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JIM Journal of Interdisciplinary Medicine

CASE REPORT

NEUROLOGY // INTERNAL MEDICINE

Recurrent Facial Nerve Paralysis: A Case report on Melkersson-Rosenthal Syndrome

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ARTICLE HISTORY

Received: February 19, 2022
Accepted: March 30, 2022

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ABSTRACT

This is a case report of a 27-year-old male who presented with recurrent facial swelling and weakness. The clinical evaluation revealed that the patient had Melkersson-Rosenthal syndrome (MRS), a rare neurological disease characterized by the triad of recurrent facial edema, recurrent facial muscle weakness, and a furrowed or fissured tongue. Accurate data regarding the incidence of MRS is challenging to obtain due to the rarity of the disorder, and early recognition and treatment can potentially reduce the probability of permanent disability.

Keywords: recurrent facial nerve paralysis, Melkersson Rosenthal Syndrome, granulomatous cheilitis

INTRODUCTION

Melkersson-Rosenthal syndrome (MRS) is a rare neuro-mucocutaneous condition with a recurrent and progressive course. It is characterized by the triad of lip swelling, also known as granulomatous cheilitis or Miescher cheilitis, a fissured tongue that is sometimes referred to as lingua plicata, and facial paralysis.

Bilateral lip swelling is the most frequent monosymptomatic clinical presentation. The diagnosis is established based on the symptoms. Although the symptoms may regress spontaneously or with medical treatment, the condition may also take a progressive course, requiring surgical treatment with facial nerve decompression.

CASE REPORT

This is a case report of a 27-year-old male with a medical history of migraines, who presented with two discreet episodes of right facial paralysis over a period of two years.

The first episode occurred three years prior, when the patient presented with right facial paralysis to a general practitioner and was treated with prednisolone

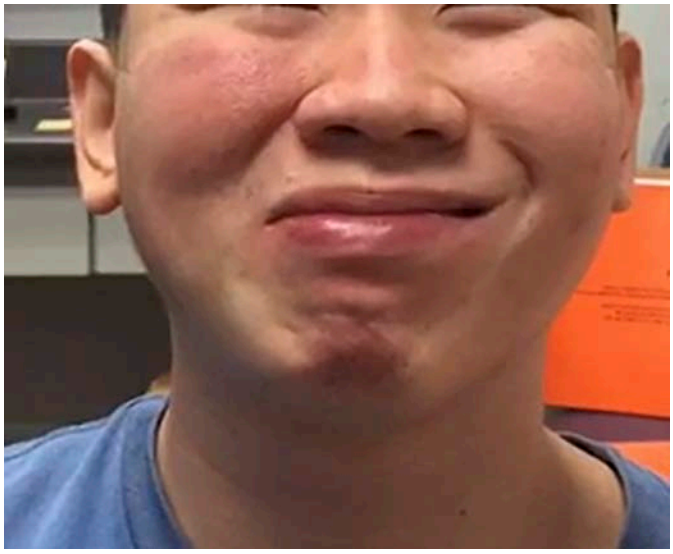


FIGURE 1. Right lower motor facial weakness



FIGURE 2. Swelling of the lower lip

for presumed Bell's palsy. He presented full recovery. However, in the following year, the patient experienced numerous episodes of swelling of his upper lip and gums, which resolved independently, without medication. He did not think much about this and attributed those episodes to a probable allergy he was uncertain of.

Regarding the current presentation, the patient woke up one morning with a significant right-sided facial weakness. He noticed his lips were swollen and numb, and he could not fully close his right eye. There were no other symptoms such as fever, rash, unintentional weight loss, nausea, vomiting, or diarrhea. He was unsure of any known allergies. Family history revealed that he was an only child; his father and uncle had a furrowed tongue just like him but nil else of note.

On clinical examination, there was a right lower motor facial weakness (Figure 1), and his lips were swollen (Figure 2). Upon inspection, his tongue was furrowed (Figure 3).

Blood investigations revealed a normal complete blood count, renal and liver function. The connective tissue disease screen was negative, and inflammatory markers were within normal limits. A nerve conduction study showed a predominant axonal type of facial nerve injury. The brain magnetic resonance imaging was normal. A biopsy was taken from his lower lip, revealing granulomas with epithelioid cells, perivascular mononuclear infiltration, non-caseating granulomas, lymphedema, and fibrosis (Figure 4).

