Melkersson-Rosenthal Syndrome (MRS), also known as orofacial granulomatosis, is a rare neurological disorder characterized by a triad of face swelling, tongue furrowing, and facial paralysis. At least two criteria from the triad are needed to make the clinical diagnosis. Flares of disease may occur intermittently and recur with days to months in between. The onset of symptoms typically occurs in young adulthood.

The cause of MRS is unknown, but there may be a genetic predisposition. Some cases of MRS have been attributed to a hypersensitivity reaction or viral infection. As a granulomatous disease, it is suspected that MRS may be the first presenting sign in a patient who will later be diagnosed with sarcoidosis or Crohn’s disease. The cause of these diseases is based in granuloma formation and underlying links have been considered.

Facial swelling or “orofacial edema” is considered the characteristic symptom of MRS and may be the first or only symptom present. This inflammation affects the facial nerve and commonly leads to swelling in the lips or “cheilitis granulomatosis,” and usually, the upper lip is affected before the bottom lip. Lips may become hard and fissured with a reddish-brown discoloration. Swelling may also occur around the mouth and eyes. Some people are mis-diagnosed as having an allergic reaction. The first episode of edema may resolve in a few hours or days. Swelling may become more severe and last longer in subsequent episodes, which can lead to permanent scarring. Isolated and persistent lip swelling is characteristic of cheilitis granulomatosis or “Miescher cheilitis.” Whereas lip swelling accompanied by facial palsy and furrowed tongue is MRS.

Furrowed tongue, also known as scrotal tongue or lingua plicata, consists of deep grooves on the top of the tongue. Fissuring may be present at birth and is typically non-painful. Tongue fissuring may lead to local infections, tongue enlargement, loss of taste buds, and an itching or burning sensation.

Facial palsy may be unilateral or bilateral and usually occurs after the first few episodes of edema. Recurrent facial paralysis occurs in 10% of cases. As the disease progresses, the duration of facial palsy becomes longer. Some may experience paralysis of other muscles in the head and neck associated with different cranial nerves.

Other neurological symptoms of MRS include migraines, dizziness, tinnitus, deafness, difficulty swallowing, and visual disturbances. Non-neurological symptoms include uveitis and diverticulitis.

Treatment is aimed at relieving swelling and decreasing inflammation. Symptomatic therapies include **intralesional steroids**, oral nonsteroidal anti-inflammatory drugs, and **antihistamines**. Other systemic therapies include antibiotics, **TNF-alpha inhibitors**, and **methotrexate**. Surgery may be an option to relieve pressure on the facial nerves. Disease severity is dependent on nerves involved, secondary symptoms, and progression to systemic granuloma formation.

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