MELKERSSON - ROSENTHAL SYNDROME
(Case Report)

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Summary

A case of Melkersson-Rosenthal Syndrome with involvement of gums is reported. The literature is reviewed briefly.

Melkersson-Rosenthal syndrome is an extremely rare condition. It is characterised by the triad of "recurring facial paresis or paralysis, non-pitting labial oedema and scrotal tongue". The first full description of this syndrome was given by Rossolino in 1901[1]. Melkersson[2] reported the association of recurring facial palsy with soft non-pitting oedema of the lips. Rosenthal[3] described the scrotal tongue in this syndrome. Miescher[4] reported 6 cases with swelling of the lips alone and progressive changes resulting in chronic permanent enlargement. According to Hornstein[5] this granulomatous chelitis, frequently associated with Miescher's name is a monosymptomatic form of Melkersson-Rosenthal syndrome.

The exact etiology of this condition is not known. Rosenthal[3] emphasised the role of genetic factors. Berger[6] described its occurrence in siblings and noted the presence of scrotal tongue in otherwise normal relations. A neurotropic etiology has been suggested on the basis of the fact that this disease has been occasionally seen in association with otosclerosis, craniopharyngioma and megacolon. Dahm and Schinko[7] suggested toxoplasmosis as a possible cause for this disease. Laymon[8] did not contribute to the earlier views that this syndrome may be a form of sarcoidosis or a reaction pattern to foreign silica in tooth pastes.

There is no satisfactory treatment for this disease. Cerimeli and Serri[9] reported good results with intralesional triamcinolone.

Case Report

A 22 year old healthy Hindu male admitted to the hospital on 6—10—1976 with the following complaints. Occasional attacks of fever with headache lasting 4 to 7 days for 9 years, swelling of the lips for 8 years and swelling of the gums for 3 years.

The first attack of fever and headache was accompanied by swelling of the upper lip which subsided along with the fever. In the early stages of these attacks of fever and headache, swelling of the lips used to subside completely. Later on some residual swelling used to persist until finally

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the lips became enlarged permanently. 3 years prior to his admission patient noticed swelling involving the cheeks and gums. He had undergone various treatments including gingivectomy without any relief. Family history revealed that patient's father had some thickening of the upper lip and scrotal tongue for 40 years. Two brothers, out of 4 siblings had scrotal tongue.

Local examination revealed diffuse, firm, nonpitting oedema of the lips, more on the right side. The adjacent areas of the cheeks were also involved to some extent (Fig.1). Tongue was moderately

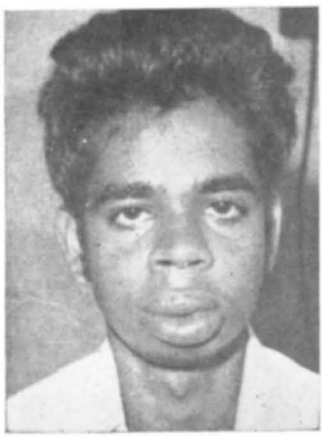

Fig. 1

enlarged and showed a deep longitudinal median groove with radiating superficial fissures (Fig. 2). The papillae were found to extend down to the full depth of the fissures. There was hypertrophy of the gums, more prominent in the anterior maxillary areas. There was no evidence of any bleeding or inflammatory changes in the gums or elsewhere. There was no evidence of facial nerve or CNS involvement. Barium meal and enema did not reveal any abnormality of the alimentary tract. Blood examinations, X-rays of the chest and skull were normal. Tuberculin test was negative. Kveim's test could not be done. Biopsy from the lower lip showed evidence of oedema of the tissues with dense pleomorphic perivascular infiltration.

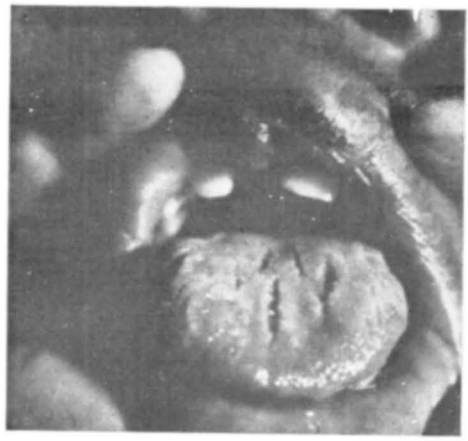

Fig. 2

Tuberculoid type of granulomatous changes were seen in one area. No evidence of salivary tissue was found. Patient was treated with cortisone, both systemically as well as by local injections, but without any remarkable improvement.

Comment

The presence of nonpitting oedema of the lips extending to the cheeks and circumoral areas which started after an attack of fever and headache with scrotal tongue suggested the diagnosis of Melkersson-Rosenthal syndrome, which was confirmed by histopathology. Our case had no facial palsy. Rook et al[10], observed facial palsy only in 30% cases. It is also possible to develop palsy at a later time. The presence of macrochelia and scrotal tongue in the father and scrotal tongue in 2 siblings are in agreement with observations made by Rook et al[10]. An unusual feature of our case is the involvement of gums. To our knowledge this feature has not been reported previously. The enlargement of gums may be due to an overgrowth of the connective tissue. Such connective tissue changes have been

reported to occur in the pharynx and respiratory tract[11].

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