgdorferi IgG testing was negative. Her antinuclear antibodies were positive.

Edema in the lower lip of the patient improved following prednisolone therapy, which was initiated at 1 mg/kg/day and gradually decreased, and valacyclovir therapy of 1000 mg two times a day. Left lower motor facial paralysis resolved almost completely within two months. Right hemifacial spasm of the patient continued. The patient refused the use of botulinum toxin and carbamazepine for the right hemifacial spasm.

The patient presented again to our clinic due to edema on the right side of the lower lip (Figure 1). Labial edema developed immediately after encountering the cold weather and eating cold yogurt. Edema was regressed following antihistamine (desloratadine) and prednisolone with the dose of 1 mg/kg/day (withdrawn by gradually decreasing).

Biopsy from the edematous lip was recommended in both presentations of the patient. However, the patient refused the biopsy.

DISCUSSION

MRS is defined as a noncaseating granulomatous disease with unexplained etiology and pathogenesis. In etiology genetics, infectious and immunologic factors may play a role. Infection, autoimmunity, neurotrophic factors, atopy and hypersensitivity to food additives may be effective in the pathogenesis of the disease, but effects of these factors have not been definitive as etiologic agents. Familial tendency has been reported. Although MRS is regarded as an autosomal dominant trait, it is yet to be proven. In our reported case, the lower motor facial paralysis was found in the twin and the other siblings had a history of atopic constitution.

Incidence of MRS is 0.08%. In some publications, this rate is reported to be higher in females. It is more common in the second and third decades of age. Clinical presentation of MRS may occur with one or more symptoms of orofacial edema, facial paralysis and fissured tongue. MRS as the triad has been reported in 10%–20% of the cases and the most common form of presentation is orofacial edema by 80%–100%. Edema is usually unilateral and most often occurs in the upper lip. Edema may be seen in cheeks, palate, gingiva, tongue, pharynx, larynx, and periorbital region. Facial paralysis, which is rarely bilateral, is observed in 47%–90% of cases and it repeats in 10% of the cases. Fissured tongue is seen in 40% of cases. Our case is the first report of hemifacial spasms accompanied by the classical triad of MRS with recurrent episodes of facial paralysis, recurrent labial edema and fissured tongue.

Association of persistent labial edema and hemifacial spasm was reported in a single case in the literature. It was reported in the labial biopsy of this case that granulomatous lesions and other inflammatory lesions were not found and lymphatic vessels were ectasic.

There is no specific diagnostic test for MRS. Computed tomography and MRI can be used for differential diagnosis. In our case, cerebral MRI examinations were normal.

Following swelling of the lips permanent indurations may develop in the lips, and these may lead to significant cosmetic defects, causing eating and speaking problems. The treatment of MRS has not been fully determined. Various treatment modalities are being tested. Corticosteroids, tetracycline, doxycycline, clofazimin, thalidomide, infliximab, and adalimumab have been tried.

MRS should be kept in mind when recurrent facial palsy, orofacial edema, and fissured tongue are seen as monosymptomatic or as a triad. Protective recommendations should be offered to patients since infection, autoimmunity, neurotropic factors, atopy, and food hypersensitivity might play roles in the etiology.
CONCLUSION

As a conclusion, we presented the case diagnosed as MRS by the symptoms of the recurrent episodes of facial paralysis, labial edema, and lingual fissure, accompanied by hemifacial spasm. This is the first such case to be reported in the literature.

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