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Case Report

Complete Melkersson-Rosenthal syndrome: an exceptional cause of facial nerve palsy

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Abstract

Melkersson-Rosenthal syndrome (MRS) is a rare disease, with unknown etiology characterized by oro-facial oedema, fissuring tongue and peripheral facial palsy. The mainstay treatment is corticosteroids.

We report the case of a 38-year old female, who presented six recurrent episodes of left peripheral facial palsy associated with simultaneous oedema of the median frontal area, treated successfully by short course oral corticosteroids with neuromuscular facial re-education. However, the recovery of the last episode was incomplete and the patient kept a left House-Brackmann grade II facial palsy at six months. The median frontal area was slightly inflammatory, being the location of simultaneous repetitive oedema and the tongue was fissured. These cardinal symptoms realise the complete triad of Melkersson-Rosenthal syndrome (MRS). The histopathological examination of the lip biopsy showed lymphocytic inflammation around the blood vessels. Since the residual facial palsy was graded as mild dysfunction, the frontal oedema fully recovered and the fissured tongue was not painful, the treatment consisted on appropriate neuromuscular re-education. The 6 months follow-up showed no recurrence with a stable grade II left facial palsy.

We present this case to supplement the rare literature data concerning the management of this rare entity. Patients should be prepared to the risk of recurrent episodes with longer duration of symptoms and more incomplete recoveries, which may indicate other therapeutic options.

Key words: melkersson-Rosenthal syndrome, recurrent facial palsy, oro-facial oedema, fissuring tongue, corticosteroids

Introduction

Melkersson-Rosenthal syndrome (MRS) is a rare disease, with unknown etiology characterized by oro-facial oedema, fissuring tongue and peripheral facial palsy. (1) The mainstay treatment is corticosteroids. But the recurrence of the disease makes the duration of symptoms longer and the recovery incomplete, indicating other therapeutic options. (2, 3)

We report a case of complete and recurrent Melkersson-Rosenthal syndrome treated in our Head and Neck surgery department of the Ibn Rochd University Hospital that supplements and supports the rare literature data concerning the management of this rare entity.

Case report

We report the case of a 38-year old female, with no particular medical history who presented six recurrent episodes of left peripheral facial palsy associated with simultaneous painless non-pitting oedema of the median frontal area since 1996. Each episode was treated by short course oral corticosteroids (1mg/kg of Prednisolone) for a week with neuromuscular facial re-education. The treatment was effective with complete resolution of symptoms and no sequelae. The last episode occurred on February 2020. The left facial palsy was graded initially as IV on House-Brackmann scale. The patient followed the same medical treatment and facial re-education with a total regression of the frontal oedema but the recovery of the facial palsy still incomplete.

We received the patient six months later for persistent left peripheral facial palsy graded presently as II on House-Brackmann scale.(Figure 1a) No cochleovestibular symptoms were reported and the otoscopic examination showed normal tympanic membranes. The clinical examination found a fissured tongue with multiple grooves on the lingual dorsum. (Figure 1b) The median frontal area was slightly inflammatory, being the location of simultaneous repetitive oedema. These cardinal symptoms realise the complete triad of Melkersson-Rosenthal syndrome (MRS). The patient also presented headaches with normal neurological examination and no other cranial nerve affection.

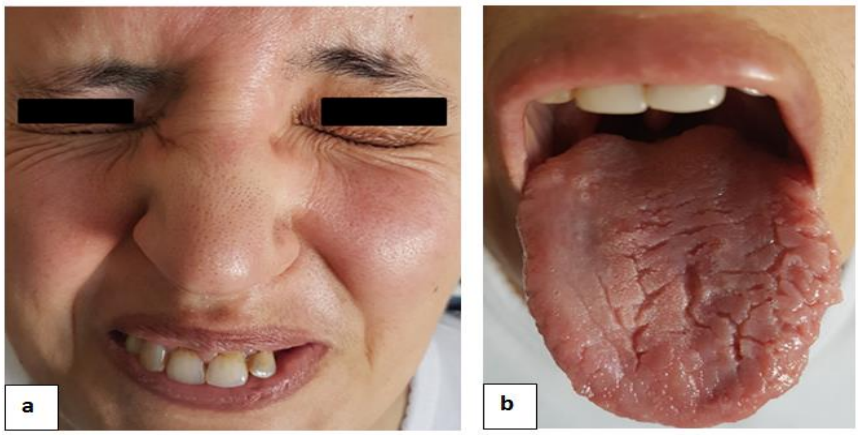

Figure 1: a. *Peripheral facial palsy graded as II on House-Brackmann scale b. Fissured tongue*

The facial nerve investigations included an MRI which showed no enlargement or source of pressure on the nerve which appeared as hypointense linear structure with no enhancement after gadolinium contrast use. The electroneuromyogram (ENMG) of the facial nerve indicated severe axonal damage on the left facial nerve, with 75% axonal loss but no active denervation. It also confirmed peripheral left hemifacial spasm. Tonal audiometry resulted on normal hearing levels, however, the acoustic reflex was absent on the left side.

