MELKERSSON-ROSENTHAL SYNDROME
(Cheilitis Granulomatosa))
Case Report

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The syndrome consists of recurrent facial swelling, recurrent facial paralysis, and fissured or scrotal tongue (lingua plicata). Any two symptoms of the triad may represent incomplete forms of the syndrome, and therefore may be acceptable for the diagnosis (Klaus & Brunsting, 1959).

Case report

The patient, a 38 year old female, first noticed diffuse swelling of the upper lip at the age of 13. Initial attacks lasted 3 months with complete resolution.

One year later, patient complained of paralysis of the right side of the face, with inability to close eyes. The palsy lasted 3 months with full spontaneous recovery. It recurred 8 years afterwards, lasted 5 months, and again there was complete recovery. The third attack occurred after another 6 year period, again with full recovery, but taking about 1 year to clear up with electrical massages.

Three years ago, the macrocheilia recurred, with severe attacks every 3 months, and mild attacks every 1½ months. Each attack lasted approximately 10 days, and progressively, there was incomplete resolution of the upper lip swelling. As this increased, it became harder in consistency, and involved the right cheek as well.

Patient's son, 13 years old, started having recurrent swelling of the upper lip with complete resolution. No fissured tongue was present.

Examination of the patient showed firm, non-pitting oedema (peau d'orange appearance) of the right cheek and upper lip.

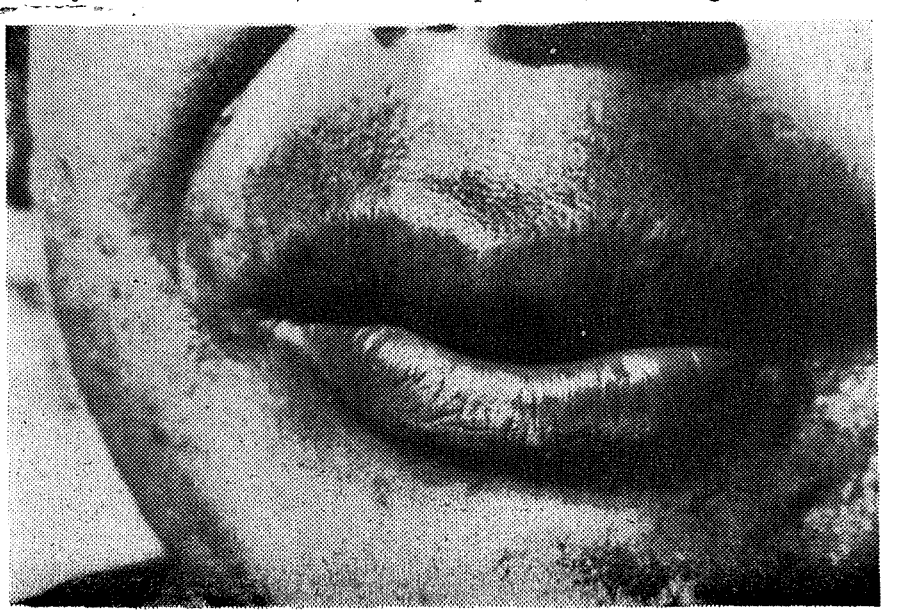

Figure 1

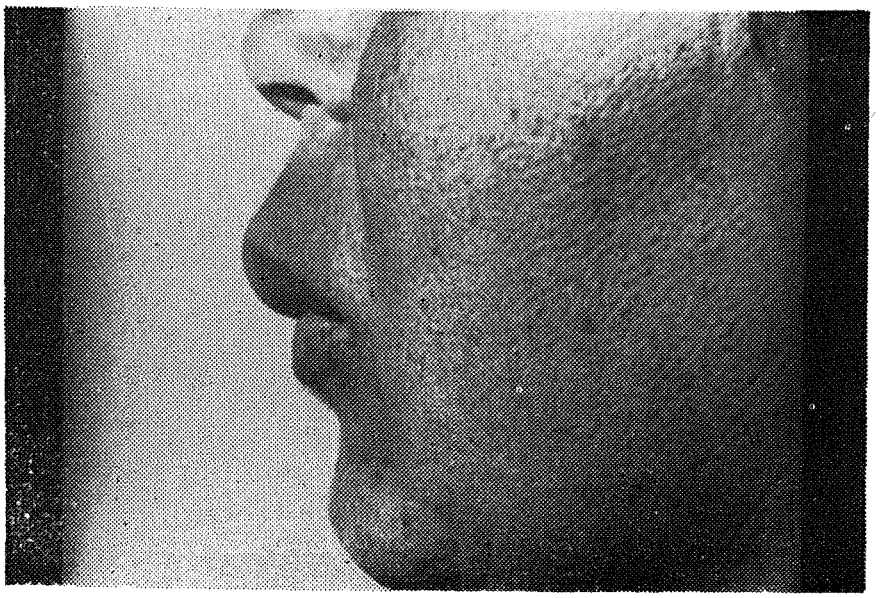

Figure 2

(Fig. 1 & 2). The dorsal surface of tongue was fissured (Fig. 3). The buccal mucosa was thrown into folds on the right side. No biopsy was taken, and all blood investigations were negative. Intralesional steroids (Depo Medrome 1 ml. subcutaneously every 3 weeks) are being tried with some success for the lip swelling.

