

**Melkersson-Rosenthal Syndrome about a Case**

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**Case Report**

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**Abstract:** Melkersson Rosenthal Syndrome (SMR) is a rare orofacial granulomatosis of controversial etiology. It combines recurrent facial palsy, upper labial edema and a plicated tongue. Through a case of SMR we recall the clinical and therapeutic peculiarities of this pathology. 36-year-old patient, who presented in our consultation for labial and gingival edema of progressive worsening, evolving by outbreaks for five years. The interrogation reveals in the antecedents the notion of transient facial paralysis. Examination shows prominent lip edema in the upper lip, edema and superior gingival hypertrophy and a plicate tongue. The diagnosis of SMR was retained. Patient was put on prolonged corticosteroid treatment in degressive dose, which led to a clear clinical improvement with disappearance of the edema not recidivism after a decline of nine months. The SMR is an orofacial granulomatosis manifested by a symptomatic triad: recurrent paralysis or facial paresis, macrocheilitis and a plicate tongue. This triad is rarely complete, and some minor signs can be observed. The diagnosis is mostly clinical and confirmed by histology. But a non-contributory histological analysis should not reject the diagnosis of SMR when the clinical symptomatology is obvious. Systemic corticosteroids are the most recommended treatment. It improves the quality of life and avoids or space recurrences. The prognosis remains good.

**Keywords:** Syndrome, edema, labial, facial paralysis, plicate tongue.

**INTRODUCTION**

Melkersson Rosenthal Syndrome (SMR) is a rare orofacial granulomatosis of controversial etiology. It consists of a triad of symptoms: plicatural or scrotal tongue, recurrent episodes of labial edema, paresis or unilateral facial palsy. The diagnosis is clinical and confirmed by histology [1].

Through a case of SMR we recall the clinical and therapeutic peculiarities of this pathology.

**OBSERVATIONS**

A 36-year-old patient who presented at our consultation for labial and gingival edema of progressive worsening, progressive in relapses for five years. The interrogation in the antecedents of the concept of transient facial paralysis.

Examination shows predominant lip edema in the upper lip (Figure-1), edema, and superior gingival hypertrophy (Figure-2) and a creased tongue (Figure-3).

The diagnosis of SMR has been made. A lingual and labial biopsy was performed, but the histological examination was negative. The view of the diagnostic clinic SMR was selected.

Patient under prolonged corticosteroid treatment in degressive dose, which resulted in a clear clinical improvement with a significant decrease of the edema. Currently, not recidivism after a decline of nine months.



**Fig-1: Front and side photo showing labial edema predominant in the upper lip**



**Fig-2: Photo showing hypertrophy and edema of the maxillary gingiva**



**Fig-3: Photo showing a plicate tongue**

## DISCUSSION

SMR is an orofacial granulomatosis manifested by a symptomatic triad: a plicate tongue, a macrocheilitis and recurrent facial paralysis or paresis, this triad is rarely complete and some minor signs can be observed.

Hubschman in 1894 described the first case without being able to establish the relationship between the three symptoms. In 1928, Melkersson made the link between macrocheilitis and facial palsy, then Rosenthal

identified the three signs that constitute the characteristic triad [1, 2].

SMR is rare. Its prevalence is estimated at 0.08% [3]. The beginning of the attack is around adolescence and then the syndrome is formed in several years. The distribution is equal in both sexes [2].

Its etiology is controversial. Some authors have referred to an autosomal dominant genetic origin with incomplete penetrance [4]. For others, it is rather a

neurological or neurovascular disorder circumscribed to an allergic or infectious stimulus [5]. The vasovasorum disorder of the small vessels of the head and neck can lead to edema of the facial nerves and subcutaneous tissue. Finally, the viral or bacterial infectious etiology (syphilis, oral infection, herpes, viral meningitis) was mentioned.

SMR has a multiple or mono symptomatic form. Miescher granulomatous cheilitis can be considered as a monosymptomatic form [6]. It evolves initially by edematous, painless, spontaneously resolute thrusts. It more frequently reaches the upper lip. The residual oedematous infiltration worsens with each push to become firm and elastic. Moderate erythema of oedematous areas can be seen [7]. The facial paralysis is of uni or bilateral peripheral type. The beginning is abrupt or progressive, the evolution is intermittent then permanent. To this can be added the involvement of the olfactory nerve, the glosso-pharyngeal nerve, the hypoglossal nerve resulting in hyposmias or parosmias, dysgeusia, and headaches. Edema can spread to other regions: the soft palate, the uvula, the tonsillar pillars, the gum or the cheek [1]. Paton [8] described frequent ophthalmic involvement associated with the syndrome (tearing, keratitis, involvement of the retinal vessels).

The diagnosis is mostly clinical and confirmed by histology. But a non-contributory histological analysis should not challenge the diagnosis of SMR when the clinical symptomatology is obvious [1]. This is the case of our patient who presented a complete symptomatology, but the histological examination was negative.

Therapeutically, no treatment claims to cure this disease. Systemic corticosteroids are the most recommended treatment. It improves the quality of life and avoids or space recurrences. The prognosis remains good [1].

In case of facial paralysis, the general corticosteroids are used at the dosage of 0.5 to 1 mg / kg / day for 10 to 20 days with progressive decrease according to the evolution of the symptoms. In the absence of recovery after one or two months, neurolysis or surgical nerve decompression can be proposed [2]. For Miescher macrocheilitis, injectable intra-lesional corticosteroids are often used iteratively as triamcinolone (kenacort Retard \*). A single injection resulted in complete remission in four cases with a follow-up of 6 months [9]. Repeated injections can be done every 3 to 6 months.

In the absence of evolution of macrocheilitis, reduction cheiloplasty is proposed by several teams. It is advisable to use pre- and post-operative corticosteroid therapy to avoid immediate relapse [1]. The plicated tongue does not require any treatment.

In all cases, given the usual benignity of the disease, it is necessary to adapt the therapy taking into account the side effects of the different treatments used.

## CONCLUSION

SMR is a rare orofacial granulomatosis. It consists of a triad of symptoms: plicature tongue, labial edema and paresis or unilateral facial paralysis. The diagnosis is mostly clinical. Treatment is mainly based on systemic corticosteroid therapy and the prognosis remains good.

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