E-ISSN:2349-3267
P-ISSN:2349-5499
Case Report
Facial Nerve

Pediatric Review - International Journal of Pediatric Research
2020 Volume 7 Number 6 July-August

Publisher
MEDRESEARCH
www.medresearch.in

Congenital unilateral absence of facial nerve with hypoplastic abducens nerve: A case report of rare Moebius syndrome

Dharmani A.[1], Kashyap S.[2], Thakur S.[3], Guleria S.[4*]

DOI: https://doi.org/10.17511/ijpr.2020.i06.13

[1] Ankur Dharmani, MD, Department of Pediatrics, Regional Hospital, Bilaspur, Himachal Pradesh, India.

[2] Sonia Kashyap, MD Internal Medicine, Deen Dayal Upadhayay Hospital, Shimla, Himachal Pradesh, India.

[3] Swati Thakur, MD, Department of Radiodiagnosis, Phoenix Diagnostic Center, Bilaspur, Himachal Pradesh, India.

[4*] Sandesh Guleria, MD, DM, Assistant Professor, Department of Pediatrics, Indira Gandhi Medical College, Shimla, Himachal Pradesh, India.

Moebius syndrome is a rare congenital neurological disorder characterized by bilateral facial and abducens nerve paralysis. The syndrome may also include involvement of the other cranial nerves and various other congenital deformities and malformations. A one-month-old male infant presented with facial asymmetry, inability to close eye, and drooling of saliva from one side of the mouth, diagnosed to have Moebius syndrome. Early diagnosis of the cases by experienced clinicians and a supportive multidisciplinary approach can help children with Moebius syndrome and prevent associated complications.

Keywords: Moebius syndrome, Abducens nerve paralysis, Congenital deformities

Corresponding Author

Sandesh Guleria, MD, DM, Assistant Professor, Department of Pediatrics, Indira Gandhi Medical College, Shimla, Himachal Pradesh, India.
Email: guleriasandesh662@gmail.com

How to Cite this Article

Dharmani A, Kashyap S, Thakur S, Guleria S. Congenital unilateral absence of facial nerve with hypoplastic abducens nerve: A case report of rare Moebius syndrome. Pediatric Rev Int J Pediatr Res. 2020;7(6):310-312.
Available From
https://pediatrics.medresearch.in/index.php/ijpr/article/view/618

To Browse

Introduction

Congenital facial nerve palsy is clinically defined as unilateral or bilateral paralysis of the facial nerve in the neonatal period. Etiology includes intrapartum injury or various developmental disorders [1]. Facial nerve aplasia is an extremely uncommon anomaly, usually associated with Moebius syndrome. Moebius syndrome is a rare congenital neurological disorder characterized by bilateral facial and abducens nerve paralysis. This Condition was originally described by Von Graefe in 1880 and by Moebius in 1888. Since then, nearly 300 cases have been described and reported in the literature by a number of authors [2–8].

An International Group of Experts at the Moebius Syndrome Foundation in 2007 formulated diagnostic criteria for diagnostic consistency, which include congenital facial diplegia or uniplegia, lower motor neuron (LMN) type in nature, and paralysis of lateral movements of eyes and strabismus due to

Manuscript Received	Review Round 1	Review Round 2	Review Round 3	Accepted
2020-08-10	2020-08-22	2020-08-24		2020-08-25

Conflict of Interest	Funding	Ethical Approval	Plagiarism X-checker	Note
No	Nil	Yes	5%	

Sixth cranial nerve palsy [2,9]. The syndrome may also involve other cranial nerves and various other congenital deformities and malformations. Weakness or paralysis of the facial muscles is one of the most common features of Moebius syndrome. The muscle weakness also causes problems with feeding that become apparent in early infancy. Dental abnormalities, including missing and misaligned teeth, are also common [10].

Case Report

A one-month-old male infant was brought to our hospital with complaints of facial asymmetry, inability to close the left eye, and drooling of saliva from the left side of the mouth. He was born at full term by normal vaginal delivery in a hospital elsewhere. He was discharged on day 3 of life, as was accepting breastfeeds well and has passed urine and meconium. On examination, he was active, alert with stable vitals. He had absent left nasolabial fold and unable to close the left eye (Figure 1). The outward gaze was affected in both eyes. The rest of the neurological and systemic examinations were normal.

Fig-1: Absent left the nasolabial fold and unable to close the left eye.

Investigations revealed normal blood counts, blood sugar, serum calcium, and electrolytes. Sleep EEG shows mature background for the age with no inter-ictal epileptiform discharges. Contrast-enhanced Magnetic Resonance Imaging (CEMRI) of the brain showed normal gray-white matter differentiation and normal signal intensity. No focal lesion, hemorrhage, or meningeal enhancement was seen. Myelination was found to be normal for age. Normal facial, as well as vestibulocochlear nerve, was seen on the left side, however, on the right side vestibulocochlear nerve was seen but the facial nerve was absent. Both abducens nerves were absent (Figure 2).

