What Is Expected of the Facial Nerve in Michel Aplasia? Anatomic Variation

Masoud Motasaddi Zarandy, M.D.,¹ Ali Kouhi, M.D.,²
Shervin Sharif Kashany, M.D.,³ Sohrab Rabiei, M.D.,⁴
Fatemeh Hajimohamadi, M.D.,⁵ and Mahtab Rabbani-Anari, M.D.²

ABSTRACT

We sought better understanding about the facial nerve anatomy in the rare inner ear Michel anomaly to help better define this aplasia and prevent potential complications in surgery on these patients. The data from computed tomography scans and magnetic resonance images of six Michel aplastic ears (three patients) were evaluated for a facial nerve course. Facial nerve course and anatomic landmarks were noted. Based on data obtained from this group of very rare patients, three different facial nerve anatomies were encountered. The first patient had normal-looking mastoid cells, normal middle ear ossicles, and a completely formed facial nerve canal through the middle ear. The second patient had pneumatized mastoid air cells despite an anomalous ossicular chain. This patient also had a facial nerve canal but not through the middle ear. In the third patient, although mastoid cells were present, neither ossicles nor a definite facial nerve canal could be detected. With guidance provided by the anatomy of the other parts of the ear, such as air cells and the ossicular chain, the danger zones posing a high probability of facial nerve injury can be predicted. Although all Michel aplasias may have aplastic petrous bone in common, there are some degrees of variation.

KEYWORDS: Facial nerve, Michel aplasia, inner ear anomaly

In 1863, Michel reported a congenitally deaf 11-year-old boy, from whose autopsy a bilateral absence of inner ear structures was diagnosed.¹ Michel named the bilateral aplasia of petrous as the *Michel anomaly*. There are some reports of this rare condition showing up in various degrees, ranging from total aplasia of petrous to more subtle defects of Michel dysplasia.^{2,3} Congenital deafness and abnormal development of the temporal bone in these patients introduced the genetic etiology and consanguinity of parents and pedigree of familial cases that helped to formulate a hypothesis proposing a

pattern of inheritance. However, perhaps due to rarity of this disease, no specific pattern of genetic inheritance has yet been proved.⁴ In any case, unilateral aplasia may indicate etiologies other than genetics.⁵

The Michel anomaly presents a spectrum of various degrees of aplasia. Some cases have a petrous bone but no inner ear structures, and others have no petrous bone at all.^{2,3} Other associated skull base anomalies may be encountered including platybasia and an aberrant course of the jugular veins.¹ The facial nerve is one of the important structures passing through the

¹Hearing Research Center, Department of Otolaryngology and Head and Neck Surgery, ²Department of Otolaryngology and Head and Neck Surgery, and ³Department of Radiology, Tehran University of Medical Sciences, Tehran, Iran; ⁴Department of Otolaryngology, Kermanshah University of Medical Sciences, Kermanshah, Iran; ⁵Department of Anesthesiology, Tehran University of Medical Sciences, Tehran, Iran.

Address for correspondence and reprint requests: Masoud Motasaddi Zarandy, Amir-A'lam Hospital, South Saadi Avenue,

Tehran, Iran (e-mail: Motesadi@sina.tums.ac.ir).

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temporal bone, and its course may be affected by this aplasia. Knowing the facial nerve direction in these patients will help to better define this anomaly and may eventually result in classification of Michel. Furthermore, as some of these patients may need ear surgery,² the anomalous course of the seventh nerve may result in the morbidity of iatrogenic trauma to facial nerve.

This study was performed to study the facial nerve's course in the ears with Michel anomaly.

CASE REPORTS

Case 1

A 3-year-old girl was diagnosed with primary bilateral profound sensorineural hearing loss (SNHL) since her fifth month. She received a hearing aid at 6 months, and she experienced no further developmental or mental problems. When she was referred to this center, auditory rehabilitation was administered. On examination, she had a bilateral lop ear deformity and a normal external auditory canal and tympanic membrane. A computed tomography (CT) scan showed a normal ossicular chain in attic. The facial nerve had near normal divisions; tympanic and mastoid portions could be distinguished (Fig. 1). The styloid process was totally absent. Unfortunately, magnetic resonance imaging (MRI) was unavailable.