Discussion

Melkersson (1928); described the recurrent facial palsy in association with labial oedema. Rosenthal (1931); stressed the genetic factors involved, and added the scrotal tongue to the syndrome. However, the full syndrome was described earlier by

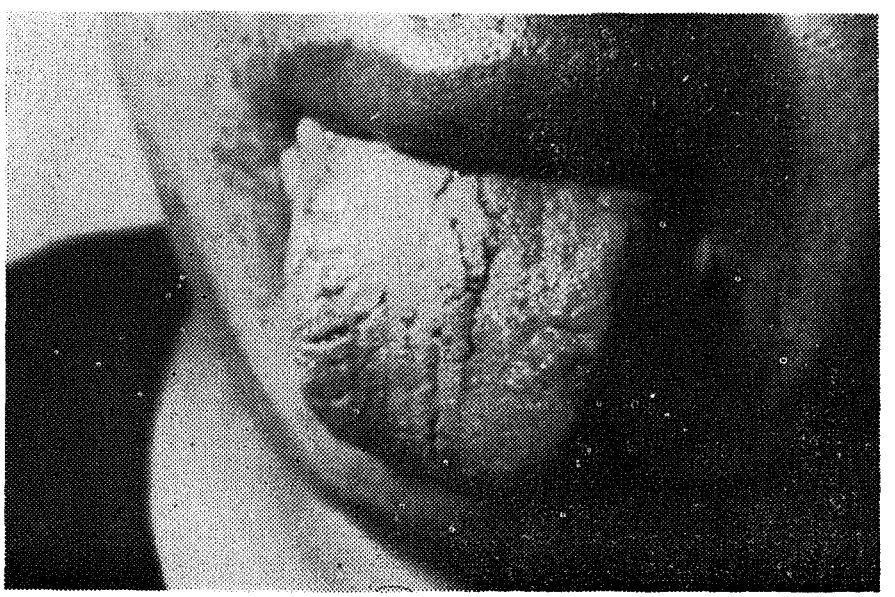

Figure 3

Rossolino (1901). Essential granulomatous cheilitis, described by Miescher (1945); is now accepted as a monosymptomatic form of the syndrome.

It may be genetically determined, as siblings have been effected, and the fissured tongue may be present in otherwise normal relatives. An infective aetiology was initially raised, but not with any convincing evidence (1971). The underlaying mechanism may be a disturbance of lymphatic drainage, like Milroy's disease, the hard oedema and granulomatous reaction resulting from the lymph seeping into the subcutaneous tissues.

The earliest manifestations usually develop in adolescents.

a) The first manifestation is cutaneous, and consists of the diffuse, non-tender, non-pitting swelling, resembling angioneurotic oedema, and affecting the upper lip, lower lip, one or both cheeks, or tongue in decreasing order of frequency. Forehead, eyelids or one side of scalp may be less commonly involved. Initially, the oedema completely subsides within a couple of days, but recurrent attacks are followed by persisting macrocheilia which increases slowly in degree. The swelling is soft at first, then becomes hard with a firm rubber consistency.

c) Facial palsy, unilateral or bilateral, has a 30% incidence, and commonly follows the oedema but may occasionally precede it. It is a lower motor neurone lesion, indistinguishable from a Bell's palsy. It may become recurrent intermittently, though permanent palsies have been recorded. Other cranial nerves may be rarely involved namely glossopharyngeal, olfactory, auditory, and hypoglossal.

a) Fissured tongue is also seen in 30% of cases. There is a 5% incidence in general population, and can be also seen in other conditions (e.g. Down's syndrome).

Histology of the oedema shows initially a perivascular lynphocytic infiltrate, but this becomes more dense with increasing attacks, forming non-caseating tuberculoid focal granulomas resembling sarcoidosis, but the ESR, Kveim test, and chest X-ray are normal.

The differential diagnoses include angioedema (in the initial stages), developmental defects involving the lips (haemangioma, lymphangioma, neurofibroma), and Ascher's syndrome consisting of swelling of lips caused by redundant salivary tissues present from childhood, together with blepharochalazia (atrophy of eyelids) (Rook et al, 1972).

Any patient with facial palsy, facial swelling or fissured tongue should be questioned and examined for the other features of the triad.

No treatment is available. Some success has been recorded with intralesional steriods. Surgical excision of the chronically enlarged tissue has also been recommended. The prognosis is bad as it is recurrent and progressive in nature. The patient is worried only about the cosmetic effect, as no systemic or local complications result from the condition.

References

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