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Radiology in Focus

Narrow internal auditory canal syndrome: parasaggital reconstruction

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Abstract
Narrow internal auditory canal (IAC) syndrome is a malformation of the temporal bone, that is defined as an IAC diameter of only 1–2 mm on high-resolution computed tomographic scans (HRCT). This syndrome is known to be caused by the absence (aplasia or hypoplasia) of the vestibulocochlear nerve. We present a case of unilateral narrow IAC syndrome which was diagnosed by HRCT. The aplasia of the vestibulocochlear nerve was confirmed using parasigittal reconstruction magnetic resonance image (MRI). The IAC was composed of two separate canals, one of which contained a facial nerve and the other was empty with aplasia of the vestibulocochlear nerve.

Key words: Temporal bone; Tomography, X-ray computed; Magnetic resonance imaging

Introduction

Computed tomography (CT) of the temporal bone is used widely to seek the possible causes of congenital sensorineural hearing loss. Narrow internal auditory canal (IAC) syndrome is a malformation of the temporal bone, which is defined as a canal of only 1–2 mm in diameter on high-resolution computed tomographic scans (HRCT).[1] This syndrome is known to be caused by the absence (aplasia or hypoplasia) of the vestibulocochlear nerve.[2] Although a narrow IAC may be visualized in the axial plane, parasagittal reconstruction magnetic resonance image (MRI) perpendicular on the course of IAC could be used for confirmation.[3]

We present a case of narrow IAC syndrome caused by aplasia of the vestibulocochlear nerve that was confirmed using MRI with parasagittal reconstruction. As far as we know, this is the first report on the narrow IAC syndrome composed of two separate canals, one of which contained a facial nerve and the other was a hollow space caused by aplasia of the vestibulocochlear nerve.

Case report

An 18-year-old girl was referred to Samsung Medical Centre for unilateral hearing loss, that had been detected five years previously. She had no history of ear infection nor head trauma, and she did not present any vestibular symptoms or tinnitus. Otological examination revealed no abnormal findings. A pure tone audiogram revealed that the right side was scaled out in all frequencies with normal hearing in the left. The vestibular function test revealed compensated vestibular weakness of the right side. A bithermal caloric test showed 100 per cent right canal paresis with normal posturographic finding. The patient did not have any facial palsy.

HRCT and MRI of IAC were obtained. HRCT showed a narrowing (1.2 mm width) of the right IAC with a normal inner ear structure, and there was no evidence of a nerve emerging from the cochlear modiolus in the axial image (Figure 1a).

3D-Fast spin echo (3D-FSE) T2 MRI showed a possibility of vestibulocochlear nerve aplasia in the right IAC on the axial image (Figure 2a). We performed parasagittal reconstruction using MRI and HRCT scan data for identification of the individual nerve in the IAC. Parasagittal reconstruction of HRCT showed two separate bony canals in the right IAC (Figure 1b). In the parasagittal reconstruction MRI, we could see complete IAC structure including facial and vestibulocochlear nerves on the left side (Figure 2c), while we found only one nerve structure, assumed to be the facial nerve on the right IAC (Figure 2b).

Discussion

Narrow IAC syndrome is defined as an IAC diameter of only 1–2 mm or less than 1.5 mm on HRCT[4] and was found in 12 per cent of congenital temporal bone anomalies. It is usually unilateral and may be associated with other malformations of the inner ear. The facial nerve is usually normal.[5]

A narrow IAC canal can be explained by the disturbance in the trophic effect that the cochlea exerts on the cochlear neurons. A disturbance in the trophic effect of the cochlea can result in the loss of too many or all neuronal fibres, and in these patients the IAC formed around the initial neuronal fibres will not develop further and eventually will be stenotic.[3]

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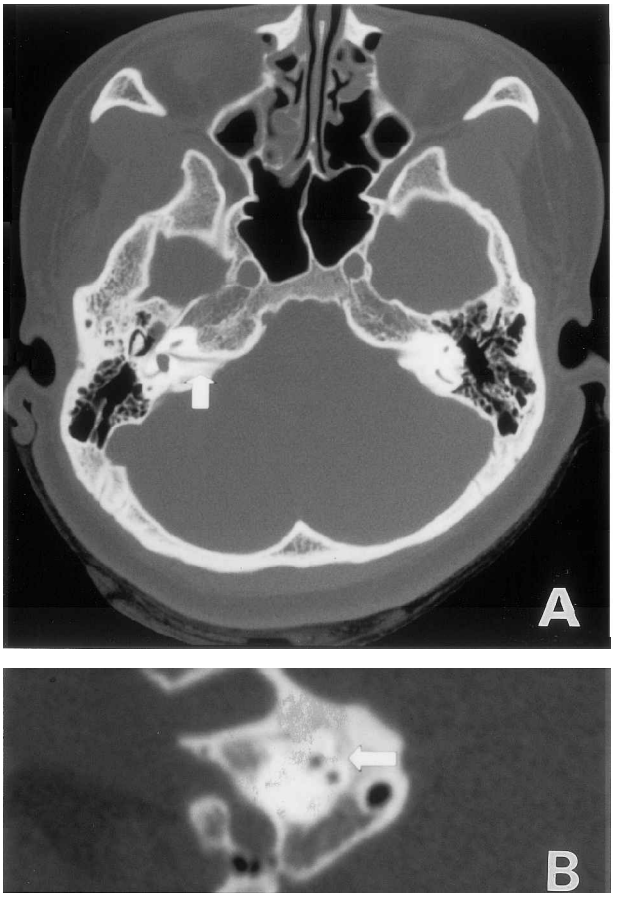

FIG. 1

Temporal bone CT images of the patient. (a) Axial CT image shows narrowing of the right internal auditory canal (IAC) (arrow) and no bony canal to the modiolus of cochlea. (b) Sagittal 2D image demonstrates anomalous separation of the right IAC into two canals by bony septum (arrow).

