American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2023; 24: e938670
DOI: 10.12659/AJCR.938670

Received: 2022.10.16
Accepted: 2022.12.27
Available online: 2023.01.17
Published: 2023.02.09

Surgical Treatment Outcome for Familial Melkersson-Rosenthal Syndrome

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Financial support: None declared
Conflict of interest: None declared

Patient: Female, 15-year-old
Final Diagnosis: Melkersson-Rosenthal syndrome
Symptoms: Fissured tongue • orofacial edema • recurrent facial nerve palsy
Clinical Procedure: —
Specialty: Otolaryngology

Objective: Congenital defects/diseases

Background: Recurrent facial nerve palsy, orofacial edema, and fissured tongue are a triad of manifestations that characterize a rare disorder named Melkersson-Rosenthal syndrome. It is important to consider this syndrome when diagnosing atypical, unilateral, or bilateral facial palsies with characteristics of familial prevalence. There is no established outcome prediction for this disease and the syndrome does not have a specific duration or prospective timeline. Recurrent facial paralysis can require surgery and a multidisciplinary approach with regular follow-up.

Case Report: We describe a 38-year-old woman presenting with a third episode of facial paralysis and discuss her pedigree chart and the treatment course chosen. After conservative treatment with oral corticosteroids, antiviral therapy, and motor physical therapy with no significant improvements, the patient underwent facial nerve decompression surgery with outstanding results. Eight months after surgery and intense postoperative physical therapy, the patient improved from grade VI to grade II palsy on the House-Brackmann Scale. The patient's older brother also presented a fissured tongue and had a history of 2 episodes of facial paralysis. The patient's son, mother, and sister also presented tongue fissuring but did not have any other clinical signs of the syndrome.

Conclusions: Despite being rare, Melkersson-Rosenthal syndrome is associated with a family inheritance and its diagnosis has prognostic implications. Therefore, it is of the utmost importance to have suspicion of this disorder in order to improve quality of care and target the treatment accordingly. Surgical treatment in these cases seems to be an excellent choice to treat current facial paralysis and prevent further episodes.

Keywords: Facial Paralysis • Melkersson-Rosenthal Syndrome • Microvascular Decompression Surgery • Tongue, Fissured

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/938670

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Background

Melkersson-Rosenthal syndrome (MRS) is a rare and idiopathic disease characterized by the triad of recurrent facial nerve palsy, orofacial edema, and fissured tongue. Two or more of these features are essential for making a clinical diagnosis. The diagnosis is challenging, mostly because it is rare to encounter all 3 characteristics present [1].

The onset is usually in childhood, and relapses are common. Cheilitis granulomatosa is a monosymptomatic variant of this condition. Facial paralysis associated with MRS occurs in 0.36 of 100 000 individuals per year. The condition happens in 33% of MRS cases and can reoccur in 70% of these patients. When recuring, MRS seems to progressively deteriorate the facial nerve [2].

There is no established outcome prediction for this disease, and the syndrome does not have a specific duration or prospective timeline. MRS has already been linked to autoimmune disorders and migraines. Other theories of possible causes, such as genetics, infection, or inflammation, remain unproven [3]. Because of the variations in the manifestations, MRS is very often underdiagnosed. Therefore, it is important to consider MRS when facing atypical – unilateral or bilateral – facial palsies [4].

Recurrent facial paralysis can require surgery and a multidisciplinary approach with regular follow-up [3]. In these cases, when patients usually have sequelae, the focus of the treatment should be to mitigate them and enhance the patient's quality of life. Hence, in this report, we present a case of MRS, highlighting its chosen treatment and outcomes.

Case Report

A 38-year-old previously healthy woman had her first episode of unilateral facial paresis at the age of 15 years, with spontaneous remission. Fifteen years later, she presented with a right-sided second facial palsy episode, which progressed to loss of movement in the hemiface, contractures, and synkinesis. At the age of 38 years, a third episode of facial paralysis occurred, this time on the left side, with a House-Brackman grade of VI (complete paralysis). The patient had a history of frequent migraines for approximately 8 years, along with nausea and dizziness.

After the last paralysis, the patient presented with intense pain throughout the topography of the facial nerve and cervical soreness. There were also clinical signs of intestinal alterations, with complaints of constipation, abdominal pain, and swelling.

The physical examination revealed bilateral asymmetrical facial palsy, with loss of facial wrinkling on the left side, facial edema, and tongue fissuring (**Figure 1A**). The patient could not close her left eye completely or smile. On the right side, she exhibited facial weakening, contractures, and synkinesis (**Figure 1B**).

