Moebius Syndrome (Moebius Sequence)

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Abstract

Moebius Syndrome is one of the rare disorder amongst the oromandibular limb hypogenesis. It is of a unknown atiology with sporadic occurrence in which there is congenital bilateral facial palsy, bilateral involvement of abducent nerve along with other cranial nerves like III, V, IX, X, XIIth and the patient having masklike expressionless face. We are reporting a case of Moebius Sequence who presented to us in the department of ENT and Head and Neck Surgery, Baroda.

The condition is one of the rare disorders amongst the disorders of oromandibular limb hypogenesis. First described by A. Graefe (1880) the syndrome goes by the name of Moebius (1888) who, while attempting to classify multiple congenital cranial nerve palsies, created a group in which VI and VII cranial nerves were combined.

Subsequently, the concept of Moebius Syndrome has been expanded into Moebius Sequence to include: masklike faces, VI and VII cranial nerve palsies usually bilateral, reductive limb anomalies (30 %), occasionally other cranial nerve involvement (II, V, IX, X, XII), Poland anomaly (15%), abnormality of Pectoralis major and ipsilateral syndactyly, and mild mental retardation (15%).

CASE REPORT

B.P., a 20 year old male studying in Xth standard presented in the ENT-OPD with complaint of difficulty in speaking since birth. He had no complaint of deafness. The patient was mainly interested in a certificate for his speech handicap. There was no history of birth trauma or any significant past or family history. On general examination, patient had mask like expressionless face. He was having bilateral facial palsy which

was complete on left side while slight movement was present near right angle of mouth (Fig.1). There was midface prominence with bilateral malar hypoplasia. Distal phalanx of left index finger was absent (Fig. 3).

Examination of oral cavity revealed bilateral atrophy of tongue with fasciculations with inability to protrude and move the tongue, suggesting bilateral hypoglossal nerve palsy. He had high arched palate and crowding of teeth (Fig. 2). Palatal movements were normal. Laryngeal examination revealed bilaterally mobile vocal cords. Gag reflex was normal. Examination of right ear showed atelectatic tympanic membrane and left

Fig 1 : Clinical Photograph of the patient showing mask like expression less face. (Bilateral Facial Nerve Palsy).

Fig 2 : Clinical Photograph of the patient showing muscular atrophy of the tongue. (Bilateral XII Nerve Palsy).

Fig 3 : Clinical Photograph of the patient showing hypodactyly (absence of distal phalanx of the left finger).

tympanic membrane showed shallow retraction pocket in posterosuperior quadrant with mild conductive deafness on both sides. Nasal cavities were normal.

Ocular examination showed bilateral IVth & VIth nerve palsy convergent squint, with normal IIIrd nerve. Bilateral epicanthal folds were also seen (Fig. 1).

Assessment of speech showed normal audibility. However, he was not able to perform articulatory acts beyond few words, which also were unintelligible. Functional efficiency of speech was zero.

DISCUSSION

A clinical diagnosis of Moebius Syndrome was put keeping in mind the congenital bilateral involvement of facial and abducent nerve along with IIIrd and XIIth cranial nerve involvement with hypodactyly and patient having characteristic mask like facies. Differentiating from other Oromandibular limb hypogenesis and other congenital syndromes like Treacher Collins, Craniofacial microsomia and Melkersson Rosenthal,

these do not have congenital bilateral facial palsy simultaneously and Dystrophia myotonica which usually presents with congenital facial diplegia does not involve the abducent nerve. The aetiology of Moebius is unknown but post-mortem studies implicate four modes of developmental pathology in the genesis of the problem. These are :

- i) Hypoplasia to absence of central brain nuclei
- ii) Destructive degeneration of central brain nuclei (most common type)
- iii) Peripheral nerve involvement
- iv) a myopathy

Recent studies suggest there may be an abnormality in chromosome 13.

TREATMENT

Reconstructive surgeries like orthognathic surgeries and static sling procedures may be of some help for such a disorder. Our patient was given the certificate for his speech handicap to enable him to apply for his job. He is also been given medical treatment for bilateral ear problem and had been advised surgery for left ear.

References

- 1) Kenneth Jones M.D. : *Smith's Recognizable Patterns of Malformation* (4th edition) 192-193, 588-589
- 2) David R James, Peter Ramsay - Baggs (1987), *Craniofacial anomalies, Scott Brown's Paediatric Otolaryngology* (Vol -6) 48-50.