Monozygotic Twin Cases of Unilateral Sensorineural Hearing loss: Elderly One Accompanied with Internal Auditory Canal Stenosis and Facial Paralysis

We introduce the case of monozygotic twins with sensorineural hearing loss (SNHL). And one of them had congenital facial paralysis (CFP) accompanied by SNHL and internal auditory canal (IAC) stenosis. Most cases of IAC stenosis are accompanied by SNHL, but the patients in our cases showed CFP also. We confirmed the agenesis of facial and vestibulocochlear nerves with the T2-weighted constructive interference in steady-state magnetic resonance imaging (CISS MRI) scans. We should consider a variety causes including genetic problems or trauma. Although the mother did not have a specific trauma history, she may have prolonged exposure to traumatic events during the first trimester of pregnancy. Clinicians who examine the SNHL, should pay attention to the history, physical and neurologic examination, and audiologic and imaging findings in a patient with or without facial nerve paralysis.

Key Words: Sensorineural hearing loss, Monozygotic twins, Facial paralysis, Magnetic resonance imaging

Introduction

Facial paralysis in the newborn is uncommon, occurring in 0.23% to 1.8% of live births, and 78% to 90% of the cases are associated with birth trauma. Congenital facial nerve paralysis (CFP) is generally considered to be of either developmental or acquired etiology. Acquired type of CFP could have infectious, traumatic, iatrogenic, neoplastic, and idiopathic causes. And there is a 90% chance of spontaneous recovery, but the developmental type occurs independently or as part of a recognized syndrome such as Möbius, Poland, Goldenhar, CHARGE, or velocardiofacial syndrome. Differential diagnosis of CFP is mandatory, because it will affect the optimal treatment and prognosis. The initial neonatal examination should thoroughly assess the severity of the facial palsy, seek physical evidence of birth trauma, and rule out the presence of associated otologic, craniofacial, or other systemic anomalies.

There is a documented association between CFP and sensorineural hearing loss (SNHL), and approximately 2–3% of patients with congenital SNHL have...
comorbid internal auditory canal (IAC) stenosis. However, almost patients with congenital SNHL have normal facial nerve, and these patients rarely exhibit functional impairments in both facial and vestibulocochlear nerves.

We report a monozygotic twin with SNHL and one of them has unilateral CFP accompanied by IAC stenosis which confirmed with CISS MRI & DTT. To our knowledge, this set of disorders has not been described in the literature.

**Case reports**

1. **Case I—twin A**
   A 7-months old female presented with persistent facial paralysis after birth. When she was crying, the mouth angle was deviated to the left accompanied by an absence of wrinkles of right eye. The patient was the elder one of a set of twins that was delivered by Cesarean section at 36th weeks of gestation due to premature rupture of membrane. The patient weighed 2,340 g at birth and had no eventful perinatal course; moreover, there was no family history of facial paralysis or other neurological deficits. Further, the mother did not have a history of disease or medication during the pregnancy.

   The patient was presumed to have normal muscle tonicity because there were normal findings following a neurological assessment and no evidence to suggest the presence of dysmorphism. There was an abnormal finding in auditory screening test performed at birth, the patient visited the outpatient clinic of otolaryngology. Brainstem-evoked response audiometry (BERA) and otoacoustic emission (OAE) indicated the presence of right-sided SNHL, and a nerve conduction velocity (NCV) showed a 75% degeneration ratio. Constructive interference in steady state magnetic resonance imaging (CISS MRI) revealed hypoplasia of the IAC, which is indicative of agenesis of facial and vestibulocochlear nerves. On the axial view, the maximum diameter of right IAC were 2 mm, but 4.5 mm on the left (Fig.1A, 1B). On the sagittal view, the vestibular and cochlear nerves in the left IAC were clearly visible, whereas those in the right IAC were irregularly shaped; there was no clear presence of the facial, vestibular, or cochlear nerve (Fig. 1C, 1D). Diffusion tensor tractography (DTT) for this patient showed normal integrity of the left facial nerve. However, right facial nerve was not reconstructed.

