대한영상의학회지

The CT and Magnetic Resonance Imaging Features of Transotic Schwannoma: A Case Report[1]

경이 신경초종의 전산화단층촬영 및 자기공명영상 소견: 증례 보고[1]

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Transotic schwannoma is a condition extremely rare, and we herein report the CT, MRI, and pathologic findings of a transotic schwannoma case in a 38-year-old woman. The lesion was identified as an expansile mass in the internal auditory canal (IAC) and a mass in the middle ear cavity (MEC) on the high-resolution, bone algorithm, temporal bone CT. It was shown as the following: the hypointense masses of the cerebellopontine angle (CPA)-IAC and MEC, and the hypointense replacement of the normal high-signal intensity fluid of the cochlea, vestibule, and semicircular canals on the 3-dimensional (3D) fast imaging, employing the steady-state acquisition (FIESTA) sequence; and the intensely enhancing masses of the CPA-IAC and MEC, and the intense enhancement of the cochlea, vestibule, and semicircular canals on the 3D gadolinium-enhanced spoiled gradient-recalled echo (SPGR) sequence. The mass was removed via a transotic approach. The pathologic findings were consistent with schwannoma. The assessment of the extent of transotic schwannoma may be enhanced by the 3D FIESTA and the gadolinium-enhanced SPGR sequences.

Index terms
Transotic Schwannoma
Tomography, X-Ray Computed
Magnetic Resonance Imaging

INTRODUCTION

Acoustic schwannoma, the most common benign neoplasm of the internal auditory canal (IAC) and the cerebellopontine angle (CPA), arises from the perineural Schwann cells surrounding the vestibular and cochlear nerves. It most commonly arises near the fundus of the IAC at the Schwann cell-glial junction, but can be also found anywhere along the nerve from the IAC to the terminal ends of the vestibulocochlear nerve, within the vestibule, cochlea, and semicircular canals (1, 2). Intralabyrinthine schwannoma, an uncommon tumor with the likely underestimated incidence rate, develops from the Schwann cells of the intralabyrinthine branches of the vestibulocochlear nerve (1, 2). Recently, the imaging identification of intralabyrinthine schwannoma has increased along with

the use of the higher field magnets and the evolving magnetic resonance (MR) imaging sequences. As the rarest form of intralabyrinthine schwannoma, transotic schwannoma is called when the neoplasm extends through the labyrinth into the IAC and the middle ear cavity (MEC). Transotic schwannoma is extremely rare. Only five cases have been reported in the English literature (1-5), while none has appeared in the Korean radiological literature. We report a case of transotic schwannoma in a 38-year-old woman along with the CT, MRI, and pathologic findings.

CASE REPORT

A 38-year-old woman presented with an aggravated tinnitus and dizziness in the recent 2 days. She had experienced a pro-

gressive hearing loss of the left ear and dizziness for 7 years, along with the intermittent tinnitus and otalgia of the left ear for 1 year. She denied otorrhea, vertigo, or headaches, and her facial expression was intact. The otoscopic examination revealed a mass behind the tympanic membrane. The axial and coronal, high-resolution, bone algorithm temporal bone CT, performed using the Sensation 16 scanner (Siemens Healthcare, Forchheim, Germany) with the slice thickness of 0.8 mm, showed an expansile mass in the left IAC and a small mass in the MEC (Fig. 1A), while the cochlea, vestibule, and semicircular canals appeared to be grossly intact (Fig. 1A). A temporal bone MRI was performed for a better assessment of the extent of the lesion by using a 3.0-T unit (Signa Excite; GE Medical System, Milwau-