Case 2

A 4-year-old girl with profound bilateral SNHL was referred. Her parents were consanguineous. An audiogram showed very high thresholds (>90 dB) for low frequencies and no response to high frequencies. She had normal facial nerve function, external ear, and tympanic membrane. Imaging indicated that mastoid air cells were well pneumatized and an anomalous incudo-malar complex was present. There was no styloid process. The vertical facial nerve canal was present but anomalous, passing directly from the cerebellopontine angle to the mastoid portion with no tympanic part. Geniculate ganglion and tympanic portions were missing. The mastoid portion was oblique, as it had been significantly displaced inferoposteriorly (Fig. 2). On T1- and T2-weighted MRI, the proximal portion of the facial nerve, as well as the vertical parts, could be seen. The cochlea-vestibular nerve was missing.

Case 3

A 3.5-year-old, normal-looking boy with profound bilateral primary SNHL was referred to this cochlear implant clinic. The parents were not consanguineous, and the patient had no history of significant prenatal

problems. An audiological evaluation (including pure tone audiometry, auditory brain stem response, and otoacoustic emission) indicated profound sensory neural hearing loss. There were no external ear and canal problems. The facial nerve function was normal and symmetric. Otoscopy revealed a normal tympanic membrane with a deformed malleus. The CT scan noted complete bilateral aplasia of the petrous bone. Mastoid air cells were pneumatized (Fig. 3). The middle ear ossicular chain was absent, as was the styloid process. In a search for the facial nerve direction or canal, no bony canal or foramen could be attributed to the seventh nerve. MRI showed the facial nerve proximal portion leaving the brain; however, the entire course could not be traced. No brain anomaly was present except for an abnormally high tentorium position and an anteromedial displacement of the cerebellar hemispheres around the pons and the medulla.

DISCUSSION

Inner ear anomalies are among the main etiologies of primary SNHL. Of these, 20% affect bone and so can be diagnosed and evaluated by CT and MRI. However, the other 80% are only membranous and therefore cannot be detected by these methods.^{3,6} One of the rare causes of inner ear anomalies affecting bone is the group of anomalies named Michel, which do not have any inner ear bony structure.¹ It should be noted that at this otology referral center, located in a highly populated country of more than 70 million, only three cases of this anomaly have been reported over a 15-year period of performing cochlear implant.

The management of these patients involves some special considerations. The absence of the cochlea-vestibular nerve makes brain stem implants the only possible choice for auditory rehabilitation. It has been shown that such patients may suffer from infectious diseases of the middle ear, due to their middle ear cavity and mastoid air cells.² Surgery may therefore be indicated in some cases, at least for infectious complications such as chronic otitis media or cholesteatoma formation. In these situations, the importance of knowing the facial nerve course cannot be overemphasized.

Regarding the degree of petrous bone formation, the variations of Michel aplasia were shown previously.^{2,3} Similarly, this study's data show that, in the six ears investigated, there were different facial nerves courses. Interestingly, some correlation was seen between middle ear structures and these patterns. In the first patient, the temporal bone consisted of normal-looking mastoid cells, normal middle ear ossicles, and a completely formed facial nerve canal. In these ears, the nerve canal was displaced inferoposteriorly, but the nerve still had a passage through the middle ear and then a curve to the oblique vertical portion. The second patient

Figure 1 Computed tomography scan showing a facial nerve originating from an opening in the inferior portion of the middle ear cavity. (a, b) Axial view, lower cuts; (c, d) axial view, higher cuts; (e, f) coronal view, anterior cuts; (g, h) coronal view, more posterior cuts.

Figure 2 Facial nerve course through the mastoid bone, without entrance to the middle ear cavity. (a, b) Axial view, lower cuts; (c, d) axial view, higher cuts; (e, f) coronal view.

Figure 3 Coronal and axial computed tomography scan showing pneumatized mastoid air cells and the absence of any passage for the facial nerve. (a, b) Axial view, lower cuts; (c, d) axial view, higher cuts; (e, f) coronal view.

had pneumatized mastoid air cells but an anomalous ossicular chain and also a facial nerve canal but not through the middle ear. The facial nerve passed directly from the brain to the mastoid, with no tympanic portion of the facial nerve present. In the third patient, although mastoid cells were present, no ossicles or definite facial

nerve canal could be seen. Seemingly, when normal middle ear structures are present, the facial nerve follows a nearly normal course, with distinct tympanic and mastoid portions. However, when there are some anomalies, the facial canal is also abnormal. With guidance provided by the anatomy of the other parts of the

ear, such as air cells and the ossicular chain, the danger zones posing a high probability of facial nerve injury can be predicted.

These data may suggest that, although all Michel aplasias may have aplastic petrous bone in common, there are still some degrees of variation. Studies involving a greater number of such patients may help to further investigate these findings and clarify why unilateral cases or different degrees of differentiation exist.

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