Medicine

Otorhinolaryngology fields

Okayama University *Year* 2004

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The Journal of Laryngology & Otology
November 2004, Vol. **118,** pp. 902–905

Abnormal direction of internal auditory canal and vestibulocochlear nerve

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Abstract
Several internal auditory canal (IAC) anomalies have been reported. To our knowledge, only one case with an abnormal direction of the IAC has been reported in an infant with Pierre Robin syndrome. In this paper, we present the first report of two non-syndromic cases with abnormal IAC direction.

Key words: Vestibulocochlear Nerve; Anatomy; Abnormalities; Nervous System Malformations

Introduction

The internal auditory canal (IAC) forms as a result of ossification of mesoderm involving the eighth cranial nerve. It is theorized that anomalies of the bony IAC are secondary to maldevelopment of the vestibulocochlear nerve.[1,2] Malformation of the IAC is a fairly rare entity. Several anomalies, including stenosis, atresia and dilatation of the IAC, have been reported.[3] Furthermore, IAC anomalies are usually associated with other inner, middle and external ear malformations. Multiple congenital anomalies have been noted in most individuals with malformation of the IAC.[4] Only one case of abnormal direction of the IAC in an infant with Pierre Robin syndrome has been reported.[5] This paper is the first report of two non-syndromic cases with unilateral abnormal direction of IAC.

Case report

Case 1

The patient was a 72-year-old woman complaining of occasional ear fullness in the left ear for three months. She did not complain of tinnitus or hearing impairment, but she had suffered a few vertigo attacks in her youth. She had never undergone otological examination previously. Her external ears and tympanic membranes were normal bilaterally. There was no malformation in the nose, oral cavity or pharynx. Furthermore, no definite anomalies, including cranio-facial malformation, malformed extremities or congenital heart disease, were found. Facial nerve palsy and nystagmus were not observed. Pure tone audiogram indicated sensorineural hearing loss in the left ear (Figure 1). To elucidate the cause of hearing loss, computed tomography (CT) and magnetic resonance imaging (MRI) scans were performed. The CT scan revealed an abnormal direction of the left IAC (Figure 2a) in contrast with the right IAC (Figure 2b). The lengths of the superior, inferior, anterior and posterior walls of the IAC and the diameters of the IAC both in the horizontal and vertical plane are shown in Figure 3. No other abnormality in the middle or inner ear was detected on CT scan. The MRI scan demonstrated the presence of the seventh and eighth cranial nerve complex even in the abnormal left side.

Case 2

A 6-year-old girl with atresia of the external auditory canal and microtia in the right ear was referred for surgical therapy. There was no history of head trauma or prenatal

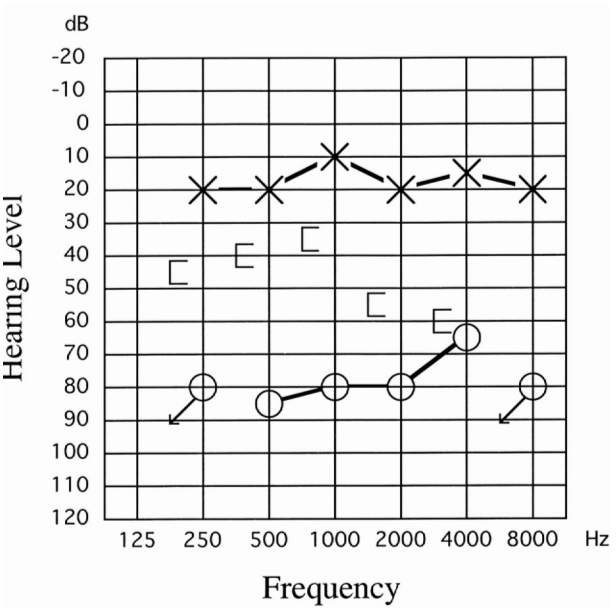

Fig. 1
Pure tone audiogram of Case 1.

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Accepted for publication: 11 June 2004.

