Case Presentation:
A 28 year old woman presented with a 10 year history of recurrent intermittent unilateral facial paralysis associated with otalgia and mild nonpitting edema. Patient denied edema of the lip, oral cavity, or eyelids. She also denied fever, hearing loss, otorrhea, or vesicles in the ear canal or auricle. Her symptoms improve after intake of steroids.
On physical exam, the patient had a House-Brackman score of 2, a Normal ear exam, no facial swelling, and no lip swelling or atrophy. She did however have lingua plicata.

Figure 1: Residual Facial Weakness

Figure 2: Presence of Lingua Plicata

Her workup included a MRI of the brain that showed no evidence of tumor in the IAM or CPA. Puretone audiometry showed normal hearing and good speech discrimination.
**Diagnosis:**
Melkersson-Rosenthal Syndrome (MRS).

**Discussion:**
MRS is described in the literature as a triad of symptoms: recurrent orofacial edema, recurrent facial palsy, and lingua plicata (fissured tongue). The complete triad is present in one-fourth of the patients (1). Although MRS is classically described as edema-dominated, recent review articles are stressing facial paralysis (FP) as the dominant factor. Review of characteristics of 11 MRS patients w/ FP revealed:

- 9 patients had the triad form
- 2 patients had non-necrotizing granulomatous infiltrations during acute edema episodes, and
- 2 had association with uveitis.

Edema was rarely persistent and did not dominate the clinical picture (2). The condition generally begins in the second decade of life and a biopsy specimens of the lip reveal non-caseating epithelioid cell granulomas surrounded by histocytes, plasma cells, and lymphocytes (1).

The cause of MRS is unknown; however, it is believed to be a variant of sarcoidosis whereby elevation of angiotensin-converting enzyme is documented in many cases. Moreover, a lip biopsy specimen showing non-caseating granulomas further supports this theory. Another possible etiology is primary vasomotor disturbance. A third etiology is of allergic cause (1). Another hypothesis concerning etiology is a deficiency in functional plasma protease C1-INH that may contribute to the orofacial swelling, since low levels of C1-INH are known to lead to recurrent edema. However, this theory was not supported by a recent review of seven MRS patients whereby only two patients were found to have low C1-INH (3). Finally, a recent case report negated the role of IgE-mediated reactions in the development of MRS after reporting a case of a woman with MRS that was found to have a markedly low serum IgE level (4).

Most commonly, treatment of MRS has been observation and steroid therapy as most of the attacks resolve. However if facial paralysis is persistent, management is indicated. There are no randomized trials to assess efficacy of corticosteroids versus surgical decompression. Cessation of the recurrent facial paralyses has been reported in several series after facial nerve decompression.

**References:**
2. Kanerva et al.08 Melkersson-Rosenthal syndrome. Otolaryngology, Head and Neck Surgery