Möbius syndrome

Sameer Vyas, Anuj Prabhakar, K. Uday Bhanu, Paramjeet Singh, Niranjana Khandelwal

Department of Radiodiagnosis and Imaging, Postgraduate Institute of Medical Education and Research, Chandigarh, India

A 6-year-old female child presented to our hospital with intellectual disability, global developmental delay, seizures, bilateral congenital squint, and features of facial weakness. On examination, the patient had lower motor facial nerve palsy and was unable to move both sides of her face and forehead. Her skin was devoid of wrinkles. She was unable to close her mouth, raise her eyebrows, or speak clearly. The patient also suffered from bilateral abducens nerve palsy with congenital internal strabismus and associated partial ophthalmoplegia. Lateral gaze paralysis was also present, indicating medial longitudinal fascicular involvement. The child also had syndactyly and small uneven-sized limbs. Magnetic resonance imaging (MRI) of the brain [Figures 1 and 2] revealed depression of the floor of the fourth ventricle, hypoplasia of the medulla, pons, and bilateral middle cerebellar peduncles with absent facial colliculi. The cisternal and canicular segments of bilateral facial nerves were absent. These features are suggestive of Möbius syndrome.

Discussion

Möbius syndrome is a relatively rare congenital cranial dysinnervation disorder.^[1] It was described by Von Graefe, Julius Möbius and was further elaborated upon by Henderson. The etiopathogenesis is multifactorial. Transient hypoxic or ischaemic insult to the fetus is considered as the most likely cause.^[2] Clinically, such

Figure 1: Axial (a) and sagittal (b) constructive interference in steady state magnetic resonance images showing the absence of facial colliculus bilaterally (arrow in a), resulting in flattening of dorsal aspect pons with hypoplasia of the pons and medulla (arrow in b)

Figure 2: Sagittal (right a and left b) constructive interference in steady state magnetic resonance images show absent facial nerves in the anterosuperior quadrants of both internal auditory canals (arrows)

patients present with facial diplegia involving the upper and lower facial muscles, impairment of eye abduction, craniofacial malformation, hypoglossia, maldevelopment of the corticospinal and corticobulbar tracts, and limb anomalies.^[2] Hearing loss and mental retardation have also been reported.^[1] Imaging findings include hypoplasia of the pons/medulla, hypoglossal prominence, facial colliculus, and the cerebellum. Calcification of the abducens nuclei in the pons and the absence of the middle cerebellar peduncle have

Address for correspondence:

Dr. Sameer Vyas, Department of Radiodiagnosis and Imaging,
Postgraduate Institute of Medical Education and Research,
Chandigarh, India.
E-mail: sameer574@yahoo.co.in

Access this article online

Quick Response Code:

Website:

www.ruralneuropractice.com

DOI:

10.4103/0976-3147.186974

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Vyas S, Prabhakar A, Bhanu KU, Singh P, Khandelwal N. Möbius syndrome. J Neurosci Rural Pract 2016;7:596-7.

also been reported.^[2] The absence or hypoplasia of the 6th, 7th, and 9th cranial nerves may also be associated.^[3] No definitive treatment is available. Imaging data in Möbius syndrome are scarce, and there are only a few reports. MRI is very useful in diagnosis by direct imaging of cranial nerves abnormalities. As illustrated in the index case, the absence of cranial nerves and brainstem changes as described are consistent with the diagnosis.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Rucker JC, Webb BD, Frempong T, Gaspar H, Naidich TP, Jabs EW. Characterization of ocular motor deficits in congenital facial weakness: Moebius and related syndromes. *Brain* 2014;137(Pt 4):1068-79.
2. Ouanounou S, Saigal G, Birchansky S. Möbius syndrome. *AJNR Am J Neuroradiol* 2005;26:430-2.
3. Wu SQ, Man FY, Jiao YH, Xian JF, Wang YD, Wang ZC. Magnetic resonance imaging findings in sporadic Möbius syndrome. *Chin Med J (Engl)* 2013;126:2304-7.