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LETTERS TO THE EDITOR

CONGENITAL UNILATERAL DEAFNESS, FACIAL PALSY AND MICROTIA

Many dermatologic disorders are related to otolaryngology and these are primarily seen in the area of the external ear which consists of the auricle (or pinna) and the external auditory canal. Deafness, either sensorineural, conductive or mixed type may be associated with a number of genetically determined diseases. Malformations of the external ear in association with deafness may occur in branchio-oto-renal dysplasia (BOR syndrome), incudostapedial abnormality and chromosomal trisomy 13-15. Recently we have seen a 45-year-old male who had deformed and small auricle of the right ear associated with congenital loss of hearing and facial palsy of the same side. He gave history of congenital deafness in his father and in two of his siblings. There was no parental consanguinity. He was born after a full-term normal delivery and neonatal period was uneventful. There was no history suggestive of purulent meningitis or otitis media in the past. Examination revealed a small and deformed right auricle (microtia). Ear lobule was not fully formed (Fig. 1) and the triangular fossa was shallow. Helix was thick and broad. He was profoundly deaf with a 50 to 110 db neural loss, most marked above 1000 cycles per second. Left ear was normal. He had features of lower motor neuron type of facial palsy on the right side (Fig. 2). Detailed systemic examination, laboratory tests and other investigations did not show any other abnormality.

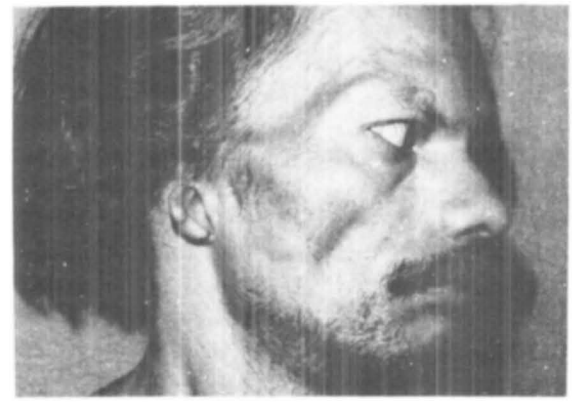

Fig. 1. Microtia and deformity of the right ear. Earlobe is not fully formed, triangular fossa is shallow and the helix is thick and broad.

Fig. 2. Paralysis of the right side of face.

Many syndromes have been reported in association with congenital deafness and malformations of the external ear. But the association of microtia, congenital sensorineural deafness and facial palsy, as observed in our patient, could not be fitted in with any of the known syndromes. Albinism, lentigenes, piebaldism, white forelock, nevus of Ota and atopic dermatitis are some of the dermatological disorders that may be associated with

hereditary deafness. To the best of our knowledge association of congenital deafness, facial palsy and microtia has not been reported and we suggest the name DFM syndrome for this condition D standing for deafness, F for facial palsy and M for microtia.

K Pavithran
Department of Dermato-Venereology
Medical College Hospital
Kottayam-686008, India.

BILATERAL NEVUS OF OTA WITH PTOSIS

Nevus of Ota or oculo-dermal melanocytosis is characterized by a diffuse bluish or grey-brown pigmentation of the skin in the areas of distribution of the ophthalmic, maxillary and rarely along the mandibular division of trigeminal nerve along with discoloration of various structures of the ipsilateral eye.[1] There may also be associated involvement of the oral and the nasal mucosa.[1] Mostly the lesions are unilateral and are confined to one side of the face. However, in a few cases the lesions of nevus of Ota are bilateral.[2,3]

During the past ten years, we have seen eight patients with nevus of Ota; all patients were females. In two, the lesions were bilateral. Due to social reasons in Libya, it has not been possible for us to have photographs of the patients.

Recently we had the opportunity of seeing yet another case of nevus of Ota in a 18-year-old female patient who was admitted into the skin wards for an unrelated eczematous eruption. There was typical slate-blue pigmentation of the right side of the face. The sclera and the hard palate were also involved. In addition, the patient had ptosis of the right upper eyelid. History revealed that the pigmentation and ptosis had been present together since about 15 years. Review of literature and records of our patients with nevus of Ota did not reveal any instance where ptosis was an associated feature. Since it is an unusual finding, we thought it worthwhile reporting. We would be interested to know if any of our dermatological colleagues have observed ptosis in their patients with nevus of Ota.

Malkit Singh, A J Kanwar, S C Bharija and M S Belhaj
Division of Dermatology, Department of Medicine, Al-Arab Medical University, Benghazi, Libya.

References

1. Mishima Y and Mevorah B: Naevus of Ota and Naevus of Ito in American negroes, J Invest Dermatol, 1961; 36: 133-154.
2. Ramesh Chandra AS: Bilateral naevus of Ota, Ind J Dermatol, 1977; 43: 206-207.
3. Nigam PK, Singh PK and Singh G: Bilateral naevus fuscocaeruleus ophthalmomaxillaris, Ind J Dermatol Venereol Leprol, 1985; 51: 287-288.