

Melkersson-Rosenthal syndrome in a patient with Hashimoto disease

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ABSTRACT

Introduction: Melkersson-Rosenthal syndrome (MRS) is a rare neurologic disease of unknown etiology. It is characterized by a triad of symptoms: relapsing peripheral facial paralysis; orofacial edema and fissured tongue. The pathological findings are varied but often characterized by the presence of noncaseating granuloma. There are few cases of MRS coexistent with Hashimoto disease in the literature.

Purpose: To present a case of MRS coexistent with Hashimoto disease.

Case presentation: We report a case of a 32-year-old woman with coexisting MRS and autoimmune thyroiditis, Hashimoto disease. Fissured tongue

and recurrent cheilitis of the upper lip were observed. In her medical history, she had facial nerve palsy. Histopathological examination of the upper lips mucose membrane revealed the presence of granulomas confirming the diagnosis.

Conclusions: Cooperation between clinicians and histopathologists is vital in diagnosing MRS. The presence of the anti-TPO antibodies in the case reported here could suggest a correlation between immunological alteration characteristics of autoimmune thyroiditis and MRS.

Key words: Melkersson-Rosenthal syndrome, cheilitis granulomatosa, Hashimoto disease, autoimmune thyroiditis, facial nerve palsy.

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INTRODUCTION

Melkersson-Rosenthal syndrome (MRS) is a rare neurologic disease that includes several symptoms: facial nerve palsy, constant or recurrent facial oedema (usually limited to upper lip – cheilitis); and lingua plicata. It was originally independently described in 1928 by Ernst Gustav Melkersson and in 1931 by Curt Rosenthal. In 1945, Miescher introduced several cases of primary interstitial inflammation of the lips and named them macrocheilitis granulomatosa. To date, the etiology of MRS remains unknown. There is, however, a critical role of infectious-inflammatory factors in pathophysiology of the disease, since the majority of patients had internal infectious foci and there has been a dermatological improvement observed after antibacterial/surgical therapy [1]. Allergies, especially those relating to cosmetics and toothpaste, and hereditary factors are being considered (familial incidence in 61% of patients). Whether there is a gene responsible for MRS remains unknown.

The course of disease is greatly variable. MRS can occur in every age. However, the first symptoms most commonly occur in the second or third decade. The beginning is sudden, and there are no prodromal symptoms. The most constant is cheilitis (which occurs in 75-100% of patients). Occasionally, migraine headaches, oral lesions (hypertrophy of the upper lip and mucosa), and salivary gland dysfunction may occur. These are the minor symptoms. They can be useful in diagnosing oligosymptomatic syndrome [2]. Recurrent edemas as well as intensity of edemas may vary in time. The main symptom is granulomatous cheilitis (Miescher) that often occurs as an isolated symptom. Histopathologically, fibrosis coexists with granulomatous inflammation. Lesions are subtle and very likely to be misdiagnosed [3].

Autoimmune thyroiditis, Hashimoto disease, can be diagnosed based on elevated anti-TPO and anti-TG antibodies concentration in patients with hypothyroidism and distinctive ultrasonographic imaging. The diagnosis can be confirmed based on a fine-needle aspiration biopsy (FNAB) of the thyroid. It occurs in 10% of the overall population [4].

CASE PRESENTATION

The 32-year-old woman has been treated ambulatorily since 2008 for autoimmune thyroiditis, Hashimoto disease. Based on anamnesis and physical examinations, the level of serum thyroid hormones, elevated anti-TPO antibodies concentration, distinctive ultrasonographic imaging and aspiration biopsy, autoimmune thyroiditis was diagnosed. The patient is under constant endocrinological care, treated with levothyroxine. In 2013, the patient was diagnosed in the

dermatological department for fixed cheilitis of the upper lip coexistent with a fissured tongue (Fig.1). For six years, there has been recurrent upper lip cheilitis, treated with corticosteroids and antibiotics with partial clinical improvement. The patient underwent facial palsy and recurrent borreliosis treated with antibiotics.



Figure 1. Fissured tongue

For several months there has been increasing inflammation of the upper lip. Suspecting orofacial granulomatosis, a tissue sample was taken. The result of histopathological examination was inflammatio chronica focalis praecipue perivascularis. There were no irregularities in laboratory tests. Patch tests and exposition tests with salicylic acid were negative. Quincke's edema was excluded based on C1-esterase level. The patient was released from the hospital with the diagnosis of a fixed upper lip edema. On a periodical endocrinological visit, suspecting MRS, there was a need to reexamine the tissue sample histopathologically, since clinical examination and medical history indicated fully symptomatic syndrome. The histopathological reevaluation under higher magnification showed small non necrotizing granulomas composed of epithelioid cells, plasma cells and lymphocytes. Both clinical symptoms and histopathological results suggest MRS (Fig. 2, 3).

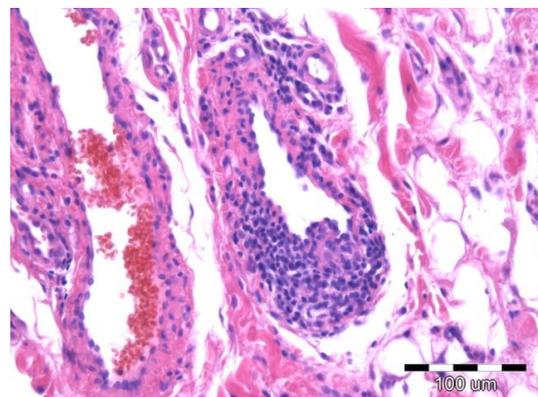


Figure 2a. Labial mucosa biopses showed hyperplastic epithelium and perivascular

lymphocytic infiltrates in connective tissue. (HE)100xmagnification.

