Case Report
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Melkersson-rosenthal syndrome

Melkersson-rosenthal sendromu

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Abstract
The Melkersson-Rosenthal syndrome (MRS) is a rare neuro mucocutaneous granulomatous disease presenting with recurrent peripheral facial paralysis, orofacial oedema, and fissured tongue. The coexistence of the classic clinical triad is rare. Clinical findings and examination make the diagnosis. It should be considered in the differential diagnosis of recurrent facial paralysis. In this case report, a 42-year-old female patient with the classical triad of MRS is presented.

Keywords: Recurrent peripheral facial palsy, orofacial oedema, fissured tongue, neuro mucocutaneous disease.

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Öz
Melkersson-Rosenthal sendromu (MRS) yineleyici periferik fasiyal paralizi, orofasiyal ödem, fissürlü dil triadı ile kendini gösteren nadir bir nöromukokütan granülomatöz hastalıktır. MRS'nin klasik klinik triadının beraber görülmesi nadirdir. Tanısı klinik bulgular ve muayene ile konulur. Yineleyici fasiyal paralizilerin ayırıcı tanısında muhakkak düşünülmesi gerekir. Bu olgu sunumunda MRS'nin klasik triadının bir arada görüldüğü 42 yaşında, kadın hasta sunulmuştur.

Anahtar kelimeler: Tekrarlayıcı periferik fasiyal paralizi, orofasiyal ödem, fissürlü dil, nöromukokütan hastalık.

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Introduction

The Melkersson-Rosenthal syndrome is an uncommon neuro mucocutaneous granulomatous disease presenting with the triad of recurrent peripheral facial paralysis, orofacial oedema, and fissured tongue. It is more common in adults. All classical triad findings are seen in approximately 25% of patients with MRS [1, 2]. Its aetiology has not been fully elucidated. It is thought that genetic predisposition and factors that subsequently affect the individual play a role in the aetiology. Many diseases, such as intracranial space-occupying lesions, head trauma, intracranial haemorrhage, leukaemia, autoimmune diseases developing after infections, allergic disorders, thyroiditis, multiple sclerosis, diabetes mellitus are included in the aetiology [2-4]. There is no agreed treatment protocol to ensure complete remission and prevent recurrences in MRS. Medical or surgical methods are used in the treatment. Facial nerve decompression can be applied surgically in cases which do not respond to medical therapy [2, 3, 5].

Case report

A 42-year-old woman was admitted to the outpatient clinic of our hospital with complaints of recurrent inability to close her right eye, swelling in the lip and the tongue and fissured tongue. The complaints started a year ago. The frequency of the complaints was variable. There was no distinctive feature in her personal and family history. In the physical examination, her vital signs were stable; she could not close the right eyelid completely, consistent with right peripheral facial paralysis. The right nasolabial sulcus was erased. Mild, painless oedema was in the right upper lip (Figure 1) and fissured tongue (Figure 2).

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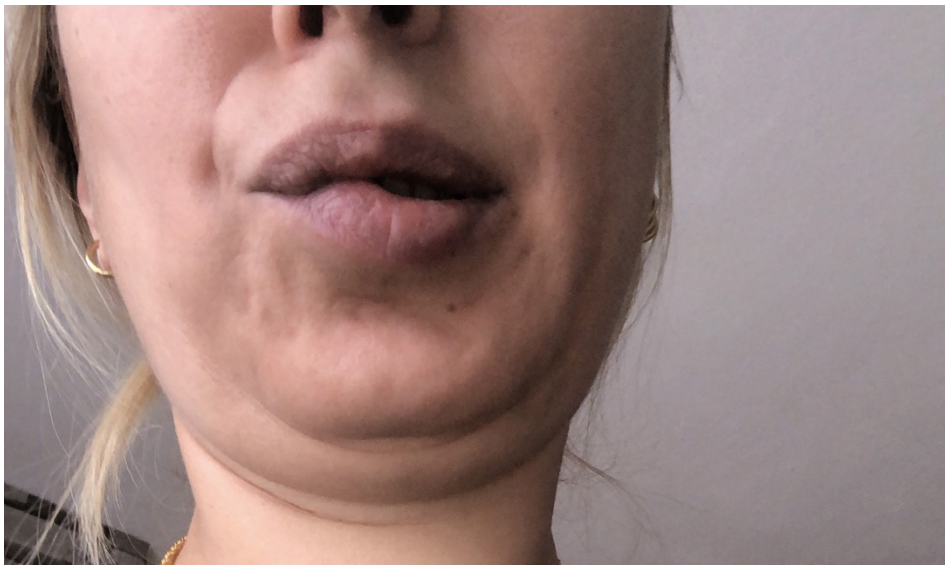

