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Case Report

# Melkersson–Rosenthal Syndrome: A Case Report

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## ABSTRACT

Melkersson-Rosenthal Syndrome (MRS) is a rare disease characterized by the triad of recurring facial paralysis, a fissured tongue, and recurring swelling of the lips and/or face. The etiology is unknown; it may be caused by hereditary factors or conditions occurring later in life. Observation of all 3 of the classic manifestations is rare; most often the symptoms are seen alone or in a pair, and they may appear at any stage of life. This report is the description of the case of a 51-year-old woman who was diagnosed with MRS based on her history and clinical features.

## INTRODUCTION

Melkersson-Rosenthal Syndrome (MRS) is a granulomatous disease characterized by recurring peripheral facial paralysis, swelling of the lips and/or face, and a fissured tongue. The median reported incidence is 0.08%,[1] and it is most frequently seen in the age range of 20 to 40 years.[2] Monosymptomatic or oligosymptomatic varieties are more often seen than the complete classic triad. The symptoms may also be observed separately in later life.

Though its etiology is not precisely known, it is thought that various infections, delayed susceptibility reactions, stress, and autoimmune and genetic factors may be related to the etiopathogenesis.[1–8] The diagnosis can be made directly based on the presence of the full triad of symptoms; when only 1or 2 are observed, a skin biopsy from the facial edema should be performed and demonstrate granulomatous cholestasis pathologically.[9]

Steroids, anti-inflammatory agents, antibiotics, and immunosuppressants may be used in treatment; however, spontaneous recovery also occurs. Reconstructive surgical procedures can be performed in the event of permanent deformities due to orofacial edema and facial paralysis.[1,10,11]

The aim of this report is to create awareness for all physicians, particularly those in the fields of otolaryngology, neurology, dermatology, and pediatrics, who may encounter patients with MRS in clinical practice.

## CASE REPORT

A 51-year-old female patient presented at the polyclinic with complaints of her mouth drooping to the left, swelling in the right half of the face, and an inability to close the right eyelid. Her history revealed that she had experienced peripheral facial paralysis 3 times before. A physical examination indicated that the light reflex on both eyes was +

/ +. Peripheral facial asymmetry was present on the right side (Fig. 1a and b) A mild degree of eye involvement was present. There was weakness in the left half of the face. No sensory loss was observed. She indicated that the facial swelling was initially widespread, though it was now localized to the right side. There were fissures in the tongue but movement was normal (Fig. 1c). Otitis and mastoiditis were not detected. Other system examinations were normal.

The complete blood count, serum biochemistry, antistreptolysin O (ASO), and immunoglobulin level results were within normal limits. A serological evaluation did not detect any abnormality. C-reactive protein and sedimentation values were slightly high. The results of audiometric examinations and a posteroanterior chest X-ray were not remarkable. Diffusion magnetic resonance imaging and temporal bone computed tomography were performed but detected no pathology. Electromyography findings consistent with facial paralysis in the subacute phase were recorded on the right side, and in the chronic phase on the left side. Oral steroid treatment was initiated.

## DISCUSSION

During the evaluation of the clinical and laboratory findings of our patient, no condition that might cause recurrent peripheral facial paralysis, including Bell's palsy, infectious mononucleosis, syphilis, herpes zoster, otitis media, multiple sclerosis, diabetes mellitus, leukemia, myasthenia gravis, Guillain-Barre syndrome, polyarteritis nodosa, or various tumors was detected.

MRS is an extremely rare disease. There is no evidence of racial distribution, but it appears to be more frequent in women.[1,4,7] The classic combination of recurring facial paralysis, a fissured tongue, and recurring swelling of the lips and/or face is observed in only 8% to 25% of MRS patients. Periodic episodes of facial swelling are the most common symptom, seen in 80% to 100% of patients. The facial swelling is generally not painful, ipsilateral, and typically does not have a lasting impact. The upper lip is the the most common site of facial edema. Less frequently, the cheek, palate, gingiva, tongue, pharynx, larynx, forehead, or periorbital regions may be involved. In our patient, this type of edema was present on the right half of the lower and upper lips.

As performed in our case, differential diagnosis should include other conditions that may cause facial edema, including hereditary angioedema, hypothyroidism, superior vena cava syndrome, dental problems, recurrent erysipelas, Crohn's disease, sarcoidosis, mayloidosis, chronic use of angiotensin-converting enzyme inhibitors, lymphangioma, hemangioma, submucosal neoplasms, eosinophilia, lymphoma, chronic herpes labialis, and Uscher syndrome. Pathological examination of the edematous tissue may reveal lymphocyte infiltration, non-caseified epithelial cell granuloma, Langerhans giant cells, mononuclear cells, and fibrosis around the vessels.[1] Etiological factors of infection and allergic reaction, as well as autoimmune and genetic factors were considered, and no related conditions were found in the patient's family history.[4]

Facial paralysis is observed in 47% to 90% of cases. It is most often unilateral, but bilateral involvement has been also reported.[5,7,9] It may resolve or in some cases it may become permanent.[1,11,12] Fissures may be found in the central region of the tongue (central chelitis), at the corners of the mouth (angular chelitis), or in other areas of the lips. In our case, fissures covered the tongue, but were most dense in the center.

