

Melkersson Rosenthal Syndrome: Oligosymptomatic Form

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The Melkersson Rosenthal Syndrome (MRS) is a rare condition characterized by the triad of recurrent lower motor neurone (LMN) facial palsy, facial edema and furrowed tongue. Cheilitis granulomatosa is now being considered an additional feature. This case is reported to emphasize the incomplete forms of manifestation of this syndrome which often goes unrecognized and which has to be differentiated from other more common causes of LMN facial palsy, particularly, Bell's Palsy. As yet, there is no reported case in Indian literature.

Case Report

A 9-year-old female born of a non-consanguineous marriage was admitted to the hospital with history of edema of the face and eyelids, inability to close the eyes com-

pletely and difficulty in opening the mouth for the past 15 days with constitutional symptoms being limited to vague myalgia. There was no history of preceding rash, fever, acute respiratory infection, otorrhea, trauma, insect bite or toothache. Her past history and family history were not contributory.

On examination, her vital parameters were normal. Positive findings on general examination included non-inflammatory, non-indurated edema of both the lips and perioral region with minimal edema of upper eyelids. Oral mucosa, tongue and gums were normal. CNS examination revealed a bilateral LMN type of facial palsy, which was more prominent on the left side. Other cranial nerves were normal as was the rest of the CNS and systemic examination.

Investigations included normal blood counts, an ESR of 29 mm at the end of 1 h, a negative test and normal CSF. Immunoglobulin studies showed total IgG 1350 mg/dl, IgM 400 mg/dl and IgA 40 mg/dl. Incisional biopsy of the lip and tongue, done 2 weeks after admission did not show any significant pathology. Electrophysiological studies were not available for lack of follow up.

The patient was treated with Paracetamol (30 mg/kg/day). The facial edema subsided within 2 weeks but the facial paresis persisted.

Discussion

The association between recurrent facial palsy and facial edema was first made by Melkersson in 1928. Fissured tongue was added to the syndrome by Rosenthal in 1931. Meischer in 1945(1) described 'cheilitis granulomatosa' in these patients and this is now considered as an additional feature of this syndrome. The triad of symptoms, facial palsy, facial edema and

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plicated tongue, constitute the classical Melkersson Rosenthal Syndrome. If only two manifestations are present, the syndrome is considered "incomplete", as seen in this case. When the incomplete or oligo-symptomatic forms are considered, the MRS may be more common than previously thought. Incidence ranges from 4-13% of analysed cases of Bell's palsy. The age of onset ranges from 2-70 yrs and the syndrome is more common in females especially in the first decade of life.

Inherited dysfunction of the autonomic nervous system(2) with vasomotor edema of *vasa nervorum* and arterioles is postulated as the possible pathogenetic mechanism for the facial edema and facial palsy. The etiologies proposed have been infection, allergic and heredo-familial. Inheritance is thought to be autosomal dominant with variable penetrance(3). The findings of biopsy from the involved areas have shown non-specific edema and dilated lymph channels in the early stages with perivascular aggregate of lymphocytes and plasma cells(4). In later phases and following recurrent attacks, non-caseating granulomas with Langhans type giant cells have been seen with increased connective tissue, for which the term "idiopathic fibroedema" was suggested by Stevens(5).

The facial palsy in MRS is typically of the LMN type, unilateral or bilateral, sudden in onset, often recurrent and in most cases clearing completely. The paralysis may be partial or complete, occasionally sensory defects of anterior 2/3 of tongue have been noted. Repeated attacks can however, cause permanent damage. The facial edema is non-erythematous, painless and non-pruritic often confined to the lips, but may involve the cheeks, eyelids and forehead. Recurrent attacks can, make the edema permanent. It has been stated that

at least one episode of paralysis ordinarily precedes the onset of edema, but the association may be coincident or it may be separated by as long as 25 years. Fissured tongue is the least important component seen in only 30% of cases(6). Gingival manifestations as described by Pindsborg and Worsaae(7) are small, irregular, edematous swellings arising from the gingival mucosa. Cheilitis granulomatosa is the name given to the non-caseating granulomas of the edematous tissues and which is now being considered as even the sole manifestation of the oligosymptomatic form of the disorder. Other associated symptoms that have been described are paraesthesia of tongue or cheeks reflecting Vth nerve involvement, ptosis or diplopia suggesting IIIrd nerve involvement, migraine-like headaches and dysfunction of autonomic nervous system reflected as aberration of lacrimation, sweating, salivation or gustatory sensation and anisocoria(8). Differential diagnosis includes all causes of LMN facial palsy, Ascher's syndrome and Heerford's syndrome(9).

Often the attack subsides spontaneously and management of facial palsy in the acute stages and good oral hygiene are all that is needed. Other modalities of treatment like irradiation, quinacrine, ultrasound, intralesional and systemic steroids have been tried with equivocal results. If attacks are very frequent and permanent damage has ensued, surgical decompression of the facial nerve, gingivectomy or wedge resection of lips have been recommended(10).

There are reports of patients with MRS having developed Hodgkins disease(11) and its potential as a probable forerunner of a malignant condition has been postulated. This becomes an important reason to identify this condition.

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Congenital Subclavian Artery Aneurysm

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It is usually assumed that if an aneurysm appears at an early age and if there is no history of trauma, systemic infection or presence of atherosclerosis, it must have developed due to some inherent weakness of the arterial wall(1). The term "congenital" aneurysm is obviously a misnomer, since it is the arterial wall weakness rather than aneurysm itself that is present at birth. It is often difficult in such aneurysms to identify the responsible congenital factor. We are reporting a case in which a congenital left subclavian artery aneurysm was diagnosed in a 11-month old child. No similar case appears to have been reported previously at this age.

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