
One of the Faces of Facial Swelling: Melkersson-Rosenthal Syndrome

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Angioedema involves rapid swelling of the skin, which typically lasts no more than 5-7 days. The affected tissues return to normal with no sequelae. Some entities with facial swelling can be misdiagnosed with angioedema. Such is the case of Melkersson-Rosenthal syndrome (MRS) [1].

MRS is a systemic disease characterized by idiopathic facial paralysis, fissured tongue, and orofacial edema that persists for weeks or months. The syndrome was described separately by Melkersson in 1928 and Rosenthal in 1931 [2].

The classic triad is rarely present, with oligosymptomatic or monosymptomatic forms being more frequent.

We describe the case of a 67-year-old woman who was referred to our Allergy Department because of recurrent facial angioedema.

She had a personal history of diabetes mellitus, dyslipidemia, hypertension, allergic rhinitis, and sensitization to *Anisakis simplex*.

At the age of 27 years, she experienced an episode of right facial paralysis, with complete recovery (no visible sequelae) approximately 10 days after treatment with corticosteroids.

In January 2011, she experienced a new episode of right facial paralysis with edema of the upper lip and without pruritus or erythema. She improved slightly with systemic corticosteroids. A month after this episode, she had a new episode of facial edema associated with another episode of facial paralysis with right ptosis, lagophthalmos, and dysgeusia. Slight facial asymmetry due to residual edema persisted (Figure).

During the following years, she had a further 6 episodes of facial edema, all of which lasted 5 to 7 days, with an incomplete response to antihistamines. She was sent to the Allergy Unit for recurrent angioedema. After the last episode, the slight facial swelling became progressively permanent. On physical examination, she presented a fissured tongue (Figure) and facial edema with no other symptoms.



Figure. Slight facial asymmetry due to residual edema and fissured tongue.

During the edema attacks, she was prescribed oral deflazacort beginning at 30 mg daily. This improved the edema, albeit without total resolution.

Laboratory examinations (complete blood count, biochemistry, sedimentation rate, antinuclear antibodies, TSH, T4, complement C3 and C4, and functional and antigenic C1 esterase inhibitor) were within normal ranges. The determination of antibodies for hepatitis virus (A, B, and C), herpes simplex virus, varicella zoster virus, and cytomegalovirus were negative or within normal limits. The result of a Mantoux test was also negative.

Histological examination of a skin biopsy from the right buccal mucosa showed epithelial hyperplasia and marked edema without granulomas. Electromyography revealed active denervation of all the muscles of the right facial nerve with distal conduction preserved, progressive acute axonal injury, and marked reinnervation across the right facial muscles. Findings on the cranial CT scan, electrocardiography, and chest radiography were normal.

Angioedema is a self-limiting, localized swelling caused by increased vascular permeability that involves subcutaneous tissues and/or mucosa. MRS is an entity where swelling is also present [3].

In 1928, Melkersson reported the association of recurrent facial palsy with labial edema, and, in 1931, Rosenthal described the presence of *lingua plicata*, or fissured tongue, as an additional manifestation that is sometimes the only characteristic. Its presence must be considered in the differential diagnosis of MRS [4]. Orofacial edema is the most dominant characteristic of the triad.

MRS can reappear at irregular intervals and differs from angioedema because it becomes permanent after a specific number of episodes. Onset can be at any age, although patients aged between 25 and 40 years are most typically affected [5].

According to Launay et al [3], during the recurrent edema phase, the syndrome may be misdiagnosed as angioedema. When edema becomes permanent or lasts more than 7 days, a diagnosis of MRS should be considered. Persistent local cutaneous edema, such as the facial swelling that is present in MRS, should not be considered angioedema [6].

The patient we report experienced recurrent episodes not only of facial edema, but also of facial palsy. More recently, the edema became progressively permanent. Streeto et al [7]

also reported a patient who had experienced 10 episodes of Bell palsy with repeated episodes of facial edema.

The full triad is uncommon. Elias et al [8] reported a series of 72 patients with facial edema and noncaseating granulomas on skin biopsy. Fissured tongue occurred in 47% of cases and unilateral or partial facial nerve palsy in only 19%. Elias et al also reported that in those cases affected by the full triad, 78% were women.

Noncaseating granulomas can be found in the biopsy specimen from a region with orofacial edema [8]; however, histologic evidence is not necessary for the diagnosis of MRS, and these findings are not present in all biopsy specimens [9].

There is no consensus on the optimal therapeutic approach to MRS, although corticosteroids have proven useful for controlling orofacial edema. Antibiotics and/or pain relievers may also be indicated. Facial reconstructive surgery, massage, and electrical stimulation could also be taken into consideration [10].

We present a case of MRS that manifested with the classic triad: idiopathic facial paralysis, fissured tongue, and orofacial edema. The syndrome was initially misdiagnosed as angioedema. MRS should be considered in the differential diagnosis of recurrent angioedema.

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Conflicts of Interest

The authors declare that they have no conflicts of interest.

Previous Presentation

Data for this patient were presented in poster form at the XXIX SEAIC congress, Salamanca, Spain, 2014.

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