The blood glucose rate was normal with hemoglobin A1c level at 6%, the C-reactive protein (CRP) level and erythrocyte sedimentation rate (ESR) were normal. Hepatitis serologies, syphilis tests and antinuclear antibodies (ANAs) were negative. The histopathological examination of the tongue and lip biopsies showed lymphocytic inflammation around the blood vessels corroborating the MRS even if frank granuloma was absent. (Figure 2)

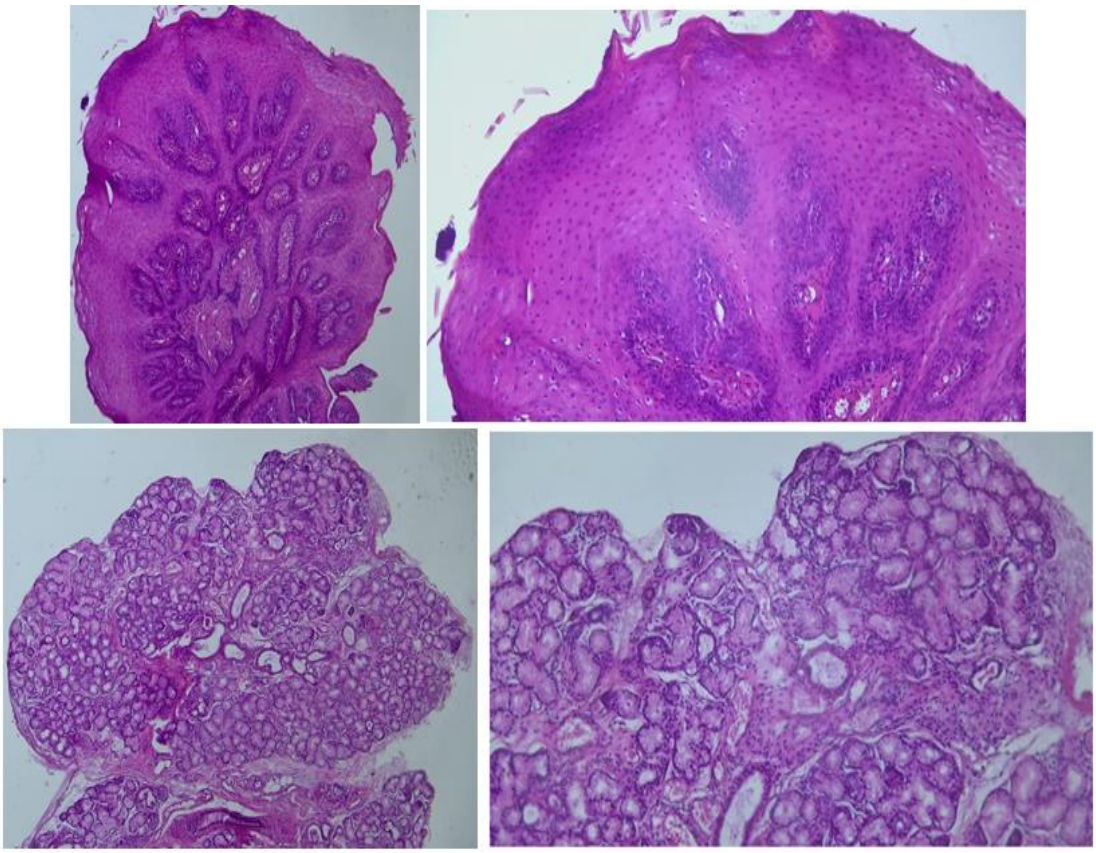

Figure 2: *Histopathological examination showing lymphocytic inflammation around the blood vessels corroborating the MRS even in the absence of frank granuloma.*

Since the residual facial palsy is graded as mild dysfunction with normal symmetry and tone at rest and complete eye closure at minimum effort, the frontal oedema is fully recovered and the fissured tongue is not painful and doesn't cause any loss of taste or secondary infections, the treatment

consisted on appropriate neuromuscular re-education. The immunosuppressant medication was not indicated regarding the absence of systemic involvement. The surgical decompression of the facial nerve was not considered since it usually concerns high grades of facial palsy with the aim to result in normal or near-normal facial mobility which is already the state of our patient.

The 6 months follow-up showed no recurrence with a stable grade II left facial palsy.

Discussion

The first description of oro-facial oedema and facial paralysis dates back to 1928 by Ernst Gustaf Melkersson in a 35-year female. In 1931, Rosenthal mentioned the fissured tongue as an additional feature completing the triad of Melkersson-Rosenthal syndrome. (4, 5)

MRS is a rare neuro-mucocutaneous syndrome with an estimated incidence of 0, 08% (6) and a female preponderance, commonly seen between the second and third decades of life. (2, 7) The pathogenesis still unknown even though an association with genetic, allergic, infectious and immunologic factors has been postulated. (8) Also, family predisposition, with autosomal dominant characteristics and incomplete penetrance has been reported. (9)

