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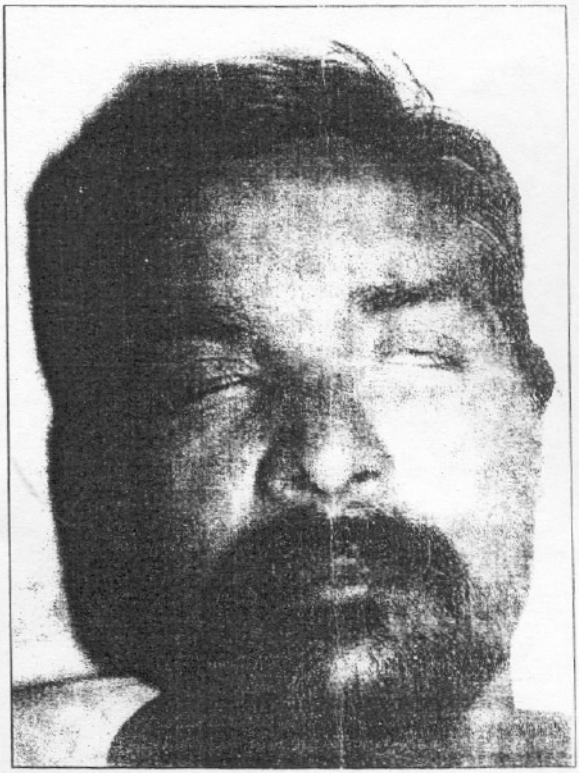
Correspondence

LEPROSY MASQUERADING AS MELKERSSON–ROSENTHAL SYNDROME

Sir,

A 45-year male patient presented himself with complaints of itching, tingling, burning sensation, pain and swelling over the upper part of the face, excessive watering of eyes, headache and nasal obstruction of 3 months' duration. Examination revealed erythema, oedema, raised skin temperature and tenderness over bilateral supraorbital and malar areas, eyelids and upper lip.

Fig. Orofacial oedema and bilateral lagophthalmos



During the course of multidisciplinary evaluation of the patient, various diagnoses were considered including frontomaxillary sinusitis (otorhinolaryngologist), preseptal cellulitis with ethmoiditis (opthalmologist), and hypothyroidism/angiooedema (physician). The results of a complete haemogram, urinalysis, X-ray of paranasal sinuses, IOPA (intraoral periapical) X-ray and cranial CT scan were normal. The physician advised oral prednisolone 40 mg OD following which oedema decreased slightly.

Opinion of the dermatologist was sought and we noticed bilateral lagophthalmos (Fig 1), which was apparently of recent onset. There was no significant nerve thickening or sensory loss. The results of oral examination was normal. Based on the clinical features of orofacial oedema and bilateral facial palsy, a provisional diagnosis of Melkersson-Rosenthal syndrome (MRS) (oligosymptomatic variant) or leprosy with type 1 reaction was considered. Slit-skin smear from the lesion was negative for AFB, while skin biopsy showed thinning of epidermis lymphocytic infiltrate around dermal appendages and neurovascular bundles, focal epitheloid cell aggregation, Langhans giant cells and negative AFB, consistent with BT leprosy. The patient was put on MB-MDT and a tapering dose of oral steroids to which he responded well.

Melkersson-Rosenthal syndrome is a rare noncaseating granulomatous disease, chracterised by a classical triad of recurrent or persistent orofacial swelling, relapsing facial paralysis and a fissured tongue (lingua plicata or scrotal tongue). The complete triad is seen only in 25%-30% of patients, while it presents more often as monosymptomatic (granulomatous cheilitis of Miescher) or oligosymptomatic variants (Rogers 3rd, 1996). Though the aetiology is largely unknown, recently mycobacteria (*M. tuberculosis* complex) have been shown to have a possible role (Apaydin *et al*, 2004).

Bilateral facial nerve palsy in leprosy has been reported in various studies (Richard & Corry, 1997). Lagophthalmos can be found in all types of leprosy (Jopling & McDougall, 1996). Bilateral lagophthalmos has been found in both multibacillary and paucibacillary cases with skin lesions, whereas only unilateral lagophthalmos has been reported in pure neuritic leprosy (Lubbers *et al*, 1994). Most of the lagophthalmos is due to type 1 reaction in the borderline (BT, BB or BL) leprosy patients (Jopling & McDougall, 1996).

Leprosy is reputed as a "great mimicker" because of its varied clinical presentations (Karat, 1978). According to the seventh report of WHO Expert Committee on Leprosy, the diagnosis of leprosy depends on the presence

of one or more of the 3 cardinal signs (WHO, 1998). However, there is evidence that clinical signs alone are unreliable and histopathology is essential for a correct diagnosis of leprosy (Ponnighaus & Fine, 1998), as happened in the present case, where clinical presentation was more in favour of Melkersson-Rosenthal syndrome for want of cardinal signs of leprosy, but histopathological evaluation established the diagnosis. The differences between leprosy and MRS are highlighted in the Table in the following page.

Table. **Differences between leprosy and MRS.**

	Leprosy	Melkersson-Rosenthal Syndrome
Aetiology	Infection (*M. leprae*)	Unknown; Proposed aetiologies: - Genetic - Infection (*M. tuberculosis* complex) - Autoimmune (Association with Crohn's disease)
Sex distribution	Male preponderance	Female preponderance
Diagnostic features	3 cardinal signs (presence of 1 or more diagnostic signs): (a) hypopigmented/reddish skin lesion(s) with definite loss of sensation, (b) peripheral nerve involvement with definite thickening and loss of sensation, (c) skin smears positive for AFB	Classical triad (however, mono/oligo symptomatic variants more common): (a) recurrent/persistent orofacial swelling, (b) relapsing facial paralysis, (c) fissured tongue (scrotal tongue/lingua plicata)
Histopathology	Depends on type; perineural/ neural tuberculoid granulomas foamy macrophages & AFB	Non-caseating sarcoidal granulomas
Treatment	Anti-leprosy therapy (MDT)	Corticosteroids (topical/intralesional/systemic); Other drugs (antibiotics, NSAIDS, clofazimine, thalidomide); Surgical

Therefore, the absence of cardinal signs does not rule out the possibility of leprosy, considering the varied and atypical presentations of the disease.

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