Case Report

Melkersson–Rosenthal syndrome: A rare cause of recurrent facial palsy – A case report

Yared Zenebe Zewde (ORCID iD: 0000-0002-6442-7148)

Abstract

Background: Melkersson–Rosenthal syndrome is a rare, neuro-mucocutaneous, granulomatous disorder of unknown etiology, clinically characterized by a triad of symptoms: recurrent facial nerve palsy, facial swelling and fissured tongue. Melkersson–Rosenthal syndrome is frequently seen in patients in their second or third decade of life. It is diagnosed based on clinical features, and it is rarely possible to observe all the classic triad symptoms at the same time. The disorder may cause recurring peripheral facial palsy that is wrongly diagnosed as recurrent Bell's palsy.

Case presentation: A 25-year-old female patient, referred from Bishoftu town in Ethiopia, was presented to the neurology clinic of Tikur Anbessa Specialized Hospital in Addis Ababa complaining of a five-day history of recurrent left-side peripheral facial weakness, facial edema and fissured tongue. Her past medical history was positive for similar symptoms, for which she was diagnosed with Bell's palsy and received oral corticosteroid treatment. Left-side lower facial swelling with flat naso-labial fold and fissured tongue were detected on examination. After excluding other mimickers, she was diagnosed with Melkersson–Rosenthal syndrome and completely recovered with a high dose of steroid treatment.

Conclusion: Melkersson–Rosenthal syndrome may present with the classic clinical triads of symptoms, but mostly it shows an oligosymptomatic pattern. So, it is usually under-recognized and often misdiagnosed as Bell's palsy, as had been done in the past in our case. Therefore, Melkersson–Rosenthal syndrome should be considered in the differential diagnosis of patients presented with recurrent peripheral facial weakness, as early detection and therapy might prevent cosmetic disfigurement from multiple relapses [*Ethiop. J. Health Dev.* 2020; 34(3): 214-216]

Key words: Melkersson–Rosenthal syndrome, facial palsy, fissured tongue, facial swelling, Bell's palsy

Introduction

Melkersson–Rosenthal syndrome (MRS) is a rare, neuro-mucocutaneous disorder characterized by a triad of symptoms: recurrent peripheral facial nerve palsy, facial and/or lip swelling and fissured tongue (1). Although the exact cause of MRS has not been recognized, genetic predisposition, allergic reaction, hypersensitivity, and autoimmune and microbial reactions have been hypothesized to contribute to its pathogenesis (2).

Patients with underlying granulomatous conditions might present with facio-labial swelling, and recurrent facial weakness could be attributed to other systemic disorders, such as diabetes mellitus and hypothyroidism. Hence, MRS is the diagnosis of exclusion.

This case study is of a young female, referred from Bishoftu town on May 30, 2019, who was presented to the neurology clinic at Tikur Anbessa Specialized Hospital in Addis Ababa with a complaint of third-time recurrence of left-side peripheral facial weakness and facial edema for a duration of five days. Her past medical history revealed similar symptoms, which were diagnosed as Bell's palsy.

Case presentation

A 25-year-old black female patient was referred from Bishoftu town and presented to the neurology clinic of Tikur Anbessa Specialized Hospital in Addis Ababa on May 30, 2019. Her main complaint was an insidious onset, left upper lip, painless swelling, which progressed to involve her left cheek. She had been unable to close her left eyelid for the previous five days. Her past medical history was positive for two episodes of same-side peripheral facial weakness. During the first attack, in 2016, her symptoms spontaneously resolved without any treatment. But when it recurred two years later, she visited a nearby clinic, and was diagnosed as a case of recurrent Bell's palsy, which showed a complete resolution following corticosteroid treatment. She did not recall any cause or inciting factor that had induced her symptoms. She had no history of trauma to the head or neck region, skin rash or sensory symptoms. She denied any recent history of infection of the ear, throat, sinus or dental structure. She had no personal history of sarcoidosis, Crohn's disease, autoimmune condition or any other chronic illness, and her family history was negative for similar conditions. She denied any history of food allergy or any other form of allergic reaction in the past.

Her physical examination demonstrated a diffuse, non-pitting swelling of the left cheek with peri-orbital fullness (see Figure 1), fissured tongue (lingua plicata) and mild left naso-labial fold flattening (see Figure 2). The clinical and laboratory evaluations for possible cause of recurrent peripheral facial palsy, including diabetes mellitus, otitis media, syphilis, Guillain–Barré syndrome, leukemia or other tumors, was non-revealing. Magnetic resonance imaging (MRI) of the head (see Figure 3) showed a normal brain parenchyma and clear course of the seventh nerve.

Department of Neurology, Addis Ababa University, Addis Ababa, Ethiopia. Email- yaredzene121@gmail.com

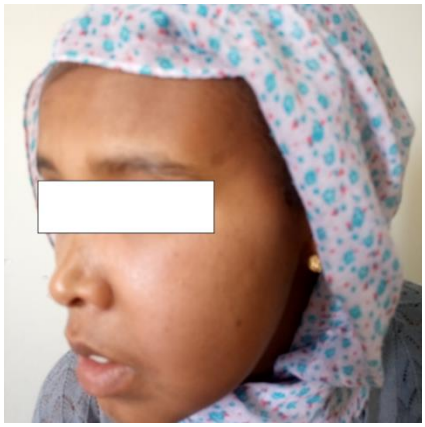

Figure 1: **Patient had a diffuse swelling of the left cheek and lower eyelid**

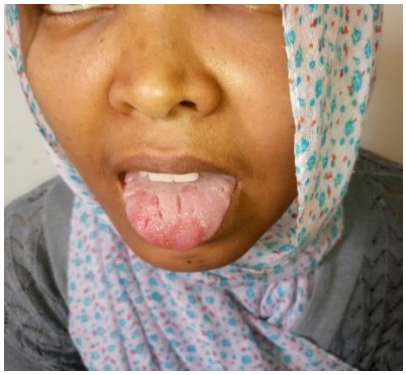

Figure 2: **Patient had a fissured tongue and mild flattening of the left naso-labial fold**