Complete MRS, only present in 8 – 25% of patients, is characterized by the triad of oro-facial oedema, recurrent peripheral facial nerve palsy and fissured tongue ("lingua plicata") that may not occur at the same time. (2, 8, 10) The oligosymptomatic MRS corresponds to only two clinical features while the monosymptomatic form corresponds to one. (1, 2) Meischer's granulomatous cheilitis is the most frequent monosymptomatic form of MRS diagnosed when there is biopsy evidence of a non-necrotising, granulomatous cheilitis in patients with isolated facial or lip oedema. (2, 11)

Peripheral facial palsy is usually the first symptom to appear preceding oro-facial oedema by months to years, involving all or a part of the facial nerve and is recurrent in 70% of the cases. Usually with spontaneous recovery, its duration increases as the disease progresses, with a tendency to become longer. (1, 2, 3) Indeed, recurrent facial palsy shows a worse prognosis since the facial nerve tends to deteriorate after repetitive episodes. Electrophysiological studies reveal loss of action potential amplitude in cases of recurrent facial palsy in contrast to those with a single attack. Also, the risk of a further episode of facial palsy increases with every recurrence, from 15% on the second episode to 50% on the fourth. (1, 12)

Orofacial swelling is the most common symptom. It is painless, non-pitting and commonly affecting the upper lip, but periorbital tissues, cheeks, nose, lower lip and chin may also be affected. Since it tends to evolve on repeated episodes at irregular intervals, partial or non-resolution of edema may lead to fibrosis and permanent disfiguration of the face. (2, 3, 8) The fissured tongue is the least common feature defined as grooves on the dorsum of the tongue which are at least 2 mm deep and 15 mm long. It may lead to secondary infections, loss of papillae, of taste, and dysesthesia. (2, 8) Other cranial nerves like trigeminal, olfactory, auditory, glossopharyngeal and hypoglossal nerves can also be involved. (2)

The histopathological analysis of the lip biopsy shows epithelioid non-caseous granulomas. However, oedema and perivascular lymphocytic infiltrates have been described as in our case. These histopathological findings can be missed if sampling is not done during the acute episodes. (2, 9)

MRS can be frequently diagnosed by clinical criteria with no need for further investigation. (7) However, some authors suggest complete blood counts, erythrocyte sedimentation rate, Angiotensin converting enzyme (ACE) levels, Anti-nuclear Antibody (ANA) testing, Antineutrophilic cytoplasmic antibody (ANCA), brainstem evoked audiometry, visual evoked potentials and nerve conduction studies of facial nerve. (2)

Corticosteroids remain the first-line treatment. Even if there are no randomized trials suggesting the optimal type and duration, corticosteroids lead to improvement in 50–80% of the cases and reduce relapse frequency by 60–75%. Oral corticosteroids are commonly used for 1 week, reserving high-dose pulse methylprednisolone for severe cases. (2, 8) Oro-facial oedema can be treated by intralesional triamcinolone acetonide (TA) injected at four sides in each lip, in the cheek and nasolabial folds following three weekly injections repeated after 6 months in case of persisting edema or recurrence. Intralesional TA is likely to be beneficial in cheilitis granulomatosa without systemic disease or refractory to oral corticosteroids. Oro-facial oedema can also be treated by intralesional betamethasone, along with oral doxycycline. (2) Non-steroidal anti-inflammatory drugs and antihistamines can be proposed. (7) Immunosuppressants had also been used in isolated cases with systemic involvement based on methotrexate, thalidomide, intravenous immunoglobulins, clofazimine, dapsone, anti-TNF therapy (infliximab), anti-histaminic drugs and

Hydroxychloroquine. (2)

Surgical management can be indicated in recurrent, high graded or persistent facial palsies after 1 – 3 months. The timing of surgery is uncertain and it consists in total or subtotal facial nerve decompression. Dai *et al.* (1) per-operative findings showed noticeable oedema of the facial nerve at the mastoid segment in all cases (100%), at the tympanic segment and geniculate ganglion in five cases (62.5%) and at the labyrinthine segment in one case (12.5%). The majority of authors advocates the benefits of surgery when electroneurography shows >90–95% degeneration of the facial nerve. (2, 12) Gantz *et al.* (13) report, after facial nerve decompression on patients with over 90% degeneration on electroneurography and no voluntary motor unit potentials on electromyography within 14 days, 91% of 19 cases with grade I or II facial palsy, versus only 42% of non-surgically treated patients. Even prophylactic decompression of the facial nerve seems to be a reasonable option to protect the nerve from potential damage. (12)

Persistent lip oedema can also be treated surgically as ultimo ratio, through Conway's technique, en-block removal of mucosa dorsal to vermillion with part of orbicularis muscle and grossly edematous tissue. Helium laser ablation is an alternative procedure for cheiloplasty. (2, 10)

Conclusion

The triad of oro-facial oedema, fissuring tongue and peripheral facial palsy represent the cardinal signs of Melkersson Rosenthal syndrome and are sufficient for clinical diagnosis, while histopathology analysis of the lip biopsy can be helpful in the oligo and monosymptomatic forms.

Patients should be prepared to the risk of recurrence with longer duration of symptoms and more incomplete recoveries. The mainstay treatment is oral corticosteroids. Other therapeutic options can be considered in front of persistent symptoms with high grades of dysfunction.

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