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Melkersson-Rosenthal Syndrome: An Atypical Presentation with Facial Diplegia (A Case Report and Literature Review)

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Abstract

Melkersson-Rosenthal Syndrome is a rare entity of unknown etiology, characterized by the triad of symptoms: recurrent face and / or lip swelling, cracked or scrotal tongue and peripheral facial palsy. This symptomatology is often incomplete. It usually begins in childhood and relapses are common. We report a case of a patient with MRS followed in the ENT and Neck Surgery department of the University Hospital August 20th of Casablanca.

Index terms— facial paralysis - fissured tongue - macrocheilia - melkersson-rosenthal- facial diplegia.

1 I. Introduction

elkersson-Rosenthal Syndrome (MRS) is an orofacial granulomatosis of unknown etiology, characterized by one or more of the following symptoms: hypertrophy of eyelids and / or lips, intermittent facial nerve palsy and fissured tongue. The diagnosis of this affection is mainly clinical and the evolution is often marked by relapses or unsightly scars. We report a very rare form of MRS and through a literature review, we discuss the diagnostic and therapeutic difficulties of this condition.

2 II. Case Report

A 29 year-old male with no noticeable medical history, presented to the ENT department of university hospital August 20th of Casablanca with a lower macrocheilia with repeated facial nerve palsy (two episodes of left-sided facial paralysis) lasting for 4 years, in a context of apyrexia and conservation condition. Physical examination found facial diplegia and hypertrophy of the lower lip (Figure **??**), there was no fissured tongue. The rest of examination of the ENT area showed no signs of infection. Neurological examination was unremarkable: no paralysis of other cranial nerves nor limbs.

Before the Facial diplegia, MRI of the brain and the cerebellar pontine angle was considered and showed no abnormalities (figure **??**). Other investigations performed in search of tuberculosis and sarcoidosis were negative. A biopsy of the lower lip has confirmed the diagnosis showing: a connective tissu with abundant noncaseating nodules rich of lymphocytes bordered by plasma and histiocytic cells in edematous and fibrotic connective tissue by place. Given these clinical, paraclinical and evolutionary findings, the diagnosis of MRS was retained. The patient was put under medical treatment, based on long-term corticosteroid cure spread over 6 weeks decreasingly. The initial dose was 1 mg / kg / day. The evolution was marked by the improvement of symptoms from the first week of treatment. Both facial paresis and labial hypertrophy resolved 3 weeks later. During 14 months of follow-up there were no recurrences.

3 III. Discussion

The association of orofacial edema and intermittent facial palsy was initially described by Melkersson in 1928. Three years later, Rosenthal has added fissured tongue as a third symptom of this affection. This triad was named and known as MRS by Lüscher in 1949 (1). However, this association is rarely complete, only found in 8-18% of all cases (2, 3), which makes its diagnosis and eventually its management difficult. This rare syndrome represents

only 0.08% of facial paralysis (1,4) with less than 300 cases reported in the literature until now. Bilateral facial palsy in MRS is even rarer. These symptoms usually occur in the second decade of life, with no racial or gender predilection (3,5), although some authors have reported female predilection with sex ratio of 1/8 (4,6). The etiology of MRS remains unknown, many theories have been suggested, including the involvement of infectious agents, allergic, autoimmune or genetic factors (4, 5, 7). MRS may be secondary when associated with a known condition as sarcoidosis, tuberculosis, Crohn's disease, a tooth abscess, a facial trauma, an insect bite, or an orofacial granuloma. (4,5). Histological examination (5,8) mostly shows lymphocytes and epithelioid granulomas without caseous necrosis with a vasculotrope character. However, at the early stage of the disease, there may be just a perivascular lymphocytes infiltrate. Treatment remains discussed, primarily based on systemic and/ or intralesional steroids, sometimes supplemented by a Cheiloplasty (5,8,9). Antileprosy (clofazimine and dapsone) may be an alternative therapy in case of failure of corticosteroids (10) or may be combined with the latter. Others are also used as cyclins, hydroxychlorine, sulfasalazine, metronidazole, methotrexate, azathioprine and cyclosporine, but with no proven results (9, **??**0).

