AUCTORES
Globalize your Research

Journal of Dermatology and Dermatitis
Justine Saucereau *

Open Access

Case Report

From Headache to Melkersson-Rosenthal Syndrome: A Case Report

Justine Saucereau [1]*, C. Legeas [1], B. Quere [2], V. Devauchelle-Pensec [2], P. Schoenlaub [1]

[1] Dermatology department, Clermont Tonnerre Instruction's army hospital, Brest, France

[2] Rheumatology department, INSERM 1227, reference center of rare auto immune diseases, Brest University Hospital, Brest, France

***Corresponding Author:** Justine Saucereau, Dermatology department, Clermont Tonnerre Instruction's army hospital, Brest, France.

Received date: December 02, 2024; **Accepted date:** February 15 2024; **Published date:** April 22, 2024

Citation: Justine Saucereau, C. Legeas, B. Quere, V. Devauchelle-Pensec, P. Schoenlaub, (2024), From Headache to Melkersson-Rosenthal Syndrome: A Case Report, *Dermatology and Dermatitis*, 10(3); **DOI:10.31579/2578-8949/135**

Abstract

Melkersson-Rosenthal syndrome is described as a syndrome which associates three cardinal signs: peripheral facial paralysis, facial oedema, and fissured tongue. These signs are rarely all present. A range of other symptoms are associated with its, making the diagnosis difficult.

We report a case of a twenty-five-year-old women presented recurrent facial paresis and clinical dry syndrome since several years. The facial paresis was secondarily accompanied by headache. After a complete dermatological examination, a Melkersson-Rosenthal syndrome has been suspected. Several treatments have been tried, reducing the symptoms, without making them disappear.

The Melkersson-Rosenthal syndrome is well known, but the characteristic triad is rarely present in its entirety. This syndrome is accompanied by a multitude of other symptoms of neurovegetative origin for the most part, and sometimes with autoimmune disorders, which can induce diagnostic delay and/or misdiagnosis.

Keywords: headache; melkersson-rosenthal; paresis; scrotal tongue; facial oedema

Introduction

Melkersson-Rosenthal syndrome is well described by its characteristic triad associating uni or bilateral peripheral facial paralysis, a facial oedema and fissured dorsal tongue [1-5] but the association of these three signs is present in only a minority of cases (8 to 45%) [1,2,4,5].

The association with a lot of other various symptom are frequent ; notably, hemicrania is sometimes noticed and can lead to a wrong diagnosis [1,2,4,6].

The first symptoms generally appear in adolescence or young adulthood.

Histological can be helpful and can sometimes put in evidence epitheloid granuloma without necrosis. However, its criterion is not pathognomonic and not always present.

The cause of this syndrome remain unclear ; but the presence of granuloma at the histological examination may suggest an infectious, allergic or genetic origin [5,7]. Also, some familiars cases do exist [8].

Due to its uncertain etiology, treatment remain difficult, and no real guidelines exist. Therapeutic are based on empirical strategies tried in the literature and adapted to treatment response.

We report a case of a patient in whom the Melkersson-Rosenthal syndrome was done in association with headache and dry syndrome.

Case presentation:

Miss M, twenty-five years old, was being followed since 2020 for a dry syndrome which was objective by different tests, such as sugar test, salivary flow test and Schirmer test and suspicion of recurrent parotidomegaly (with abnormality grade 2 OMERAVT in the submandibular glands), seemed to be transient [9].

Accessory salivary gland biopsy and parotid gland biopsy didn't find any formal histological argument in favor of Sjogren disease syndrome (i.e., no granuloma or no lymphocytoma infiltration were objective; three foci were found but with a focus score <1) [10].

The search for specific antibodies in the blood came back negative. Oedema of the left side of her face were associated every week, for three days.

A consultation with dermatologists has been requested due to the apparition of a left face oedema and an eyelid ptosis that suddenly appeared, occurring during an homolateral headache that had been lasting

for seven days. Cerebral scanner was performed in front of this unhabitual headache and ptosis but remained normal.

Clinical examination shows a left face unilateral impasto, especially of the left cheek (figure 1), an eyelid ptosis and a hemifacial paresis. She also presented a scrotal and geographical tongue (figure 2).

Clinically, the Melkersson-Rosenthal syndrome diagnosis was evoked. A cheek deep biopsy was performed coupled with tongue biopsy during a new crisis. The cheek biopsy shows moderate and diffuse mononuclear inflammatory infiltration, predominantly perivascular, without vasculitis or granuloma. The tong biopsy showed a very thickened epithelium, of accentuated relief, papillomatous, irregular, with sometimes a parakeratotic layer.