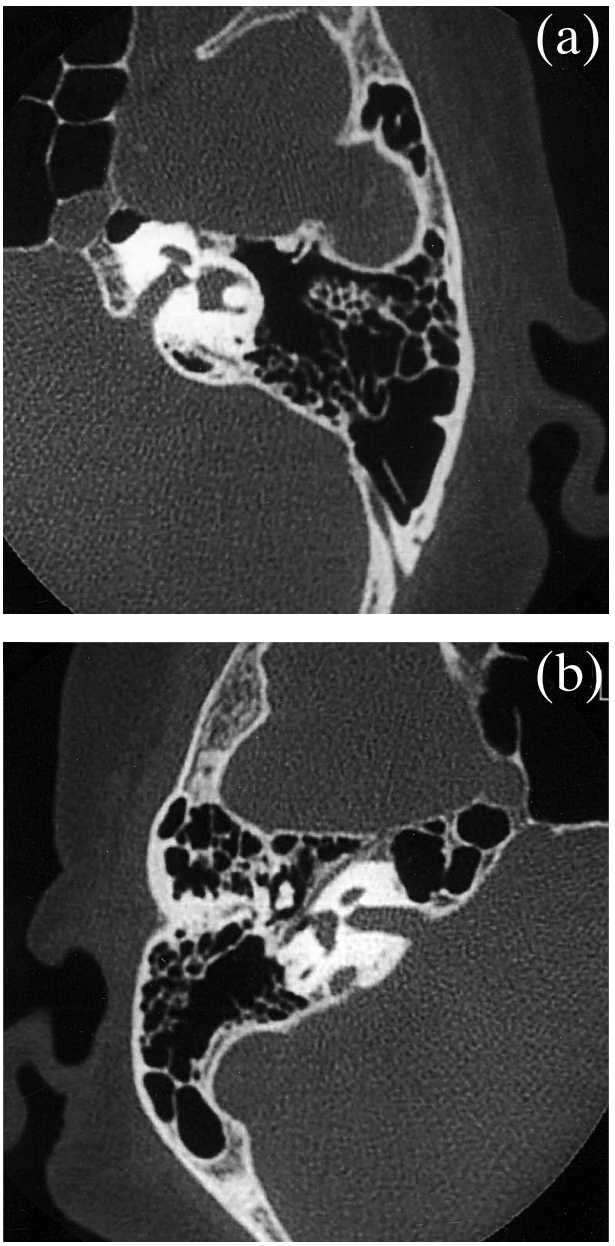

FIG. 2

(a) Axial CT scan at the level of the lateral semicircular canal in the left ear of case 1, showing an abnormal direction of the internal auditory canal, with a remarkable inclination to the front. Malformation of the cochlea, semicircular canal and middle ear were not detected in the left ear. (b) Axial CT scan at the level of the tympanic portion of the facial nerve in the right ear of case 1 shows normal findings.

or postnatal infection, such as otitis media or meningitis. In addition, the patient had no tinnitus or dizziness. The external ear and tympanic membrane in the left ear were normal. No other craniofacial deformities, such as cleft palate, micrognathia or congenital ptosis, were detected. Furthermore, systemic developmental malformations, including cardiac septal defects, polycystic kidney, skeletal anomalies and duodenal canal atresia, were not found. The patient had had normal mental and physical development throughout her life. Pure tone audiogram revealed a mixed-type hearing loss in her right ear, in contrast with a normal hearing level in the left ear (Figure 4). A CT scan showed normal findings in the left ear (Figure 5a), whereas in the right ear there was an abnormal direction of the IAC

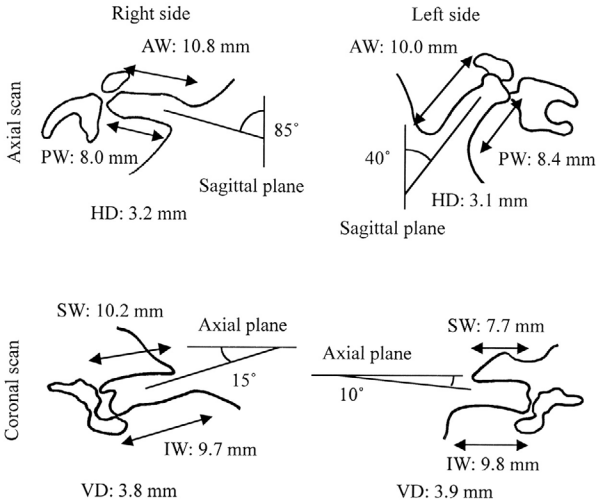

FIG. 3

Schematic drawing of the internal auditory canal of Case 1. AW = anterior wall; PW = posterior wall; SW = superior wall; IW = inferior wall; HD = horizontal diameter; VD = vertical diameter.

and poor development of mastoid pneumatization (Figure 5b). The lengths of each part of the IAC are shown in Figure 6. No other cochlear malformation in either ear was detected on CT scan.

