Available online at www.scientiaresearchlibrary.com

Scientia Research Library

ISSN 2348-0416
USA CODEN: JASRHB

Journal of Applied Science And Research, 2020, 8 (1):53-59

(*http://www.scientiaresearchlibrary.com/arhcive.php*)

Facial edema revealing Melkersson-Rosenthal syndrome: a case report

J. OUJABER (1), B. NISRINE, Z. CHAHBI, S. HADDOURI, S. QACIF, M. ZYANI (2), K. TOURABI (3), R. SIRBOU* (1)

(1): medico-surgical emergency service, Marrakech Avicenna military hospital.

(2) : service de médecine interne, hôpital militaire avicenne Marrakech.

(3): plastic and restorative surgery service,Avicenna military hospital Marrakech.

CASE REPORT

ABSTRACT

Melkersson-Rosenthal syndrome (SMR) is a rare entity. It is a granulomatosis Facial affecting both sexes with a triad of bucco-facial manifestations: mucocutaneous edema of the face, recurrent peripheral facial paralysis and a plicature tongue. . 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 macrochemistry and facial paralysis, then Rosenthal identified the three signs that constitute the characteristic triad). Through this observation, we present the clinical and therapeutic features of this pathology 46 year old patient consults in the emergency for the sudden appearance of edema of the face, a feeling of numbness and a swelling affecting the labial region, attributed to a food allergy according to the patient. An edema covering the middle floor of the face and the right and left latero-mandibular region The charles Bell negative sign, negative eyelash sign, no hypoesthesia The endo buccal examination reveals a pleated appearance of the tongue, gingival inflammation, the absence of dental infectious . Objective joint osteo examination of diffuse myalgia, poly arthralgia of large inflammatory joints. A salivary gland biopsy confirmed the diagnosis, finding a histological appearance compatible with chronic granulomatous epitheloid and giganto-cellular sialdenitis in the absence of caseous necrosis, on an inflammatory lymphocytic background.. A protocol adopting a monthly intra-lesional injection of 40 mg of triamcinolone acetonide (Kénacort Retard®) in each lip was undertaken giving satisfactory results.

INTRODUCTION

Melkersson-Rosenthal syndrome (SMR) is a rare entity. It is a granulomatosis

Facial affecting both sexes with a triad of bucco-facial manifestations: mucocutaneous edema of the face, recurrent peripheral facial paralysis and a plicature tongue. (1,2).

The triad of SMR is often incomplete and certain minor signs are frequently observed.

There is a fairly characteristic histology, it is obliterating epitheloidendovascularitis

Several theories have been advanced suggesting an infectious cause, a genetic predisposition or an

allergic cause.

Recently, researchers identified a monoclonal lymphocyte expansion in lesions of orofacial granulomatosis (GOF) and suggested that it could be secondary to chronic antigenic stimulation (4).

Its treatment is mainly medical, sometimes surgical.

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 macrochemistry and facial paralysis, then Rosenthal identified the three signs that constitute the characteristic triad (3).

Through this observation, we present the clinical and therapeutic features of this pathology.

MATERIALS AND METHOD

OBSERVATIONS

46 year old patient consults in the emergency for the sudden appearance of edema of the face, a feeling of numbness and a swelling affecting the labial region, attributed to a food allergy according to the patient.

The clinical examination on admission to the emergency room found a patient stable on the respiratory hemodynamic neurological level GS = 15/15, TA = 14/8, spo2 = 100% FR = 24cpm FC = 105bpm

The patient was apyretic to the touch .

The examination of the face: objective a homogeneous swelling of the 2 painless lips

Elastic, occupying the vermilion and the red lip an interesting edema on palpation this swelling has a firm and fibrous consistency without dysphagia or odynophagia.

An edema covering the middle floor of the face and the right and left latero-mandibular region

The charles Bell negative sign, negative eyelash sign, no hypoesthesia .

