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MELKERSSON-ROSENTHAL SYNDROME COMPLICATED WITH FACIAL HEMISPASM AND INTRACRANIAL HYPERTENSION.

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ABSTRACT.

Since it was first described in 1928 by neurologist Ernest Melkersson¹¹, and complemented in 1931 by Curt Rosenthal¹², the Melkersson-Rosenthal Syndrome (SMR) has been shown to be a rare disorder and still in need of specific treatment^{13,14}. We describe the case of a young patient, who at 23 years old had the first episode of peripheral facial paralysis on the right. At the time, it was conducted in a protocol manner, with prednisone, paracetamol and eye care, as well as pertinent physiotherapy. There was an improvement in facial asymmetry, when in 2006 she presented a new peripheral facial paralysis, this time on the left, which was conducted in a similar way. Then, she presented 2 more episodes of facial paralysis in the interval of 3 years, already with aesthetic sequelae. In 2009, in the 5th episode of paralysis, she associated facial edema and cheilitis, when biopsy of the upper eyelid was requested, which was unspecific. In 2012, after the 8th facial paralysis, she underwent a new biopsy, in which an inflammatory infiltrate was demonstrated, consistent with clinical suspicion of SMR. Imaging exams (skull resonance and angioresonance), face electroneuromyography - for prognosis, and CSF study, without abnormalities, were performed. That year she began to experience involuntary and rhythmic contractions of the orbicularis musculature of the eyes and mouth on the right, in addition to risory and platysma, featuring facial hemispasm, treated every 4 to 6 months with 100 IU Onabotulinum. Throughout the treatment period, alternating use of 10mg prednisone with deflazacort 6mg, and in 2019, due to persistent headache, she underwent a new resonance with arterial and venous cranial angioresonance, which brought elements suggestive of Intracranial Hypertension (IH). Currently, the patient is undergoing surgical programming for facial nerve decompression bilaterally and undergoing treatment for IH.

Keywords: facial hemispasm, cheilitis, intracranial hypertension, facial edema, facial paralysis.

Introduction.

Facial paralysis and edema were first described by Melkersson in 1928¹¹. Three years later, Rosenthal associated these symptoms with a cleft tongue¹². his triad was called Melkersson-Rosenthal syndrome by Lüscher in 1949¹. However, some authors argue that the complete syndrome is rare, being the most common monosymptomatic or sequential presentation².

The Melkersson-Rosenthal syndrome has an estimated incidence of 0.08% and an unknown etiology³. According to Ang and Jones, 2002, there is a slight predilection for the female sex, usually around the second decade of life⁴, there is no racial predilection⁵ and there are no reports that associate S.M.R with malignant transformation⁶. The syndrome has a classic triad, consisting of recurrent or persistent orofacial edema, recurrent facial paralysis and plicate or scrotal tongue, which occurs very rarely⁷. Frequently, SMR manifests itself in its oligosymptomatic (combination of 2 signs) or monosymptomatic forms, such as Miescher's granulomatous macrocheilitis, which can be a diagnostic difficulty^{5,7}.

The first manifestation is, predominantly, acute diffuse edema involving the upper lip (75-100%), and less the lower lip, which completely regresses in hours or days⁷. O Edema develops through outbreaks, sometimes unilateral, for days or weeks, at irregular intervals, with an increase in volume that can reach two to three times the

normal size until a permanent infiltration is established⁷. The involvement of the intraoral mucosa may also appear, namely edema of the gums, palate, oral, sublingual mucosa, tongue and even pharynx and larynx^{5,7}.

Peripheral facial paralysis occurs as an initial manifestation in 30 to 50% of patients with SMR. Clinically, it is indistinguishable from Bell's Palsy¹⁵, usually having a sudden onset or, rarely, gradually over 24 to 48 hours, with complete resolution after a few weeks⁷. Although at first it is intermittent, it can become persistent⁷. Facial paralysis often develops after granulomatous cheilitis, but there are reports of cases in which it precedes it for several months or years or occurs simultaneously⁷. In most cases it is unilateral, corresponding (although not always) alongside orofacial infiltration and, rarely, it is bilateral, it can be partial or complete⁷.

