Melkersson–Rosenthal syndrome (MRS) is a rare neuro-mucocutaneous granulomatous disorder characterized by a triad of orofacial swelling, recurrent facial palsy, and lingua plicata (fissured tongue).1 The complete triad of symptoms presents in a minority of patients, reported between 8% and 18% of cases, but the monosymptomatic and oligosymptomatic forms are the most common presentations.2 We present a case of a 71-year-old man who presented with the classic triad of symptoms of MRS and underwent surgical management for his orofacial edema.

CASE PRESENTATION

A 71-year-old man with a medical history of recurrent squamous cell carcinoma of the lower lip, lagophthalmos of the left eye, and coronary artery disease was evaluated at an outside hospital 3 years ago for a 2-year history of upper lip swelling. He was diagnosed with orofacial granulomatosis by punch biopsy by dermatology. Pathology at that time was negative for active disease. His lip swelling was refractory to systemic steroids, corticosteroid injections, doxycycline, TNF-alpha inhibitors, and hydroxychloroquine. He was subsequently referred to plastic surgery and underwent liposuction of the abdomen with autologous fat grafting to the lower lip to restore symmetry to the mouth. Additionally, he had seen ophthalmology numerous times for recurrent bilateral lagophthalmos and bilateral mechanical ptosis of the brow.

He then presented to our plastic surgery clinic, now with 5 years of persistent upper lip swelling. On physical examination, the patient demonstrated upper lip enlargement with prominence of the vermillion border and cephalad lip displacement (Fig. 1). He also presented with redness and tearing of the left eye, consistent with his history of lagophthalmos. A deep central fissure groove was present on the dorsal surface of the tongue, consistent with lingua plicata, and fulfilling the characteristic triad of Melkersson–Rosenthal syndrome. Considering his refractory response to medical management, surgical debulking of the upper lip was planned.

Surgical debulking was performed via the Conway method.3,4 A transverse incision was made approximately 1 cm dorsal to the vermilion border and cephalad lip displacement (Fig. 1). He also presented with redness and tearing of the left eye, consistent with his history of lagophthalmos. A deep central fissure groove was present on the dorsal surface of the tongue, consistent with lingua plicata, and fulfilling the characteristic triad of Melkersson–Rosenthal syndrome. Considering his refractory response to medical management, surgical debulking of the upper lip was planned.

Surgical debulking was performed via the Conway method.3,4 A transverse incision was made approximately 1 cm dorsal to the vermilion border. Dissection was carried out through the mucosa to the level of the orbicularis. A superior plane consisting of skin and dermis and an inferior flap consisting of oral mucosa were dissected, allowing for isolation of the orbicularis oris. Redundant mucosa was excised along with a small redundant portion of the orbicularis oris, with careful consideration to leave the bulk of the muscle intact to preserve oral continence and function.

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A multilayered closure was performed to minimize tension on the mucosa. Finally, Kenalog-40 was injected into the upper lip to reduce inflammation and edema postoperatively. Final pathology showed multinucleated giant cells, noncaseating granulomas, and perivascular inflammation, confirming the diagnosis of MRS (Fig. 2).

At the 2 week and 5 month follow-up visits, there was clear reduction in upper lip volume and minimized vermilion show (Figs. 3, 4). Oral continence and labial sensation were fully intact on examination.

**DISCUSSION**

The diagnosis of MRS is difficult due to its rarity and its varied presentations. The characteristic triad of symptoms presents in up to one fourth of patients, but the mono- and oligo-symptomatic forms of the disease are more common. Hornstein et al note that the true incidence of MRS is likely underreported in the literature. The treatment of MRS is equally challenging, owing in part to the wide spectrum of specialists who encounter these patients. Patients with facial paralysis more often present to otolaryngology, whereas those with facial edema typically present to dermatology or plastic surgery. In fact, our patient had been evaluated separately by both dermatology and ophthalmology for two seemingly independent complaints of lip swelling and periorbital facial paralysis.

The current understanding of the progression of orofacial edema in MRS identifies two phases: an initial, relapsing inflammatory phase and an eventual, quiescent noninflammatory phase. Worsaae et al considered the disease to be inactive and in the noninflammatory phase when no exacerbation of symptoms had occurred for at least 1 year. Medical management may best address the inflammatory phase of the disease. Therapies for orofacial edema in MRS include corticosteroids, nonsteroidal anti-inflammatory drugs, clofazimine, antihistamines, and antibiotics. Radiation of the lip has also been reported. Notably, our patient had failed multiple medication trials over several years before presentation to our clinic.
Surgical management, typically reduction cheiloplasty, is considered when the orofacial swelling is persistent and refractory to noninvasive treatments. Numerous surgical approaches have been reported, focusing on restoring lip symmetry with conservative resection of affected mucosa. In the Conway procedure, a transverse mucosal incision is made dorsal to the vermillion border to resect the affected mucosa. The Conway method can be modified with a central wedge excision with or without a Z-plasty. Mouly’s approach preserves the natural central depression of the upper lip. Cederne et al later demonstrated a fleur-de-lis pattern resection with transmodiolar sutures acting as a labial splint. In our case, the Conway method was performed. Notably, a small portion of the orbicularis oris was resected due to muscular redundancy; however, oral continence remained intact postoperatively.

Postsurgical recurrence of orofacial edema in MRS has only been reported in small case series. Vistnes et al reported two patients with recurrence. In a later case series of 13, local recurrence occurred in only one patient. Notably, this patient had developed acute exacerbation of labial swelling 1 week before surgery, suggesting that he was in the inflammatory phase of the disease. In contrast, our patient did not report any recent exacerbations and a biopsy 2 years before surgery was negative for active disease.

CONCLUSIONS
The case presented highlights the difficulty in diagnosis and treatment of Melkersson–Rosenthal syndrome as our patient had been evaluated by multiple specialists for different symptoms of the classic triad. To treat these patients effectively, it is of utmost importance to recognize the diagnosis of MRS and note their disease state based on symptom progression, response to treatment, and histological findings. Surgical excision should be considered when medical therapies have failed, and exacerbations have not occurred for at least 1 year.

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