SÍNDROME DE MELKERSSON-ROSENTHAL

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MELKERSSON-ROSENTHAL SYNDROME

ABSTRACT – The Melkersson-Rosenthal Syndrome is composed of the Triad: lingua plicata, intermittent facial palsy and orofacial swelling. Usually, the dominant and earliest finding of the syndrome is the orofacial swelling. Therefore, it’s frequently the dermatologist the first professional to have contact with the patient. Yet, the condition has been few times described in dermatological literature. We present a classical case of the complete triad.

KEY-WORDS – Melkersson-Rosenthal Syndrome; Clofazimine; Tongue, fissured; Facial paralysis.

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INTRODUCTION

Merkelsson-Rosenthal Syndrome is a rare condition composed of the triad: recurrent orofacial swelling, lingua plicata e intermittent facial nerve palsy^{1,2}. The Syndrome was first described by Melkersson in 1928 in its incomplete form. In 1931 Rosenthal added lingua plicata to the syndrome. Since 1949 the triad is known as Melkersson-Rosenthal syndrome^{1,3}. We describe a case of this condition in its complete form.

CASE REPORT

Female, 22 years old with a four year history of recurrent inferior lip swelling. Firstly the swelling was intermittent but it developed to persistency with periodic exacerbations (Fig. 1). She complained also of an episode of facial palsy one year earlier. The patient denied the use of medications, gastrointestinal symptoms and cough. Physical examination showed edema of the inferior lip mainly on its right portion and the presence of lingua plicata and macroglossia (Fig. 2). There were no infectious signs on the dental area. The eletroneuromyography showed evidence of previous right facial nerve palsy with signals of aberrant reinnervation. The
Caso Clínico

A histopathological examination added to the diagnosis as it uncovered the presence of lymphocytic granulomata in the skin of the inferior lip. We began treatment with clorfazimine 100mg/day which she used for two years with great clinical improvement. At the moment she is with no treatment for a year and shows no signs of remission (Fig. 3).

DISCUSSION

We present a classical case of Melkersson-Rosenthal Syndrome. The condition is more frequent in young adults and has similar distribution among the sexes. The dominant clinical feature of the triad is facial swelling and it happens mostly on the lip in the form of granulomatous cheilitis. Nevertheless, the complete triad happens only in 13%-25% of cases. The diagnosis is often difficult since the symptoms seldom present simultaneously. The histopathological examination shows non-caseating granulomata. However, it is necessary clinical correlation since diseases like sarcoidosis and Crohn’s disease have similar histopathology. Clinically should be excluded gastrointestinal alterations that may suggest Crohn’s disease. The patient should also be queried about the presence of chronic cough which would suggest sarcoidosis, histoplasmosis and tuberculosis. The use of medications such as ACE inhibitors and calcium channel blockers, as well as other diseases like Rosacea and Angioedema are differential diagnosis too.

The treatment remains a challenge and it will depend on the severity of clinical manifestations. The majority of patients with granulomatous cheilitis benefit from the use of topical corticosteroids such as triamcinolone or clobetasol in orobase. More extensive cases may respond to intralabial triamcinolone or to short courses of systemic glucocorticoids. Clorfazimine, hydroxychloroquine or sulfasalazine are attractive alternatives to the last. In case of disfiguring or recurrent swelling of the lip the surgical treatment is sometimes essential. On the other hand, facial palsy treatment is hard and none of the available options seems to change its evolution. Clorfazimine was chosen to the management of this patient leading to a well succeeded treatment.

REFERENCES