However, the biopsy's nonspecific aspect was compatible with the diagnosis of Melkersson-Rosenthal syndrome.

Treatment with Doxycycline 200mg/day permitted a decrease of the headache and oedema frequency [11]. With this treatment, the symptomatology occurred only once during the next 3 months.

Then, in view of more frequent recurrence of episode, a treatment by Clofazimine was tried [4], and then was relayed by PLAQUENIL because of the poor efficacity of Clofazimine [12].

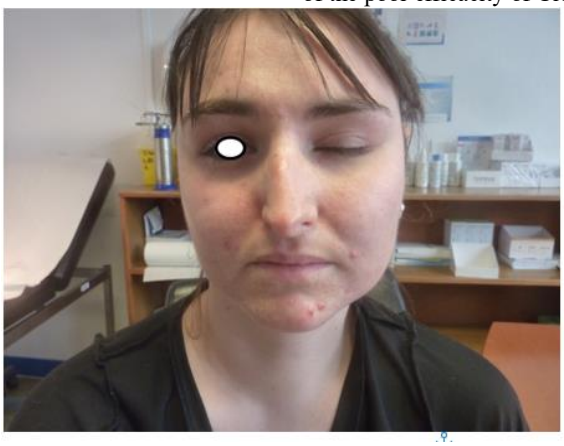

Figure 1: left face oedema and an homolateral eyelid ptosis.

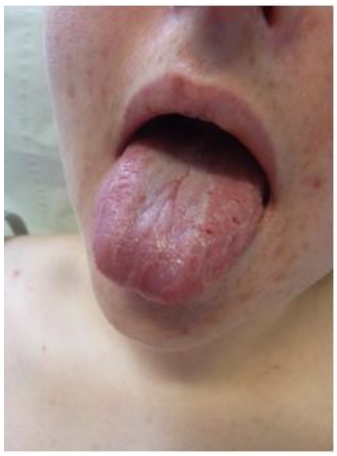

Figure 2: scrotal and geographical tongue.

Discussion:

The Melkersson-Rosenthal syndrome is a rare neuro-muco-cutaneous syndrome, affecting more frequently young adults and women [13], which is characterized by recurrent facial swelling, recurrent facial paralysis, and fissured tongue. It can be accompanied by other symptoms or illness such as dental (periodontal disease [1]), digestive (abdominal pain, Crohn's disease [5]), ophthalmic (uveitis, keratitis, blepharitis [4]) or sensitive disorders (neurovegetative disorder, trigeminal neuralgia, paresthesia [2], dysgeusia [14]), and headache [1,2,4]. Geographical tong seems to be frequently associated with the Melkersson-Rosenthal syndrome [2], as it was present in this case.

In this observation, the presence of headache resulted in a diagnosis delay. This associated symptom is poorly described in the literature and may take part in "minor symptom" of Melkersson-Rosenthal syndrome which can help to do the right diagnosis. Furthermore, here only a paresis was notice and not a complete facial palsy, as it is sometime described too [15,16].

The physiopathology of this syndrome is poorly understood; genetic, infectious and allergic factors seem to influence the pathology [4,7].

Associations with auto immune disorder such as lupus or scleroderma were described [1,17], as well as neurovegetative disorder [4,11] that can explain dry syndrome presented by our patient. However, the diagnosis of Sjogren was not confirmed and the follow up would be interesting.

Moreover, these varied manifestations lead to a multiplication of medical interveners (ENT, dermatologist, neurologist, rheumatologist, dentist) hence the need for a transversal knowledge of the numerous possible symptoms of this pathology.

Histological analysis can guide the diagnosis when it shows small epithelioid granulomas, but it is not a formal criterion. A continuum has been described with a non-specific appearance on histology at the beginning of the disease, then the appearance of granuloma and finally fibrosis when the disease has been present for several years [1].

Initially was observed a good improvement by corticoid treatment at the begin of the manifestation, notably for parotidomelgaly's episode. Treatment by DOXYCYCLINE allowed a clear regression of the attacks. Therefore, CLOFAMIZINE, which showed some efficacity in the treatment of Melkersson-Rosenthal syndrome [18], was introduced, even if there is no consensus standard of care for Melkersson-Rosenthal syndrome [13,19]. Other authors purpose immunosuppressive therapeutic and sometimes surgery is needed for facial palsy and/or lip swelling [13]. Finally, some purpose IgIV therapeutic for recurrent case [14] according to a possible link with immune dysregulation.

