일측성 감각 신경성 난청과 내이도 협착을 동반한 안면 신경 마비를 보이는 일란성 쌍생아

영남대학교 의과대학 소아청소년과¹, 재활의학과²

서규현¹ · 손수민² · 김세윤¹

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 audiology and imaging findings in a patient with or without facial nerve paralysis

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

Kyoo Hyun Suh, MD¹, Su Min Son, MD², Saeyoon Kim, MD¹

¹Department of Pediatrics, ²Department of Physical Medicine and Rehabilitation, College of Medicine, Yeungnam University, Daegu, Korea

Submitted: 23 August, 2016

Revised: 15 September, 2016

Accepted: 15 September, 2016

Correspondence to Saeyoon Kim, MD
Department of Pediatrics, College of Medicine,
Yeungnam University, 170 Hyeonchung-ro, Nam-gu,
Daegu 42415, Korea
Tel: + 82-53-620-3533, Fax: +82-53-629-2252
E-mail: sysnow88@hanmail.net

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 trauma¹⁾. Congenital facial nerve paralysis (CFP) is generally considered to be of either developmental or acquired etiology²⁾. Acquired type of CFP could have infectious, traumatic, iatrogenic, neoplastic, and idiopathic causes¹⁾. 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^{3,4)}. 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 anomalies²⁾.

There is a documented association between CFP and sensorineural hearing loss (SNHL)⁵⁾, and approximately 2-3% of patients with congenital SNHL have

comorbid internal auditory canal (IAC) stenosis⁶⁾. However, almost patients with congenital SNHL have normal facial nerve, and these patients rarely exhibit functional impairments in both facial and vestibulocochlear nerves^{3,4,6)}.

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 fibers⁷⁾. Its motor fibers originate from caudal border of the pons, run along the anterolateral side of the pons, and then exit from the cere-

Fig. 1. (A) (B). Axial view of the T2-weighted constructive interference in steady-state magnetic resonance imaging (CISS MRI) scans of the brain of the elder of the twins. (A) The facial nerve (arrow) runs anterior to the vestibulocochlear nerve (arrowhead) in the left internal acoustic canal (IAC). (B) The right IAC has a maximum diameter of 2 mm, which is suggestive of hypoplasia (circle). There are no visible nerve fibers in the right IAC. (C) (D). Sagittal view of the T2-weighted CISS MRI scans of the brain of the elder of the twins. (C) In the left IAC, the facial nerve (arrow) is located superior to the cochlear nerve (arrowhead) and anterior to the superior portion of the vestibular nerve (curved arrow). (D) Relative to the left IAC, the right IAC has an irregular shape and the facial, vestibular, and cochlear nerves are poorly visualized.

belliopontine 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 posteriorly⁷.

When a child was born with CFP, we have to consider the etiologies are traumatic or developmental². 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, hemotympanum or ecchymosis are suspected of having traumatic CFP^{6,8}. 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 significance².

To differentiate these diagnoses, clinicians should perform electromyography, NCV, blink reflex, or electroneuronography tests⁹ as well as an evaluation of the auditory brainstem reflex⁹. 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 canal¹⁰, 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^{11,12}. 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 nerve⁷. On an axial view, the facial nerve runs anterior to the vestibulocochlear nerve⁷.

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^{3,4,6}. 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.

Fig. 2. (A,B) Axial view of T2-weighted constructive interference in steady state magnetic resonance imaging (CISS MRI) scans of the brain in the younger of the twins. The right and left IAC have a maximum diameter of 5.3 and 4 mm, respectively. In addition, the facial and vestibulocochlear nerves are well visualized in both the right and left IAC. (C,D) Sagittal view of T2-weighted constructive interference in steady state magnetic resonance imaging (CISS MRI) scans of the brain in the younger of the twins. (C) The facial (arrow), cochlear (arrowhead) and superior and inferior vestibular nerves (curved arrow) are well visualized in the right IAC. (D) There are no cochlear nerves at the anterior inferior portion of the left IAC. In addition, the facial (arrow) and superior and inferior vestibular nerves (curved arrow) are well visualized in the left IAC.

요약

저자들은 일측성 감각 신경성 난청을 보이는 일란성 쌍생아에서 이중 한 명에게서 선천성 안면 마비와 내이도 협착을 보이는 증례를 소개하고자 한다. 내이도 협착을 보이는 대부분의 환자에서 감각 신경성 난청을 동반되는데, 본 증례에서는 한 명의 쌍생아에서 감각 신경성 난청과 안면 신경 기능 이상 소견을 보였고, 다른 한 명은 내이도 협착은 없었으나 감각 신경성 난청 소견을 보였다. 안면 신경 이상을 보이는 환자에서는 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 환자에서 안면 신경의 마비의 유무에 상관 없이 주의깊은 병력청취 및 이학적 검사와 청력검사 및 영상학적 검사 등을 세심하게 고려해야 할 것으로 생각된다.

References

- 1) Shargorodsky J, Lin HW, Gopen Q. Facial nerve palsy in the pediatric population. *Clinical Pediatrics* 2010;49:411-7.
- 2) Shapiro NL, Cunningham MJ, Parikh SR, Eavey RD, Cheney ML. Congenital unilateral facial paralysis. *Pediatr* 1996;97:261-5.
- 3) Lin KM, Huang CC, Leung JH. Congenital unilateral facial palsy and internal auditory canal stenosis. *Pediatr Neurol* 2008;39:116-9.
- 4) Terzis JK, Anesti K. Developmental facial paralysis: a review. *J Plast Reconstr Aesthet Surg* 2011;64:1318-33.
- 5) Ozkiris M. Unilateral sensorineural hearing loss and facial nerve paralysis associated with low-voltage electrical shock. *Ear Nose Throat J* 2014;93:62-6.
- 6) Baek SK, Chae SW, Jung HH. Congenital internal auditory canal stenosis. *J Laryngol Otol* 2003;117:784-7.
- 7) Jäger L, Reiser M. CT and MR imaging of the normal and pathologic conditions of the facial nerve. *Eur J Radiol* 2001;40:133-46.
- 8) Nakamura K, Koda J, Koike Y. Stenosis of the internal auditory canal with VIIth and VIIIth cranial nerve dysfunctions. *ORL J Otorhinolaryngol Relat Spec* 1999;61:16-8.
- 9) Renault F. Facial electromyography in newborn and young infants with congenital facial weakness. *Dev Med Child Neurol* 2001;43:421-7.
- 10) Casselman JW, Offeciers EF, De Foer B, Govaerts P, Kuhweide R, Somers T. CT and MR imaging of congenital abnormalities of the inner ear and internal auditory canal. *Eur J Radiol* 2001;40:94-104.
- 11) Cho YS, Na DG, Jung JY, Hong SH. Narrow internal auditory canal syndrome: parasagittal reconstruction. *J Laryngol Otol* 2000;114:392-4.
- 12) Rothschild MA, Wackym PA, Silvers AR, Som PM. Isolated primary unilateral stenosis of the internal auditory canal. *Int J Pediatr Otorhinolaryngol* 1999;50:219-24.