CASE REPORT

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Melkersson-Rosenthal Syndrome

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ABSTRACT

Melkersson-Rosenthal syndrome (MRS) is manifested by recurrent edema in the region of the face or oral cavity, paralysis of facial muscles and fissured tongue. It is a rare, granulomatous disease of unknown cause. We report a case of a patient with the complete clinical triad of MRS symptoms.

KEY WORDS: Melkersson-Rosenthal syndrome, nervus facialis, facial edema, fissured tongue

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Introduction

Melkersson-Rosenthal syndrome (MRS) or Miescher-Melkersson-Rosenthal syndrome is a rare neurological disease manifested by a triad of symptoms: recurrent (unilateral or bilateral) peripheral facial nerve palsy, relapsed swelling of one side of the face, eyelids or lips and fissured tongue.

It was first described in 1928 by Ernst Gustav Melkersson who described a paralysis and facial edema, whilst in 1932 Curt Rosenthal added lingua plicata or fissured tongue as an additional symptom to the previously mentioned relapsing symptoms¹. In 1945 Miescher introduced several cases of primary interstitial inflammation of the lips, along with the abovementioned symptoms, and named them macrocheilitis granulomatosa.²

Case Report

A 32-year-old female patient (she gave a written authorization to use her medical information and photographs) was hospitalized at the Neurology Clinic, University Hospital – Clinical Centre of Banja Luka because of the recurrent bilateral peripheral facial palsy, swollen lower lip (Figure 1), fissured tongue (Figure 2) and a headache.

Anamnestically we received the information that she had left-sided peripheral facial nerve palsy first time at the age of 25, during her pregnancy, but it withdrew spontaneously without neurological deficit. Five years later (at the age of 30) she had a right-sided peripheral facial nerve palsy, which was treated with antibiotics and corticosteroids, and this, together with the physical therapy, led to the partial improvement. Eighteen months later right-sided peripheral facial nerve palsy recurred and six months after that the left-sided peripheral facial nerve palsy also recurred, this time followed by taste disorder, pronounced salivation and tearing, as well as the periodic headache on left side of migraine character. The last attack occurred three months before she was hospitalized in our clinic. After oral corticosteroid therapy and physical treatment there was a small improvement of the weakness of the mimic muscles. The intensity of a headache significantly decreased in the last recide of the disease with the use of tricyclic antidepressants, and after a month it completely stopped, while in the same period, both hypersalivation and epiphora withdrew, and the sense of taste got normalized. Swelling of the lower lip occurred on several occasions, regardless the weakness of the mimic musculature, and was sometimes accompanied by the feeling of general weakness and febrility, and after the second recide there was a more voluminous lower lip of harder consistency then the previous. Fissured tongue was noticed in early childhood.

Figure 1. More voluminous lower lip

Figure 2. Melkersson-Rosenthal syndrome-fissured tongue

The family anamnesis gave significant information regarding the father who, at the age of 40, had a peripheral facial nerve palsy, which didn't occur again, and he is also the only one in the family, apart for our patient, with a fissured tongue. The personal anamnesis showed neither previous diseases, operations, injuries nor allergic reactions.

Within the diagnostic process recommended by the authorized neurologist, laboratory analysis were conducted at the medical clinic where complete blood count was made on several occasions, as well as the biochemical analyses and inflammation parameters, which were normal. IgM and IgG antibodies to *Borrelia burgdorferi* in serum by Western blot method were negative, and the level of serum angiotensin-converting enzyme was normal. Otorhinolaryngological, ophthalmological and the internist's tests were all normal on several occasions.

Biochemical blood analysis, thyroid hormonal status, cytochemical analysis of the cerebrospinal fluid and immune analysis of serum done during the hospitalization at our institution were within the limits of the reference values and negative. Abdominal ultrasound examination and x-ray of the heart and lungs were NAD. Electromyoneurographic findings showed a hard lesion of the facial nerve to the right of the foramen stylomastoideum with the damage of the fibers m. orbicularis oris and m. orbicularis oculi of strong degree and in the same regions on the left side, the changes of a modest degree were registered. Visual evoked potentials showed normal results, while brainstem auditory evoked potentials showed normal results after stimulation to the left side and possible disturbance in the area of the brainstem after stimulation to the right side. MR endocranium and cervical spine MR were conducted two times in 2012 and in 2013 and showed normal findings.

The neurological examination of the patient (7 years after the first symptoms) found a bilateral peripheral facial nerve palsy more on the right side with the presence of Bell's phenomenon and the synkinetic movements in the form of the removal of the right nasolabial fold while closing eyes, and opening of an eye while showing teeth, on the same side. A weak contraction of the platysma is bilaterally registered, more on the right side. Tongue examination showed a fissured tongue with dominant groove/fissure in the middle and pronounced transverse grooves, as well as the discrete tremors on protrusion. The lower lip is more voluminous, dominant in the central area, with a discrete hyperpigmentation in that area, and also of harder consistency in response to palpation, in comparison to the upper lip. The rest of the neurological finding was normal.

Discussion

MRS is a rare neurological disease that affects both sexes, most commonly in the second or third decade of life, but there have been cases when patients had their first symptoms in childhood or when they were older than 50.² Information on incidence is inconsistent but is about 0.3 per 100 000 patients per year.³

It is manifested by a triad of symptoms: recurrent (unilateral or bilateral) peripheral facial nerve palsy, recurrent swelling of one side of the face, eyelids or lips (cheilitis granulomatosa-Miescher cheilitis) and fissured tongue (lingua plicata). In only 25% of cases all three symptoms are present at the same time, which, with the variation of the symptoms mentioned through the time (from several days to several years), makes the diagnostic process much harder.⁴ So far, the etiology is not clear enough, but it is assumed that it may be a genetic predisposition, infectious cause or an immune response to some of the agents from the outside.