Figure 1. Right peripheral facial paralysis and oedema in the lip

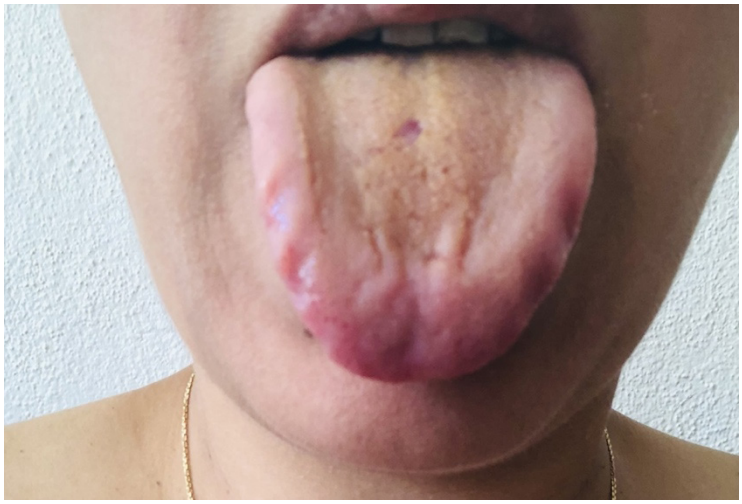

Figure 2. Fissured tongue

In laboratory examinations, complete blood count (CBC), blood glucose, glycated haemoglobin (HbA1c), serum electrolyte levels, liver and renal function tests, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), vasculitis panel tests including antineutrophilic cytoplasmic antibody (ANCA), serum complement 3 (C3) and 4 (C4), folic acid, vitamin B12, homocysteine, free thyroxine (fT4), thyroid stimulating hormone (TSH), antithyroglobulin (anti-Tg) and anti thyroperoxidase antibodies (anti-TPO), serum angiotensin-converting enzyme (ACE), methylmalonic acid levels were normal. Serum and urine immunofixation electrophoresis tests showed no monoclonal gammopathies. Viral serology tests, including anti-human immunodeficiency virus (HIV), were negative.

The C1 esterase inhibitor test result did not suggest hereditary angioneurotic oedema.

Cranial and bilateral temporal bone magnetic resonance imaging (MRI) with gadolinium revealed no abnormalities. The bilateral facial nerve electrodiagnostic test with needle electromyography (EMG) demonstrated neither acute nor chronic pathologies.

The dermatology department examined the patient regarding possible Behcet's disease and angioedema. Serum immunoglobulin E (Ig-E) level was normal. Findings and examination did not suggest Behcet's disease or hereditary angioneurotic oedema. In terms of possible underlying rheumatological pathologies, a salivary gland biopsy was taken

from the lip. No pathology was found. The rheumatology department evaluated her with the biopsy results and vasculitis panel tests. No rheumatological pathology was detected. The electrocardiogram (ECG) was in normal sinus rhythm. Transthoracic echocardiography (TTE), cardiac stress test, and coronary computed tomography (CT) scan were within the normal range. The cardiology department assessed this patient with the previously mentioned cardiac tests, and cardiac pathologies were ruled out. Since the bilateral facial nerve electrodiagnostic test with needle EMG and the temporal bone MRI with contrast demonstrated no pathologies, she was consulted to the otorhinolaryngology department, and the examination was within the normal range. Otorhinolaryngological pathologies were excluded.

