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Case report

Magnetic resonance imaging in congenital facial palsy

Masayuki Sasaki [a,*], Yoshihiko Imamura [a], Noriko Sato [b]

[a] *Department of Child Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry (NCNP), Kodaira 187-8551, Japan*
[b] *Department of Radiology, National Center Hospital for Mental, Nervous and Muscular Disorders, NCNP, Japan*

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Abstract

We report magnetic resonance (MR) findings in a patient with congenital unilateral facial palsy and a patient with atypical Moebius syndrome. MR imaging showed a complete deficiency of right facial nerve in the patient with congenital unilateral facial palsy and bilateral, thin proximal facial nerves in the Moebius syndrome patient. Three-dimensional constructive interference in steady state (3D-CISS) MR imaging, especially reconstructions perpendicular to the bilateral internal auditory channel, was very useful when diagnosing patients with facial palsy due to the associated facial nerve abnormalities.

Keywords: Facial nerve; Moebius syndrome; 3D-CISS; Diagnosis

1. Introduction

Congenital facial palsy is not uncommon in children. The most frequent cause of unilateral congenital facial palsy is birth injury [1]. Pressure on the temporal bone during birth can induce transient facial nerve dysfunction. Usually this injury recovers completely within several weeks. However, congenital unilateral facial nerve palsy without birth injury is rare, and usually of unknown etiology. Conventional neuroimaging does not contribute to understanding the pathogenic mechanism of this condition [2], except in the case of a large pontine lesion [3]. In contrast, congenital bilateral facial nerve palsy is usually accompanied by other congenital disorders, such as Moebius syndrome or Goldenher syndrome. Recently, congenital facial nerve absence was reported in some patients with Moebius syndrome by using three-dimensional constructive interference in steady state (3D-CISS) magnetic resonance imaging (MRI) sequence [4,5]. We applied this method to a patient with isolated unilateral congenital facial nerve palsy and an atypical patient with Moebius syndrome, and obtained important diagnostic evidence in both patients.

2. Case reports

2.1. Patient 1

This 12-month-old boy was born to healthy and unrelated parents after an uneventful delivery at full term, not involving forceps. Right facial palsy was noticed immediately right after birth by the laterality in his crying face. He was completely healthy except for the right facial paralysis, which did not recover.

He was referred to our hospital for the right facial palsy. At that time, he spoke a few words and walked with support. Motor and mental development was within normal limits, and there were no abnormal neurological findings except for his right facial palsy. The right part of his face did not move at all even when crying or smiling, and there were no wrinkles on his right

* Corresponding author. Tel.: +81 42 341 2711; fax: +81 42 344 6745.
E-mail address: masasaki@ncnp.go.jp (M. Sasaki).

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forehead when looking upward. He could close his right eye when he slept. No other cranial nerve symptoms were noted. He was diagnosed with isolated right facial nerve palsy.

Neurophysiological studies showed normal results in auditory brainstem reaction (ABR), somatosensory evoked potentials (SEP), and visual evoked potentials (VEP). The blink reflex was not induced at all on the right side, but was normal on the left side.

2.2. Patient 2 [6]

This 9-month-old girl was born to healthy and unrelated parents after an uneventful delivery at full term. She did not cry at birth and she was diagnosed with mild neonatal asphyxia. She had difficulty with swallowing and needed tube feeding. Her clinical course was followed in a hospital neonatal intensive care unit. She showed delayed motor development, repeated aspiration pneumonia, and showed chronic hypoxia necessitating oxygen inhalation. Movements in her extremities were extremely weak and infrequent. She was suspected of having congenital myopathy based on the severe motor delay and lack of facial expression. She was referred to our hospital for motor retardation and swallowing difficulty. She could not move her face, tongue, or swallowing muscles. She had facial and glossopharyngeal nerve palsy, with no head control. Despite the absence of abducens nerve palsy, she was tentatively diagnosed as having atypical Moebius syndrome. She underwent tracheostomy because of the repeated aspiration pneumonia and chronic hypoxia. This returned her oxygen saturation levels in the peripheral blood to normal range and activated her body movements, as well as anti-gravity movements in her extremities. The aspiration pneumonia has not recurred.

3. MR findings

MRI was performed on both patients with a 1.0 T MR unit (Siemens, Harmony, Germany). Sequences included conventional T1- and T2-weighted images and 3D-CISS: TR/TE/NEX = 11.6/5.8/1, 70° flip angle, 180 × 180-mm FOV, 39.2-mm slab thickness, 256 × 224 matrix, 56 three-dimensional partitions, on

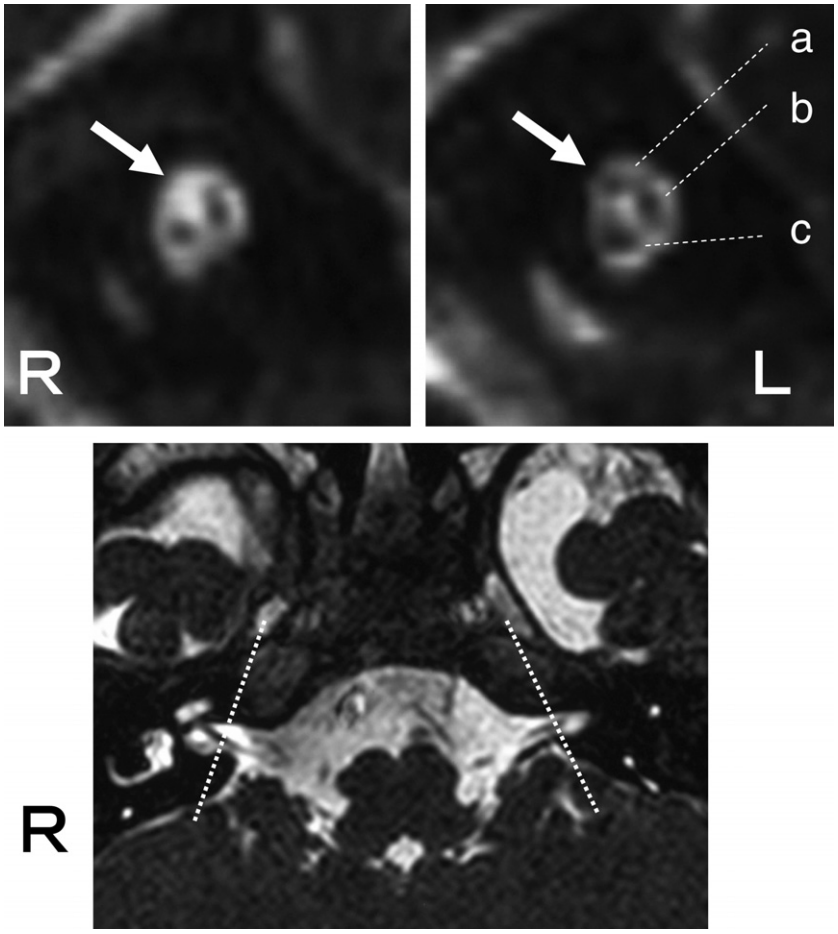

Fig. 1. 3D-constructive interference in steady state (CISS) sequences with reconstructions perpendicular to the bilateral internal auditory channel in patient 1 show complete absence of the right facial nerve. Left side is normal, indicating facial nerve (a), vestibular nerve (b), and acoustic nerve (c).