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Abstract

Melkersson-Rosenthal syndrome (MRS) is a rare disease characterized by a triad of relapsing or persistent orofacial edema, recurrent lower motor neuron facial nerve palsy and fissured tongue. Acute, painless, non-erythematous orofacial edema is observed in all patients. We report a case of a 13-year-old girl who presented with a 2-year history of swelling of the upper lip, facial paralysis, and fissured tongue; she was treated successfully with dapsone.

Keywords: dapsone; treatment; Melkersson-Rosenthal syndrome

A 13-year-old girl was referred to our clinic because of swelling of upper lip and face. The dermatological examination revealed severe asymmetric facial edema (Figure 1). She had a fissured tongue (Figure 2). Although we did not observe facial palsy during examination, she had a history of facial paralysis three times in the last two years. MRI of the brain and neck were unremarkable. She had the characteristic triad of MRS that consists of relapsing orofacial swellings, recurrent lower motor neuron facial nerve palsy, and fissured tongue.
When she presented to our clinic, she had not taken oral prednisone for a month. She did not have symptoms of MRS during oral prednisolone treatment but when oral prednisolone was stopped, the patient suffered a relapse of symptoms. Because of the recurrent episodes, a safe and long term treatment was required. Therefore, we prescribed a single daily dose of dapsone 50 mg, instead of corticosteroids. From the first month of treatment, a significant improvement of facial edema was seen (Figure 3). We observed no reactivation of orofacial edema and facial paralysis during the six-month follow up.
MRS is a rare disease characterized by a triad of relapsing or persistent orofacial edema, recurrent lower motor neuron facial nerve palsy, and fissured tongue. Acute, painless, non-erythematosus orofacial edema is observed in all patients. Asymmetrical edema involves mainly lips, but also the cheek, nose, eyelids [1].

Various therapeutic methods were described for the treatment of MRS but most of them are inadequate. Corticosteroids, clofazimine, dapsone, sulfasalazine, sulfapyridine, hydroxychloroquine, antihistamines, antibiotics, methotrexate, infliximab, and surgery are the treatment options [2-4].

Corticosteroids are usually preferred as an initial treatment but the route of administration is controversial. Some authors recommend systemic corticosteroids, whereas others recommend topical, intralesional, or subcutaneous corticosteroids. Although intralesional triamcinolone was postulated to be efficient and safe in orofacial granulomatosis, its effect is unknown in patients with involvement of facial nerve palsy [4]. Recently, Qudhriri et al. showed that intralesional betamethasone and doxycycline could be a useful alternative therapy [5]. Corticosteroids are quite effective but they have high risk of side effects with long-term usage. Therefore, other treatment choices for MRS are needed. Sussman et al. observed complete remission of five patients with clofazimine treatment [6], and Ratzinger et al. reported an association of cheilitis granulomatous and Crohn disease and showed a good response to clofazimine and infliximab [7]. In a few reports, successful treatment with dapsone was also seen. Van der Kooi et al. reported a patient who was treated successfully with dapsone and triamcinolone injections [8]. Rozen presented a patient with MRS and headache who responded to dapsone (100 mg per day) [9].

Dapsone (diaminodiphenylsulfone) is a sulfone antibiotic used for the treatment of leprosy. Because of its anti-inflammatory reactions, it has been used for the treatment of several dermatological diseases. These include neutrophilic and/or eosinophilic dermatoses, such as dermatitis herpetiformis, subcorneal pustular dermatosis, pyoderma gangrenosum, Sweet disease, erythema elevatum diutinum, and eosinophilic pustular folliculitis.

In our patient, dapsone has been very effective. Therefore, if there are recurrent episodes, we suggest that dapsone can replace corticosteroids because it is safer for long term treatment.

REFERENCES


