case report

Melkersson-Rosenthal syndrome with partial oculomotor nerve palsy

Mehmet Ufuk Aluclu,[a] Ugur Keklikci,[b] Aslan Guzel,[c] Kaan Unlu,[b] Mehmet Tatli[c]

From the Departments of [a]Neurology, [b]Ophthalmology, [c]Neurosurgery, Medical School, Dicle University, Diyarbakir, Turkey

Correspondence and reprints: Dr. Mehmet Ufuk Aluclu · Department of Neurology, Dicle University, Medical School, Diyarbakir 21280, Turkey · aluclu@dicle.edu.tr · Accepted for publication July 2007

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Melkersson-Rosenthal syndrome (MRS) is a rare granulomatous disease characterized by orofacial edema, facial nerve palsy, and furrowed tongue (lingua plicata).[1,2] This is the classic triad which defines the syndrome, although it is accepted that the presence of two manifestations or one with a granulomatous cheilitis in the eyelid biopsy, is sufficient to make the diagnosis.[3] Facial paralysis, most commonly unilateral, may also occur as well as a congenital fissuring of the tongue. It was first described in 1928 by Melkersson as a syndrome of recurrent facial palsy and edema, and Rosenthal later added the third feature of the syndrome, furrowing of the tongue, in 1931.[4] The etiology of MRS is unknown, although both genetic and acquired factors have been implicated.[2,5] Isolated eyelid edema, particularly bilateral involvement, is also rare, and as such it is usually misdiagnosed as orbital tumor, orbital pseudotumor, thyroid ophthalmopathy, or blepharochalasis.[1,6,7] In this study, we report a case of MRS that presented with diffuse facial swelling, left facial palsy and restricted muscle movement in the dextroelevation of the left eye.

CASE

A 15-year-old male, whose family history was unremarkable, was suffering with persistent upper facial edema and diplopia. The patient had peripheral facial palsy (lower motor neuron facial palsy) 3 months previously and received treatment with prednisolone, but there was no improvement. The patient's general condition was good. On neurological examination, bilateral lids in both the upper and lower regions were involved, with edema and chemosis and the presence of lingua plicata (Figure 1, 2). His best-corrected vi-

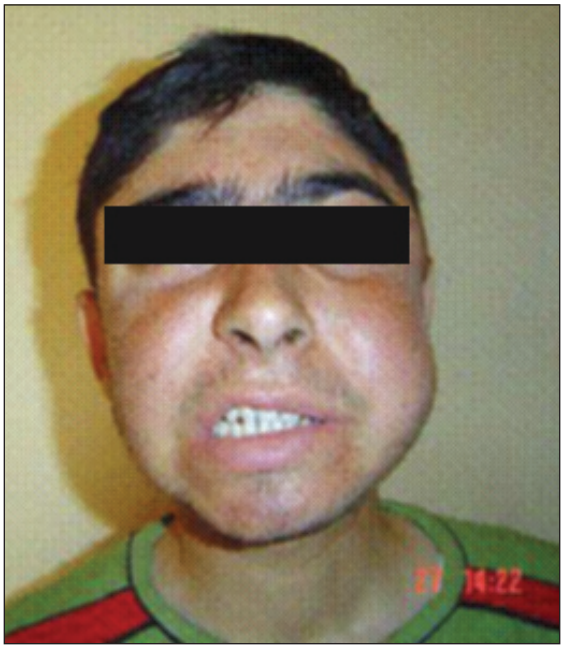

Figure 1. Left facial edema with facial nerve in paresis.

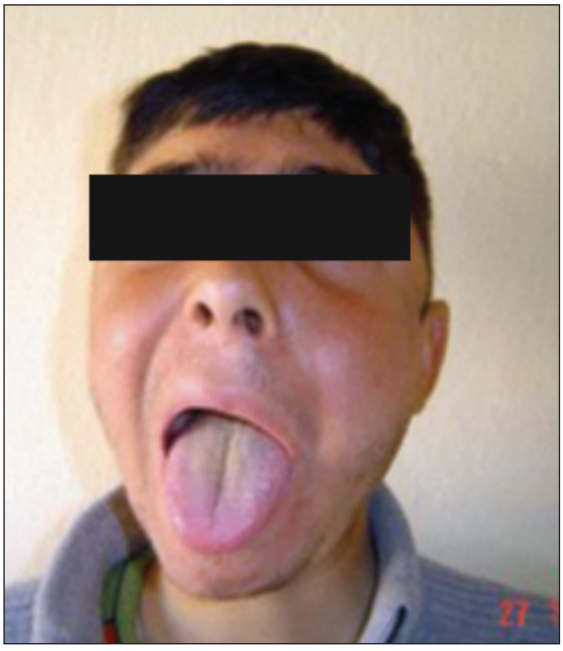

Figure 2. Furrowed tongue of the patient.