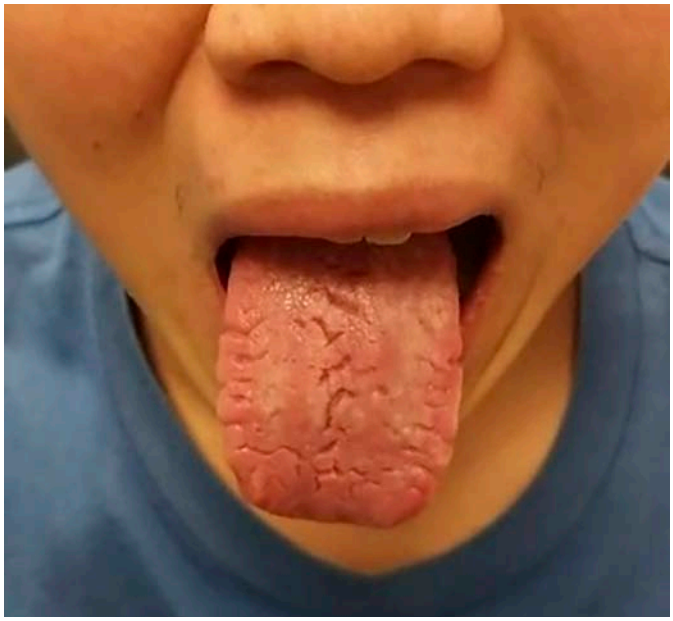


FIGURE 3. Furrowed tongue

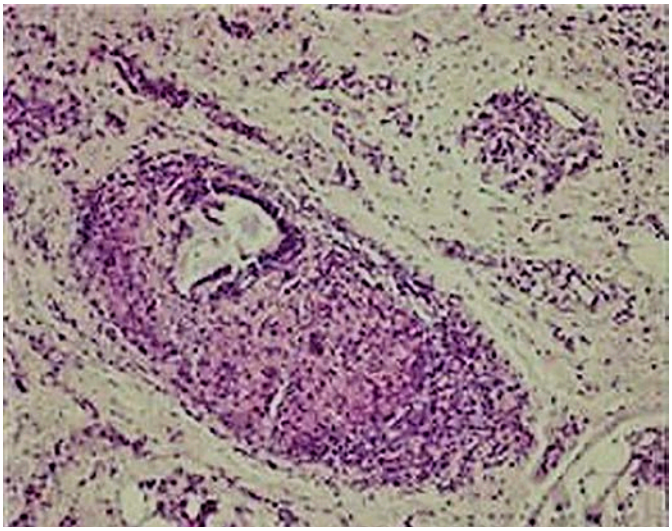


FIGURE 4. Histopathology showing non-caseating granuloma, epithelioid cells, and perivascular mononuclear infiltration

We established a diagnosis of Melkersson-Rosenthal syndrome, and the patient was started on oral prednisolone 1 mg/kg/day for one week, tapered over the following two weeks. Intralesional triamcinolone acetonide was used for the orofacial edema. The patient was also prescribed a benzoate-free diet and vitamins including thiamine, niacin, riboflavin, pyridoxine, ascorbic acid, and vitamin E in the form of a multivitamin tablet as part of an ancillary treatment. He showed good recovery in the following three months, without disability.

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

DISCUSSIONS

Melkersson-Rosenthal syndrome is a rare neurological disease characterized by the triad of recurrent facial edema, particularly affecting the lips, one or both cheeks, or one side of the scalp,[1] recurrent facial muscle weakness, and a furrowed or fissured tongue.[2] Most frequently, the affected patients present with only one or two of the listed symptoms. The syndrome was initially described in 1928 by Ernst Gustaf Melkersson in the case of a 35-year-old woman, who presented with orofacial swelling and facial paralysis. Since then, more clinical and case reports have emerged, providing a better understanding of this syndrome.

MRS can affect individuals of all ages, but it is most commonly seen in young adulthood. Most data indicate that MRS tends to be diagnosed more in female patients. Evidence also suggests that MRS may be hereditary because familial transmission has been observed in several patients.[3] A few cases of MRS have been accredited to various bacterial or viral infections including herpes viruses. It is also supposed that MRS could be part of the secondary involvement of other systemic conditions such as Crohn's disease or sarcoidosis. However, no strong associations have been identified between MRS and these diseases.

The clinical diagnosis of MRS can be established when at least two of the triad's symptoms are present. There are no specific radiological findings and diagnostic tests. However, a nerve conduction study can be done, and it typically shows a predominant axonal type of facial nerve injury. A biopsy may be necessary when a clinical diagnosis cannot be established, but also to exclude other possible causes such as infections, recurrent angioedema, Crohn's disease, sarcoidosis, or cancers. The histopathology examination usually reveals non-caseating granulomas.[4]

There are no standard therapeutic measures for the treatment of MRS. Although it may resolve with no treatment, episodes may become more frequent and with longer duration if no treatment is administered.

Therapeutic agents that can be administered in MRS consist in intravenous corticosteroids, nonsteroidal anti-inflammatory drugs, antihistamines, and antibiotics. In subjects with other systemic disorders, immunosuppressants are sometimes considered.5 The reduction of abnormally swollen lips may require surgery and/or radiation. Also, in case of persistent and repetitive facial paralysis, surgical treatment for facial nerve decompression may be needed.

MRS may reappear intermittently after the first presentation; the patient may experience recurrent facial weakness, which can improve as the nerve regains its function. However, it can become a long-standing clinical issue as each attack results in less adequate recovery.

CONCLUSION

Accurate data regarding the incidence of MRS is difficult to obtain due to the rarity of this condition. Therefore, it is probably an underdiagnosed condition, as its symptoms may be attributed to other common disorders due to the overlapping clinical presentation.

CONFLICT OF INTEREST

The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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