From the results of pure tone audiogram and the vestibular function test, absence of vestibulocochlear nerve function could be assumed and the cause of the empty canal might result from an incompletely developed vestibulocochlear nerve that eventually regressed.

Narrow IAC syndrome is most definitely visualized in the axial image of thin thickness HRCT. Information on structures smaller than 1 mm such as the vestibulocochlear and facial nerves can be obtained by MRI using various techniques (3D MP-RAGE, 3D CISS, T2 gradient echo).[6,7] A narrow IAC does not contain the vestibulocochlear nerve, but a hypoplastic IAC can contain a vestibulocochlear nerve with abnormal nerve function. In the latter case, comparison of width between facial nerve and vestibulo-cochlear nerve can be performed using MRI parasagittal reconstruction for confirmation.[3]

Patients with congenital profound sensorineural hearing loss and various inner ear malformation can benefit from a cochlear implant, but bilateral narrow IAC as seen on CT or MRI is not accepted as a indication for a cochlear implant.[8] Patients with narrow IAC syndrome who received a cochlear implant cannot respond to electric stimulation because of the absence of the cochlear nerve in spite of having normal labyrinthine structure.[9] For bilateral patients, a hearing-aid would not help but vibrotactile devices may be useful.[1] HRCT and MRI could now be used in candidates for cochlear implantation to confirm the presence of a normal cochlea and whether they have a normal IAC and cochlear nerve.

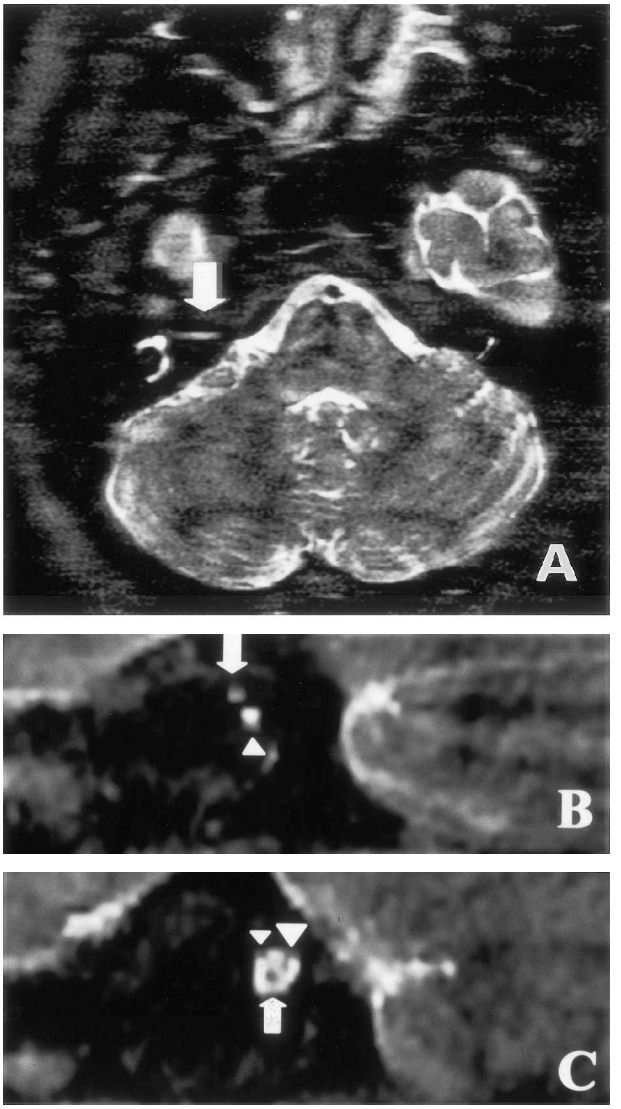

FIG. 2

3D-FSE T2-weighted MR images of the patient. (a) Narrowing of the right IAC is demonstrated on axial image (arrow). (b) Parasagittal reconstruction image obtained perpendicular to the course of the nerve of the right IAC. The facial nerve fills the upper canal (arrow), while the vestibulocochlear nerve is not seen in the lower canal (arrowhead). (c) Parasagittal reconstruction image of the patent left IAC. Facial nerve (small arrowhead), common vestibular nerve (large arrowhead), and cochlear nerve (arrow) are seen in the IAC.

Approximately 20 per cent of patients with congenital SNHL will have demonstrable abnormalities on HRCT.[4] If narrow IAC syndrome was found on HRCT, aplasia of the vestibulocochlear nerve could be expected and additional MRI would not give more information for diagnosis itself. But we think that MRI with parasagittal reconstruction has some meaning for evaluating the state of the nerves in the IAC, which would help to understand the patient's symptoms and determine future rehabilitation plans.

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