Despite the benign nature of its various symptoms, MRS course remains random often marked by frequent relapses with dreadful unsightly complications (5).

4 IV. Conclusion

The rarity of MRS limits the researches which still required to understand the mechanism generating this condition and try to find effective remedies.

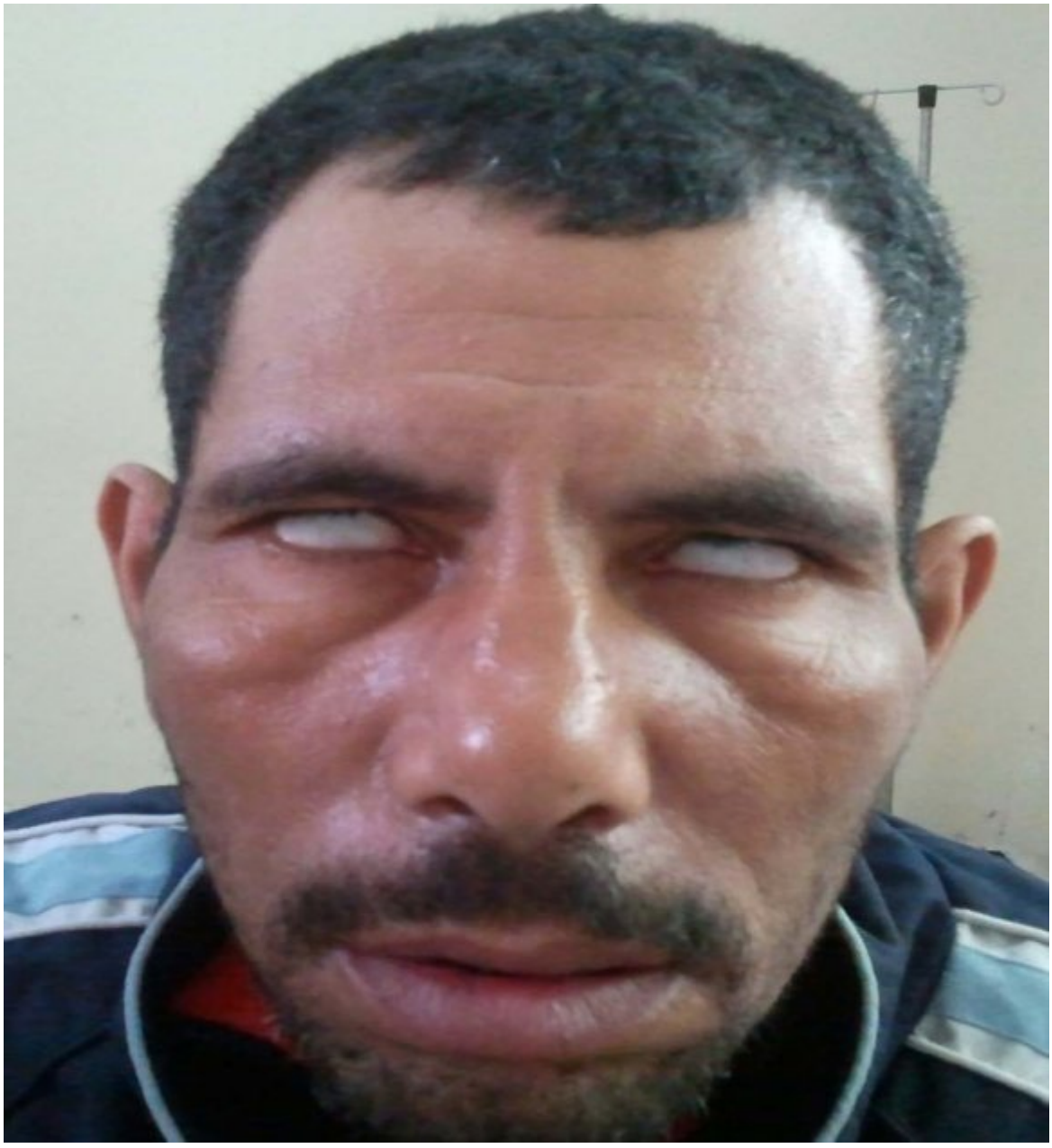

Figure 1:

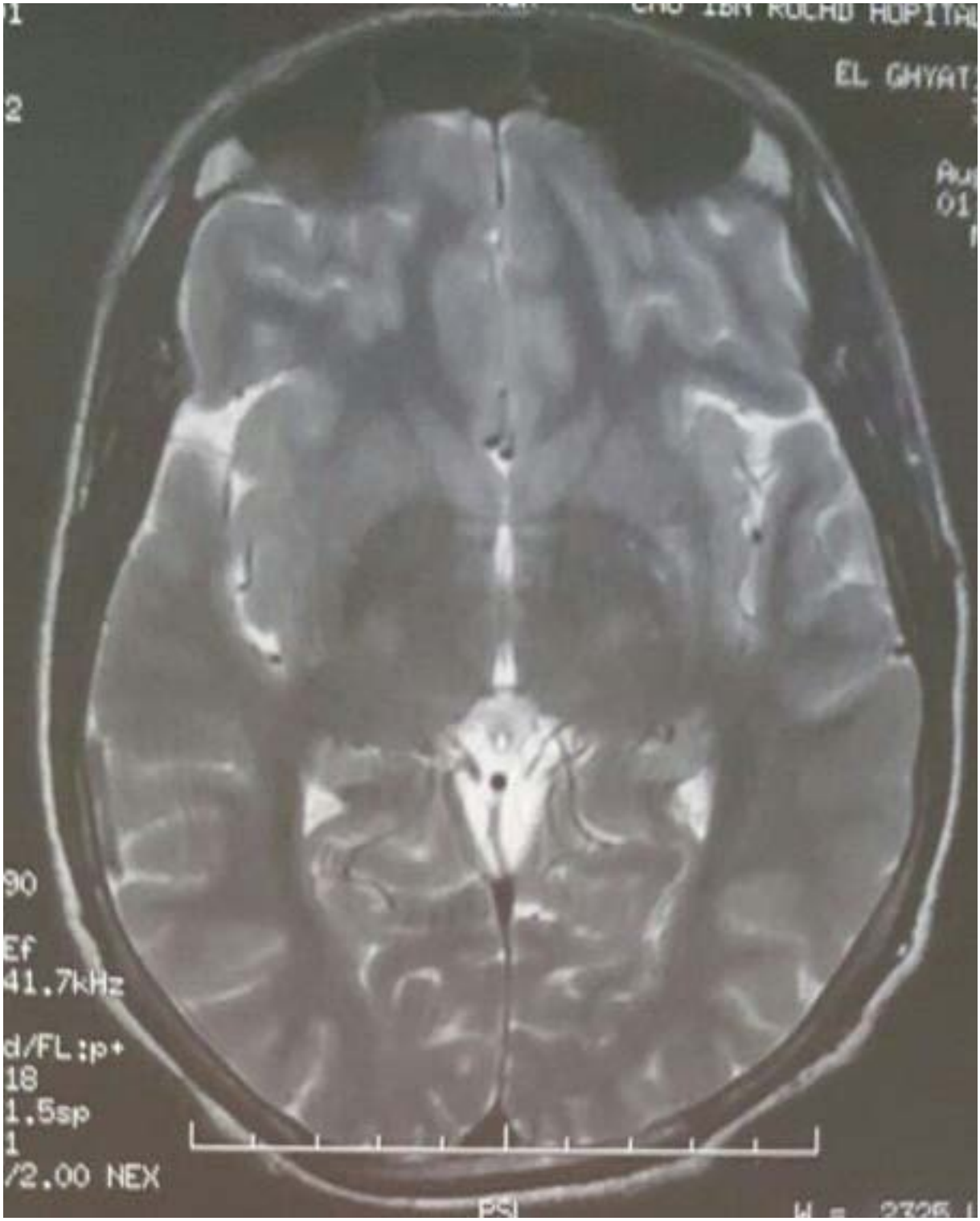

Figure 2:

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