The diffusely fissured tongue (plicated or scrotal tongue) is described in 20 to 60% of patients with SMR and may be associated with a burning sensation, edema, loss of taste and decreased salivary secretion⁷. Some signs and symptoms are defined as minor criteria that are also part of the Melkersson-Rosenthal syndrome⁸. The involvement of other cranial pairs, migraine and dysfunctions of the salivary and lacrimal glands and pupillary motricity constitute these minor criteria, in addition to the presence of hyperhidrosis, hyperacusis, acroparesthesia, epiphora, hypergeusia and multiple ophthalmological findings such as lagophthalmos, keratitis by exposure,

blepharocalasia, retrobulbar neuritis, anomaly of the retinal veins and paralysis of the medial rectus muscle.

The histopathology of cutaneous or mucous biopsies in patients with SMR may show, in an initial phase, only a nonspecific lympho-plasmacytic and histiocytic infiltrate, predominantly perivascular, which may represent the primordial stage of granuloma formation⁷. However, even in complete forms, this granulomatous infiltrate is not observed in more than half of the cases and its absence should not exclude the diagnosis of SMR⁷.

The involvement of unusual locations in the orofacial region and the presence of minor isolated manifestations often precede the development of typical clinical manifestations. This fact, associated with the underestimation of symptoms by the patient himself, makes, in most cases, difficult to make a correct diagnosis early or even probable^{7,14}. This way, SMR has been shown to be a rare disorder and still in need of specific treatment^{13,14}. In the present report, we describe a case not yet described in the literature, in which the patient initially treated as Bell's Palsy, evolved with facial edema, cheilitis, right facial hemispasm and Idiopathic Intracranial Hypertension Syndrome (IIH).

Case description.

SSL, 23 years old, domiciled in São Mateus, ES, presented peripheral facial paralysis of the right hemiface in 2005 and was treated as Bell's Palsy with corticosteroid and analgesic. There was an improvement in asymmetry on the face, when in 2006 she presented a new paralysis, this time on the left, which

was conducted in a similar way. Then, she presented 2 more episodes in the interval of 3 years, already with aesthetic sequelae. In 2009, in the 5th episode of paralysis, she associated facial edema and cheilitis, when the SMR hypothesis was suggested, but with an upper eyelid biopsy that was unspecific. In the period from 2009 to 2012, the patient had conductive and sensorineural hearing loss in her left ear and had several episodes of pain in the face refractory to clinical treatment that made her unable to work.

In 2012, after the 8th facial paralysis, she underwent a new biopsy, this time showing an inflammatory infiltrate consistent with clinical suspicion. Arterial resonance and angioresonance of the skull without abnormal findings at the time, electroneuromyography of the face - for prognosis, which revealed signs of neuropathic involvement of the right and left facial nerves with moderate / moderately severe axonal injury data, slightly more intense on the left, and a CSF study.

, without abnormalities. Later that year, she evolved with facial hemispasm, treated every 4 to 6 months with Onabotulinum 100 IU.

During the entire treatment period, alternating use of 10mg prednisone with deflazacort 6mg, and in 2019, due to persistent headache and visual blurring, she underwent a new resonance with arterial and venous cranial angioresonance, with elements suggestive of Intracranial Hypertension (IH). Currently, the patient is undergoing surgical programming for facial nerve

decompression bilaterally and undergoing treatment for IH.

Material and methods.

A bibliographic review of the Melkersson-Rosenthal Syndrome was carried out, a medical record survey and interview with the patient by telephone contact and message application, since the patient lives in the interior of Espírito Santo.

Discussion.

SMR is characterized by the recurrent orofacial edema and facial paralysis and plicated tongue¹⁶, characteristics present in the case. We did not find, in the literature, cases associating SMR with facial hemispasm and IH. We emphasize the importance of insisting on biopsy, in order to document the syndrome and institute appropriate therapy, based on prolonged corticosteroid therapy. Unresponsive cases are referred for surgical treatment of facial nerve decompression, bilaterally¹⁶. We believe that the patient's weight gain (BMI of 27.1 to 30.8 after 15 years of treatment), resulting from prolonged use of corticosteroids, may have led to the Intracranial Hypertension syndrome.

