Melkersson-Rosenthal Syndrome and Hashimoto’s Thyroiditis: A Case Report

Melkersson-Rosenthal Sendromu ve Hashimoto Tiroidit’li Bir Olgu

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Abstract
Melkersson described a syndrome that consists of peripheral facial palsy and swelling of the lips. Its classical form is characterized by recurrent paralysis of the facial nerve, swelling of one or both lips, and lingua plicata (also defined as “scrotal tongue” or “fissured tongue”). In our case, rheumatoid arthritis, Parkinson’s disease, Hashimoto’s thyroiditis, hypertension and pleural effusion were found together with MRS. Whether it is only a co-incidence or there is a causal relationship between MRS and these disorders, especially rheumatoid arthritis and Hashimoto’s thyroiditis, merits further investigation. Türk Jem 2009; 13: 40-2

Key words: Melkersson-Rosenthal Syndrome, Rheumatoid Arthritis, Hashimoto’s Thyroiditis

Özet

Anahtar kelimeler: Melkersson-Rosenthal Sendromu, Romatoid Artrit, Hashimoto Tiroiditi

Introduction
Melkersson described a syndrome that consists of peripheral facial palsy and swelling of the lips, in 1928. Later, in 1931, Rosenthal included the presence of a fissured tongue as a third symptom (1). So, this triad has been called Melkersson-Rosenthal syndrome (MRS). Its classical form is characterized by recurrent paralysis of the facial nerve, swelling of one or both lips, and lingua plicata (also defined as “scrotal tongue” or “fissured tongue”). Although these signs define the classic syndrome, the classical triad is not common. On the other hand, the presence of two manifestations or one with a granulomatous cheilitis in the eyelid biopsy is considered to be sufficient for the diagnosis (3). Usually syndrome manifests during adolescence and has rarely occurred in early childhood or in individuals older than 50 years (2). Prevalence of MRS is estimated to be about 0.08%. It is not as rare as previously considered; rather some cases are not appropriately diagnosed, so prevalence of the disorder might be a little higher (4). The syndrome is seen approximately three times more frequently in females than males. Although both genetic and acquired factors have been implicated, the exact etiology of MRS remains unclear. It has been suggested that genetic predisposition is involved in the pathogenesis of the disease, because some cases occur within a family. Subsequently, a gene defect at chromosome 9 p11 has been discovered as related with MRS, so there is an evidence for autosomal dominant inheritance (5). On the other hand, genetic factors do not account for all cases, and infections (viral, bacterial), autoimmunity and hypersensitivity all have been implicated as etiologic factors with reasonable evidence (6,7). Generally, the first and most common manifestation of MRS (75% of cases) is a soft, nontender, nonpitting, and painless facial edema,
which is generally unilateral and without pruritus. The edema is located in the perioral area and the upper lip is most often involved (8). It has an acute onset, which lasts from hours to several weeks. Facial palsy, which cannot be distinguished from Bell’s palsy, might occur months before or after the beginning of facial edema. The paralysis is usually self-limiting and complete recovery is very often, but rarely it can remain with residual signs and symptoms. Lingua plicata, the third factor of the triad, is seen in almost 40% of cases, and is congenital. Furthermore, other minor signs including dysgeusia/ageusia, neuralgic pain, limnitis, vertigo, sudden deafness, anosmia, hyperesthesia, paresthesia, migraine headaches, nausea, vomiting, facial tics or spasms, can be found in the patients (9). The course of the syndrome is generally chronically recurrent or chronically progressive in most of the patients. Besides, the episodes become less severe and frequent with increased age.

