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CASE REPORT

Endoscopic trans-canal facial nerve decompression in Melkersson–Rosenthal syndrome: A novel approach

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Key Clinical Message

Melkersson–Rosenthal syndrome can cause recurring bilateral facial paralysis. When steroids fail, surgical decompression of facial nerve is recommended, with endoscopic trans-canal decompression as a safe, minimally invasive, and effective option.

Abstract

Melkersson–Rosenthal syndrome (MRS) is a rare neuro-mucocutaneous disorder, clinically diagnosed by a triad of orofacial swelling, recurrent facial palsy, and fissured tongue. Due to the lack of a comprehensive understanding of MRS, there is no accepted standard of care. In this study we report a 30-year-old female patient, who was referred to the otolaryngology clinic of Rasool Akram Hospital, with classical triad of MRS that was managed by endoscopic trans-canal facial nerve decompression. Bilateral endoscopic trans-canal facial nerve decompression was done when we did not find any improvement with systemic steroids. Endoscopic trans-canal facial nerve decompression could be a safe, reliable minimal invasive treatment of facial paralysis in MRS patients. It needs no external incision or temporal bone drilling which makes this method more convenient for patients with shorter recovery time.

KEYWORDS

Bell's palsy, endoscopic trans-canal facial nerve decompression, Melkerson–Rosenthal syndrome, MRS, recurrent facial palsy

1 | INTRODUCTION

Melkersson–Rosenthal syndrome (MRS) is a rare neuro-mucocutaneous disorder, clinically diagnosed by a triad of orofacial swelling, recurrent facial palsy, and fissured tongue. A minority of patients—between 8% and 18% of cases—present with the complete triad of symptoms, but the most frequent presentations are monosymptomatic and oligosymptomatic. This disorder may cause recurring unilateral or bilateral peripheral facial palsy that is wrongly diagnosed as recurrent Bell's palsy. Orofacial edema is the first symptom of MRS and its most prevalent. About 20%–40% of those who are affected have a fissured tongue, which may have been present since birth;

30% of the patients experience facial paralysis. Usually swelling occurs in the upper lip, but may also be seen in lower lip, tongue, and buccal. Facial swelling has no pain and pitting. When swelling is partial or non-resolved, it may cause to long-lasting deformation of the face and fibrosis.[1]

Histopathology may be used to rule out other disorders including sarcoidosis and Crohn's disease even though MRS is a clinical syndrome and does not require histological evidence to be diagnosed.[1] Multinucleated Langhans-type giant cells, perivascular mononuclear inflammatory infiltration, edema, noncaseating epithelial cell granulomas, and fibrosis are among the distinctive histological features.[2]

According to current knowledge, there are two phases in the development of orofacial edema in MRS: an early, recurrent inflammatory phase and an eventual, quiescent noninflammatory phase. When there had been no worsening of symptoms for at least a year, Worsaee et al. considered the disease to be inactive and in the noninflammatory phase.[3] The inflammatory stage of the disease might be best managed medically. Corticosteroids, nonsteroidal anti-inflammatory medications, clofazimine, antihistamines, and antibiotics are treatments for orofacial edema in MRS.[2] Surgery is other procedure that may be done in recurrent facial palsies. Total facial nerve decompression could be done by combined trans-mastoid and middle cranial fossa approach. The facial nerve is decompressed from internal auditory meatus to stylomastoid foramen. Conductive hearing loss may happen in this approach. In this study we herein, with permission from the patients and under institutional review board exemption, report a case of MRS with recurrent facial paralysis, had failed multiple medication trials over several years, that was managed by endoscopic trans-canal facial nerve decompression.

2 | CASE HISTORY AND EXAMINATION

A 30-year-old female patient was referred to the otolaryngology clinic of Rasool Akram Hospital complaining of recurrent bilateral peripheral facial paralysis, facial swelling, and fissured tongue. She had the history of facial paralysis in right side 10, and 2 years ago and in left side 3 years and 5 months ago (Figure 1A). She was diagnosed as recurrent Bell's palsy and received 1 mg/kg oral prednisolone for 10 days in each paralysis attacks. In last attack she also received acyclovir 800 mg five times a day for 5 days. None of these treatments were successful. On physical examination, her face movements showed facial paralysis grade IV and V House–Brackmann (HB) on right and left side consequently.

3 | DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS, AND TREATMENT

We exclude other causes of recurrent facial paralysis by physical exam, CSF analysis, chest X-ray, serum calcium, angiotensin converting enzyme, fluorescent treponemal antibody test, Lyme titer, and human immunodeficiency virus screen. We performed bilateral endoscopic trans-canal facial nerve decompression when systemic steroid therapy failed to produce any improvement. Facial nerve decompression was done from geniculate ganglion to distal part of mastoid segment near to stylomastoid foramen endoscopically. The mastoid segment was explored via transcanal scutum and posterior canal drilling. The facial nerve showed significant edema and swelling bilaterally when lateral of fallopian canal was removed about 180 degrees (Figure 2). The canal defect was reconstructed by tragal cartilage graft.

