Pathology of the Vestibulocochlear Nerve

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KEYWORDS

- Internal auditory canal • Vestibulocochlear nerve
- Hearing loss

This article covers congenital deformity of the internal auditory canal (IAC), neoplastic and pseudoneoplastic lesions with special detailed emphasis on schwannoma of the eight cranial nerve (acoustic neuroma), non-neoplastic IAC/cerebellopontine angle mass (CPA) pathology, including vascular loops and numerous additional differential diagnostic entities with particular emphasis on non-neoplastic meningeal disease.

CONGENITAL DEFORMITY OF THE INTERNAL AUDITORY CANAL

The caliber of the internal auditory canal (IAC) depends upon the development of the vestibulocochlear nerve.[1] This is consistent with findings noted in the evaluation of the other skull base foramina, virtually all of which develop in response to the presence of the regional nerve or artery. As such, congenital vestibulocochlear nerve deficiency commonly is associated with a hypoplastic IAC containing only the facial nerve (**Fig. 1**). Although measurements are somewhat arbitrary, the IAC is considered hypoplastic when less than 2 mm in diameter. This usually is appreciated best upon examination in the axial and coronal plane; however, high-resolution thin-section oblique sagittal T2W MR imaging not only allows demonstration of this small canal but also depicts to best advantage absence/hypoplasia of the vestibulocochlear nerve (**Fig. 2**).[2] The cochlear nerve is defined as hypoplastic when similar in size or smaller than either of the vestibular nerves.

Cochlear nerve deficiency in the presence of a normal-sized internal auditory canal implies that it is acquired after formation of the IAC. This has been observed most commonly in patients who have labyrinthitis ossificans (**Fig. 3**).

Abnormality of vestibulocochlear nerve caliber has crucial implications for patients requiring cochlear implantation.[3] Absence of the cochlear nerve is the only complete contraindication to this procedure. In addition to the IAC caliber, the imaging specialist must evaluate the status of the cochlear nerve aperture at the fundus of the IAC. Stenosis of this aperture consistently is associated with cochlear nerve deficiency also (**Fig. 1**B).[4] Additional considerations regarding cochlear implantation are discussed later in this article.

Narrowing of the IAC may occur secondary to exostoses or osteomas. These bony lesions are rare, but they may clinically mimic vestibular schwannomas by compression of the vestibulocochlear nerve. Hearing loss and vestibular symptoms including dizziness and vertigo may improve after surgical removal of the bony lesion. Compromise of the IAC also may occur as a complication of several of the otodystrophies, including Paget's disease and fibrous dysplasia.

A large IAC (greater than 9mm) may be incidental. Bilateral IAC enlargement is seen with neurofibromatosis (NF)-2 secondary to bilateral acoustic tumors. NF also may result in IAC widening because of dural ectasia without neoplasm.

The large IAC also has other important implications. Special attention to the fundus (lateralmost aspect) of the canal may reveal an unusually wide neural aperture leading to the cochlea or vestibule. This implies increased pressure within the perilymphatic space and predisposes to abnormal flow of perilymph (gusher) upon stapes manipulation. This can occur with various congenital inner ear malformations but is a classic manifestation of x-linked progressive mixed deafness (XLPMD) (**Fig. 4**).[5] These male patients present with mixed hearing loss possibly masquerading

