SÍNDROME DE MELKERSSON-ROSENTHAL

Daniel Holanda Barroso[1], Camila Pinon Medeiros Zoby[2], Ana Carolina Depes Perdigão e Vasconcelos[1], Silvana Maria de Morais Cavalcanti[2], Emmanuel Rodrigues de França[3], Márcia Almeida Galvão Teixeira[2]
[1]Residente de Dermatologia Universidade de Pernambuco, Recife, Brasil/Resident Dermatology, University of Pernambuco, Recife, Brazil
[2]Dermatologista, Doutora em Medicina Tropical, Universidade de Pernambuco, Recife Brasil/Dermatologist, Doctor in Tropical Medicine, University of Pernambuco, Recife, Brazil
[3]Dermatologista, Doutor em Dermatologia, Universidade de Pernambuco, Recife, Brasil/Dermatologist, Doctor in Dermatology, University of Pernambuco, Recife, Brazil

RESUMO – A síndrome de Melkersson-Rosenthal é composta pela tríade: língua plicata, paralisia facial intermitente e edema orofacial. O achado dominante e mais precoce é o edema orofacial. Desta forma, o dermatologista é frequentemente o primeiro profissional a ter contato com o paciente. A condição tem sido pouco descrita em revistas dermatológicas. Apresentamos um caso clássico da tríade completa.

PALAVRAS-CHAVE – Síndrome de Melkersson Rosenthal; Clofazimina; Língua fissurada; Paralisia facial.

MELKERSSON-ROSENTHAL SYNDROME

***ABSTRACT** – The Melkersson-Rosenthal Syndrome is composed of the Triad: linqua 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, tissured; Facial paralysis.*

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.
No conflicts of interest.
Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.
No sponsorship or scholarship granted.
Direito à privacidade e consentimento escrito / ***Privacy policy and informed consent***: Os autores declaram que pediram consentimento ao doente para usar as imagens no artigo. *The authors declare that the patient gave written informed consent for the use of its photos in this article.*

Recebido/*Received* - Maio/*May* 2014; Aceite/*Accepted* – Junho/*June* 2014

Correspondência:
Dr. Daniel Holanda Barroso
Rua Amapá nº: 77 Apartamento: 602 Cep
52050390 Recife, Pernambuco, Brasil
E-mail: danielhbarroso@gmail.com

Caso Clínico

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

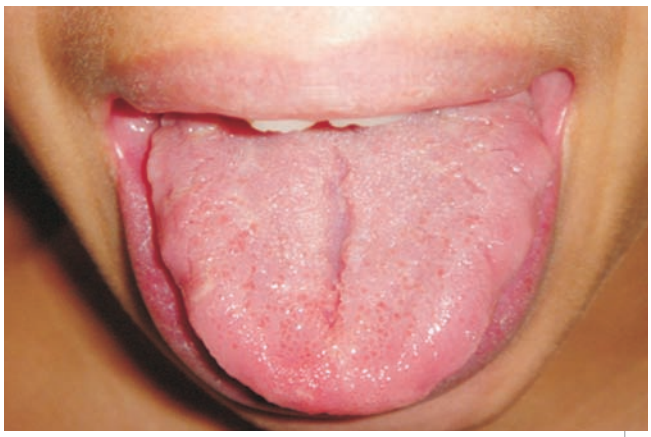

Fig 2 - Lingua *plicata*.

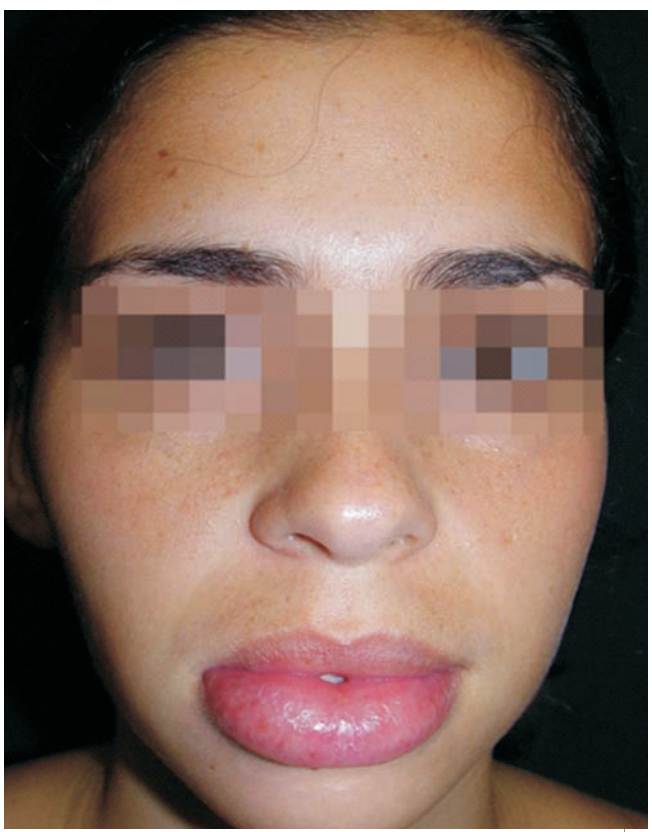

Fig 1 - Important inferior lip swelling.

