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Melkersson's syndrome

When multiple sclerosis, sarcoidosis, leprosy, recurrent cranial polyneuritis, and other causes of recurring and sometimes bilateral lower motor neuron facial pareses have been considered there is a curious but rare group of cases which fit the picture of Melkersson's syndrome. In addition to facial palsy these patients have recurrent facial oedema, and about 25% have a congenitally horizontal fissuring of the tongue—lingua plicata. The first possible reference to this syndrome was that of Rossolimo in 1901 who described a woman with recurring facial palsy associated with attacks of migraine; but, this case may have been one of facial migraine.

Melkersson's paper,¹ translated from Swedish,² described a man of 35 who at the age of three had a left facial palsy which lasted for two weeks. At 14, facial palsy recurred on the same side, and again he recovered completely in three weeks. At 18 a third attack lasted about two weeks, but left residual weakness; he could not close the left eye tightly, and the left corner of his mouth did not move properly. At the age of 35 another attack was followed by severe and persisting paresis. After the age of 14, transient swelling occurred on the upper or lower lip. At first the swelling appeared with the facial palsy. When both facial palsy and swelling were present he felt unwell.

From the age of 35 his facial countenance was grotesque; the left half of his face was totally paralysed . . . The left cheek was atrophic. Lower lip was cyanotic and intensely swollen and doughy, especially in the right half.

Melkersson thought a vasomotor dysfunction likely. He did not describe the lingua plicata, which is a condition affecting 0.5% of the population as an incompletely penetrant dominant trait. Rosenthal, a German neurologist, three years later referred to an abstract of Melkersson's paper in German, and noted lingua plicata in an arthritic family who also had Melkersson's syndrome of recurrent facial palsy and facial oedema.³ A familial occurrence has been confirmed in several subsequent reports.

J M S PEARCE
304 Beverley Road,
Anlaby,
Hull HU10 7BG

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