# Non granulomatous Melkersson–Rosenthal syndrome: A Case Report

Case Report

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## ABSTRACT

**Introduction:** Melkersson–Rosenthal syndrome is a rare condition that is of unknown or unclear etiology , neuro_mucutanous characterized by a triad of orofacial edema, facial paralysis, and a fissured tongue. In this article we present a rare case that shows the absence of granulomatous histological features which is a usual findings.

**Case report:** A 17-year-old Sudanese boy presented to our clinic at Sharq Alnil Hospital complaining of upper lip and facial swellings of one month duration. Clinical examinations revealed severe lip and facial edema, 'fissured tongue and unilateral facial palsy. Histopathological investigation did not reveal any granulomatous formation. The patient was treated successfully with systemic steroids.

**Conclusion:** Although Melkersson–Rosenthal syndrome may present without granulomatous histopathlogical features it still respond to systemic steroid in its management. Evaluation and treatment of Melkersson-Rosenthal syndrome require multidisciplinary approach by an allergologist-immunologist, neurologist, dentist, maxillofacial surgeon and histopathologist. Regular follow-up of these patients is necessary.

**Key Words:** Facial palsy, Lip swelling, Fissured tongue.

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## INTRODUCTION

Melkersson–Rosenthal syndrome is rare disease characterized by a triad of symptoms: orofacial swelling, facial palsy and fissured tongue, firstly was described by Melkersson in 1928 in a 35-year female with oro-facial edema and facial paralysis, later Rosenthal in 1931 added the third feature of fissured tongue[1, 2]. The etiology of MRS remains idiopathic, but genetics, infectious and immunologic factors may contribute to its pathogenesis[3]. Diagnosis can be made clinically based on the presence of the three symptoms; when only 1or 2 are observed, a biopsy from the facial edema should be performed and demonstrate non-caseating granulomas[2].

Oral corticosteroids are the first line in the treatment of MRS followed by immunosuppressants drugs for more resisted cases while surgeries in form of facial nerve decompression and Cheiloplasty are reserved for medically non responding recurrent facial nerve palsies and facial edema[4].

We report here a case of MRS with a classic triad of orofacial edma, facial palsy and fissural tounge histopathology proved absence of non – caseating granuloma treated successfully with short dose of oral prednisolone.

## CASE REPORT

A 17-year-old male Sudanese patient presented to the department of oral maxillofacial surgery at Sharq Al Nil hospital complaining of a sudden onset of facial swelling especially the lips and left side facial paralysis, he reported a recent history of viral infection treated by topical A acyclovir ointment for one week without response then he tried a traditional therapy which was stopped after week after developing gastrointestinal upset. On physical examination, edema of both upper and lower lips was observed. Facial asymmetry with complete lower motor neuron type left facial nerve paralysis was observed involving the left eyes and eyebrows. Intraoral examination revealed a fissured tongue. Laboratory analysis revealed normal blood count, urine general and stool general, immunoglobulin level and CRP were within the normal range. The chest x-ray, temporal and cranial magnetic resonance imaging no pathological tissue or

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abnormalities were detected. A biopsy taken from lower lip revealed inflamed connective tissue with hyperplastic oral epithelium and no granulomas. Patient started treatment by prednisolone 40 mg/day for one week then tapering for next week .the orofacial swelling decrease in size and facial palsy disappeared ( Figure 1 and 2 and 3 )