The hearing thresholds were normal in the audiogram, with a discrete lowering in the frequency of 6 kHz in the right ear (**Figure 2**). Laboratory test results were unremarkable. Serological tests for Lyme disease, syphilis, toxoplasmosis, herpes simplex virus, and herpes zoster virus were negative, ruling out these diseases. Brain magnetic resonance imaging without contrast showed no significant changes. Nerve electrophysiological study confirmed a loss of action potential amplitude bilaterally, which was worse on the left side. The test revealed

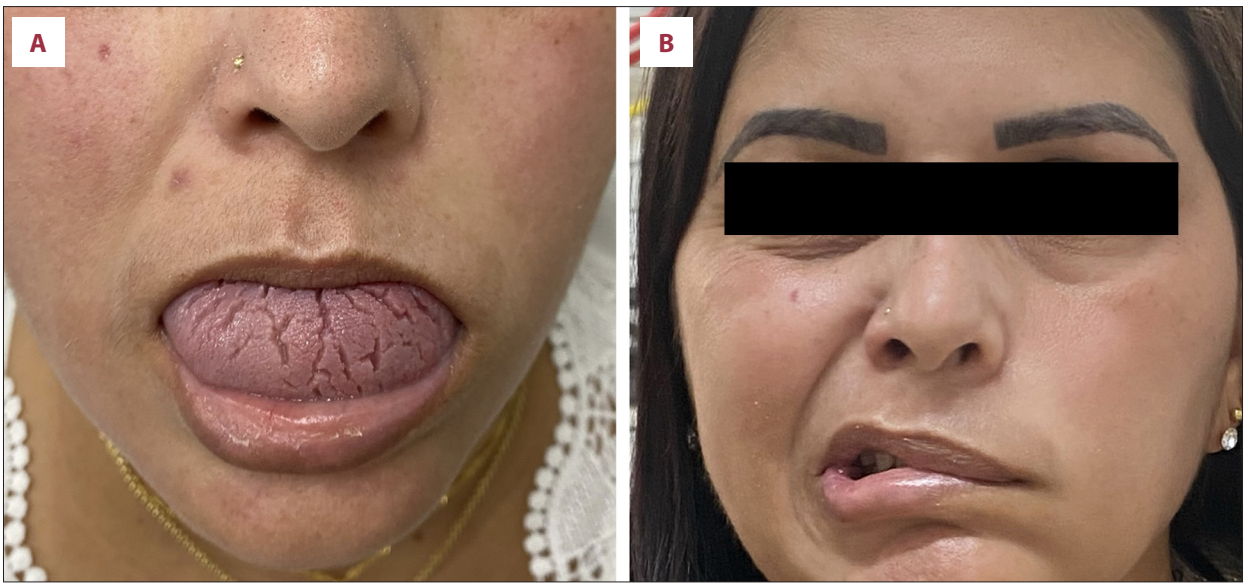

Figure 1. Melkersson-Rosenthal syndrome presenting with (**A**) facial edema and tongue fissuring and (**B**) peripheral facial paralysis.

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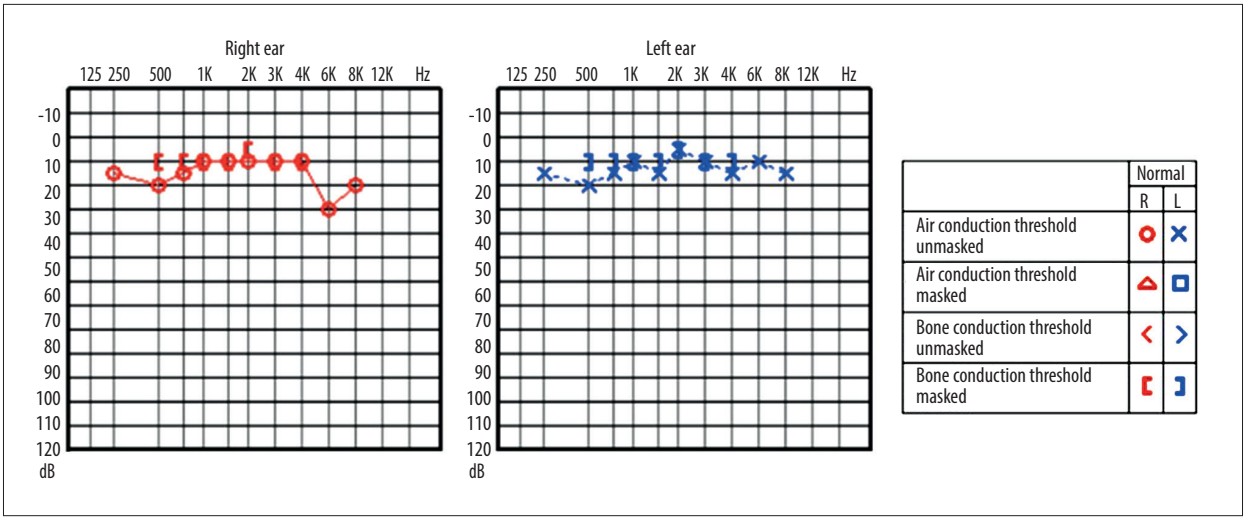

Figure 2. Pure tone audiometry.

