



UNILATERAL MOEBIUS SYNDROME: A RARE CASE REPORT FROM HILLY STATE OF NORTH INDIA

Ophthalmology

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ABSTRACT

Moebius syndrome is a very rare congenital neuromuscular disorder which is classically defined as combined nerve palsies of the VI (abducens) and the VII (facial) cranial nerves. Very few cases have been reported in literature with average incidence of 0.002% with geographic variation. In India, less than ten cases have been reported and our case, Moebius syndrome with dermatological manifestations in hilly state of Himachal Pradesh is probably the first one to be reported.

KEYWORDS

Moebius syndrome, abducens nerve, facial nerve, hilly state, oculofacial diplegia, dermatological manifestations

INTRODUCTION

The Moebius syndrome has been earliest reported in history in 1880 by Von Graefe as congenital facial diplegia.¹ The syndrome was reviewed and defined further by Paul Julius Moebius, a German neurologist, in 1888 and 1892.² Because of these contributions, Moebius is now the eponym used to describe the syndrome. This name is also synonymous with congenital oculo-facial paralysis. It may involve one or both the eyes. These nerves are involved in the control of facial and the ocular muscles. It may, or may not be associated with involvement of other cranial nerves like III, V, VI, IX, X and XII cranial nerves. The most obvious symptom of this syndrome is loss of facial expression. Ocular presentation varies from bilateral ptosis, restricted temporal movement of the globe, strabismus, inability to close the lids and chronic conjunctivitis. Mild mental retardation has been associated with syndrome and it is probably over diagnosed due to mask like face and drooling of saliva. In addition, in extremely rare instances, skin manifestation like café-au-lait pigmentation, webbing of the axilla, and an absence of subcutaneous tissue may also be seen.

CASE REPORT

A 3-year-old girl was brought in the department of Ophthalmology by her parents with complaints of watering from the right eye since birth. The parents had consulted various local practitioners in the past and were prescribed various drops and ocular massage which they religiously did but had no relief of symptoms. Further history revealed that she had born to parents without any consanguineous marriage and had full term normal vaginal delivery. Mother denied any history of illness or intake of any drugs during gestation. On examination, head posture was normal. The position of both the eyelids were normal. On performing Hirschberg test; unilateral esotropia of about 45° of right eye was noticed. She was directed to close her eyes when inability to close right eyelid was observed as a result of which epiphora was observed. At the same time deviation of angle of mouth to left side was noted. Also, decreased blinking was evident in right eye. Further detailed examination revealed inability to wrinkle the forehead and raise the eyebrow on right side. When an attempt was made to forcibly close the lids, Bells phenomenon was observed. When test to elicit VII nerve palsy was done, the child was not able to retain air in mouth. No skeletal abnormality was detected in either of upper or lower limbs. Her rest of neurological examination was normal.

On the basis of history and clinical examination, unilateral VI and VII nerve palsy; a diagnosis of Moebius syndrome was made.



FIG 1: INABILITY TO CLOSE RIGHT EYE



FIG 2: UPWARDS TURNING OF RIGHT EYEBALL ON ATTEMPTED EYE CLOSURE



FIG 3: CAFÉ AU LAIT SPOT

DISCUSSION

As an extremely rare disorder, the various theories postulated are equally debatable. However, the common factor of all hypothesis remains that there is some hypoxic or ischemic insult to the embryonic brain stem during first trimester of gestation when rapid development of brain occurs. Most common drug associated with this is misoprostol.³ It is a prostaglandin analogue easily available over the counter drug. It is clinically used for cervical ripening and induction of labor and unfortunately used for termination of pregnancy by expectant mothers. Apart from this, maternal infections, alcohol abuse and drug abuse also lead to manifestation of Moebius syndrome due to same etiology. One report describes bilateral calcifications of the basal ganglia on the brain CT scans of 2 siblings with classic Moebius syndrome.⁴ The absence or hypoplasia of CN VI and CN VII may be the most common radiologic features in sporadic Moebius syndrome, and hypoplasia of CN IX may be an associated feature.⁵ The syndrome is listed as Online Mendelian Inheritance in Man (OMIM) Number 15700, with a gene map locus of 13q12.2-q13.⁶ Sporadic mutations in PLXND1 and REV3L genes have also been identified in a number of patients.⁷

The children with Moebius syndrome are best managed by a multidisciplinary team, often in a craniofacial center. Involved specialists include: pediatricians; neurologists; plastic surgeons; otolaryngologists; orthopedists; dental specialists; speech pathologists; audiologists; ophthalmologists and other healthcare professionals.

The management includes training of the parents regarding patching of eye at night to prevent exposure keratitis and use of artificial tears during morning hours. Some of the Corrective procedures include temporalis tendon transfer, cross-facial nerve graft, the smile operation, surgical correction of strabismus and lagophthalmos.⁸ These operations have shown remarkable results in correcting lagophthalmos, speech, facial mobility and self-esteem.

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