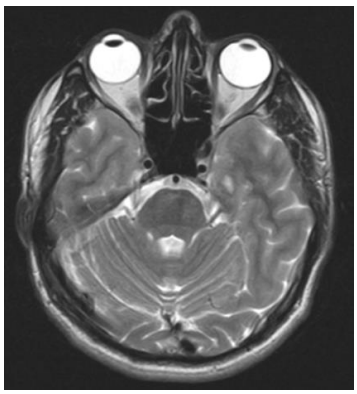

Figure 3: **A T2-weighted axial brain MRI showed a normal brain parenchyma with a clear course of the facial nerve intra-cranial segment**

She was initiated on oral prednisolone therapy at 1 mg/kg/day, with gradual tapering for three weeks. Her facial edema completely subsided by the third day of treatment and facial weakness recovered by the second week. Therefore, a final diagnosis of MRS was made and the patient was appointed for outpatient follow-up care.

Discussion

MRS is a neuro-mucocutaneous syndrome belonging to the orofacial granulomatosis group. The condition was first reported by Melkersson in 1928, where a woman with intermittent peripheral facial palsy and lip edema was documented (3). Later, in 1931, Rosenthal completed the triad of symptoms of the syndrome by adding the presence of fissured tongue (4).

MRS is a rare syndrome with an estimated incidence of 0.08% in the general population. Disease onset is more frequent in young adults, between the second and third decades of life, and a relatively higher rate is seen in females (5). Our case was of a young female in her early 20s during symptom onset.

The etiology of MRS remains uncertain, but it has been postulated that genetics, infectious and immunologic factors may play an etiologic role (6). There are reports of MRS associated with viral and other bacterial infections, autoimmunity, neurotropic factors, atopy and hypersensitivity reactions to various antigens, including food additives such as monosodium glutamate, which may have a pathogenesis effect but has not been confirmed as an etiologic agent (1).

The classic triad of recurring facial paralysis, fissured/scrotal tongue and recurring orofacial and/or lip edema is observed in only 8% to 25% of MRS patients (1). Recurring lip and/or orofacial edema is the most common presenting symptom, in 80% to 100% of patients (7). The facial edema is generally painless, unilateral, and most often occurs in the upper lip. Less frequently, edema may involve cheeks, palate, gingiva, tongue, pharynx, larynx, and periorbital region (8). Facial paralysis is observed in 47%-90% of cases and can be transient or sometimes permanent. It is most often unilateral but rarely can be bilateral, and the recurrence rate is around 10% (6). Oligosymptomatic forms are reported in around 50% of cases, while the combination of orofacial edema and facial palsy was seen by Chan *et al* in 22% of cases. (9). In 40% of MRS patients, fissured tongue (lingua plicata) is identified (10). The classic triad symptoms are observed in less than a quarter of MRS patients, so this make clinical diagnosis very difficult. In the present case, the woman was wrongly diagnosed, in the past, with recurrent Bell's palsy.

The diagnosis of MRS is difficult, as there are no acknowledged diagnostic criteria or biomarkers to test. Many authors agree that MRS is a clinical syndrome that is diagnosed by a constellation of sign and symptoms with no need for further investigation. But in oligosymptomatic cases, the diagnosis is confirmed by histopathological examination showing non-caseating granulomas (11). However, as shown in our case, subtle features – such as fissured tongue and a complete resolution of symptoms – support the diagnosis of MRS after possible mimickers were excluded following extensive investigation.

The differential diagnosis of MRS includes a broad spectrum of heterogeneous conditions, mainly represented by other granulomatous disorders, such as allergic reactions and contact dermatitis, sarcoidosis, Crohn's disease, Wegener's vasculitis and amyloidosis. Causes of recurrent facial palsy, including hypothyroidism, lymphangioma, erysipelas, lymphoma and herpes simplex labialis, should be considered in the differential diagnosis; Bell's palsy, orofacial herpes, rosacea, and allergic reactions should also be considered (6-8).

Although there is no definite treatment for the disease and recurrences are unavoidable, we have options of therapy, including antihistamines, methotrexate, systemic and/or intra-lesional steroid administrations, which can improve or cure orofacial edema and are considered as initial choices (12). Surgical options are available for refractory facial edema with significant deformity of the face (13).

Conclusion

To the best of the author's knowledge, this is the first case of MRS reported in Ethiopia. It highlights the importance of considering MRS in the differential diagnosis of recurrent peripheral facial nerve paralysis, and to look for other subtle clinical features such as tongue fissures, since most MRS patients present with an oligosymptomatic pattern. Early detection and timely management can reduce relapse and prevent deformity.

Declarations

Ethical approval and consent to participate

Written informed consent for publication of patient's clinical details and images was obtained from the patient. A copy of the consent form is available for review by the editor of this journal.

Consent for publication

The patient gave written informed consent for participation in this case report and publication of her clinical details and images. The signed consent is available on request.

Availability of data and materials

All data are available in the manuscript.

Competing interests

The author declares that he has no competing interests.

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Author's contribution

The author evaluated, diagnosed and managed the patient. He was also involved in the data interpretation, draft of the manuscript and collection of the pictures, and read and approved the manuscript for publication.

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