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A Child without A Smile : Moebius Syndrome – A Case Report

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ABSTRACT

Background: *Moebius syndrome is a rare congenital neurological disorder. It is characterised by Facial Palsy accompanied by unilateral or bilateral Abducens nerve palsy. It may be accompanied by weakness of III, IV or XII cranial nerves andoro-facial/ limb deformities.*

Case Characteristics: *A 9 months old girl child presented with absence of a smile and a drropy right eyelid. On examination, she had bilateral facial weakness, right sided abducens nerve palsy and right sided ptosis.*

Outcome: *The child had no other congenital deformity. Her developmental milestones were normal as per age.*

Message: *Moebius syndrome, because of the facial paralysis, usually masquerades as developmental delay. But the patients usually have normal development and normal intelligence*

Background

Moebius Syndrome is a very rare (2 to 20 per million) congenital neurological disorder. The basic features are congenital facial paralysis (usually bi-lateral) and Abducens nerve weakness (unilateral or bilateral). This may be accompanied by other cranial nerve weakness leading to ptosis, palatal or lingual palsy and hearing loss. Other variable features include orofacialdysmorphism and limb malformations. The syndrome was first described by Graefe (1880), and by Moebius (1888)(1,2).

The cause is unknown. Most of the cases are sporadic, but few families have been identified with Moebius syndrome inherited with either autosomal dominant or autosomal recessive

pattern. The pathology may be either because of an abnormality in the nerve nucleus or a peripheral nerve abnormality (2).

Moebius syndrome can be diagnosed soon after birth usually because of the associated cranial nerve palsies (IX, X, XII) which manifest as poor sucking and/or noisy breathing. Later on, it is noted that the child has an immobile face, does not move her face while crying, and does not smile. The facial paralysis is usually bilateral and incomplete, involving the upper face more. These children may have dysarthria because of the involvement of hypoglossal nerve. The combination of a mask like face, absence of a smile and dysarthria usually leads to them being misdiagnosed as cases of developmental delay and they are dubbed as mentally retarded(3,4).

In the West, self help groups have been formed with excellent online resources to help these patients with information and equipment (5).

Case Report

9 month old girl, weighing 8.5 kg, presented to us with a droopy right eyelid and inability to smile (Pic 1). The girl was born through an uneventful normal delivery, with normal perinatal and post natal period. She was born of a non-sanguineous marriage and there was no family history of similar deformities. There was no history of sucking difficulty or noisy breathing.

The girl was found to have bilateral facial weakness, right sided lateral gaze palsy, and right sided ptosis. She did not move her face while crying or grimacing and could not smile. She had a normal suck and swallow; she made normal sounds and could hear sounds in both ears normally as per her age. Her gross and fine motor developments were as per her age. She had no other orofacial or limb deformity. Her systemic examination was normal.

Follow up

We sent her for a fundoscopy examination which was reported to be normal. Herthyroid profile was sent and no abnormalities were detected. We will follow up the child for her social and adaptive development and her speech pattern.

Discussion

Moebius syndrome is a very rare condition. But isolated congenital facial paralysis is more common. These children, because of their mask like appearance and associated cranial nerve abnormalities, are very frequently mis-diagnosed as mentally retarded. But usually they have normal intelligence, and guided properly, can have excellent outcome. Various surgical and non surgical measures can improve their appearance (5).

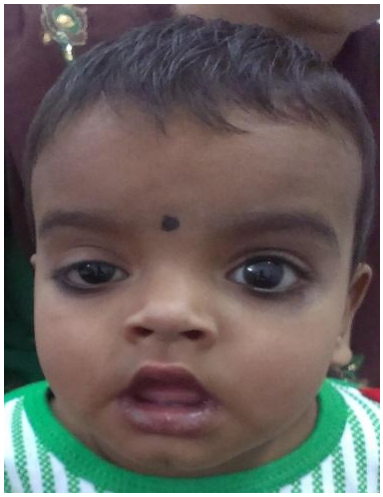

Figure I: Moebius Syndrome

Acknowledgement

Dr S Patowary, senior ophthalmologist, kindly did the fundus examination of this patient.

Bibliography

1. Kliegman R M, Stanton F B, Schor N F, St. Geme J W, Behrmam R E: Editors. Nelson Textbook of Pediatrics. Edition 19. Elsevier. 2012. P 2006, 2160.
2. OMIM. % 157900. MOEBIUS SYNDROME, MBS: accessed on May 4, 2014.
3. Kumar D. Moebius Syndrome. J Med Genet 1990; 27: 122-126 (full text in pdf in Pubmed)
4. Khalid AA, Yazeed A A, Abdulrhman I A.Ann Saudi Med. 2009 Nov-Dec; 29(6): 482–484. (Full text in HTML in Pubmed)
5. Moebius Syndrome Foundation. www.moebiussyndrome.com. Accessed on May 4, 2014.