Moebius Syndrome

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Moebius syndrome presents as congenital, nonprogressive unilateral or bilateral facial and horizontal gaze palsy.^[1,2] A 16-year-old male presented to our center with restricted eye movements with turning of head sideways to look at, inability to blow out the cheeks, and accumulation of food between teeth and cheeks since childhood. There was no history of drooling and he didn't report any recurrent childhood ear infections. On examination, patient had gaze-evoked nystagmus, horizontal gaze restriction with bilateral medial and lateral rectus palsy with preserved vertical movements. There was micrognathia, bilateral facial palsy (predominant lower face involvement), tongue atrophy, and tongue fasciculations. Intelligence quotient of the patient was normal. MRI of the brain revealed brainstem hypoplasia and straightening of the floor of the fourth ventricle [Figure 1].

The pathogenesis and etiology of the Moebius sequence appeared to be multifactorial. It is postulated to be due to



Figure 1: (a) Tongue atrophy. (b) Restriction of right lateral gaze (c) Restriction of left lateral gaze (d) Brainstem atrophy (e) Flattening of the floor of 4th Ventricle at the level of the inferior colliculus. (f) Inability to puff out cheeks suggestive of bilateral facial palsy

vascular disruption in the brain during prenatal development leading to hypoplasia or agenesis of the cranial nerve nuclei during fetal development. Sporadic mutations in *PLXND1* and *REV3L* genes have also been identified in a number of patients and confirmed to cause a constellation of findings consistent with Moebius syndrome when introduced in animal models. In rare cases, familial patterns have been reported. Most likely, Moebius syndrome is multifactorial, which means that both genetic and environmental factors play some causative role.

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Conflicts of interest

There are no conflicts of interest.

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