Motor nerve conduction study

Site	Latency	Amplitude	Area
Facial, R			
Nasalis	4.92 ms	470.00 uV	1.23 mVms
Frontalis	3.9 ms	780.00 uV	2.47 mVms
Orb. oris	4.2 ms	281.00 uV	275.80 uVms
Facial, L			
Nasalis	5.14 ms	219.00 uV	276.50 uVms
Frontalis	3.84 ms	462.00 uV	1.89 mVms
Orb. oris	3.9 ms	110.00 uV	128.00 uVms

ENM findings summary

Muscle/side	Ins. Act.	Fibs.	Pos. Wave	Fasc.	MYO. Disch	Normal MUP	Poly	Low Amp.	High Amp.	Dur	Recruit	Int. Patt
Frontalis L	Incr.	+1	+1	0	0	0	++	0	+2	Long	Reduce	2
Frontalis R	Normal	+1	0	0	0	0	++	0	+1	Long	Reduce	3

Blink

Stim. Site		R1	R1	Diff	R1 Amp
Left	Ipsi.	10 ms	39.2 ms	3.3 ms	19 uV
	Contra.		35.9 ms		
Right	Ipsi.	13.9 ms	38.6 ms		31 uV
	Contra.				

Figure 3. Facial nerve electrophysiological study.

neuropathy with chronic axonal impairment of both facial nerves with signs of active denervation on the left (**Figure 3**).

The patient was treated with corticosteroid therapy of prednisone 60 mg once a day (1 mg per kilogram per day) for 10 days, followed by a weaning dose, and received antiviral therapy with acyclovir 400 mg every 4 h for 10 days, without improvement. For pain management, trazodone 50 mg was taken at night and pregabalin 75 mg 3 times a day. The patient was also referred to early motor facial physical therapy, consisting of massage, exercises to enhance coordination between both sides of the face, exercises to help with eye and lip closure, and orientations to prevent synkinesis.

The diagnosis of MRS was made because of the patient's history and clinical findings of recurrent peripheral paralysis, facial edema, and fissured tongue. With no significant improvements after initial 10 days of drug therapy and 90 days of physical therapy following the third episode of facial palsy, left facial nerve decompression surgery was indicated. During the surgery, the nerve was released from its bone canal. Intraoperative findings revealed hemorrhagic areas on the

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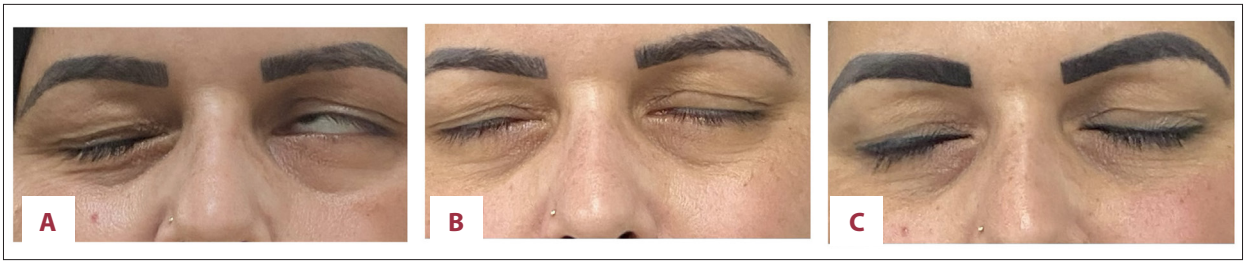

Figure 4. Images of the upper face: (**A**) initial status exhibiting a House-Brackmann grade VI palsy; (**B**) improvement of motor facial function after 1 month of treatment with a grade III palsy; and (**C**) improvement of motor facial function after 8 months of treatment with a grade II palsy.

Figure 5. Facial motor function improvements: (**A**) initial status exhibiting a House-Brackmann grade VI palsy; (**B**) after 1 month of treatment, grade III palsy; and (**C**) after 8 months of treatment, grade II palsy.

left facial nerve, and no other abnormalities were identified in its sheath. We then incised the nerve sheath, decompressing its fibers. At this time, we did not remove any fragment for histopathological purposes.

After the surgery, the patient continued with the facial rehabilitation program. Excellent results in the facial palsy were detected over time. Follow-up 1 month after the surgery revealed an improvement of the facial motor function from grade VI to grade III in the House-Brackmann scale, and after 8 months, the patient improved to a grade II palsy (**Figures 4, 5**). Today, it has been 2 years and 2 months since the surgery. No other episodes of facial paralysis occurred to date, and the patient maintains House-Brackmann grade II. During this time, she presented intermittent self-limited orofacial edema on 3 occasions.