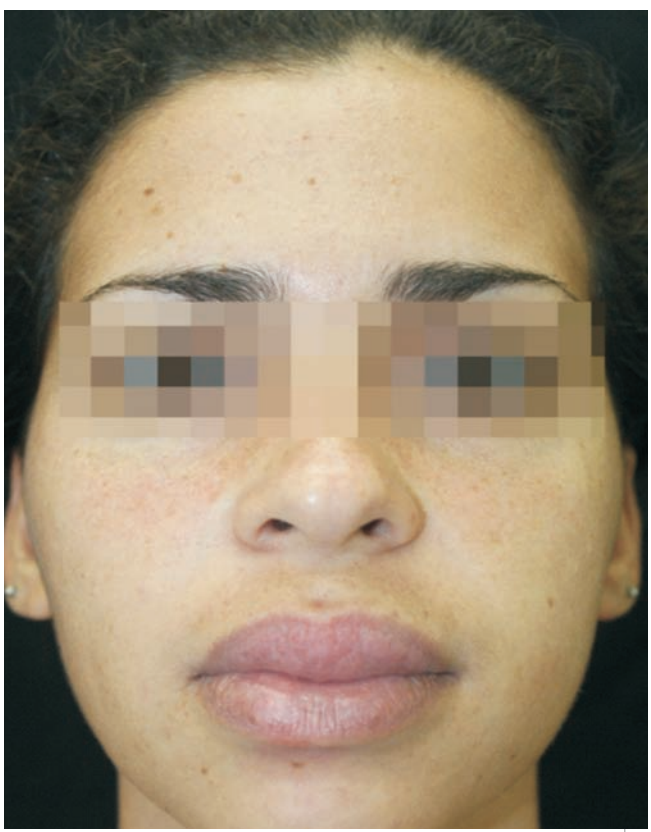

Fig 3 - Aspect of the lips one year after the completion of treatment.

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[4]. The Dominant clinical feature of the triad is the facial swelling and it happens mostly on the lip in the form of granulomatous cheilitis[1]. Nevertheless, the complete triad happens only in 13%-25% of cases[1,2]. The Diagnosis is often difficult since the symptoms seldom present simultaneously[1].

The histopathological examination shows non-caseating granulomata. However, it is necessary clinical correlation since diseases like sarcoidosis and Crohn's disease have similar histopathology[4]. 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[5].

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 intralesional triamcinolone or to short courses of systemic glucocorticoids. clofazimine, hydroxychloroquine or sulfasalazine are attractive alternatives to the last[6]. In case of disfiguring or recurrent swelling of the lip the surgical treatment is sometimes essential[7]. On the other hand, facial palsy treatment is hard and none of the available options seems to change its evolution[2]. Clorfazimine was chosen to the management of this patient leading to a well succeeded treatment.

REFERENCES

1. Greene RM, Rogers RS 3rd. Melkersson-Rosenthal syndrome: a review of 36 patients. J Am Acad Dermatol. 1989 ;21(6):1263-70.
2. Elias MK, Mateen FJ, Weiler CR. The Melkersson--Rosenthal syndrome: a retrospective study of biopsied cases. J Neurol. 2013;260(1):138-43.
3. LUSCHER E. Syndrom von Melkersson-Rosenthal. Schweiz Med Wochenschr. 1949;79(1):1-3.
4. Rogers RS 3rd. Melkersson-Rosenthal syndrome and orofacial granulomatosis. Dermatol Clin. 1996;14(2):371-9.
5. Critchlow WA, Chang D. Cheilitis Granulomatosa: A Review. Head Neck Pathol. 2013 [*in press*].
6. van der Waal RI, Schulten EA, van der Meij EH, van de Scheur MR, Starink TM, van der Waal I. Cheilitis granulomatosa: overview of 13 patients with long--term follow-up - results of management. Int J Dermatol. 2002;41(4):225-9.
7. Kruse-Lösler B, Presser D, Metze D, Joos U. Surgical treatment of persistent macrocheilia in patients with Melkersson-Rosenthal syndrome and cheilitis granulomatosa. Arch Dermatol. 2005;141(9):1085-91.