



Idiopathic Melkersson-Rosenthal Syndrome: A Case Report

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Abstract

A 33-year-old Caucasian Italian female was admitted at the Campus Bio-Medico in Rome for a lip swelling evaluation of increased in last 4 days associated orofacial edema, yellowish vesicles, erosion over lips. In addition, she presented asymmetry of the face with paralysis sign on the region of VIIth nerve distribution and angle mouth deviation. Anamnesis reported abdominal pain and diarrhea 4-5 times/day since last month, severe muscle pain, fatigue and dryness eye. We supposed Melkersson-Rosenthal syndrome (MRS) based on clinical manifestation and we considered, as differential diagnosis angioneurotic edema, cheilitis glandularis, lymphangioma, sarcoidosis, tuberculosis and crohn's disease.

Keywords: Cheilitis granulomatosa; Granulomatous cheilitis; Melkersson-Rosenthal syndrome; Lip swelling

Learning Points

In patients with facial and lip edema, Melkersson-Rosenthal syndrome, Angioneurotic edema, Cheilitis Glandularis, Lymphangioma, Sarcoidosis, Tuberculosis and Crohn's disease have been considered in differential diagnosis – MRS is a diagnostic and treatment challenge.

Case Presentation

In the early days of July, a 33-year-old Caucasian Italian female was admitted at the Dermatology Department of the Campus Bio Medico in Rome for a clinical evaluation of lips edema increased in last 4 days associated orofacial edema, yellowish vesicles, erosion over lips (Figure 1). In addition, she presented asymmetry of the face with paralysis sign on the region of VIIth nerve distribution and angle mouth deviation. She reported fever (37.4°C), headache without any other symptoms. Her tongue had a normal appearance and there was no tenderness over the maxillary, ethmoid or frontal sinuses. Palpation showed hardening of the lips and mild lymph adenopathy was noted. The patient was treated previously with oral acyclovir due to incorrect diagnosis by general practitioner of herpes simplex infection. Her therapy was suspended after a day of administration.

Anamnesis reported abdominal pain and diarrhea 4-5 times/day since last month, severe muscle pain, fatigue and dryness of eyes. The patient had no history of similar complaints, angioedema or anaphylaxis, although she reported vaginal dermatitis from infancy. She had no history of skin cancer or plastic reconstruction on the lips and no pharmacological therapy before the initial of edema signs.

We supposed Melkersson-Rosenthal syndrome based on clinical manifestation and we considered, in the differential diagnosis, other illnesses as angioneurotic edema, cheilitis glandularis, lymphangioma, sarcoidosis, tuberculosis and crohn's disease.

Screening was done for crohn's disease and autoimmune pathology because MRS may present alone or in conjunction with these pathologies suspected based on symptoms, although there was no personal or family history.

Laboratory data, blood tests, thyroid hormone dosage, urine and feces exams were all surprising within normal data. Anti-*Saccharomyces cerevisiae* Antibodies (ASCA), Antineutrophilic Cytoplasmic Antibody (pANCA and cANCA) and Angiotensin Converting Enzyme (ACE test) were negative.

Patient was treated with antibiotic therapy for a week (amoxicillin-clavulanic acid), paracetamol

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Figure 1: Patient with lips edema before the treatment.



Figure 2: Lips edema after two days of treatment.



Figure 3: Lips edema in remission after one week of treatment.

and topical tetracycline, was followed up twice a week for two weeks, and had complete resolution of signs and symptoms in ten days (Figure 2 and 3). Complete resolution of fatigue and diarrhea in ten days.

Discussion

The etiology of MRS remains unknown and diagnosis may be difficult. The causes proposed in medical literature include irritants (climatic, mechanical, caustic agents), chronic infectious, autoimmune mechanisms [1] but in some cases, it may also be idiopathic. In this case, we investigated all possible etiological causes through anamnesis and laboratory exams. MRS may present alone or spectrum of various other underlying diseases [2]. Association has been seen in many other chronic disorders including crohn's disease, sarcoidosis, tuberculosis, hashimoto's thyroiditis, psoriasis, rosacea,

diabetes mellitus, leprosy and Down syndrome can be associated with this syndrome. Moreover, negative ACE Test excluded the presence of sarcoidosis.

Thyroid hormone levels were measured to search correlation with autoimmune thyroid disease but they were within normal range as well as inflammatory index. We suggested a biopsy on the lip, but the patient denied due to aesthetic issue.

As known, histological evidence is not essential to diagnosis [3] and various treatments have been reported in literature including antibiotics like tetracycline and amoxicillin, oral and intralesional corticosteroids, and surgical resection in severe cases [4,5].

In order not to compromise laboratory tests we did not administer corticosteroid at first. Edema and vesicles disappeared after ten days of treatment based on topical tetracycline antibiotics and oral amoxicillin-clavulanic acid. *Lactobacillus fermentum* prescription was needed to counteract acute diarrhea. Considering the patient improvement following a few days of therapy and observing the cheilitis resolution, we decided to not administer corticosteroid to reduce furthermore the inflammation.

Conclusion

Certainly, MRS represents a challenge in terms of diagnosis and treatment. The clinical manifestation in this case was compatible with MRS after intensive investigation through amnesia and laboratory tests, the case was classified as idiopathic. We suspect that an infectious issue or an inflammatory bowel process, involved in a granulomatous process that manifested with diarrhea and cheilitis after being asymptomatic for a long time, was the cause of her clinical manifestations.

In conclusion, this case report underlines the importance of not underestimate lip edema etiology, including investigation about gastrointestinal symptoms, which may alert the clinician to suspect undiagnosed crohn's disease, as well as inquire about a chronic cough, which may indicate sarcoidosis or tuberculosis, considering that very often the cause is idiopathic.

References

1. Samimi M. Cheilitis: Diagnosis and treatment. *Presse Med.* 2016;45(2):240-50.
2. Okudo J, Oluyide Y. Melkersson-Rosenthal Syndrome with orofacial swelling and recurrent lower motor neuron facial nerve palsy: A case report and review of the literature. *Case Rep Otolaryngol.* 2015;2015:214946.
3. Chatzistefanou I, Zikos P, Venetis G, Vahtsevanos K, Antoniadis K. Melkersson-Rosenthal syndrome. *World J Stomatol.* 2015;4(1):8-11.
4. Aldeolivas-Casillas N, Piteiro-Bermejo AB, Trasobares-Marugán L. Lip swelling in an immunocompetent man. *Actas Dermosifiliogr.* 2015;106(5):430.
5. Innocenti A, Innocenti M, Taverna C, Melita D, Mori F, Ciancio F, et al. Miescher's cheilitis: Surgical management and long term outcome of an extremely severe case. *Int J Surg Case Rep.* 2017;31:241-4.