Duplication Anomaly of the Internal Auditory Canal

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Congenital abnormalities of the internal auditory canal (IAC) are rare. The only report of the radiological features of this anomaly is by Curtin and May [1], who described a duplication of the IAC, which they studied with pleuridirectional tomography. We report a case of a unilateral duplication of the IAC studied with MR imaging and CT.

Case Report

A 45-year-old man presented with slowly progressive left-sided hearing loss of 5 years duration. Examination revealed bilateral sensorineural hearing loss, more severe on the left, with poor speech discrimination on the left. Brainstem-evoked-response audiometry demonstrated borderline prolongation of interpeak latencies and poor early wave identification on the left side. A retrocochlear lesion was considered a possibility.

MR imaging was performed on a GE Signa unit (Milwaukee, WI) at 1.5 T. Images were acquired before and after IV administration of gadopentetate dimeglumine (Magnevist, Berlex). The cerebellopontine angle and brain parenchyma were normal. There was no enhancement to suggest an acoustic neuroma. Adjacent to the fundus, the more posterior canal, was more prominent than the right, but was isointense with CSF (Fig. 1A). The fundus of the left IAC was more prominent than the right, but was isointense with CSF (Fig. 1B).

A CT scan (GE 9800, Milwaukee) was obtained to evaluate the possibility of an osseous anomaly. Contiguous 1.5-mm axial scans taken through the temporal bone using a bone algorithm revealed two canals extending from the cerebellopontine angle cistern to the large fundus (Figs. 2A and 2B). The anterior canal was narrower than the more posterior canal and slightly superior to it (Figs. 2A, 2B, 2D, and 2E). The two canals joined at the medial aspect of the fundus of the IAC, 1 cm medially to the cochlea (Fig. 2B). The fundus was patulous, and larger than its counterpart on the right. From the fundus, the labyrinthine portion of the left facial nerve canal followed a normal course to the geniculate turn. The tympanic facial nerve canal, second genu, and descending facial nerve canal appeared to be normal. The right temporal bone was normal.

Additional physiological testing was then done to assess the functional integrity of the nerves that might traverse the anomalous canal. Electroneuronography of the facial nerve demonstrated normal responses bilaterally, confirming the clinical impression of normal facial nerve function. Similarly, electronystagmography revealed normal vestibular function on the side of the anomalous temporal bone.

Eight months after the initial evaluation, hearing and brainstem-evoked audiometry were tested again. The findings were unchanged.

Discussion

The seventh (facial) and eighth (vestibuloacoustic) nerves arise together from the facioacoustic primordium, a cluster of cells derived from the neural crest [2, 3]. This primordium, which can be identified in embryos as small as 5 mm, has an attachment to the metencephalon just rostral to the otocyst [4].

The facioacoustic primordium begins to divide by the 8–10 mm stage [4]. Separation into two distinct nerves, the facial and vestibuloacoustic, is complete by the time the embryo is 14 mm [4, 5].

The otic capsule first appears at about the 5-mm stage (fifth week of gestation) as mesenchyme condensing around the developing labyrinth (otocyst) [3]. By the beginning of the sixth week, this mesenchyme surrounds the entire otocyst except for a gap medially, through which the primitive seventh and eighth nerves pass. This gap will become the IAC.

The primitive mesenchyme of the developing otic capsule is replaced first by embryonic cartilage (precartilage) and then by true cartilage. At both these stages, the IAC containing the facial and acoustic nerves can be identified.

The cartilage of the otic capsule ossifies from 14 separate ossification centers, which appear at specific times beginning at approximately the end of the 16th week [2]. Ossification does not begin until each corresponding component of the inner ear has attained adult size [3]. Four ossification centers (the fifth, sixth, seventh, and ninth to appear) form the bony walls and roof of the IAC [3]. The facial nerve occupies the anterior superior portion of the mature osseous IAC.

Although surgical proof is lacking, it seems most likely that the anterior of the two canals in our patient contains the facial nerve or a portion of it. However, the accessory canal could contain the seventh or eighth cranial nerve, or a portion of either.

Anomalies of the IAC portion of the facial nerve are rare [1].

Received December 28, 1990; revision requested February 24, 1991; revision received March 26, 1991; accepted April 22, 1991.

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More often, it is the facial nerve distal to the IAC that follows an atypical course through the temporal bone. In a histopathologic study of 100 cadaveric temporal bones with congenital inner and middle ear anomalies, Sando et al. [6] found that the middle ear structure most often anomalous was the facial nerve. The most common facial nerve anomaly is a course more anterior than normal [7, 8]. This can be thought of as "migration toward the target organ" [1], which, in the case of the facial nerve, would be the muscles of facial expression. Normal development of the walls of the tympanic and mastoid portions of the canal and perhaps the cochlea may inhibit such anterior migration of the facial nerve [1]. These structures were normal in this patient.

Theoretically, early or abnormal separation of the facioa-
coustic primordium might result in an aberrant course of fibers of the seventh or eighth nerves. For example, a delay in condensation of the mesenchymal precursor of the walls of the IAC might permit the facial nerve to migrate away from the acoustic nerve [1]. Two canals could then develop instead of one, one canal around the facial nerve and one around the vestibuloacoustic nerve.

If anterior deviation of the facial nerve is the result of "migration toward the target organ," the first region that should deviate anteriorly is the geniculate turn, which would be "pulled" by the greater superficial petrosal nerve. There is no branch leaving the intracanalicular segment that could provide the "pull" toward the abnormal position that was seen in this patient. Migration may therefore be an effect of the overall expansion and growth of the osseous and soft-tissue structures rather than the result of an actual "pulling." The exact mechanism of facial nerve migration is poorly understood. Apparent migration may also be the result of an anomalous ossification center or anomalous ossification of one of the centers that contributes to the walls of the IAC.

Because the patient with a duplicated IAC previously described by Curtin and May [1] exhibited clinical facial nerve weakness, we specifically tested for subclinical facial nerve abnormality with electroneuronography. Facial nerve function in our patient was normal.

Duplication of the IAC is an important radiologic observation that could alter the surgeon's approach if exposure of the IAC becomes necessary. The findings presented here indicate that although an osseous anomaly can be detected or suspected on an MR study of the temporal bone, CT with thin sections and bone algorithms provides much more detailed information. Because of its clear delineation of osseous anatomy, CT is the examination of choice.

REFERENCES