Case Report

A 73-year-old woman presented with a left-sided facial edema. Her illness repeated 6 times in last three years and resolved without any treatment approximately in a week. She was complaining from dysgeusia which was accompanying the swelling of the face. Furthermore, she had experienced a right peripheral facial palsy 20 years ago. She did not state a family history related with this condition. She has had rheumatoid arthritis for 20 years, Parkinson’s disease for 15 years, hypothyroidism for 8 years and hypertension for 7 years. Her medications for these diseases included: methotrexate, ibuprofen, amlodipine and L-thyroxine. Physical examination revealed diffuse edema located in the left upper lip and cheek. A mass was palpated in the swollen cheek, which was firm, painless and 15 mm in diameter. There was no evidence of peripheral facial palsy, and bilateral otoscopic and ophtalmoscopic examinations were normal. She had tremor of the hands, rigidity in both arms and hypomimia, which are the characteristic features of Parkinson’s disease (Figure 1). We observed minimal left pleural effusion. Pleural fluid analysis was as follows: glucose level 38 mg/dl, rheumatoid factor 380 U/L, protein level 3.9 mg/dl, white blood cell count 1400/mm³, and pleural fluid pH was 7.31. These results indicate an exudative effusion because of rheumatoid arthritis. Other neurological examination and circulatory and gastrointestinal evaluations were all normal. Furthermore, we observed the deformities in her hands and in wrists including ulnar deviation and loss of extension; all were consistent with her known diagnosis, rheumatoid arthritis. Her complete blood count; erythrocyte sedimentation rate; C-reactive protein; biochemical tests including glucose, renal function tests, amylase, liver and muscle enzymes; thyroid function tests; complement [C3, C4, C1q] and Ig E levels were within normal ranges. In parotid ultrasonography, there were lesions, which were interpreted as granulomas. Therefore, we evaluated the patient to rule out granulomatous disorders. Chest radiography, angiotensin converting enzyme and calcium levels were normal. Because of her rheumatoid arthritis and swollen parotid gland, we considered the possibility of Sjogren’s syndrome, so that we looked for anti-Ro and anti-La antibodies. As a matter of fact, she had no complaints associated with Sjogren’s syndrome such as dry mouth, dry eye, etc. High rheumatoid factor 430 U/L (0-16) and anti-microsomal antibodies 1100 IU/mL (Immulfite, Diagnostic Products, Los Angeles, CA; reference range, <35 IU/mL) levels were the only positive findings that we found. Thyroid ultrasonography revealed heterogeneity in the parenchyma and there were no nodular formation in ultrasonography. With her signs and symptoms, we considered MRS as a final diagnosis, so we planned a parotid biopsy, which revealed diffuse mononuclear cell infiltration with epithelioid cell aggregates, a finding compatible with granulomatous cheilitis.

Discussion

MRS is a relatively rare disease with an unknown etiology. MRS is a systemic neuro-mucocutaneous granulomatous disease and is characterized by a triad, which includes orofacial edema, peripheral facial paralysis and lingua plicata.

The first and most common manifestation of the syndrome is a painless swelling of the upper lip, which was the presentation in our patient. Angioedema, acute allergic reaction and parotitis should be considered in differential diagnosis. In our case, complement, Ig E levels and amylase levels were normal, and history and physical examination were not correlated with these disorders. Because she had rheumatoid arthritis, one should think the possibility of Sjogren’s syndrome in this clinical scenario. She had no complaints related to Sjogren’s syndrome, and ANA, Anti-Ro and Anti-La tests were all negative, so we excluded this disorder.

The second element of the triad, peripheral facial nerve palsy, is seen in 30% to 90% of the patients with MRS that cannot be differentiated from Bell’s paralysis; our patient also had experienced it about 20 years ago (10). Lingua plicata, the third sign of the triad, was not found in our patient.

The diagnosis of MRS is difficult because the syndrome does not occur with all three signs of the classic triad. The disease more usually manifests itself in oligosymptomatic or monosymptomatic forms (11). Our case came with oligosymptomatic form of the disease. Therefore, we obtained a biopsy to confirm the diagnosis. The biopsy revealed nonnecrotizing granulomatous inflammation which was consistent with MRS. Because of this finding, MRS has been thought to be a variant of sarcoidosis (12). For this reason, we evaluated the patient for the presence of sarcoidosis by examining chest X-ray and serum ACE levels.
MRS, commonly observed in the second and third decade of life, is rarely seen in individuals older than 50 years; our patient is one of the oldest cases among the patients reported in the literature (13). In the literature, there were many disorders associated with MRS, including Crohn’s disease (14), uveitis (15), multiple sclerosis (16), Ehlers-Danlos syndrome (17), lymphoma (18), arthritis (19) and Hashimoto’s thyroiditis (20).

In our case, rheumatoid arthritis, Parkinson’s disease, Hashimoto’s thyroiditis and hypertension were found together with MRS. Whether it is only a co-incidence or there is a causal relationship between MRS and these disorders, especially rheumatoid arthritis and Hashimoto’s thyroiditis, merits further investigation.

References