Melkersson-Rosenthal Syndrome

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Abstract

Melkersson - Rosenthal syndrome was described by Melkersson and Rosenthal separately in the year 1928 and 1931 respectively. It is supposed to be a rare syndrome of bilateral alternating recurrent facial paralysis alongwith fissured tongue and oedema of the lips, face and eyelids. A case of Melkersson - Rosenthal syndrome is reported with all the classic findings which is a rarity. In this case there was alternating facial paralysis to begin with followed by bilateral paralysis third time, along with oedema of lips and face, fissured tongue, and dialation of sigmoid colon with absence of haustrations.

CASE REPORT

A 17 year old female was admitted on Sept. 4, 1995 with history of swelling left side of face. On examination clinically there was infranuclear facial paralysis alongwith oedema of left side of face, more so on left upper lip. She had fissured tongue. There was no skin granuloma. She was treated conservatively with corticosteroids and supportive therapy and got releived of her symptoms. Again after one year of gap in August, 1996 she was admitted with weakness of right side of face, swelling lips, eyelids and fissured tongue. She was treated conservatively with which she improved. She again reported on September 11, 1998 and was admitted with history of bilateral facial weakness, which was partial and more on right side with fissured tongue, oedema of lips, face and eyelids. Interestingly all the three episodes of facial paralysis occured during the month of August and September. There was facial paralysis grade III on first two occasions, and grade IV paralysis third time on right side and grade III on left side (Fig. I and II). On barium enema there was dialatation of sigmoid colon alongwith absence of haustrations. All the other investigations were within normal limits.

Discussion

Melkersson-Rosenthal syndrome is a rare association of recurrent facial Palsy, recurrent oedema of the lips, face, eyelids alongwith a fissured tongue in one third of the cases. However the classic triad is not frequently seen in its complete form. Monosymptomatic and oligosymtomatic forms are more common. The histological findings of sarcoid like granulomas

Fig 1. Melkersson Resenthal Syndrome (Fissored Tongue)

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in skin or mucosal biopsy specimens support the diagnosis (Nifos IG et al 1997). However Dhar S & Kanwar AJ (1995) failed to find any granuloma from lip biopsy in five out of six reported cases. Treatment is difficult but intralesional or systemic corticosteroids may be helpful, (Daoud MS et al 1995). Thalidomide 100mgs daily for 3 to 6 months has been tried (Sofa G et al 1995) but role of this therapy remains questionable. In recent years, novel therapeutic approaches involving either oral clofazimine or laser beam acupuncture according to traditional Chinese medicine has proved to be successful in some cases of

Melkersson-Rosenthal syndrome (Hornstein OP 1997, Dhar S et al 1995).

Most cases of Melkersson-Rosenthal syndrome begins before the age of 16 years, recurrences are frequent and facial oedema usually involves the distribution of motor part of facial nerve and usually affects the upper lip. There may be bilateral involvement of facial nerve. The etiology of this condition remain obscure. The course is chronic but benign. The syndrome is often associated with migraine, megacolon, facial swelling and stress which suggest the vasomotor instability fundamental to autonomic dysfunction. There may be involvement of other cranial nerves. Most of the series of facial Palsies describe this syndrome as a rarity and treatment is identical to that of Idiopathic Palsies. Some suggestions in the literature point to the complete facial nerve decompression as a prophylactic measure against further recurrences but this has so far not been substantiated by surgical results (Melkersson & Rosenthal 1928, 1931).

Summary

A case of melkersson-Rosenthal syndrome is reported with classic features of recurrent alternating facial paralysis, oedema of face lips, fissured tongue and megacolon.

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