The patient's older brother, who is currently 40 years old, also presented a fissured tongue and had a history of 2 episodes of facial paralysis. His first episode of facial palsy took place 24 years ago, and, now, he experiences sequelae. The patient's 21-year-old son and the patient's 62-year-old mother also presented tongue fissuring but did not have any other clinical signs of the syndrome. The patient's 42-year-old sister also presented a fissured tongue with no facial palsy. However, she had a history of frequent migraines. These features are presented in **Figure 6**.

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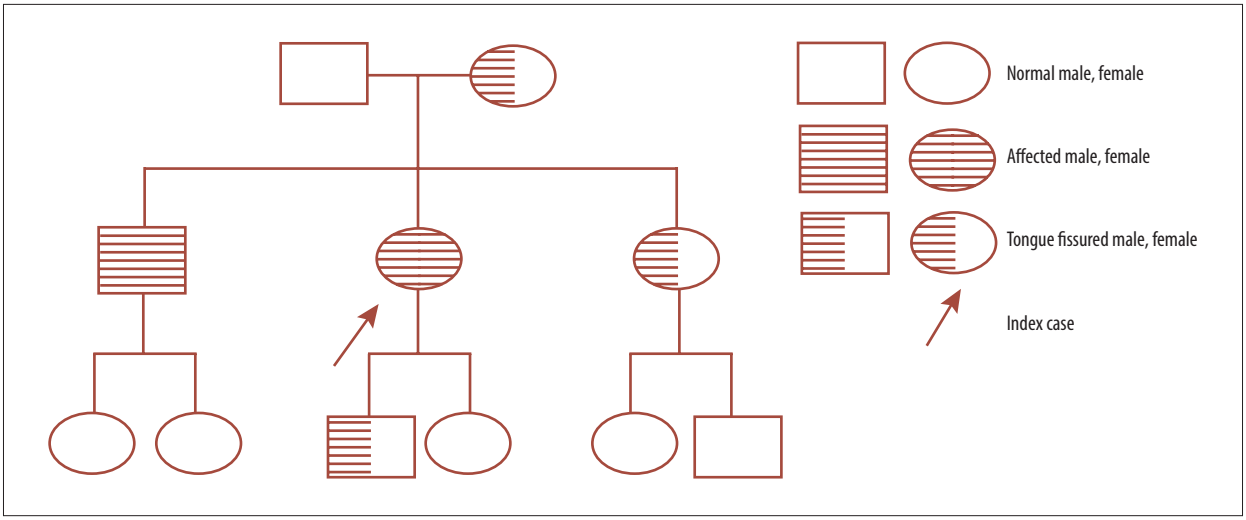

Figure 6. Pedigree chart.

Discussion

MRS is an uncommon otoneurologic disorder that is often underdiagnosed owing to the variability of its clinical signs [3]. However, when facial palsy comes with other infrequent symptoms, it leads to the suspicion of other possible etiologies, indicating the need to investigate syndromes such as the one presented.

The cause for this syndrome is still unidentified, but some hypotheses have been raised. Infectious agents, allergic reactions to various food additives, and autoimmune and genetic factors have been proposed [3,5]. Some researches point to an autosomal dominant relationship in its heritance. The association of MRS with other chronic diseases has not yet been completely elucidated [3].

Since there are no etiological hypothesis proven yet, the target of the treatment focuses on reducing the patient's symptoms. As described in this case, complete facial nerve paralysis can be the final outcome after several recurrent episodes, along with other sequelae, such as synkinesis. Assuming that the pathophysiological explanation for the palsy is nerve swelling, the treatment course can require facial nerve decompression [2,3,6].

Berania et al [6] suggested that delayed decompression could be used to prevent severe continuous facial paralysis for House-Brackmann grades of IV or higher in patients who had not undergone previous surgical procedures. Decompression surgery can still be beneficial after 90 days of symptoms. According to Tan et al [2], evidence shows that total facial nerve decompression can also prevent future episodes of facial paralysis in cases of MRS. Thus, we present a positive outcome after surgical decompression treatment for MRS and reinforce this treatment option as a preferable choice for more severe cases, with optimistic improvements in prognosis. Also, we point out the importance of examining other family members with similar features, which facilitates the diagnosis and allows the identification of undiagnosed generations [5].

Conclusions

MRS is often underdiagnosed, which directly affects prognosis. Atypical cases of facial palsies must be investigated, allowing specialized treatment and proper follow-up. Surgical treatment in these cases seems to be an excellent choice to treat current facial paralysis and prevent further episodes. Future research should include controlled trials comparing conservative treatment versus surgery and also focus on exploring the long-term outcomes after surgical treatment.

Acknowledgements

All of the authors contributed to this manuscript. No financial and other material support was used to its production. The authors declare no conflicts of interest regarding the publication of this paper.

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