



## Case Report

# Melkersson-Rosenthal syndrome associated to Hashimoto's thyroiditis

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### ABSTRACT

Melkersson-Rosenthal syndrome (MRS) is a rare neuromucocutaneous granulomatous disorder classically defined by the triad: fissured tongue (lingua plicata), recurrent orofacial edema, and relapsing facial paralysis. Association with other dysimmune disorders was reported suggesting an immunological origin to this syndrome. The association with autoimmune thyroiditis remains exceptional and unusual.

We report the original case of an MRS associated with Hashimoto autoimmune thyroiditis in a 39-year-old Tunisian man with favorable outcome under thyroxine and systemic glucocosteroids.

A dosage of thyroid hormones and a screening for anti-thyroid antibodies would be useful in patients with an MRS.

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## 1. INTRODUCTION

Melkersson-Rosenthal syndrome (MRS) is a rare neuromucocutaneous granulomatous disorder classically defined by the triad: fissured tongue (lingua plicata), recurrent orofacial edema, and relapsing facial paralysis [1, 2].

The pathophysiology of this syndrome is still unknown. Association with other dysimmune disorders such as systemic lupus erythematosus, multiple sclerosis, Crohn's disease, sarcoidosis, and anterior uveitis was reported suggesting a dysimmunitary/immunological origin to this syndrome [2, 3]. The association with autoimmune

thyroiditis, however, remains exceptional and unusual [3-7].

We report the original case of an MRS associated with Hashimoto autoimmune thyroiditis.

## 2. CASE REPORT

A 39-year-old man with no pathological medical history, was referred to our department for exploration of alternating unilateral peripheral facial nerve palsy (two episodes of left facial paralysis and one episode of right facial paralysis in two years).

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**Figure 1:** Macroglossia with fissured dorsal tongue (*lingua plicata*)

Somatic examination noted a macroglossia with a large dorsal and central fissure of the long (Figure 1) and teeth marks on tongue edges (scalloped tongue) (Figure 2). Biopsy of accessory salivary glands objectified noncaseating granulomas. Screening for systemic granulomatosis, connective tissue diseases, vasculitis, infections, and cancers was negative.



**Figure 2:** Teeth marks on tongue edges (*scalloped tongue*).

Free thyroxine was at 2 pmol/l and thyrotropin (TSH) at 120  $\mu$ UI/ml. Anti-thyroperoxidase antibodies (anti-TPO) was strongly positive at 2180 UI/ml and anti-thyroglobulin antibodies at 328 UI/ml leading to the diagnosis of Hashimoto's thyroiditis associated to MRS. Patient was treated with thyroxine in progressive doses

normalizing TSH, and systemic glucosteroids with a favorable outcome. No recurrence has been noted for five years now.

### 3. DISCUSSION

Originally described by Ernst Gustav Melkersson in 1928 [8] and identified as an autonomous entity in 1932 by Curt Rosenthal [9], MRS is a rare neuro-mucocutaneous disorder of unknown etiology. Its pathophysiology is multifactorial involving genetic predisposition, infections, inflammation, stress, immune deficiency, and food intolerance [10, 11].

A dysimmune origin seems very likely given the frequent association with other autoimmune diseases, the frequent detection of immunological markers in the sera of patients with MRS, and the effectiveness of systemic and local corticosteroid therapy [3-11].

In its complete form, this syndrome combines the classic triad: orofacial edema, recurrent facial paralysis and *lingua plicata* [10-12]. This complete triad is noted only in 8-25% of patients with MRS [10-12]. *Lingua plicata* also called "fissured tongue" or "scrotal tongue" is the least common feature of this syndrome [13].

These symptoms may occur simultaneously or gradually associate over time [3-12]. Oligosymptomatic or monosymptomatic forms are far more frequent and represent a real diagnostic challenge for clinicians [12].

This syndrome is extremely rare with an incidence estimated at 0.08% and is particularly frequent in women and during the second and third decades of life [10-12].

The combination of MRS with autoimmune thyroiditis of Hashimoto remains exceptional and unusual; indeed the review of the literature finds only five cases: Scagliusi P et al. 2008 [3], Karadurmus N et al. 2009 [4], Aksu K et al. 2013 [5], Lee YJ et al. 2014 [6], and Frackowiak L et al. 2014 [7]. No case of this association was noted in the large series of Elias MK et al. 2013 of the Mayo Clinic collecting 72 patients with MRS over a period of 40 years, among them 28 had comorbidities [2]. Our observation is to our knowledge the sixth reporting this association.