2. **Case II—twin B**
   The younger one of the twins weighed 2,240 g at birth and had an uneventful perinatal course; however, she exhibited abnormal findings on auditory screening at birth. In this patient, BERA and OAE revealed left sided SNHL; however, otolaryngological and physical examinations yielded normal findings. She did not have the symptom of facial paralysis clinically. CISS MRI revealed hypoplasia of the left cochlear nerve, but the clear presence of both facial nerves, left vestibular nerve, and right vestibulocochlear nerve (Fig. 2) without hypoplasia of the IAC. DTT showed normal integrity of the both facial nerves.

**Discussion**

The facial nerve branches into motor and sensory fibers. Its motor fibers originate from caudal border of the pons, run along the anterolateral side of the pons, and then exit from the cere-
bellopontine angle. Otherwise, sensory fibers run along the vestibulocochlear nerve and joins the motor fibers. The vestibulocochlear nerve branches into the cochlear and vestibular nerve fibers. The cochlear nerve originates from the spiral ganglion of the cochlea, while the vestibular nerve originates in the vestibular ganglion of the IAC. The vestibulocochlear nerve passes the IAC; in the IAC, the facial nerve runs along the vestibular nerve anteriorly and the cochlear nerve posteriorly. When a child was born with CFP, we have to consider the etiologies are traumatic or developmental. Patients with anomalies like manifestations of craniofacial dysmorphism or multisystemic syndromal pathology are often suspected of having developmental CFP, while patients with a history of complicated delivery, hematotympanum or ecchymosis are suspected of having traumatic CFP. Diagnostic evaluation of a child born with facial paralysis is a multistep process with the initial neonatal assessment being most important. The distinction not only provides prognostic information to guide proper management but may be of medicolegal significance.

To differentiate these diagnoses, clinicians should perform electromyography, NCV, blink reflex, or electroneuronography tests as well as an evaluation of the auditory brainstem reflex. In the present series, both computed tomography (CT) and MRI were useful diagnostic modalities. High spatial resolution CT scans can provide standard imaging for temporal bones, including the IAC and facial canal, while T2-weighted gradient echo imaging and CISS MRI are useful for confirming the facial and vestibulocochlear nerves. In the present study, the IAC had a mean length of 12 mm and a mean vertical diameter of 4 mm (range: 2–8 mm); a narrow IAC is defined as having a diameter of <2 mm. On T2-weighted MRI, the facial nerve has a hypointense line, and is located anterior to the superior portion of the vestibular nerve and superior to the cochlear nerve. On an axial view, the facial nerve runs anterior to the vestibulocochlear nerve.

In the current case, the patient initially showed IAC stenosis on MRI; therefore, temporal bone CT scans were not performed. According to a review of the literature, there are rare cases of hypoplasia in both the facial and vestibulocochlear nerves that may have been due to different embryological origins, but only a few have been reported in this case series. Therefore, we should consider a variety causes including genetic problems or trauma, even though the mother had no specific history of trauma.

This is the rare case with SNHL in the twin and one of them has facial nerve palsy with IAC stenosis occurred which proven by CISS MRI and DTT. The present case suggests that clinicians should make a differential diagnosis between traumatic and developmental CFP in infants who initially present with related symptoms and also consider the histories. Additionally, a meticulous examination of the head and neck region and the central nervous system should be performed.

요약
저자들은 일측성 감각 신경성 난청을 보이는 일란성 쌍생아에서 이 중 한 명에서 선천성 안면 마비와 내이도 협착을 보이는 증례를 소개하고자 한다. 내이도 협착을 보이는 대부분의 환자에서 감각 신경성 난청을 동반하는데, 본 증례에서는 한 명의 쌍생아에서 감각 신경성 난청과 안면 신경기능 이상 소견을 보였고, 다른 한 명은 내이도 협착은 없으나 감각 신경성 난청 소견을 보였다. 안면 신경 이상을 보이는 환아에서는 CISS MRI에서 안면 신경(7번 뇌신경)과 내이 신경...
(8번 뇌신경)의 이상소견을 확인하였다. 원인을 명확하게 확인할 수 없으나, 유전적인 문제 및 외상에 의한 것 등을 고려해야 하겠다. 임상 의들은 청력소실을 보이는 환아에서 안면 신경의 마비의 유무에 상관 없이 주의深い 병력청취 및 이학적 검사와 청력검사 및 영상학적 검사 등을 세심하게 고려해야 할 것으로 생각된다.

References