

Melkersson-Rosenthal Syndrome



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FIGURE 1. Patient with lip edema.

A 9-year-old girl presented with lip swelling, drooling, and deviation of the angle of her mouth to the left side for 3 days. She had a similar episode 2 years back. There was no history of trauma, fever, or similar illness in the family. On examination there was edema of the lower lip (Figure 1) with fissuring of the tongue and left side lower motor neuron facial palsy (Figure 2). A diagnosis of Melkersson-Rosenthal syndrome was made and she was started on a tapering dose of prednisolone for 3 weeks. The facial palsy was completely resolved and there was no recurrence at 7-month follow-up. Melkersson-Rosenthal syndrome was described separately by Melkersson and Rosenthal in 1928 and 1931, respectively. It is characterized by recurring

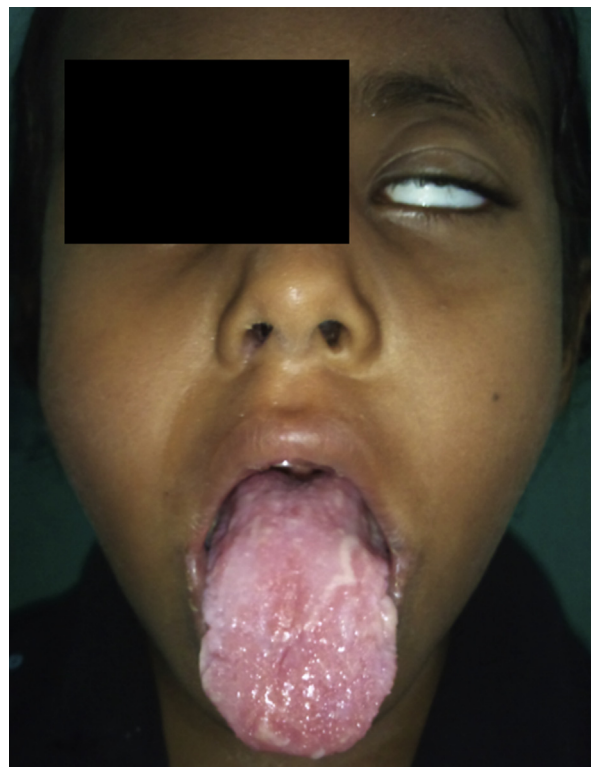


FIGURE 2. Patient with fissured tongue and lower motor neuron facial palsy.

facial paralysis, swelling of the face and lips, and the development of folds and furrows in the tongue.¹⁻³ The facial nerve weakness may occur after months or years of facial swelling, may present on one or both sides of the face, and can become permanent after multiple episodes.¹ Although genetics, infections, family history, food allergies, or inflammation have been implicated, the evidence of a cause or underlying pathophysiology is lacking.^{2,3} It may present alone or in conjunction with other chronic diseases such as Down's syndrome, sarcoidosis, Crohn's disease, psoriasis, leprosy, multiple sclerosis, HIV, autoimmune thyroiditis, ocular palsies, and diabetes mellitus.^{2,3} Although not always necessary, a biopsy may be required to differentiate the syndrome from other conditions, especially Crohn's disease. The histopathology shows intra- and extravascular clusters of histiocytic-epithelioid cells and noncaseating sarcoid granulomas.⁴ A multidisciplinary team approach involving otolaryngologists, dermatologists, and neurologists is required to manage these patients. Most patients are managed with steroids and nonsteroidal anti-inflammatory drugs,¹⁻³ and other drugs that have been tried are danazol and sulfasalazine.^{2,3} Facial nerve decompression may become necessary for patients with recurrent facial paralysis.¹

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