MRS may occur even in hypothyroid patients properly treated with clinical and biological euthyroidism. High levels of anti-thyroid antibodies, particularly anti-TPO, are associated with these MRS recurrences/relapses [4,7]. These findings reinforce the immunological hypothesis of MRS. This hypothesis is reinforced by the publication of Karadurmus N et al. where the patient simultaneously presented multiple dysimmunitary diseases: MRS, Hashimoto's thyroiditis, and rheumatoid arthritis [4].

### 4. CONCLUSION

As rare as it is, the association MRS and Hashimoto's thyroiditis deserves to be known by health professionals.

To the best of our knowledge, only five cases have been previously reported. Our observation is the sixth reporting this association.

This association seems to be far from a mere coincidence. A common immunological hypothesis is evoked. Thus a dosage of thyroid hormones and a screening for anti-thyroid antibodies would be useful in patients with an MRS.

## 5. REFERENCES

1. Rivera-Serrano CM, Man LX, Klein S, Schaitkin BM. Melkersson-Rosenthal syndrome: A facial nerve center perspective. *J Plast Reconstr Aesthet Surg.* 2014;67(8):1050-4. doi: 10.1016/j.bjps.2014.04.014.
2. Elias MK, Mateen FJ, Weiler CR. The Melkersson-Rosenthal syndrome: a retrospective study of biopsied cases. *J Neurol.* 2013;260(1):138-43. doi: 10.1007/s00415-012-6603-6.
3. Scagliusi P, Sisto M, Lisi S, Lazzari A, D'Amore M. Hashimoto's thyroiditis in Melkersson-Rosenthal syndrome patient: casual association or related diseases? *Panminerva Med.* 2008;50(3):255-7.
4. Karadurmus N, Fiahin M, Kahraman E, Yilmaz S, Taflçi C, Inal S, et al. Melkersson-Rosenthal syndrome and Hashimoto's thyroiditis: A case report. *Turk Jem.* 2009;13:40-2.
5. Aksu K, Kurt E. A case of Melkersson-Rosenthal syndrome associated with autoimmune hypothyroidism. *Asthma Allergy Immunol.* 2013;11:131-3.
6. Lee YJ, Cheon CK, Yeon GM, Kim YM, Nam SO. Melkersson-Rosenthal syndrome with Hashimoto thyroiditis in a 9-year-old girl: an autoimmune disorder. *Pediatr Neurol.* 2014;50(5):503-6.
7. Frąckowiak L, Wroński K, Kańczuga-Koda L, Koda M. Melkersson-Rosenthal syndrome in a patient with Hashimoto disease. *Prog Health Sci.* 2014;14(1):269-72.
8. Melkersson E. Ett fall av recidiverande facialispares i samband med angioneurotiskt ödem. *Hygiea.* 1928;90:737-41.
9. Rosenthal C. Klinisch-erbbiologischer beitrug zur konstitutionspathologie Gemeinsames auftreten von (rezidivierender familiärer) facialislähmung, angio-neurotischem gesichtsödem und lingua plicata in arthritismus-familien. *Z Neurol Psychiatr.* 1931;131:475-501.
10. Bakshi SS. Melkersson-Rosenthal Syndrome. *J Allergy Clin Immunol Pract.* 2017;5(2):471-2. doi: 10.1016/j.jaip.2016.09.034.
11. Wehl G, Rauchenzauner M. A Systematic Review of the Literature of the Three Related Disease Entities Cheilitis Granulomatosa, Orofacial Granulomatosis and Melkersson - Rosenthal Syndrome. *Curr Pediatr Rev.* 2018;14(3):196-203. doi: 10.2174/1573396314666180515113941.
12. Basman A, Gumusok M, Degerli S, Kaya M, Toraman Alkurt M. Melkersson-rosenthal syndrome: a case report. *J Istanbul Univ Fac Dent.* 2017;51(1):42-5. doi : 10.17096/jiufd.96279
13. Lin TY, Chiang CH, Cheng PS. Melkersson-Rosenthal syndrome. *J Formos Med Assoc.* 2016;115(7):583-4. doi: <http://dx.doi.org/10.1016/j.jfma.2015.08.011>