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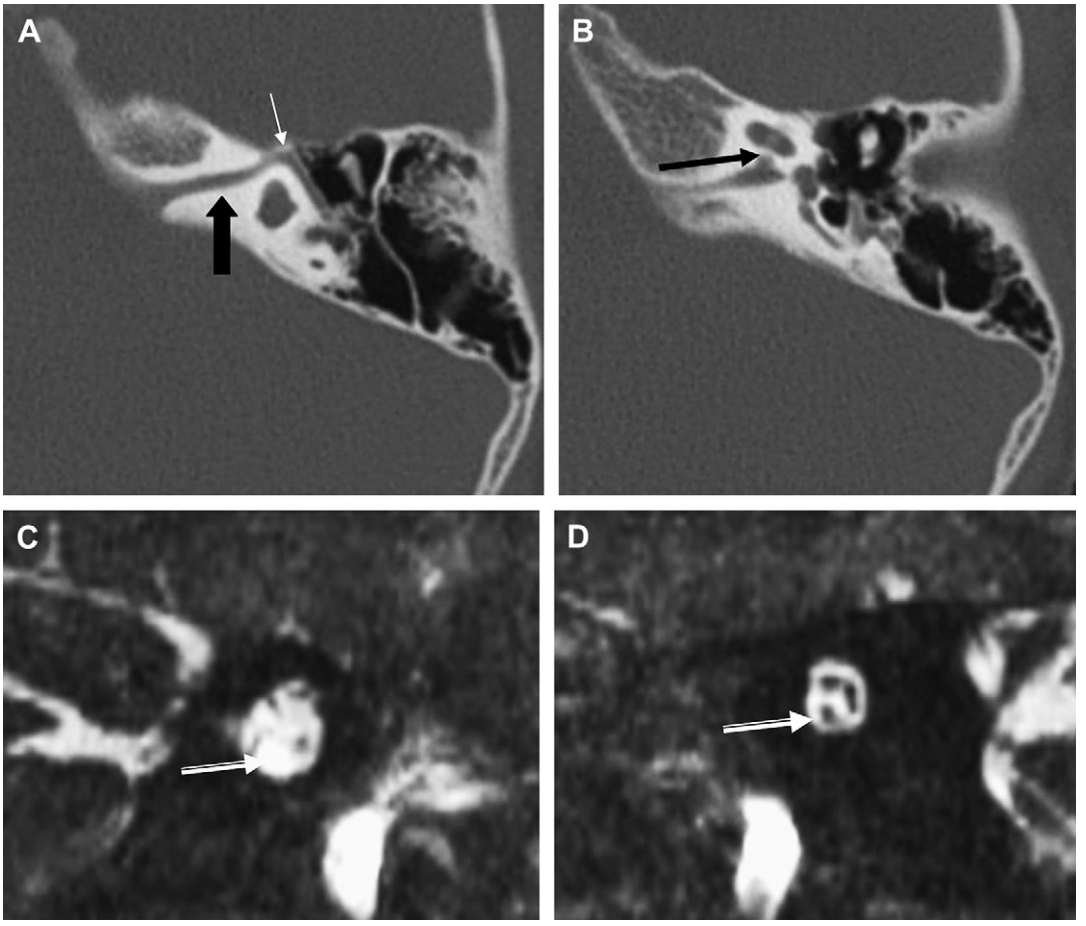

Fig. 1. Hypoplastic internal auditory canal (IAC), occluded cochlear neural foramen. (*A,B*) Axial CT images of the left ear reveal an extremely narrow internal auditory canal (*thick black arrow*) contiguous with the anterior genu of the facial nerve canal (*white arrow*) with a bony plate occluding the cochlear neural foramen (*long black arrow*). (*C*) Sagittal T2WI of the left ear reveals a vacant anteroinferior fundal IAC quadrant (*white arrow*) indicating that the cochlear nerve is absent. (*D*) Sagittal T2WI of the right ear reveals a normal cochlear nerve (*white arrow*). (*Courtesy of* C.D. Phillips, MD, Charlottesville, VA.)

as otosclerosis clinically. If the deformity is recognized before middle ear exploration, the imaging specialist can provide a potentially life-saving service.

NEOPLASTIC AND PSEUDONEOPLASTIC

The differential diagnosis of the IAC/cerebellopontine angle mass (CPA) is fairly clear-cut in that there is universal agreement regarding the four most common entities. Schwannoma of the eighth cranial nerve (acoustic neuroma) is the most common, followed by meningioma, arachnoid cyst, and epidermoid (in that order). The next several paragraphs summarize the most important clinical and imaging findings of these entities. Subsequent paragraphs will focus on the less common diagnostic entities.

Schwannoma of the Eighth Cranial Nerve (Acoustic Neuroma)

Overview

Schwannoma of the eighth cranial nerve (acoustic neuroma) is by far the most common CPA neoplasm.[6–9] They are noncalcifying, slow-growing, well-encapsulated lesions of midlife that account for 6% to 10% of all intracranial tumors and 60% to 90% of CPA tumors. They are somewhat more common and tend to be larger in women. There are strong indications that pregnancy may accelerate the growth of this tumor. Schwannomas most commonly present as a combined IAC/CPA lesion; however, they may be entirely intracanalicular in nature. Interestingly, this latter subgroup of lesions is more common in men. Schwannomas arising from the extracanalicular portion of