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Case Report **ENT**

Melkersson-Rosenthal Syndrome: A Case Report

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Abstract

Melkersson-Rosenthal syndrome (MRS) is defined by the triad of labial edema, peripheral facial paralysis, and plicated tongue. It is a complex entity with clinical polymorphism and unknown etiopathogenesis. A better understanding of the pathophysiological mechanisms would improve the management of these patients, who often present with significant aesthetic damage. We report a rare case of this syndrome with a plicated tongue and sequellar lagophthalmos.

Keywords: Melkersson-Rosenthal syndrome, labial edema, plicated tongue, facial paralysis, diagnosis, histology.

INTRODUCTION

Melkersson-Rosenthal syndrome (MRS) is a neuro-muco-cutaneous disease classically defined by the triad of labial edema-peripheral facial paralysis (PF)-plicated tongue. It is a complex entity whose management is difficult because of its clinical polymorphism and the absence of identified etiopathogenesis.

CASE REPORT

A 51-year-old patient presents in his history a Guillain-Barré syndrome, three years later, he presents a tilting intermittent peripheral facial paralysis. The clinical examination showed a swelling of the lower lip, a plicated tongue with a scrotal aspect, and a defect of palpebral closure sequelae of facial paralysis (Figure 1).

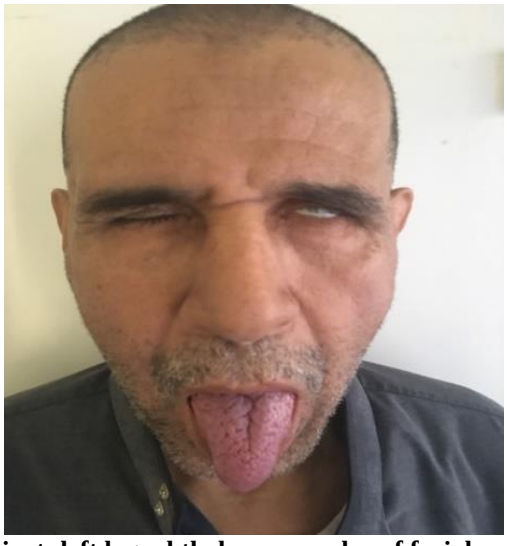

Figure 1: Clinical aspect of the patient: left lagophthalmos sequelae of facial paralysis, plicated tongue and edema of the upper lip

Lip biopsy revealed an epithelial-giganto-cellular granuloma without caseous necrosis with obliterating epithelioid endolymphangitis (EEO), a pathognomonic sign of MRS (Figure 2).

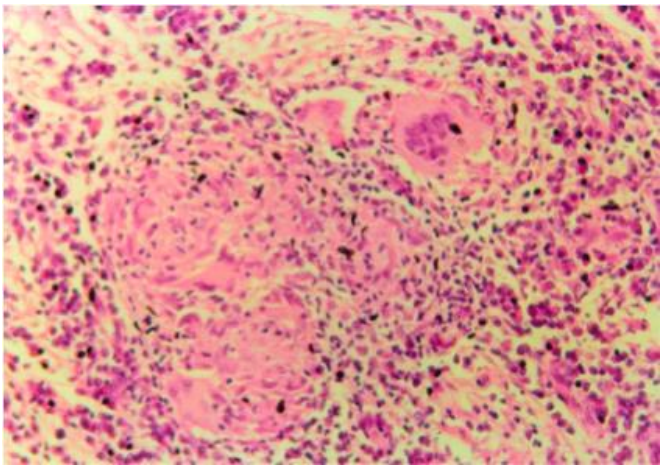

Figure 2: Histological appearance: Epithelio-gigantocellular granulomas without caseous necrosis (Hematein-eosin, 400)

The diagnosis of Melkersson-Rosenthal syndrome is retained on the association of facial paralysis, scrotal tongue, and macrocheilia. The patient is treated with boluses of methylprednisolone with a slight improvement.

DISCUSSION

MRS is an orofacial granulomatosis that manifests as a symptomatic triad: plicated tongue, macrochelitis, and recurrent facial paresis or paralysis [1].

In 1928, Melkersson made the connection between macrochelitis and facial paralysis, then Rosenthal identified the three signs that constitute the characteristic triad [2].

Its etiology is controversial. Some authors have mentioned a genetic origin. For others, it is rather a circumscribed neurological or neurovascular disorder resulting from an allergic or infectious stimulus. Finally, the viral or bacterial infectious etiology (Syphilis, oral infection, herpes, viral meningitis) was mentioned [3].

The MRS takes a multi or mono-symptomatic form. Miescher's granulomatous cheilitis can be considered a monosymptomatic form [4].

It more frequently affects the upper lip, achieving a tapir-lip appearance. Facial paralysis is a uni or bilateral peripheral type. The onset is sudden or gradual, the evolution is intermittent and then permanent.

The positive diagnosis is histological. According to several authors, the histological images of the tongue or the lips are identical. We discern a subepithelial tissue with loose and diffuse edema. One frequently but inconstantly finds the histological aspect of gigantoepithelioid granulomas without caseous necrosis, with vascular tropism with obliterating epithelioid endovascularitis [5].

A non-contributory histological analysis should not reject the diagnosis of MRS when the clinical symptoms are evident.

Therapeutically, no treatment claims to cure this disease. The symptomatic treatments offered are simply intended to avoid or space out recurrences to relieve patients and improve their quality of life.

In the case of facial paralysis, systemic corticosteroid therapy is used at a dosage of 0.5 to 1 mg/kg/day for 10 to 20 days with a gradual reduction depending on the evolution of symptoms [6]. In the absence of recovery after one or two months, neurolysis or surgical nerve decompression can be proposed [3, 6].

For Miescher's macrochelitis, intralesional injectable corticosteroid therapy is often used repeatedly. In isolated cases, hydroxychloroquine (Plaquenil), thalidomide, and metronidazole have been effective [7]. In the absence of scalability of the macrochelite, reduction cheiloplasty is proposed by several teams [8]. It is advisable to use corticosteroid therapy pre and postoperatively to avoid immediate relapses.

The plicated tongue does not require any treatment. In all cases, given the usual mildness of the disease, it is necessary to adapt the therapy taking into account the side effects of the different treatments used.

CONCLUSION

SMR represents a diagnostic and therapeutic challenge. A better understanding of the pathophysiological mechanisms would improve the management of these patients, who often present with significant aesthetic damage.

CONSENT:

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

Competing Interests: The authors declare that they have no competing interests.

Author's Contributions: All authors have read and agreed to the final manuscript.

ABBREVIATIONS:

MRS: Melkersson-Rosenthal syndrome

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