Conclusion:

We report on twenty-five-year-old women who presented with recurrent facial paresis and clinical dry syndrome. The facial paresis was secondarily accompanied by headache. After a complete dermatological examination, a Melkersson-Rosenthal syndrome was suspected, and several treatments tried that reduced the symptoms but the symptoms did not completely disappear.

We conclude that the Melkersson-Rosenthal syndrome is well known but the characteristic triad is rarely present in its entirety, and some other symptom can be associated with it.

Consent: The patient is aware of this article and has given us informed consent.

Conflicts of interest: none.

References:

1. Elias MK, Mateen FJ, Weiler CR. (2013). The Melkersson-Rosenthal syndrome : à retrospective study of biopsied cases. J Neurol, 260 :138-143.
2. Stosiek N, Birolleau S, Capesius C, Hornstein OP. (1992). [Chronicity and diagnostic doubts of Melkersson-Rosenthal syndrome. Analysis of developing ways in 5 cases]. Ann Dermatol Venereol, 119 :635-638.
3. Gavioli CF, Florezi GP, Lourenço SV, Nico MM. (2021). Clinical Profile of Melkersson-Rosenthal Syndrome/Orofacial Granulomatosis : A Review of 51 Patients. J Cutan Med Surg, 25 :390-396.
4. Tausch I, Sönnichsen N. (1992). [Experiences with clofazimine therapy of Melkersson-Rosenthal syndrome]. Hautarzt, 43 :194-198.
5. Jasinska D, Boczon J. (2015). Melkersson-Rosenthal syndrome as an early manifestation of mixed connective tissue disease. Eur J Med Res, 20 :100.
6. Rozen TD. (2001). Melkersson-Rosenthal syndrome presenting as a new daily persistent headache: relief with dapsone. Cephalalgia, 21 :924-925.
7. Fantacci C, Mariotti P, Miceli Sopo S, Ferrara P, Rendeli C, Chiaretti A. (2018). Intravenous immunoglobulins in Melkersson-Rosenthal syndrome : A clinical and neuroimaging study. Pediatr Allergy Immunol, 29 :881-883.
8. Hentati H, Hammami S, Oualha L, Mannaï I, Selmi J. (2008). Syndrome de Melkersson-Rosenthal à caractère familial : à propos d'un cas. Med Buccale Chir Buccale, 14 :29-34.
9. Zhang X, Feng R, Zhao J, Wang Y, He J, Liu L, et al. (2021). Salivary gland ultrasonography in primary Sjögren's syndrome from diagnosis to clinical stratification a multicentre study. Arthritis Res Ther, 23 :305.
10. Segerberg-Konttinen M, Konttinen YT, Bergroth V. (1986). Focus score in the diagnosis of Sjögren's syndrome. Scand J Rheumatol Suppl, 61 :47-51.
11. Dhawan SR, Saini AG, Singhi PD. (2020). Management Strategies of Melkersson-Rosenthal Syndrome : A Review. Int J Gen Med, 13 :61-65.
12. Caccavale S, Vitiello P, Tancredi V, Sica A, Ronchi A, Franco R, et al. (2022). A case of Melkersson-Rosenthal syndrome successfully treated with hydroxychloroquine. Dermatol Ther, 35 :15409.
13. Liu R, Yu S. (2013). Melkersson-Rosenthal syndrome : A review of seven patients. Journal of Clinical Neuroscience : Official Journal of the Neurosurgical Society of Australasia, 20.
14. Gaitan-Quintero G, Camargo-Camargo L, López-Velásquez N, González M. (2021). Presentation of Bilateral Facial Paralysis in Melkersson-Rosenthal Syndrome. Case Rep Neurol Med, 6646115.
15. Pathania YS. (2022). Current treatment modalities in granulomatous cheilitis. Int J Dermatol, 61 :755-759.
16. S S, A R, T F, A L, Am EP, G F, et al. (2019). Melkersson−Rosenthal Syndrome in Child Hood : Report of Three Paediatric Cases and a Review of the Literature. International Journal of Environmental Research and Public Health, 16.
17. Gavioli CFB, Nico MMS, Florezi GP, Lourenço SV. (2020). The histopathological spectrum of Melkersson-Rosenthal syndrome : Analysis of 47 cases. J Cutan Pathol, 47 :1010-1017.
18. Azaz B, Nitzan DW. (1984). Melkersson-Rosenthal syndrome. Oral Surg Oral Med Oral Pathol, 57 :250-253.
19. Rossi F, Tsakadze N, Rossi EM, Hoffmann M. (2019). Teaching NeuroImages: Melkersson-Rosenthal syndrome with permanent bilateral facial weakness. Neurology, 92 :81.

DOI:10.31579/2578-8949/135