A Surgical Approach to Persistent Orofacial Swelling: A Rare Case of Melkersson-Rosenthal Syndrome

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Introduction: This is a case of a patient who presented with a years-long history of upper lip swelling, which had previously been diagnosed as orofacial granulomatosis.

Case Presentation: A 71-year-old man with coronary artery disease and lagophthalmos of the left eye presented with a five-year history of progressive upper lip enlargement. Three years ago, he was diagnosed at an outside hospital with orofacial granulomatosis by punch biopsy. He later underwent liposuction of the abdomen and fat grafting to the lower lip in an attempt to restore symmetry to the mouth.

On presentation to our Plastic Surgery clinic, physical examination revealed dramatic enlargement of the upper lip with prominence of the vermillion border. Additionally, he was noted to have a fissured tongue. With these two features and his history of recurrent facial palsy in the setting of lagophthalmos, his presentation fit the clinical criteria of Melkersson-Rosenthal syndrome.

Final Diagnosis: The final diagnosis was Melkersson-Rosenthal syndrome, a rare neuro-mucocutaneous syndrome consisting of a clinical triad of orofacial swelling, relapsing facial palsy, and lingua plicata. All three of these features were seen in this patient.

Management/Outcome: Melkersson-Rosenthal syndrome often presents in two phases: an initial inflammatory phase, during which medical management is indicated, and a subsequent noninflammatory phase, at which time surgical intervention can be considered. Of note, this patient had previously been treated with intralesional corticosteroid injections and TNF-alpha inhibitors. Unfortunately, his lip enlargement continued despite medical management.

Surgical debulking of the upper lip was performed via the Conway method, in which a mucosal incision was made 1 cm dorsal to the vermillion border with resection of the affected mucosa and partial excision of the orbicularis oris. Two weeks postoperatively, the patient demonstrated significant reduction in upper lip volume as well as minimized vermillion show. Oral continence and labial sensation were preserved.

Learning Objectives:

- Recognize the characteristic findings of Melkersson-Rosenthal Syndrome as well as identify the need for surgical intervention in patients with orofacial swelling which has progressed to fibrosing lymphedema
- Implement a proper treatment strategy based on the two phases of the disease, with medical management indicated for the initial inflammatory phase, and surgical treatment indicated during the subsequent stable phase of the disease