MELKERSSON-ROSENTHAL SYNDROME: A CASE REPORT

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SINDROM MELKERSSON ROSENTHAL PRIKAZ SLUČAJA

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ABSTRACT

This paper presents a case of a 60-year-old patient with an oligosymptomatic form of Melkersson-Rosenthal syndrome. This syndrome is characterized by three symptoms: swelling of the lips and face, unilateral facial palsy, and lingua plicata. The literature often includes information on monosymptomatic forms of the syndrome and granulomatous cheilitis or the existence of only two symptoms of the classic triad. The anamnesis, clinical and histopathological findings in this case confirmed the existence of chronic granulomatous inflammation confined to the lower lip, indicating an oligosymptomatic form of Melkersson-Rosenthal syndrome. The treatment included prednisone application at a dose of 25 mg daily, histamine H1 and H2 antagonists, and the antibiotic ofloxacin for urinary tract infection.

Keywords: *Melkersson-Rosenthal syndrome, granulomatous cheilitis, prednisone.*

SAŽETAK

U ovom radu je prikazan slučaj bolesnika starog 60 godina sa oligosimptomatskom formom Melkersson Rosenthal Sindroma. Ovaj sindrom karakterišu trias simptoma: otok usne i lica, unilateralna pareza facijalisa, uvećan i izbrazdan jezik-lingua plicata. U literaturi se često navode podaci o monosimptomatskom sindromu i to je granulomatozni helitis, ili postojanje samo dva simptoma. Anamneza, klinička slika i Ph analiza ukazuju na postojanje hroničnog granulomatoznog zapalenja donje usne, oligosimptomatske forme Melkerson-Rosentalov-og sindroma. U terapiji primenjen je pronison u dozi od 25 mg, uz H1 i H2 blokatori, antibiotska terapija (Visiren) za urinarnu infekciju.

Ključne reči: *Melkerson-Rozentalov sindrom, granulomatozni heilitis, prednizon.*

INTRODUCTION

In 1928, Melkersson first described a case of recurrent facial paresis with mouth edema. A few years later (in 1931), Rosenthal added a third symptom, lingua plicata, to the description, and Melkersson-Rosenthal syndrome was established (1). It usually occurs in adulthood and middle age in both sexes. The etiology of Melkersson-Rosenthal syndrome is unknown, and many believe that there is a genetic predisposition.

This syndrome is characterized by three symptoms: swelling of the lips and face, unilateral facial nerve palsy, and fissured tongue. There are numerous case reports on monosymptomatic variants of the syndrome with granulomatous cheilitis or the existence of only two symptoms. In addition to the swelling of the lips and facial region, patients also lose taste and smell, secretion from the salivary and lacrimal glands is reduced, and mobility of the lips is decreased. Sometimes, the patients have difficulties speaking (1,2,3).

Treatment of the syndrome is based on the local administration of triamcinolone every 4 to 6 months. Using clofazimine 100 mg twice daily for 10 days followed by twice a week for 4 months is also recommended. Surgical treatment along with the previously mentioned therapy is also possible (4,5).

THE CASE REPORT

The patient was a 60-year-old male who was admitted to the hospital for the evaluation of chronic swelling of his lower lip. From the patient history, the first incidence of swelling and redness of the lower lip occurred in 1989. He was treated then with both local and systemic antibiotics, but there was no improvement. After a tooth extraction, the swelling spontaneously subsided but returned in 2007 and persisted until the current presentation, regardless of treatment. He had attempted to treat himself with local antibiotics and antihistamines. He had no hives, rashes, or swelling of other soft tissues. He denied the existence of other diseases and allergies to food, medication, insect bites, and surgery. His family history was unremarkable.

The patient was conscious during the hospital admission, properly oriented, eupneic, and without fever. His body shape and weight were normal, and there were no signs of peripheral lymphadenopathy or hemorrhagic syndrome. The excessive swelling and redness in the area of the lower lip were the most prominent findings (Figures 1 and 2); the tongue was coated with whitish skim, dry and lightly scarred. The heart rhythm was regular, with clear tones; the arterial blood pressure was

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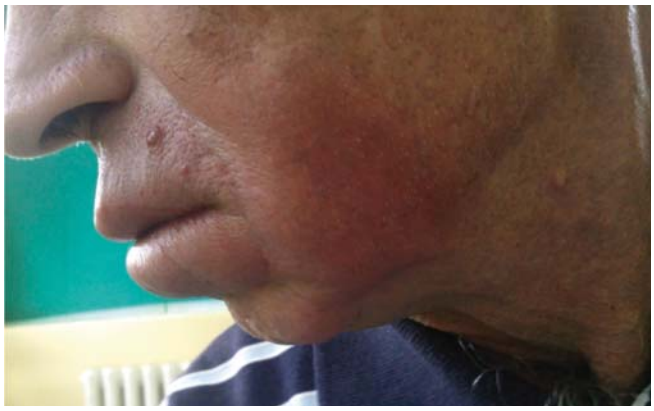

Figure 1. Granulomatous inflammation of the lower lip: side view.