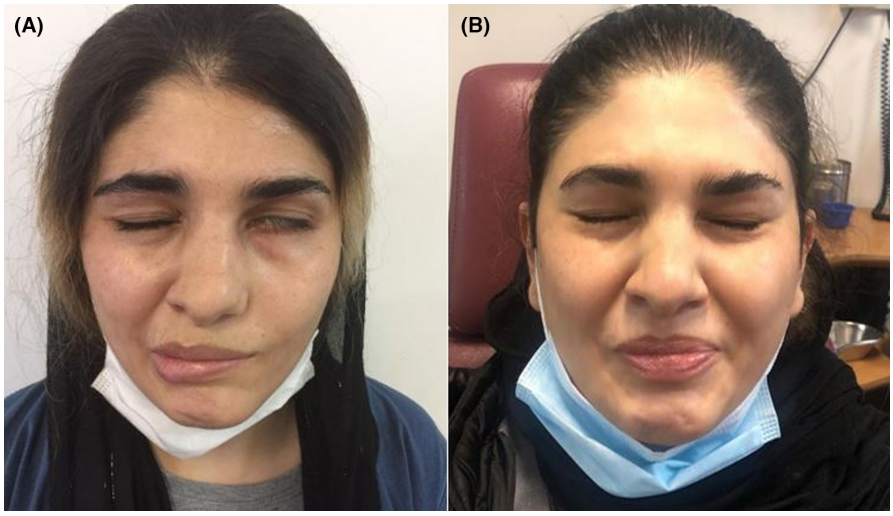

FIGURE 1 Endoscopic trans-canal facial nerve decompression, before surgery (A) and after 3 months (B).

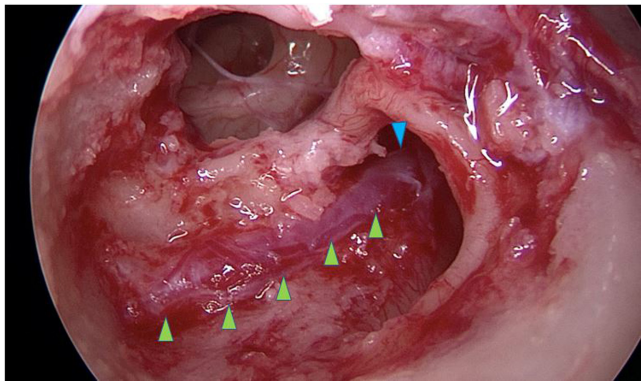

FIGURE 2 Endoscopic view, swelling, and edema of the facial nerve in tympanic and mastoid segment (green arrows). Geniculate ganglion (blue arrow).

4 | OUTCOME AND FOLLOW-UP

Left and right side of her face improved to grade III and II HB consequently within 3 months (Figure 1B). There is no relapse of facial paralysis attack, hearing loss or any ear problems after 20 months follow up.

5 | DISCUSSION

Recurrent facial paralysis is very common in MRS, occurring for unknown reasons that may improve by treatment or spontaneously. Unfortunately, after repeated paralysis attacks, the function of the facial nerve gradually weakens.[4] Surgical decompression has shown gross edema in the course of facial nerve. Irreversible facial edema in boney fallopian canal may be the main reason of steroid treatment failure in some cases. In these cases, surgical decompression may be effective treatment.

According to an electrophysiological study, action potential range is diminished in frequent cases of facial paralysis compared to those with an attack. Additionally, the risk of developing facial paralysis rises with each recurrence, from 15% in the second incident to 50% in the fourth. It is still controversial whether surgical intervention should be used to treat facial paralysis in MRS. The majority of research indicated that surgery is beneficial if evoked electromyography (E-EMG) reveals more than 90%–95% facial nerve degeneration. In Fisch's study, Bell's palsy patients had a less than 50% probability of recovery if there were more than 95% of non-irritating fibers present on day 14, while 78.8% of those who underwent surgical decompression showed good improvement.[5] In a study, Dai and colleagues studied patients who had more than 95% of facial nerve degeneration and had a history of prednisolone treatment. After surgery, 87.5% recovered to grade I or II HB, and three of them recovered completely.[6]

In the surgery, by the transmastoid approach, facial nerve decompresses to the labyrinthine section from the stylomastoid foramen. This surgical procedure was proposed by Yanagihara et al.[7] provisionally the incus is removed and then repositioned entirely to expose the labyrinthine section and geniculate ganglion. This method is considerable for preventing damage to the stapes and the inner ear, but there is still a risk of mild conductive hearing loss.

This is the first case of endoscopic trans-canal facial nerve decompression for patients with MRS, even though surgical decompression of the facial nerve had previously been documented as a therapy for refractory facial paralysis and post-traumatic facial nerve paralysis.[4] Our investigation showed that the left side improved significantly more than the right side. It may be explained by the short duration of last left paralysis. Both facial nerves were explored from Geniculate ganglion to mastoid part near the stylomastoid foramen. The facial nerve showed gross swelling and inflammation in its course. Although there is risk of conductive hearing loss in this approach, by carefully preserving ossicular chain, we preserved the hearing of both sides. In MRS when there is no improvement of facial paralysis within 3 months after systemic steroid treatment, facial nerve decompression may improve the paralysis. This surgical approach also could be done in traumatic facial nerve paralysis.

6 | CONCLUSION

In MRS patients with facial paralysis, endoscopic transcanal facial nerve decompression may be a safe, effective noninvasive treatment. This procedure is more practical for patients with shorter recovery times because it doesn't require an external incision or drilling of the temporal bone.

AUTHOR CONTRIBUTIONS

Alimohamad Asghari: Conceptualization; supervision; visualization; writing – original draft; writing – review and editing. **Yaser Nasoori:** Data curation. **Ahmad Daneshi:** Supervision; validation; writing – review and editing. **Mojgan Kianiasiabar:** Supervision; validation. **Fatemeh Dehghani Firouzabadi:** Visualization; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare that they have no competing interests.

DATA AVAILABILITY STATEMENT

All data generated during this study will be made available on request.

ETHICS STATEMENT

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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