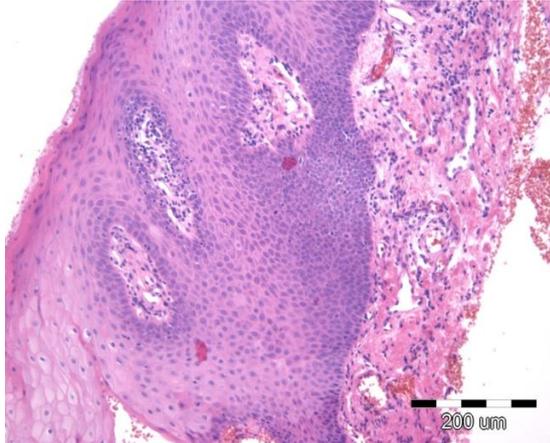


Figure 2b. Labial mucosa biopses showed hyperplastic epithelium and perivascular lymphocytic infiltrates in connective tissue. (HE) 200x magnification.

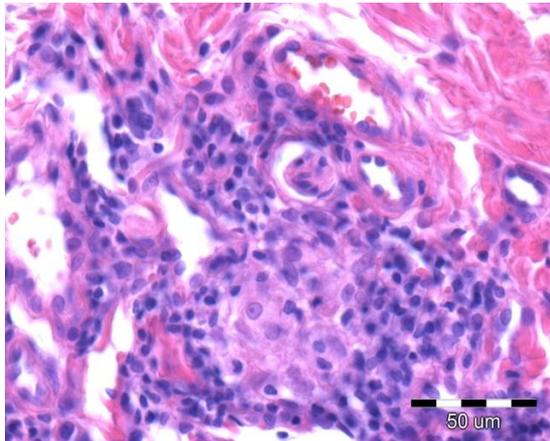


Figure 3a. Small non necrotising granulomas composed of epithelioid cells, plasma cells and lymphocytes. (HE 400 x magnification).

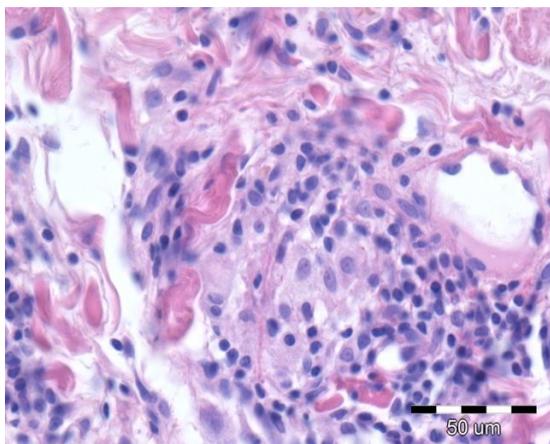


Figure 3b. Small non necrotising granulomas composed of epithelioid cells, plasma cells and lymphocytes. (HE 400 x magnification).

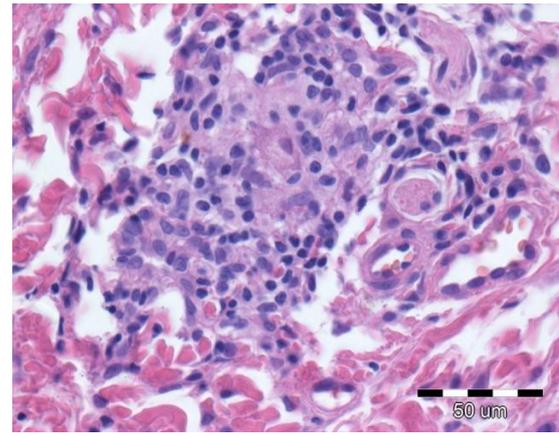


Figure 3c. Under higher magnification biopses showed small non necrotising granulomas composed of epithelioid cells, plasma cells and lymphocytes. (HE 400 x magnification).

DISCUSSION

Hashimoto disease commonly links with autoimmune diseases, such as: rheumatoid arthritis; Sjogren syndrome; psoriasis; celiakia; and multiglandular autoimmune syndromes. MRS is often coexistent with Crohn's disease and sarcoidosis. There are few cases of MRS and Hashimoto disease coexisting [5].

In both cases, there is autoimmune background considered. The presence of the anti-TPO antibodies in the case reported here could suggest a correlation between immunological alteration characteristics of autoimmune thyroiditis and MRS. The connection between MRS and immunological disorders, such as sarcoidosis, Crohn's disease, and multiple sclerosis is documented in the literature.

A case of a 42-year-old woman with psoriatic arthritis and neurological incidents connected to MRS after anti-TNF treatment (Etanercept) was described [6]. Though there is a distinctive triad of symptoms linked to MRS [5], there is often a need to verify the diagnosis histopathologically [3]. After combining clinical and histopathological data the patient can be diagnosed. Despite many therapeutical attempts, unacquaintance of a pathologic factor makes it impossible to introduce targeted therapy; patients are treated symptomatically. Treatment with systemic corticosteroids is the most effective in the short term, especially using high doses and in the early stage of the disease. Antibacterial therapy and eradication of infectious focuses is effective (regression of edema, long-term remission) in a low percentage of patients. An alternative is to use leprosy drugs (Clofazimine), which has a 94% effectiveness rate. This method of treatment appears to be safe [7].

CONCLUSIONS

The diagnosis is complicated. Cooperation between clinicians and histopathologists is vital in diagnosing MRS. The presence of the anti-TPO antibodies in the case reported here could suggest the correlation between immunological alteration characteristics of autoimmune thyroiditis and MRS.

Conflicts of interest

The authors declare that they have no competing interests in the publication of the manuscript.

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