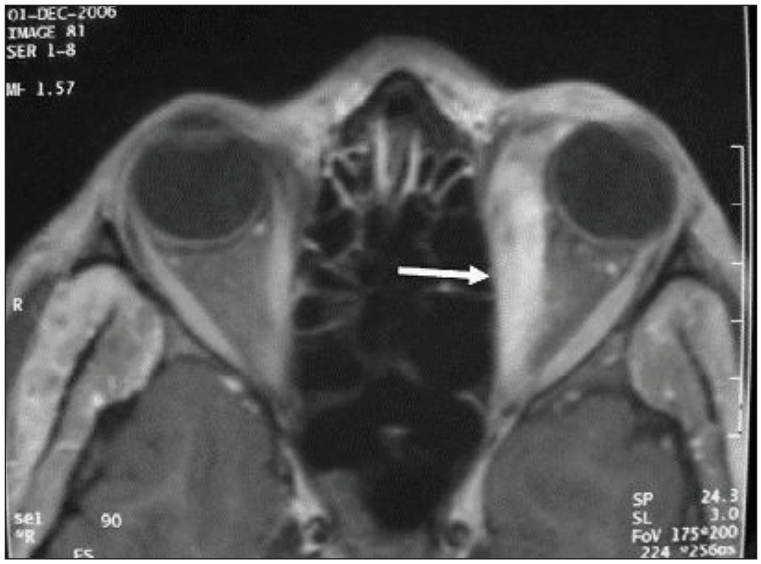

Figure 3. MRI of the orbit showing an increase in signal (arrow) on the left medial rectus muscle.

sual acuity was 20/20 OU. There was a restriction of muscle movement in the dextroelevation of the left eye. Laboratory investigations, including a complete blood cell count with differential cell count, electrolytes, erythrocyte sedimentation rate, thyroid function panel, antinuclear antibody, rheumatoid factor, and serology for syphilis, were within normal limits. An MRI of the head was normal, but the orbital MRI demonstrated a hyperintense lesion extending from the left globe in the surrounding orbital tissue and abutting the left medial rectus muscle. After administrating gadolinium, a strong heterogeneous enhancement was determined in the left medial rectus muscle (Figure 3). We administered prednisolone (80 mg/d for 5 days followed by 70 mg/day for 5 days, and followed by 10 mg reduction for each week), which reduced the eyelid edema. However, there was restricted improvement in movement of the left medial rectus muscle.

DISCUSSION

MRS is a rare granulomatous disease.[1,2,8,9] The incidence of this syndrome was reported as 0.08 %.[10] Male and female patients are equally affected and adolescence is the most common time of presentation.[4,11] The pathogenesis of MRS remains uncertain, but several theories have been proposed including heredity, chronic infection, allergy and hypersensitivity to bacteria.[12] The diagnosis of MRS is often difficult.[13] All three features of the classic triad are rarely encountered at the time of presentation, which makes the diagnosis of this rare entity more difficult.[5] Our case presented with the complete clinical triad of MRS, consisting of bilateral upper eyelid edema, facial palsy, and a furrowed tongue.

The dominant feature of this syndrome is facial edema.[8,14] The swelling in the current case did not involve the lips or cheeks. Isolated eyelid edema is uncommon.[1,7] The major differential diagnoses with isolated lid edema is angioedema, thyroid orbitopathy and blepharochalasis syndrome. The eyelid swelling of angioedema is usually pruritic, migratory, and resolves relatively quickly; additionally, abnormal serum levels of C1 esterase inhibitor and of components of the complement cascade may be present in affected patients. In thyroid orbitopathy, eyelid retraction and lagophthalmos often accompany bilateral eyelid edema. The skin may be erythematous, but is not thickened. Blepharochalasis is characterized by remitting and relapsing eyelid edema.[13]

Facial nerve palsy has been reported in 30% to 90% of MRS cases and may be seen months to years before or after the onset of facial swelling and may not be distinguished from Bell's palsy.[14,15] The palsy may be unilateral or bilateral and simultaneous with the facial swelling in 13% to 50% of cases.[4,16] The usual rate of recurrences is about 10%.[17] In the present case, left facial nerve palsy had occurred three months before the onset of eyelid swelling and it recovered partially. The other factor in the triad, lingua plicata, is not specific to the syndrome and may be congenital (40%).[4,5]

CT and MRI may be useful in the differential diagnosis, such as thyroid orbitopathy.[12] In the literature, MRI findings of the facial nerve in patients with MRS have been described as enhancement in the intratemporal part of the nerve.[18] Although the facial nerve findings of our case were normal in the head MRI, the orbital MRI demonstrated an increase in signal extending from the left globe in the surrounding orbital tissue and abutting the left medial rectus muscle. After gadolinium, the left medial rectus muscle showed strong heterogeneous enhancement on T1-weighted section.

Rarely, other cranial nerves including the olfactory, oculomotor, auditory, glossopharyngeal, vagus and hypoglossal nerves may also be affected.[5,20] The course of the paralysis may be unilateral or bilateral, and the paralysis may be partial or complete.5,20 In our case partial third cranial nerve palsy was due to diplopia and the restriction of movement of the left medial rectus muscle. If the third cranial nerve is involved, there is ptosis, or paralysis of gaze. Associated ophthalmic findings include retrobulbar neuritis, lagophthalmos, corneal opacity, and keratoconjunctivitis sicca.[15]

The management of MRS is difficult. Reported therapies include corticosteroids, minocycline, clofazimine, sulfasalazine, hydroxychloroquine, methotrexate and

thalidomide, but their efficacy is not proven.[2,13,19] Rare cases of spontaneous remissions have been reported.[21] Our case showed some improvement; the swelling was reduced after the initiation of oral corticosteroid.

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