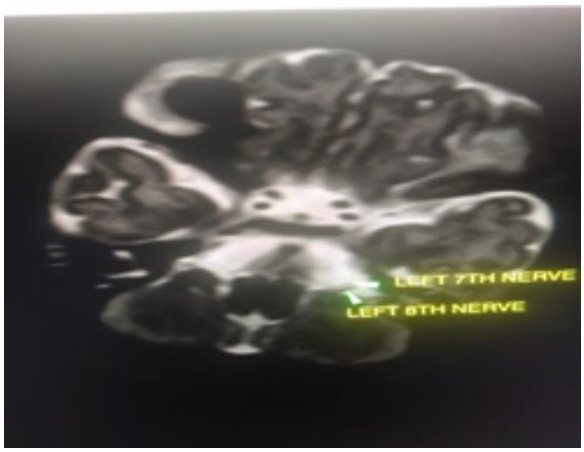

Fig-2

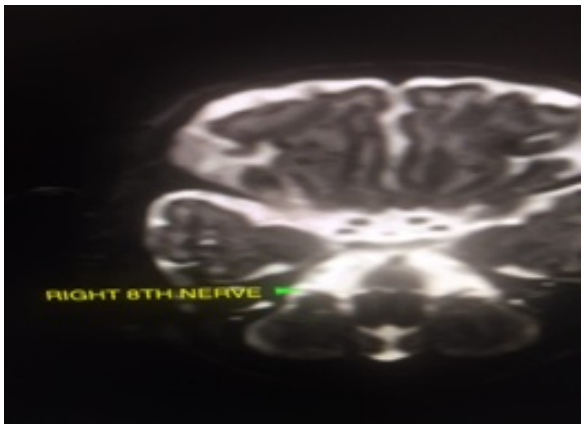

Fig-3

Fig-2 and 3: Contrast-enhanced Magnetic Resonance Imaging (CEMRI) of the brain.

So with LMN facial nerve palsy, bilateral abducens nerve palsy left preauricular tag, and findings of MRI brain, diagnosis of Moebius syndrome was made. Parents were counseled regarding the disease, planned regular follow up, and multidisciplinary management.

Discussion

Moebius syndrome is an extremely rare congenital dysinnervation disorder. There is diminished facial expression resulting in poor parental bonding. Autism spectrum disorders have widely been reported in the patients, amounting to around 30%–40% according to a study by Ana et al [5]. Physical findings entirely depend on the case definition of Moebius syndrome. By using the most commonly accepted definition [4,11], the typical phenotypic appearance is an immobile facial feature with various gaze palsies. In our case, the left LMN facial

Nerve palsy with bilateral abducens nerve palsy and left preauricular tag was present. These findings are consistent with Moebius syndrome and the MRI brain further corroborated these.

No definite treatment for this syndrome is available. The surgical goal in Moebius syndrome is far more modest and differs in patients with unilateral developmental facial paralysis. It is impossible to restore a true smile in these masks-like expressionless faces. Ocular surgeries, orthognathic surgeries, and surgical corrections for other associated abnormalities may be tried. Medical care is only supportive and symptomatic. Complications depend on the severity of the patient's deficits. These may include aspiration pneumonia, corneal ulceration/ abrasion, dysphagia, and poor nutrition. Consultations may be required from pediatricians, pedodontists, oral physicians, orthopedics surgeons, ophthalmologists, psychologists and speech therapists [1,9,12,13].

Conclusion

Early recognition of this condition and a supportive multidisciplinary approach can help children with Moebius syndrome and prevent associated complications.

Reference

01. Kumar I, Verma A, Ojha R, Aggarwal P. Congenital facial nerve aplasia- MR depiction of a rare anomaly. Indian J Radiol Imag. 2016;26(4)517-520.
[Crossref]

02. Pitner SE, Edwards JE, Mccormick WF. Observations on the pathology of the Moebius syndrome. J Neurol Neurosurg Psychiatry. 1965;28(4)362-374.
[Crossref]

03. Raju MS, Suma GN, Prakash R, Goel S. Moebius syndrome- A rare case report. J Indian Acad Oral Med Radiol. 2011;23(3)267.
[Crossref]

04. Palmer C. Möbius syndrome. Medscape [internet]. 2012.
[Crossref]

05. Ana SC, Tatiana VB, Vera CLSR, Saul Martins PM, Isabela PA. Moebius syndrome: a case with oral involvement. Cleft Palate-Craniofacial J. 2008;45(3)319-324.
[Crossref]

06. Mobius PJ. Uber infantilen kernschwund, Munch. Med Wochenschr. 1892;39;41-43.
[Crossref]

07. Jones KL, Jones MC, Campo MD. Smith's recognizable patterns of human malformation. 7th ed- Philadelphia, Saunders. 2013.
[Crossref]

08. Huebner O. UberangeborenenKernmangel (infantile Kernschwund, Moebius). Charité-Ann. 1900;25;211-243.
[Crossref]

09. Miller G. Neurological disorders, The mystery of the missing smile. Sci. 2007;316(5826)826-827.
[Crossref]

10. Briegel W, Schimek M, Knapp D, Holderbach R, Wenzel P, Knapp EM. Cognitive evaluation in children and adolescents with Möbius sequence. Child Care Health Dev. 2009;35(5)650-655.
[Crossref]

11. Lin KJ, Wang WN. Moebius syndrome- Report of case. ASDC J Dent Child. 1997;64(1)64-67.
[Crossref]

12. Ouanounou S, saigal G, Birchansky S. Mobius syndrome. Am J Neuroradiol. 2005;26(2)430-432.
[Crossref]

13. Terzis JK. Mobius and Mobius-like patients. Clinc Plastic Surg. 2002;29(4)497-514.
[Crossref]