

Case Report

A Rare Case of Melkersson–Rosenthal Syndrome

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ABSTRACT

Melkersson–Rosenthal syndrome (MRS) is a rare clinical syndrome. The onset of illness usually occurs in the second decade of life. It is characterized by idiopathic facial paralysis and/or fissured tongue with oro-facial swelling, mainly lip edema. The diagnosis is mainly clinical as the biochemical marker is not ascertained. The isolated facial palsy can mimic various other clinical entities including Bell palsy. The treatment guidelines for MRS are not mentioned in the literature, due to the paucity of randomized clinical trials, which could not happen because of its rarity. However, steroids are effective in this condition. This case report highlights the case of MRS with the typical triad of features, which is quite rare.

KEYWORDS: Facial paralysis, fissured tongue, lip edema, Melkersson–Rosenthal syndrome

INTRODUCTION

Melkersson–Rosenthal syndrome (MRS) is a rare clinical disorder; its onset mainly occurs in the adolescent age group. The true incidence is unknown because of its rarity. The diagnosis is based on clinical manifestations as there are no specific biomarkers or laid criteria.^[1] It is a clinical diagnosis. The either of facial paralysis or plicated tongue and orofacial edema is suggestive of the diagnosis of MRS.^[1] In this presentation, we intend to present a case of MRS with the complete triad. The facial palsy was recurrent. This case has been reported as a clinical revisit for physicians.

CASE REPORT

A 17-year old female, 2 months before developed acute onset left-sided facial weakness. She had a deviation of angle of mouth on the right side, obliteration of nasolabial fold on the left side accompanied by persistent widened palpebral fissure, and loss of forehead wrinkles on the same side. She also complained of dysgeusia on the left side.

The history was negative for weakness in limbs, joint pains, arthritis, rash anywhere on the body, dry mouth, and difficulty in breathing. She denied features of tinnitus, ear discharge, vertigo, or vesicular eruptions over the ear cavity. There was an absence of fever, hemoptysis, and cough with expectoration or loss of

weight. The general examination was unremarkable. The neurological examination revealed a left-sided lower motor neuron type of facial palsy. Bell's phenomenon was intact. Ocular examination was normal. The motor, sensory, cerebellar, and extrapyramidal assessment was normal.

Investigations done in this patient were normal which include hemogram, liver function test, renal function, and thyroid evaluation. The autoantigen markers, anti-nuclear antibody (ANA), anti-double stranded DNA antibody, and rheumatoid factors showed negative results. The workup for sarcoidosis showed a negative study. The viral markers in serum were negative. The computerized tomography of cranium ruled out any structural disorder.

At this juncture, Bell's palsy was entertained and was started oral prednisolone 1 mg/day body weight in tapering dosage for a month along with facial exercises. Initially, she has been advised acyclovir therapy for 7 days. After follow-up at 15 days, her facial weakness was partially improved.

After a month, she reported recurrence of facial palsy. It was a lower motor neuron type of facial

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weakness, now on the right side [Figure 1a]. At this time, few investigations were repeated. The hemogram, blood glucose, complete blood counts, erythrocyte sedimentation rate, and C-reactive protein revealed normal results. The liver function test, renal assessment, and thyroid study exhibited normal parameters. The auto-immune markers which include ANA, antineutrophilic cytoplasmic antibody, and extractable nuclear antigens antibody were negative. The viral markers for herpes viruses, Epstein-Barr virus, cytomegalovirus, and dengue were unremarkable. The investigations related to tuberculosis showed normal results. The angiotensin-converting enzyme level was within the normal range, 49 U/L (normal values: 12–60 U/L). The computerized scan of the chest was done, and it did not reveal hilar lymphadenopathy or other abnormal features. This ruled out sarcoidosis. The investigations for Crohn's disease did not demonstrate any abnormality. The abdominal ultrasound did not demonstrate features of stenoses and abscesses. Full ileocolonoscopy was performed, and it did not suggest Crohn's disease.

The patient had lip edema and was swollen [Figure 1b]. Her tongue revealed a fissured plicated tongue [Figure 1c]. With this clinical triad, MRS was confirmed. The patient was re-administered steroids, and she completely recovered in 1 month.

DISCUSSION

MRS is an extremely rare clinical disorder. The clinical hallmarks of this disease are recurrent facial paralysis, intermittent episodes of orofacial edema, and fissured tongue. This condition is mainly reported in females as hormonal triggers for episodes of the fissured tongue or facial paralysis have been postulated.^[2]



Figure 1: (a) Right-sided lower motor neuron facial palsy, (b) The picture of lip swelling, (c) The tongue revealed fissured plicated tongue

Our patient had all three features at the time of the presentation. The clinical manifestations are variable, and all patients did not present with all the features. The etiology is unknown, and various theories have been postulated which include infective agents, allergic reactivity, autoimmune factors, and genetic predisposition.^[3]

The mutation at gene related to fatty acid transportation protein formation has been implicated as a causal gene for MRS. The genomic theories are at an early stage of development. The literature is not conclusive about familial predisposition.^[4] The histopathology of biopsy samples taken at an active stage of edema revealed lymphoepithelioid granulomas in addition to mononuclear infiltrates in perivascular areas.^[5]

The literature suggested clinical overlap with oral Crohn's disease sharing pathological features of noncaseating granuloma, but Crohn's disease is conspicuous by the absence of facial paralysis or fissured tongue.^[6] The guidelines for effective therapy could not be established because of its rare occurrence. However, steroids, nonsteroidal anti-inflammatory agents and antibiotics have been tried in this condition. The steroids proved to be effective in MRS.^[6] The facial nerve decompression was done in few patients with successful outcomes.^[7]

This case has been reported due to its rarity and will increase the understanding of treating physicians.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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