Discussion

Malformations of the IAC are often accompanied by multiple congenital malformation syndromes such as Pierre Robin syndrome, trisomy 13 syndrome and trisomy 18 syndrome, and frequently present in association with minor abnormalities of the bony labyrinth.[4] In our cases, no distinct anomalies in the cochlea or the vestibule were found on CT scan. It is well known that the dimensions of the IAC vary within the population.[6] Although symmetry of the canals in the individual is important, severe asymmetry in paired ears is rare.[7] In the normal temporal

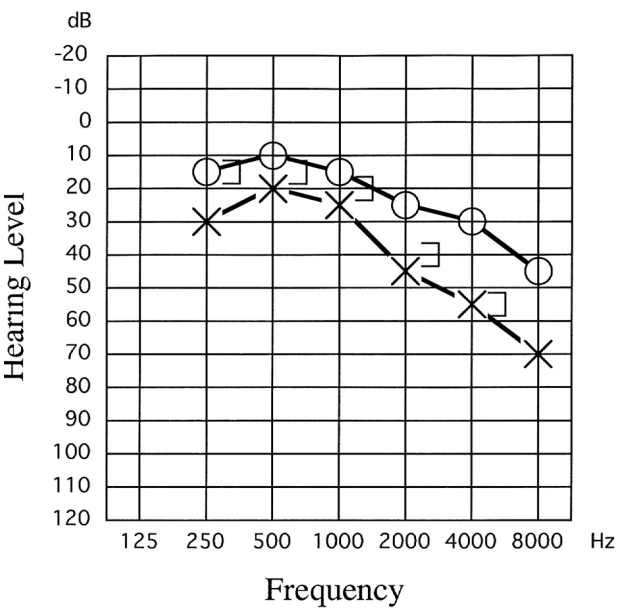

FIG. 4.

Pure tone audiogram of Case 2.

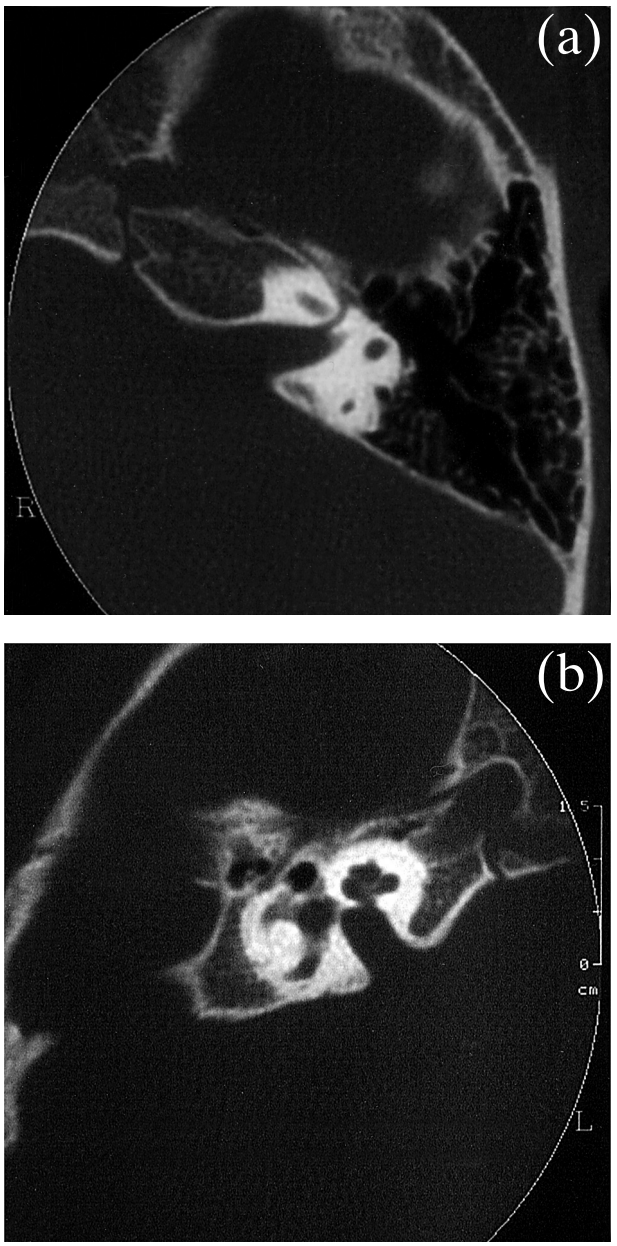

FIG. 5

Axial CT scan of case 2: (a) shows normal findings of the internal auditory canal at the level of the genu of the facial nerve in the left ear, while (b) shows abnormal direction of the internal auditory canal, which makes a 30º angle with the transverse biauricular plane, and poor development of mastoid pneumatization at the level of the lateral semicircular canal in the right ear.