In patients with the sign of a fissured tongue it is assumed to be a congenital disease since this characteristic is present in several members of the same family and without other symptoms.⁵

It is often coexistent with Crohn's disease and sarcoidosis, as well as with Hashimoto's thyroiditis.⁶ Histopathologically, it is a granulomatous angiitis with perivascular inflammatory cells and accompanying tissue edema.⁷

The diagnosis is made based on the clinical features and by excluding other autoimmune and infectious diseases with similar symptoms and, in cases that are still not clear, it can include a biopsy and pathohistologic analysis of the affected tissue and the genetic analysis.⁸

MRS has a chronic course accompanied by the relapses and remissions, with occasional spontaneous recovery, aided by certain symptomatic procedures.⁹

Since the etiology is not known enough, the therapy includes a local and/or systemic use of corticosteroids, analgesics, in rare cases immunosuppressives and antibiotics, as well as the physical treatment.

There have been successful examples of treating swelling of the lips by the local parenteral application of betamethasone and doxycycline with the previous application of anesthetics, once a month, during the period of three years, as well as by the use of adalimumab in the recent times.¹⁰ If the swelling of the eyelid is severe, does not react on therapy, functionally disturbs a patient or if it is unacceptable in cosmetic terms, a surgical treatment like aesthetic correction can be applied, but also a facial nerve decompression especially in the cases in which, by neurophysiological methods, we can detect the place of compression and when there is a denervation.^{11,12}

With this report we wanted to indicate the MRS as one of the possible causes of bilateral, peripheral damage of the facial nerve, which is necessary to distinguish from some other possible causes of this disorder such as multiple sclerosis, neuroborreliosis, neurosarcoidosis and acute polyradiculoneuritis. Apart for the normal findings of laboratory and neuroradiological analyses, a characteristic triad of symptoms points to the MSR as an independent entity.

It is also important to pay attention to this disease because it is highly likely that, apart for the neurologists, there are other doctors of different fields of specialty participating in its diagnosis and treatment (ORL specialists, pediatricians, ophthalmologists, dermatologists, rheumatologists, physiatrists) and that, given the low incidence, unclear etiology and uncertain outcome of the natural course of the disease, the early detection and the start of the symptomatic therapy is of great importance for the better functional recovery.

References

1. Rana AP. Orofacial granulomatosis: A case report with review of literature. *J Indian Soc Periodontol.* 2012;163:469-74.
2. Frąckowiak L, Wroński K, Kańczuga-Koda L, Koda M. Melkersson-Rosenthal syndrome in a patient with Hashimoto disease. *Prog Health Sci* 2014; 4:269.
3. Kanerva M, molilanen K, Vholainen S at al. Melkersson-Rosenthal syndrome. *Otolaryngol Head Neck Surg* 2008; 138:246-51.
4. Pierre-Filho Pde T, Rocha EM, Natalino R, Cintra ML, Caldato R. Upper eyelid oedema in Melkersson-Rosenthal syndrome. *Clin Experiment Ophthalmol* 2004;32:439-440.
5. Chen C, Huilgol SC, James C, Selva D. Melkersson-Rosenthal syndrome presenting with upper lid edema and facial palsy. *Can J Ophthalmol.* 2002;37:361-363.
6. Scagliusi P, Sisto M, Lisi S, Lazzari A, D'Amore M. Hashimoto's thyroiditis in Melkersson-Rosenthal syndrome patient: casual association or related diseases?, *Panminerva Med* 2008;50(3):255-7.
7. Cockerham KP, Hidayat AA, Cockerham GC, Depper MH, Sorensen S, Cytryn AS, et al. Melkersson-Rosenthal syndrome: new clinicopathologic findings in 4 cases. *Arch Ophthalmol* 2000;118:227-232.
8. Kajal B, Harvey J, Alowami S. Melkersson-Rosenthal Syndrome, a rare case report of chronic eyelid swelling. *Diagn Pathol* 2013; 8:188.
9. Ates O, Yoruk O. Unilateral anterior uveitis in Melkersson-Rosenthal syndrome: a case report. *J Int Med Res* 2006;34:428-432.
10. Oudrhiri L, Chiheb S, Marnissi F, Zamiati S, Benchikhi S. Successful treatment of Miescher's cheilitis in Melkersson-Rosenthal syndrome with betamethasone injections and doxycycline. *Pan Afr Med J* 2012; 13: 75.
11. Glickman LT, Gruss JS, Birt BD, Kohli-Dang N. The surgical management of Melkersson-Rosenthal syndrome. *Plast Reconstr Surg* 1992; 89:815-821.
12. Feng S, Yin L, Jadong H, et al. Melkersson - Rosenthal syndrome: a retrospective study of 44 patients. *Acta Oto-Laryngologica* 2014; 134:977-981.

Melkersson-Rosenthal sindrom

SAŽETAK

Melkersson-Rosenthal sindrom (MRS) manifestuje se rekurentnim edemom u regiji lica ili usne duplje, paralizom mišića lica, izbrazdanim jezikom. To je rijetka granulomatozna bolest nepoznatog uzroka. Prikazali smo slučaj pacijentice sa kompletnom kliničkom trijadom simptoma MRS-a.

KLJUČNE RIJEČI: Melkersson-Rosenthal sindrom, nervus facialis, edem lica, izbrazdan