The endo buccal examination reveals a pleated appearance of the tongue, gingival inflammation, the absence of dental infectious foci.

Objective joint osteo examination of diffuse myalgia, poly arthralgia of large inflammatory joints.

The pleuro pulmonary examination was without particularity.

Ophthalmological and ENT examination without abnormality.

The rest of the examination was normal, in particular there was no dry eye or mouth syndrome, no facial paralysis or neurological signs.

The medical history of the patient finds a left peripheral facial paralysis dating from a year preceding the onset of macro-cheilitis of the 2 lips and having regressed under medical treatment without leaving any after-effects.

In addition, the patient does not report any feeling of chronic fatigue or any intestinal problem that could suggest an inflammatory bowel disease.

Faced with the association of the facial paralysis triad, machro cheilitis and fissured aspect of the tongue, the diagnosis of SMR was retained.

The patient is hospitalized in the internal medicine department in order to eliminate other possible etiologies (sarcoidosis, crohn's disease, tuberculosis, leprosy syphilis, systemic mycosis…)

A salivary gland biopsy confirmed the diagnosis, finding a histological appearance compatible with chronic granulomatous epitheloid and giganto-cellular sialdenitis in the absence of caseous necrosis, on an inflammatory lymphocytic background.

Biological examinations (NFS, TP, TCA, sedimentation rate, renal, hepatic function, calcium phosphorus assessment, LDH, EPP, thyroid assessment, c2 c4 complement level) are normal

The serologies (HIV, HVB, HVC, TPHA, VDRL) are negative, the spirometry was normal the cerebral and thoracoabdomino pelvic scanner and the colonoscopy found no abnormality.

A protocol adopting a monthly intra-lesional injection of 40 mg of triamcinolone acetonide (Kénacort Retard®) in each lip was undertaken giving satisfactory results

RESULTS AND DISCUSSION

In 1928, Melkersson described facial edema and facial paralysis in a 35-year-old patient and suggested that there was a relationship between the two symptoms. Rosenthal added, in 1931, a new symptom, the plicatured tongue. The triad will be designated, in 1949, under the name of SMR (5)

SMR occurs in young adults, most often after the end of the 2nd decade, without predominance of sex or race (6).The pathogenesis of SMR remains unknown.

Stosiek et al. (1992) propose an inflammatory theory: the presence of non-specific circulating antigens would cause vasomotor disorders of the small vessels of the cephalic end, giving subcutaneous edema and nerve sheaths (7).

Other mechanisms have been mentioned: anomalies in lymphatic drainage, allergic phenomena (food, cosmetics, certain components of products used for oral hygiene), bacterial or viral origin (herpes, toxoplasmosis, syphilis, etc.), role of dental infectious foci and periodontal (5.7).

Indeed the elimination of infectious foci of dental origin often present in the carriers of SMR can bring a sometimes spectacular improvement of the edema, which seems to plead in favor of the infectious theory in particular bacterial, but there is no evidence real scientists (6).

The SMR could represent a particular form of sarcoidosis or Crohn's disease. Currently, many authors consider that there is a constitutional or hereditary predisposition.

The SMR takes a pluri or monosymptomatic form. The granulomatous cheilite of Miescher can be considered a monosymptomatic form [8]. It evolves initially by oedematous, painless, spontaneously resolving attacks. It reaches the upper lip more frequently, producing a lip-like appearance. Residual edematous infiltration worsens with each push to become firm and elastic. Moderate erythema of the edematous areas can be seen [9].

The facial paralysis is of a uni or bilateral peripheral type. The beginning is abrupt or progressive, the evolution is intermittent then permanent.

May be added the involvement of the olfactory nerve, the glosso-pharyngeal nerve, the large hypoglossal nerve causing hyposmia or parosmia, dysgeusia, and headache. The edema can spread to other regions: the soft palate, the uvula, the tonsillar pillars, the gum or the cheek. Paton [10] described frequent ophthalmic disorders associated with the syndrome (lacrimation, keratitis, damage to the retinal vessels).