bone, the bony IAC makes an angle of about 45º with the longitudinal axis of the petrous pyramid. Since this, in turn, is directed at an angle of 53º to the sagittal plane, the general axis of the internal auditory meatus makes an angle of 8º with the transverse biauricular plane.[8] Portmann *et al*. described the position of the IAC fundus as being lower than that of the IAC porus, and reported the angle between the IAC and the axial plane as being 15º.[9] In addition, the angle between the IAC and the sagittal plane is 80–90º. In both our cases, the angles between the IAC and the axial plane were within normal limits, but the angles between the IAC and the sagittal plane in the left ear of case 1 and in the right ear of case 2 were 40º and 30º, respectively. In addition, the length of the superior wall of the left IAC in case 1 and the length of the right IAC in case 2 were short in comparison with the normal data for adults[10] and children,[11] respectively.

- **Internal auditory canal abnormalities have been previously reported, including abnormal alignment of the canal in a patient with Pierre Robin syndrome**
- **Abnormal internal auditory canal directions were found in two non-syndromic patients**
- **Findings are discussed in light of the embryological development of this area**

According to Gulya and Schuknecht, atresia or stenosis of the IAC is secondary to aplasia or hypoplasia of the vestibulocochlear nerve.[12] At 37 days' gestation, the fibres of the vestibulocochlear nerve have grown to contact the developing otocyst.[13] Abnormal direction of growth of the vestibulocochlear nerve may be due to an unusual otocyst position, which may cause the malformation of the IAC seen in our cases.

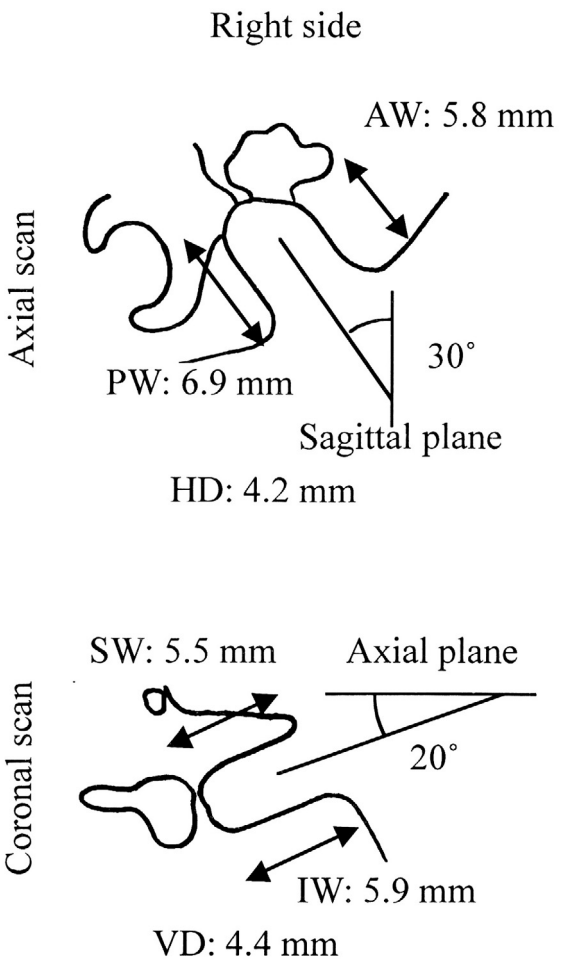

FIG. 6.

Schematic drawing of the internal auditory canal of case 2. AW = anterior wall; PW = posterior wall; SW = superior wall; IW = inferior wall; HD = horizontal diameter; VD = vertical diameter.

Acknowledgement

The authors thank Sebahattin Cureoglu, M.D., and Patricia A. Schachern, B.S., for their great support for this case report.

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Dr Shin Kariya takes responsibility for the integrity of the content of the paper.
Competing interests: None declared