

Peripheral Facial Palsy in the Setting of Melkersson-Rosenthal Syndrome – Case Report

Paralisia Facial Periférica no Contexto de Síndrome de Melkersson-Rosenthal – Relato de Caso

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ABSTRACT

Melkersson-Rosenthal syndrome is characterized by recurrent peripheral facial palsy, recurrent or persistent orofacial edema and a fissured tongue. However, this clinical triad occurs only in a minority of cases; mono or oligosymptomatic forms are much more common. The authors describe a case report of a 39-year-old woman with Melkersson-Rosenthal syndrome, discuss the evolution of the disease and treatment, highlighting the need of a high clinical suspicion index and a regular follow-up to reduce progression of deficits.

Keywords. Peripheral Facial Palsy, Melkersson-Rosenthal Syndrome, Blepharospasm.

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RESUMO

A síndrome de Melkersson-Rosenthal é caracterizada por episódios de paralisia facial recorrente, edema orofacial persistente ou recorrente e língua fissurada. Todavia, esta tríade clínica apenas está presente numa minoria de casos; formas mono ou oligossintomáticas são muito mais comuns. As autoras descrevem um caso clínico de uma mulher de 39 anos com síndrome de Melkersson-Rosenthal, discutem a evolução da doença e tratamento, colocando em destaque a necessidade de elevada suspeita clínica e de um acompanhamento regular para reduzir a progressão dos défices.

Unitermos. Paralisia Facial Periférica, Síndrome de Melkersson-Rosenthal, Blearospasmo.

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INTRODUCTION

Melkersson-Rosenthal syndrome is a noncaseating granulomatous disease characterized by recurrent peripheral facial palsy, recurrent or persistent orofacial edema and a fissured tongue¹⁻⁶. The three typical clinical manifestations of this syndrome can occur either simultaneously or at different times⁷. However, the full clinical triad occurs only in a minority of cases¹⁻³. Mono or oligo-symptomatic forms are much more common^{1-4,6,7}, so this syndrome is frequently under diagnosed⁸.

Its etiology remains unknown, though genetic, allergic and immune factors as well as infectious and traumatic causes have been implicated. It is sometimes described associated to systemic diseases such as Crohn disease⁵.

Although it can affect people of any age or gender, it is more common in women and in the second decade of life^{5,8}.

Its diagnosis is mainly clinical, suggested by history and physical examination. However, the definitive diagnosis is obtained with electrophysiological and imaging studies and biopsy of facial swelling areas.

CASE PRESENTATION

A 39-year-old woman with a past medical history of a recurrent left peripheral facial palsy was referred to Neurology consultation because of blepharospasm in the right eye. Health Ethic Committee of Hospital São João (#2011-07-07).

Besides blepharospasm in the right eye and residual facial asymmetry resultant from peripheral facial palsy, she complained of involuntary movements of the face and both eyes which were aggravated by stress and anxiety and had a negative psychological impact.

She had the first episode of peripheral facial palsy when she was 11 years-old and did physical therapy. She also had recurrent episodes of orofacial edema since she was 19 years-old. On physical examination, she presented slight left peripheral facial palsy (House-Brackmann scale grade II), blepharospasm in the right eye, spontaneous and brief involuntary muscle contractions in the left eye and left lower face with the appearance of wormlike rippling of the muscle, consistent with myokimias, a slight upper lip edema with local skin changes (figure 1) and

a fissured tongue. She had neither compromise of other cranial nerves nor systemic complaints.

She underwent an extensive analytic study, including serologies (EBV, Borrelia, syphilis), viruses markers (HBV, HCV, HIV) and immunological tests, which were within normal limits. Brain MRI and thoracic radiography were normal. She also performed an upper lip biopsy that demonstrated a granulomatous cheilitis. In electrophysiological studies, she demonstrated a pattern compatible with myokimias in electromyographic study of *orbicularis oris* bilaterally and signs of chronic axonal damage of both facial nerves, consistent with a disease affecting both sides of the face.

She was treated with anti-inflammatory drugs and topical application of hyaluronic acid to alleviate the skin lesions and with oral oxcarbamazepine to decrease myokimias of face and eyes. Injections of botulinum neurotoxin type A were administered in the right *orbicularis oculi* muscle. This treatment resulted in a reduction of her blepharospasm.

DISCUSSION

Melkersson-Rosenthal syndrome is a hypothesis that should be always considered in the differential diagnosis of recurrent peripheral facial palsy.

In this syndrome, peripheral facial palsy can occur from months to years either before or after the beginning of orofacial swelling. It can be unilateral or bilateral, partial or complete. It appears suddenly and it is clinically indistinguishable of Bell's palsy^{7,9}. Facial weakness can be accompanied by taste or hearing disorders, otalgia or



Figure 1. Slight sequelae of left peripheral facial palsy and upper lip edema with local skin changes.

salivary hyposecretion³. Orofacial edema is the dominant feature of Melkersson-Rosenthal syndrome, affecting mainly the upper lip⁹. It can be either unilateral or bilateral, occurring in the same side of the facial palsy. After several episodes, it can persist indefinitely, causing facial deformities. Fissured tongue (also called *lingua plicata* or scrotal tongue) is the least frequent clinical feature, is not pathognomic³ occurring in general population without meaning disease^{3,10}. Other findings associated with Melkersson-Rosenthal syndrome include: trigeminal neuralgia, ocular palsies, paresthesias, blepharospasm, lachrymal and salivary gland dysfunction, epiphora, keratitis, psychotic episodes and migraine^{7,11}.

Initially, facial palsy tends to complete recovery within three weeks even without medicines (systemic corticoids like prednisone, which can help reduce facial nerve edema and consequently symptoms)¹². As disease progresses, facial palsy episodes increase in frequency and duration and may cause residual paresis or sinkinesias. After several relapses, permanent deficits of facial mobility may lead to difficulties in communication or feeding; esthetic changes may appear with potential negative psychological impact¹³. Therefore, it is necessary to rapidly reestablish facial mimic through physical therapy¹⁴, including neuromuscular proprioceptive facilitation techniques and therapeutic modalities such as orofacial massage, ice massage, facial muscle exercises in front of mirror (biofeedback) and strengthening of facial muscles – essentially the same treatment patterns used in Bell's palsy. Surgery can be an option for patients with recurrent or persistent palsies in whom medical and physical treatments are ineffective, but there is only very low quality evidence¹⁵.

Worth mentioning in this case report is that besides presenting the typical clinical triad, the patient had focal dystonia in the right eye, blepharospasm and myokimias of the left face due to facial nerve damage progression.

Melkersson-Rosenthal syndrome has to be considered in the differential diagnosis of recurrent peripheral facial palsy. These patients need a regular follow-up, given the chronic and progressive character of the disease.

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