The positive diagnosis is histological. According to Pindborg, Chomette and Auriol [8/9], the images Histological of the tongue or lips are identical. We discern a sub epithelial tissue With loose and diffuse edema. We find the histological appearance of giganto-epitheloid granulomas without caseous necrosis, with vascular tropism with endo vascularitis epithelioid obliterans.

Fibrosis lesions actually correspond to the scarring course of inflammation Lymph-nodular granulomatous.

A non-contributory histological analysis should obviously not reject the diagnosis of SMR when the clinical symptomatology is obvious.

The management of patients with SMR remains difficult. There is no basic or specific treatment [5]. It is essentially symptomatic and depends on the aesthetic repercussions and clinical signs (6) In case of facial paralysis, corticosteroid therapy

Générale is used at a dosage of 0.5 to 1 mg / kg / day for 10 to 20 days with progressive reduction depending on the evolution of symptoms. In the absence of recovery after one or two months, neurolysis or surgical nerve decompression may be proposed (11).

The surgical indication can be guided by electromyographic monitoring.

For Miescher macrochelitis, corticosteroid injection intra-lesion is often used iteratively in the form of triamcinolone (kenacort Retard *). A single injection allowed complete remission in four cases with a 6-month follow-up [12].

Repeated injections can be made every 3 to 6 months. Dermocorticoids are often tried with mostly null results.

Clofazimine (Lamprene) has often been used in this disease, in particular in Miescher's macrochelitis [13]. Its effectiveness is not constant. Recurrences are frequent when treatment is stopped. In isolated cases hydroxychloroquine (Plaquenil) [14], thalidomide and metronidazole have been effective. In the absence of scalability of the macrochelite, a reduction cheiloplasty is proposed by Several teams [15].

It is advisable to use corticosteroid therapy before and after surgery to avoid immediate relapse. The plicatured tongue requires no treatment.

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

CONCLUSION:

Melkerssonrosenthal syndrome is a rare condition characterized by the association of facial paralysis and oral manifestations: plicated tongue, labial macrocheilitis. The etiopathogenic mechanisms have not yet been elucidated. The management of patients with SMR remains difficult. No treatment exists. It is essentially symptomatic and depends on the aesthetic repercussions and clinical signs.

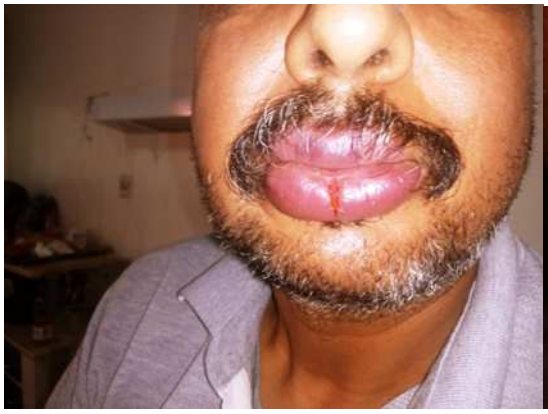

Figure 1 : Front view. Edema on the upper floor Middle of the face and the mandibular region.