To evaluate sarcoidosis and tuberculosis, a thoracic CT scan was performed. There were no lesions in the thoracic CT scan.

She received oral methylprednisolone 64 mg/day treatment. In addition, to avoid any gastrointestinal haemorrhage, pantoprazole sodium 40 mg/day was added to the treatment. The dose of the corticosteroid was gradually tapered. After 26 days of oral methylprednisolone, her complaints and symptoms decreased; however, they did not disappear. A whole-body dual-energy x-ray absorptiometry (DEXA) scan was performed to examine the adverse effects of the corticosteroid treatment. Osteopenia in the left femur was detected. Due to this side effect, the methylprednisolone treatment was discontinued, and an anti-inflammatory non-steroidal agent, naproxen sodium 500 mg/day, was commenced. The frequency and severity of her complaints have decreased to almost non-existent.

Discussion

The Melkersson-Rosenthal syndrome is an infrequent neuro mucocutaneous granulomatous disease presenting with the triad of recurrent peripheral facial paralysis, orofacial oedema, and fissural tongue. It is more common in adults. All classical triad findings are seen in around 25% of MRS patients, making diagnosing the syndrome arduous [1, 2]. Its aetiology has not been fully elucidated. It is thought that genetic predisposition and factors that subsequently affect the individual play a role in the aetiology. Many diseases such as intracranial space-occupying lesions, head trauma, intracranial haemorrhage, leukaemia, autoimmune diseases developing after previous infections such as adenotonsillitis, allergic disorders, thyroiditis, multiple sclerosis, diabetes mellitus are included in the aetiology [2-4].

All the clinical findings might be seen simultaneously or separately. The presence of not less than one of the findings of idiopathic facial paralysis or fissured tongue at once with permanent or recurrent orofacial oedema is enough for MRS diagnosis [6-9].

The typical symptom of MRS is diffused, painless, acute orofacial oedema [6, 10-12]. Oedema usually recurs and lasts a few hours to weeks [13, 14]. MRS may mimic angioedema; however, it differs from angioedema by lasting longer and lacking response to antihistaminic treatments [15]. The second component of the triad of MRS is peripheral facial paralysis, which might be seen in almost 0.3 of the patients [16]. Paralysis might be both transient or permanent. Permanency is usually observed as the disease duration increases. Peripheral facial paralysis may be partial, bilateral and unilateral. Facial paralysis is found to be associated both with pressure due to oedema on the facial nerve within the temporal bone and its granulomatous infiltration [17, 18]. Our patient had a history of transient intermittent peripheral facial paralysis. The third symptom of MRS is the fissured tongue, a common finding in the population [19]. Therefore, it is slighter crucial in the diagnosis. Our patient also has a transient recurrent fissured tongue.

There is no agreed treatment protocol to ensure complete remission and prevent recurrences in MRS. Medical treatments such as corticosteroids, anti-inflammatory non-steroidal agents or surgical methods are used in the treatment. Facial nerve decompression can be applied surgically in cases that do not respond to medical therapy [2, 3, 5].

The patient showed all the elements of the characteristic triad. The corticosteroid and anti-inflammatory non-steroidal choices have worked in this patient. The frequency and severity of the symptoms have decreased to almost non-existent. Even though MRS might be under

control with the treatments, reoccurrence is always possible [20].

In conclusion, the diagnosis of MRS is arduous due to its infrequency. The typical symptom is orofacial oedema. Medical doctors should remember this syndrome, especially with patients with recurrent, transient or permanent swelling in the orofacial area and peripheral facial paralysis.

Conflict of interest: No conflict of interest was declared by the authors.

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Informed consent: An informed consent form was obtained from the patient to conduct this case report and use the photographs.