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OLGU SUNUMU CASE REPORT

A case of Melkersson-Rosenthal syndrome associated with autoimmune hypothyroidism*

Otoimmün hipotiroidi ile ilişkili Melkersson-Rosenthal sendromu olgusu

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ABSTRACT

Melkersson-Rosenthal syndrome (MRS) is a rarely encountered clinical entitiy characterized with the triad of facial nerve palsy, recurrent episodes of orofacial oedema and fissured tongue. The etiology of MRS is not yet clearly understood. We report a case of MRS with classical triad who also received diagnosis of autoimmune hypothyroidism pointing out that investigating autoimmune diseases including autoimmune hypothyroidism in patients with MRS is important.

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Key words: Hypothyroidism, Melkersson-Rosenthal syndrome, autoimmunity

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ÖZ

Melkersson-Rosenthal sendromu (MRS) fasiyal sinir paralizisi, tekrarlayan orofasiyal ödem atakları ve fissüre dil triadı ile karakterize nadir görülen bir klinik durumdur. MRS etyolojisi halen tam olarak anlaşılmamıştır. Klasik triad ile presente olan ve aynı zamanda otoimmün hipotiroidi tanısı alan bir MRS olgusu sunulmakta ve MRS tanılı hastalarda otoimmün hipotiroidi de dahil olmak üzere otoimmün hastalıkların araştırılmasının önemi vurgulanmaktadır.

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Anahtar kelimeler: Hipotiroidizm, Melkersson-Rosenthal sendromu, otoimmünite

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INTRODUCTION

Melkersson-Rosenthal syndrome (MRS) is a rarely encountered clinical entity characterized with the triad of facial nerve palsy, recurrent episodes of orofacial oedema and fissured tongue. Most cases are oligo- or monosymptomatic rather than being presented with all three symptoms of the clinical triad. However the complete clinical triad is only seen in around one-fourth of the cases, rather most cases are either monosymptomatic or are presented with sequential occurence of the symptoms[1,2].

CASE REPORT

We report a 57-year-old female patient admitted to allergy clinic complaining of persistent swelling of upper lip for the last two years. She did not describe accompanying rush or pruritus or swelling in any other part of the body. She had a history of facial nerve palsy recurred for several times since her mid-twenties. She did not report any adverse drug or food reactions and she was not on angiotensin-converting enzyme inhibitor treatment. Previously she was treated with intralesional corticosteroid for swollen lip which revealed partial response. In her first degree relatives fissured tongue was present but the family history was negative for any other related symptoms. Physical examination revealed prominently swollen upper lip and fissured tongue with no other findings (Figure 1). The laboratory examinations including complete blood count, C-reactive protein, renal and liver function tests, blood glucose and serum electrolytes, urinalysis, C3 and C4 levels were all within normal limits. Skin prick tests with aeroallergens and food allergen extracts as well as patch tests with European baseline series came out to be negative. Chest tomography evaluation, pulmonary function test and carbon monoxide diffusion capacity showed no abnormalities. No pathologic findings were detected in opthalmologic and gastroentorologic evaluations. Based on the presentation with all of the three findings of MRS the patient received the diagnosis of MRS. The patient also received diagnosis of autoimmune hypotyroidism based on elevated tyroid stimulating hormone level and anti-TPO antibody positivity.

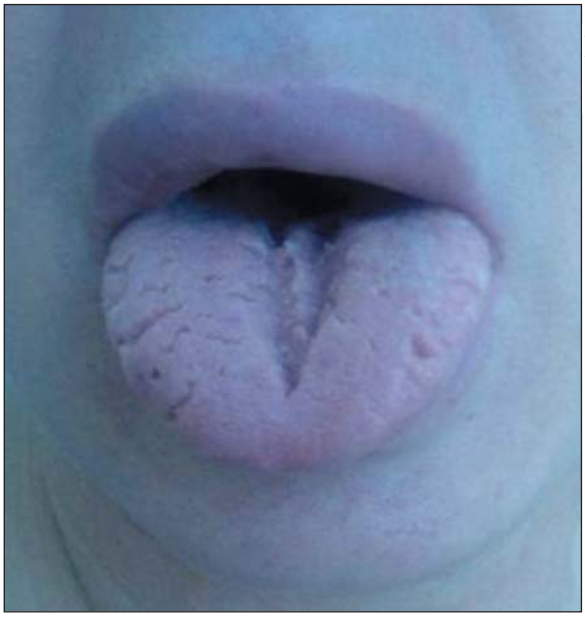

Figure 1. Swollen upper lip and fissured tongue in a patient with Melkersson-Rosenthal syndrome.

DISCUSSION

The etiology of MRS is not yet clearly understood though autosomal dominant inheritance is reported in some cases[3]. Some factors possibly involved in the pathogenesis are infections, atopy, contact allergy and hypersensitivity to food additives[2,4-6]. Autoimmunity may also be important in the pathogenesis since some cases are associated with Crohn's disease and sarcoidosis[7-9]. Previously in 2008, Scagliusi et al. reported Hashimoto's thyroiditis in a patient with MRS suggesting an association between these two clinical entity, possibly due to an immunologic alteration[7]. That is the only case reported upto now MRS associated with autoimmune hypothyroidism in the literature. With the present paper we report a case of MRS with classical triad who also received diagnosis of autoimmune hypothyroidism which is the second case in the literature to the best of our knowledge and it points out that MRS should be suspected in cases presented with angioedema. It also emphasizes that investigating auto-

immune diseases including autoimmune hypothyroidism in patients with MRS is important and suggests that further epidemiologic studies are required to investigate possible immunological association between autoimmune hypothyroidism and MRS.

The diagnosis of MRS is based on features of the classical triad,and there is no spesific radiological or histopathological findings[10]. The whole clinical triad is seen in about 25% of the cases[1]. If the presentation is oligo- or monosymptomatic then for the definitive diagnosis histopathological demonstration of granulomatous inflammation is also needed[11]. Accordingly, in the present case histopathological evaluation was not performed. Persistent characteristic of swollen lip, partial response to intralesional steroid therapy, absence of swelling in any other site of the body or any urticarial lesions or pruritus, no history of food or drug inducing the occurence of the complaints together with laboratory data including C4 revealing no abnormalities excludes hereditary, acquired or idiopathic angioedema. Normal findings in the radiological evaluation of the lungs and no clinical findings supporting tuberculosis and sarcoidosis excluded these two granulomatous diseases.

Since the etiopathogenesis of MRS is not well understood there is no spesific treatment modalilty that ensures complete cure of the disease. Especially in the early stages of the disease the symptoms are self limited and spontaneous remission is frequent. Systemic or intralesional corticosteroid therapy are the most commonly used modalities for facial paralysis and orofacial swelling. Short term recovery of the symptoms is provided by therapy but disease usually recurs in the follow-up[10,12]. Radiotherapy, anti-histamines, salazosulfapyridine, danazol, methotrexate, infliximab, clofazimine, minocycline and metronidazole are other therapeutic agents which are reported to be beneficial. Surgical nerve decompression is also an effective option when needed[2,11,12].

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