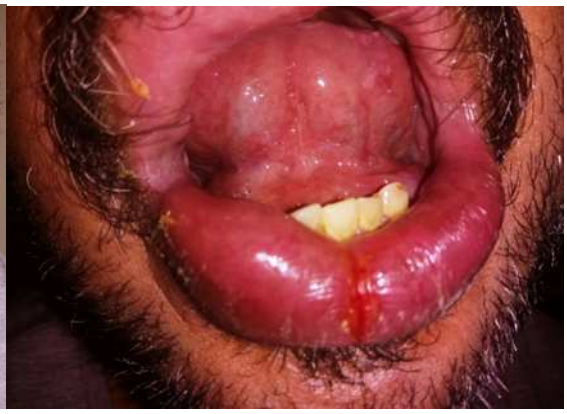

Figure 2 : filling of the anterior maxillary vestibule

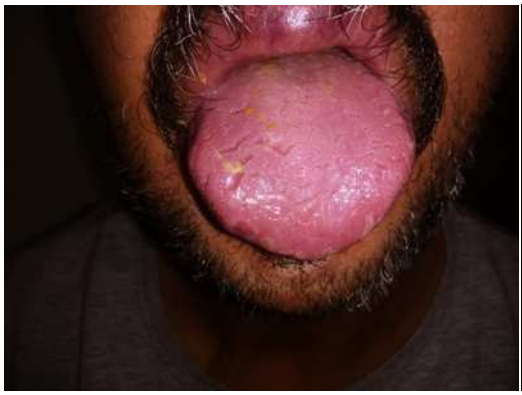

Figure 3 : Plication tongue.

Figure 4 : Kénacort Retard® intra-lesional injection

REFERENCE

[1] Elias MK, Mateen FJ, Weiler CR. The Melkerrson-Rosenthal syndrome: a retrospective study of biopsied cases. *J Neurol*, 260 (1): 138–143, **2013**.

[2] Sciubba JJ, Said-Al-Naief N. Orofacial granulomatosis: presentation, pathology and management of 13 cases. *J Oral Pathol Med*., 32: 576–85, **2003**.

[3] Hentati H, Hammami S, Oualha L, Mannai I, Selmi J. Melkersson-Rosenthal syndrome of a family nature: on a case. Med Buccale Chir Buccale, 13: 29-34, **2007**.

[4] Lim SH, Stephens P, Cao QX, Coleman S, Thomas DW. Molecular analysis of T cell receptor beta variability in a patient with orofacial granulomatosis. Gut, 40: 683–6, **1997**.

[5] Benyahya I, Mbirik K, El Kabli H. Syndrome de Melkersson-Rosenthal: about a case. actual.

Odonto-Stomatol **2004**; 227: 217-24.

[6] Cherkaoui A, Sahnoun I, Benrachadi L, Benzarti N. Melkersson-Rosenthal syndrome: oral aspects. Medical Hope. Dental Special **2004**; 52: 21-4.

[7] Stosiek N, Birolleau S, Capesius C, Hornstein OP. Chronicity and diagnostic uncertainties of

Melkersson-Rosenthal syndrome. *Ann Dermatol Venerol* **1992**; 119: 635-8.

[8] Pindborg JJ Atlas of diseases of the oral mucosa. Medsiedit. Paris **1981**.

[9] L. Vaillant. Granulomatouscheilites. *Ann Dermatol Venerol*. **2000**; 127: 149-53.

[10] Paton D. The Melkersson-Rosenthal syndrome: a case report. *Ann J Ophtalmol*. **1966**; 59, 705.

[11] G. LE TOUX. CheilitisGranulomatosa, Facial Neuropathy, OrofacialEdema .Current data and case report. *Ann Oto-Laryng*. **1989**; 106: 44-46.

[12] SAKUNTABHAI A, MAC LEOD RI, LAWRENCE CM. Intralesional steroid injection after nerve block anesthesia in the treatment of orofacial grnaulomatosis. *Arch Dermatol*. **1993**; 129: 477-480.

[13] POMODORE P, BURROWS D. Clofamizine: year effective treatment for Melkersson-Rosenthal syndrome or Mieascher's cheilitis. Clin Exp Dermatol. **1986**; 11: 173-178.

[14] ALLEN CM, CARMINA, HARNZEH S, STEPHEN L. Cheilitis granulomatosa: report of six cases and review of the literature. *J Am Acad Dermatol*. **1990**; 23: 444-450.

[15] ELLITSGAARD N, ANDERSSON AP, WORSAAE N, MEDGYESI S. Long-term results after surgical reduction cheiloplasty in patients with Melkersson-Rosenthal syndrome and cheilitis granulomatosa *Ann Plast Surg*. **1993**; 31: 413-420.