Though not seen in our case, possible pathologies associated with MRS include migraine, trigeminal neuralgia, otosclerosis, craniopharyngioma, congenital megacolon, syphilis, Crohn's disease, ulcerative colitis, and psychosis.[11]

There is no single treatment for MRS that will be effective for all patients. A fissured tongue usually does not require

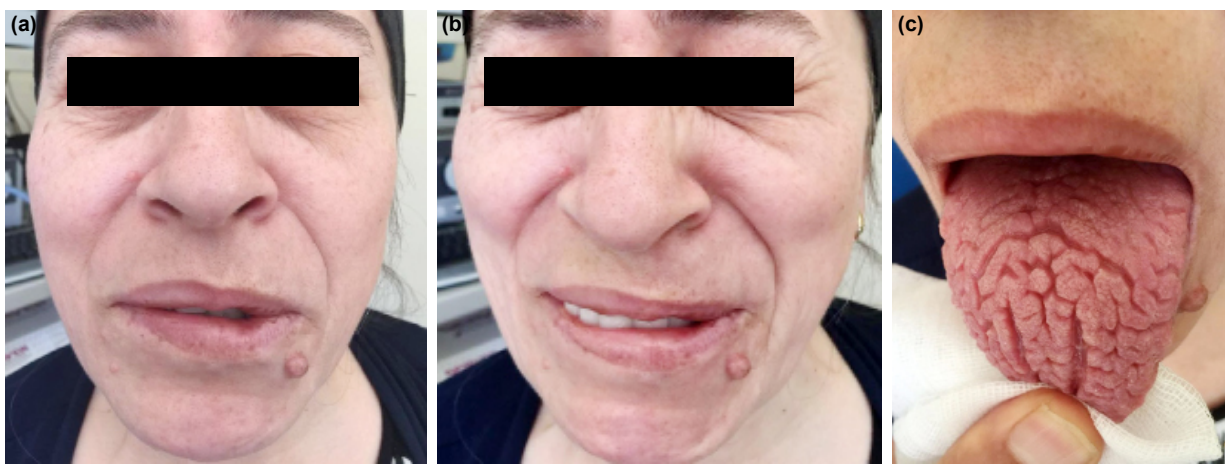


**Figure 1. (a, b)** Peripheral facial paralysis on the left side of the patient's face. **(c)** Fissured tongue with edema of the lip and the right side of the face.

treatment, but short-term systemic steroid treatment may be provided to cure episodes of facial nerve paralysis. Antibiotics and anti-inflammatory agents may be useful for facial swelling, but none have been effective in maintaining full control of the disease.[10] Reconstructive surgery may be considered for patients who develop a permanent deformity; however, surgical interventions should be postponed until after an attack has subsided.

### Informed Consent

Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

### Peer-review

Internally peer-reviewed.

### Authorship Contributions

Concept: S.A..; Design: S.A.; Data collection &/or processing: S.A., S.Ö., S.Ç., A.F., T.Ç.; Analysis and/or interpretation: S.A., S.Ö.; Literature search: S.A., S.Ç., A.F., T.Ç.; Writing: S.A., S.Ö.; Critical review: S.A.

### Conflict of Interest

None declared.

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## Melkersson–Rosenthal Sendromu: Olgu Sunumu


Melkersson-Rosenthal sendromu tekrarlayan fasiyal paralizi, fissürlü dil ve tekrarlayan dudak ve/veya yüzde şişlik triyadı ile kendini gösteren, etiyolojisi tam olarak bilinmeyen ve nadir görülen bir hastalıktır. Kalıtsal veya akkiz etkenlerin yol açabileceği düşünülmektedir. Klasik triadın görülmesi nadir olup çoğunlukla tek ya da iki semptom birlikte görülmektedir. Bulgular hayatın farklı dönemlerinde ayrı ayrı da ortaya çıkabilmektedir. Bu yazıda, öykü ve klinik özellikleriyle 51 yaşında kadın hastada Melkersson-Rosenthal sendromu tanısı alan olgu sunuldu.



**Anahtar Sözcükler:** Fasiyal ödem; fissürlü dil; Melkersson-Rosenthal sendromu; tekrarlayan fasiyal paralizi.