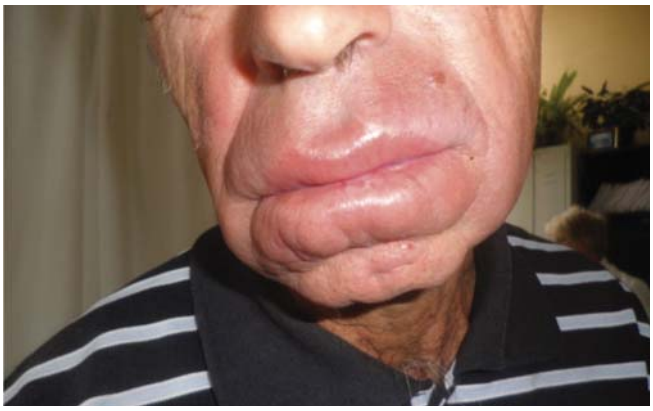

Figure 2. Granulomatous inflammation of the lower lip: frontal view.

180/120 mmHg. His anterior abdominal wall was above chest level when the patient was lying on his back, insensitive to palpation and without swelling. There were no deformities or venous varices on the lower extremities. The laboratory findings were as follows: erythrocyte sedimentation rate 6 mm in the first hour, fibrinogen 3.1 g/l, leukocyte count 5.89 x 10^9/l (lymphocytes 1.38 x 10^9/l, monocytes 0.54 x 10^9/l, eosinophils 0.29 x 10^9/l, and basophils 0.04 x 10^9/l), erythrocyte count 4.39 x 10^{12}/l, hemoglobin 147 g/l, hematocrit 0.439, MCV 97 nL, platelet count 255 x 10^9/l, glucose 4.1 mmol/l, creatinine 74 µmol/l, total bilirubin 29 µmol/L, direct bilirubin 16.4 µmol/L, total blood proteins 73 g/L, albumin 43 g/L, cholesterol 5.23 mmol/L, triglycerides 1.25 mmol/L, potassium 3.7 mmol/L, sodium 138 mmol/L, calcium 2.39 mmol/L, AST 36 IU/L, ALT 41 IU/L, ALP 70 IU/L, gamma-GT 12 IU/L, and LDH 517 IU/L. The immunological results were as follows: antinuclear antibodies 0, anti-neutrophil cytoplasmic antibodies 0, anti-mitochondrial antibodies 0, anti-smooth muscle antibodies 0, anti-liver kidney microsomal antibodies-1 0, IgG 13.9 g/L, IgA 2.72 g/L, IgM 0.76 g/L, C4 0.115 g/L, C3c 1.32 g/L, CRP 0.44 nM/L, IgE1 204 mg/L, CH50 12.78 IU/ml, alternative pathway 2.29 g/L, C3 1.41 g/L, C4 0.3 g/L, C1 kv 0.33 g/L, C1 funk 46.2%, and C1q 0.081 g/L.

Microbiological culturing of the patient's urine revealed the presence of *Klebsiella* and *Enterobacter* spp. Culturing of the throat and nose swabs revealed only normal microbial flora. Skin prick tests with standard respiratory and nutritive allergens were negative. Neurological examination showed only cheilitis, and there were no other abnormalities. After consulting with an ear-nose-larynx specialist, the lower lip was biopsied. Histologic examination of the biopsied specimen revealed fragments of mucosa with signs of chronic inflammation and a moderate number of rare, poorly formed epithelioid granulomas without caseous necrosis.

Based on the abovementioned findings, the existence of chronic granulomatous inflammation of the lower lip was confirmed, and the diagnosis of the oligosymptomatic form of Melkersson-Rosenthal syndrome was made.

The patient was treated with oral prednisolone 25 mg daily, H1 and H2 blockers, and the antibiotic ofloxacin for a urinary tract infection. The patient was advised to go to a dentist and a gastroenterologist for further treatment.

DISCUSSION

Melkersson-Rosenthal syndrome is characterized by a triad of symptoms: swollen lips, facial nerve paresis, and an enlarged and grooved tongue. The majority of the reports in literature describe monosymptomatic forms of this syndrome (3,5). Our patient is another example of monosymptomatic syndrome characterized by granulomatous cheilitis. The data from the patient's history and clinical and histological analysis indicated the existence of chronic granulomatous inflammation of the lower lip, without the other two characteristic elements of Melkersson-Rosenthal syndrome. After unspecific anti-inflammatory therapy (prednisolone 25 mg daily, H1 and H2 blockers), the inflammation of the lips subsided, with satisfactory aesthetic results.

Because Melkersson-Rosenthal syndrome can be associated with Crohn's disease, physicians should be aware of this association (6) and refer their patients with the syndrome for further diagnostic evaluation. Moreover, sarcoidosis may have similar clinical features as Melkersson-Rosenthal syndrome (6); thus, further evaluation by a pulmonologist is also warranted.

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