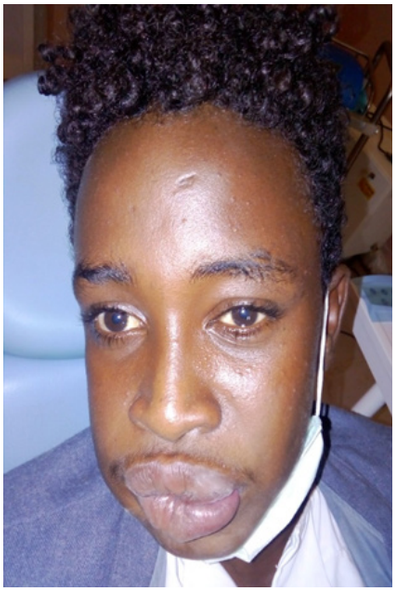

**Figure 1:** shows lip swelling

**Figure 2:** shows facial palsy at the left side

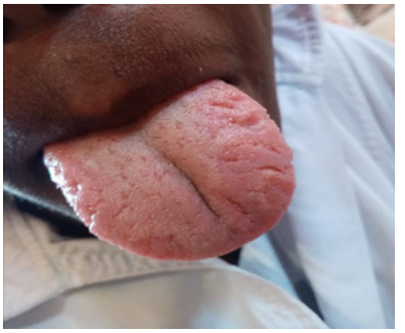

**Figure 3:** shows fissure tongue.

## DISCUSSION

Melkersson–Rosenthal syndrome is rare disease characterized by a triad of symptoms: orofacial swelling, facial palsy and fissured tongue, firstly was described by Melkersson in 1928 in a 35-year female with oro-facial edema and facial paralysis, later Rosenthal in 1931 added the third feature of fissured tongue [1, 2]. Incidence is estimated to be between 0.2 and 80 in 100,000 per year [5] The etiology of MRS remains idiopathic, but genetics, infectious and immunologic factors may contribute to its pathogenesis [3]. Recently a report had documented a case of classic triad symptoms associated with COVID-19 infection which was considered as triggering factor [6] In this case, the patient had a history of intraoral vesicles preceded the onset of the disease and a dermatologist prescribed an anti-viral drug to treat that vesicles which augments the argument of infectious disease as an etiological factor in pathogenesis of MRS studies has agreed in the minority of cases presented with the classic triad of recurring facial paralysis , fissured tongue and recurring orofacial and/or lip edema which is estimated to be not more than 25 % of MRS patients[7]. Our case has shown the full spectrum of the symptoms. Although the association of MRS and non-necrotizing granuloma is uncommon, it is still used as a confirmatory investigation, especially in patients with one or two characters of the symptomatic triad[2, 4, 8, 10]. Histopatholgy of a biopsy taken from the lower lip of our presented case showed the absence of the frank granuloma (Figure 4)
which is justified by Dhawan etal as the histopathological findings may be missed if sampling is not done during the acute episodes[4]. We encourage the concepts of routine performing of histopathlogical invistigatin even in the cases of classic triad to exclude other granulomatous diseases.
Corticorsteriods in the form of oral prednisolone and intra lesion injections of triamcinolone remain the cornerstone of treatment of MRS. They lead to improvement in
50 – 80 % of patients and reduce relapse frequency

by 60 – 75 %. 7. for a patient who presents with oro-facial swelling and facial paralysis, oral corticosteroids are the mainstay while intralesional injections are introduced if oro-facial edema alone is present or is refractory to oral corticosteroid[4. 6, 11]. Our patient was treated successfully (Figure 5) by introducing prednisolone tabs 40 mg/day for one week, then tapered for the next week, and after six months of regular follow-up, we did not encounter any relapse. Others Immunosuppressant therapy such as thalidomide, azathioprine, and methotrexate and antibiotics (tetracycline and dapsone) have been reported to treat some cases of MRS acceptably[11]. After long term of failure of corticosteroids, tumors necrosis factor-α blockers (adalimumab ) were successfully used to control all the symptoms of MRS[12, 13]. In our opinion, this type of treatment should be taken after exhausting other nonsurgical options. Facial nerve decompression and chillioplasty represent the surgical aspect of the treatment of MRS. They are usually spared for medically refractory recurrent cases[4].

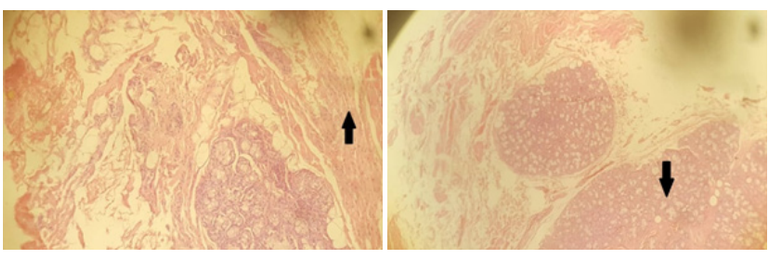

**Figure 4:** shows chronic non-specific inflammation with epithelial hyperplasia

**Figure 5:** shows patient post- treatment.
( The facial palsy has been subsided after corticosteroids treatment.)

## CONCLUSION

Although the presentation of the triad of orofacial edema, facial paralysis, and fissured tongue symptoms is diagnostic for Melkersson-Rosenthal syndrome, it is crucial to take a biopsy to exclude other granulomatous diseases. This case highlights the importance of conducting a deep study to investigate the reasons behind the variety of histopathology pictures among MRS cases, but in spite of these varieties, corticosteroids are still the first line of treatment.

## CONFLICT OF INTEREST

The authors declare no conflict of interest.

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