Conclusion.

In the present case, we highlight the treatment of one of the late complications of peripheral facial paralysis, which consists of facial hemispasm, currently amenable to symptomatic

treatment with intramuscular injections of botulinum toxin in the most reactive muscles. We also emphasize the importance of monitoring indirect causes of chronic corticosteroid therapy, such as IH resulting from overweight and obesity.

References.

- 1) Lüscher E. Syndrom von Melkersson-Rosenthal. Schweiz Med Wschenschr; 79:1-3, 1949.
- 2) Nossa, L.M.B.; Costa, A.L.; Marback, R.L. Melkersson-Rosenthal syndrome: a clinical-pathological study of a case. Arq. Bras. Oftalmol., Vol.64, nº6, São Paulo, Nov./Dec. 2001.
- 3) Glickman, L.T.; Gruss, J.S.; Birt, B.D.; Kohli-Dang, N. The Surgical Management of Melkersson-Rosenthal Syndrome. Plast. Reconstr. Surg., 89:815-21, 1992.
- 4) Ang, K. L.; Jones, N. S. Melkersson-Rosenthal syndrome. J. Laryngol. otol., London, v. 116, n. 3, p. 386-388, May. 2002.
- 5) Greene, R.M.; Rogers, R.S. Melkersson-Rosenthal syndrome: A review of 36 patients. J. Am. Acad. Dermatol., 21: 1263-9, 1989.
- 6) Vilela, D.S.A.; et all. Melkersson-Rosenthal syndrome: case report and literature review. Rev. Bras. Otorrinolaringol., vol.68, nº5, São Paulo, Ouc. 2002.
- 7) Soares, E.C.S.; et all. Melkersson-Rosenthal syndrome: case report. Rev. Cir. Traumatol. Buco-Maxilo-Fac., Camaragibe, v.6, nº.1, p. 29 - 36, jan./mar. 2006.
- 8) Alexander, R.W.; James, R.B. - Melkersson-Rosenthal Syndrome: review of literature and report of case. J. Oral Surg., 30: 599-604, 1972.

- 9) Cairns, R.J. Melkersson Rosenthal Syndrome. Proc. R. Soc. Med., 54: 217, 1961.
- 10) Hornstein, O.P.; Stosiek, N.; Schönberger, A.; Meisel-Stosiek, M. Classification and clinical variation of Melkersson-Rosenthal Syndrome. Rev. Z. Hautkr; 62: 1453-1475, 1987.
- 11) Melkersson, E. A case of recurrent facial paresis in connection with angioneurotic edema. Rev. Hygiea, 90: 737-41, 1928.
- 12) Rosenthal, C. Clinical-hereditary contribution to constitutional pathology: common occurrence of facial paralysis (recurrent family member), angioneurotic facial edema and plicated tongue in families with arthritis. Rev. Z Neurol Psychiatr, 131: 475-501, 1931.
- 13) González-García, A.; Barbolla, I. D.; Sifuentes, W.A. G.; Patier-de la Peña, J.L. Miescher syndrome: a rare cause of lip swelling. Rev. Reumatol Clin., 13:363-364, 2017.
- 14) Martínez, M.L; et al. Granulomatous cheilitis. Presentation of 6 cases and review of the literature. Rev. Actas Dermosifiliogr., 103: 718-24, 2012.
- 15) Carvalho, V.A.S.; et al. Unilateral facial palsy: clinical aspects and main treatments. Rev. Brazilian Journal of Health, Curitiba, v. 3, n. 2, p.1761-1765 mar./apr. 2020.
- 16) Gonçalves, D.U.; et al. Granulomatous cheilitis associated with Melkersson-Rosenthal syndrome. Rev. Bras Otorrinolaringol, 73(1):138-9, 2007.