kee, WI, USA). The axial 3-dimensional (3D) fast imaging employing the steady-state acquisition (FIESTA) sequence [repetition time (TR)/echo time (TE) = 4.6/1.4; flip angle = 45; matrix number = 384 × 256; number of excitation = 1; slice thickness = 1.0 mm] revealed an approximately 12 × 22 × 10-mm-sized, hypointense mass in the left CPA-IAC, a small hypointense mass in the MEC, and a hypointense replacement of the normal high-signal intensity fluid of the cochlea, vestibule, and semicircular canals (Fig. 1B). The gadolinium (Gd)-enhanced axial and coronal 3D spoiled gradient-recalled echo (SPGR) sequence (TR/TE = 8.2/3.3; flip angle = 20; matrix number = 256 × 224, number of excitation = 2; slice thickness = 1.2 mm) demonstrated the heterogeneous, intensely enhancing masses in the CPA-IAC and
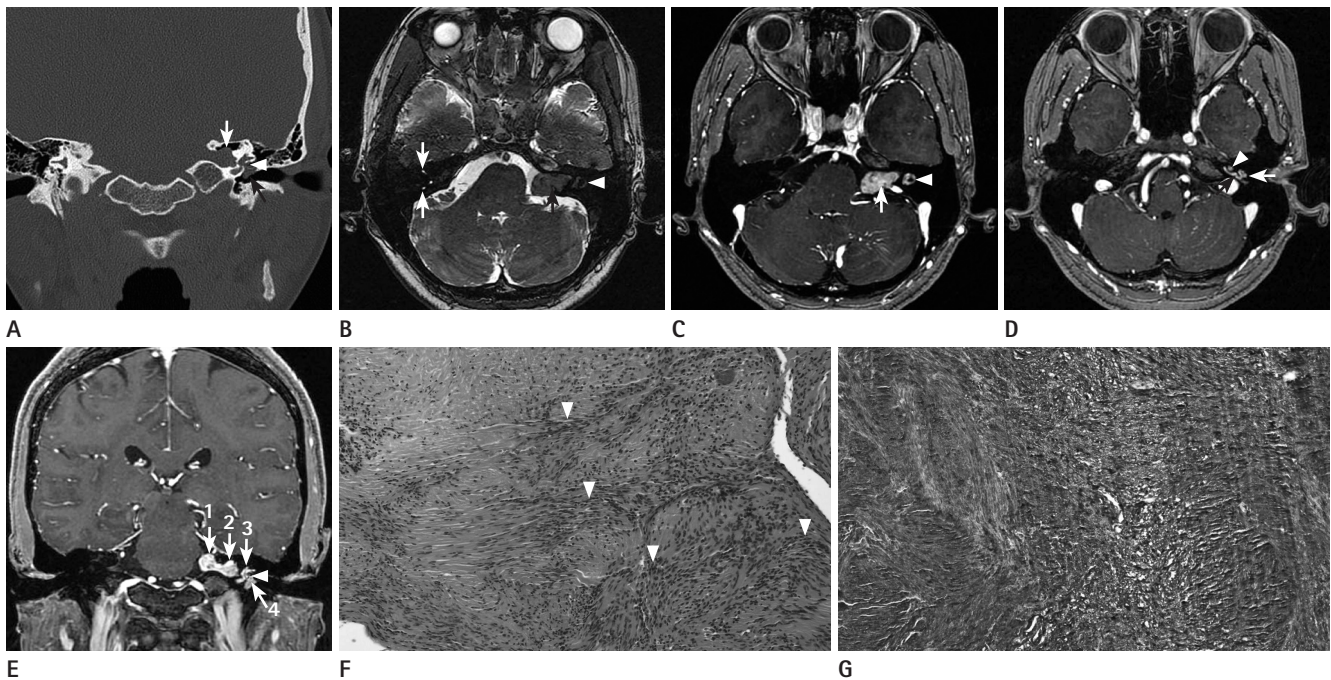

Fig. 1. CT, MRI, and pathologic features of transotic schwannoma in a 38-year-old woman.
A. A coronal high-resolution temporal bone CT image shows an expansile mass in the left internal auditory canal (IAC) (white arrow), and a mass in the left middle ear cavity (MEC) (black arrow) extending from the inner ear through the oval window (arrowhead).
B. An axial 3-dimensional (3D) fast imaging employing the steady-state acquisition (FIESTA) image reveals a hypointense mass in the left cerebellopontine angle (CPA)-IAC (black arrow), and hypointense replacement of normal high signal intensity fluid of the left lateral semicircular canal (arrowhead). Note normal high signal intensity of the right superior and posterior semicircular canals (white arrows).
C. A gadolinium (Gd)-enhanced axial 3D spoiled gradient-recalled echo (SPGR) image demonstrates heterogeneous, intensely enhancing mass (arrow) in the left CPA-IAC, and intense enhancement of the left lateral semicircular canal (arrowhead).
D. A Gd-enhanced axial 3D SPGR image shows an enhancing mass in the left MEC (white arrow) extending from the inner ear through the round window (black arrow). Also noted is intense enhancement of the left cochlea (arrowhead).
E. A Gd-enhanced coronal 3D SPGR image shows the components of transotic schwannoma consisting of a dumbbell-shaped mass of the left CPA (arrow 1)-IAC (arrow 2), intense enhancement of membranous labyrinth (arrow 3), and a mass in the MEC (arrow 4) extending from the inner ear through the oval window (arrowhead).
F, G. A photomicrograph of histological examination (F) of the specimen obtained by surgical resection demonstrates proliferation of the spindle cells in short bundles and nuclear palisading (arrowheads) (hematoxylin-eosin, × 200), and a photomicrograph of immunohistochemical staining (G) reveals strong positivity for S-100 protein in the cytoplasm and nuclei of tumor cells (original magnification, × 200), which are consistent with schwannoma.

MEC, and an intense enhancement of the cochlea, vestibule, and semicircular canals (Fig. 1C-E). The coronal Gd-enhanced 3D SPGR sequence revealed a dumbbell-shaped mass involving the CPA and IAC (Fig. 1E). The extension of the mass from the labyrinth to the MEC was thought to occur through the round and oval windows, which was excellently demonstrated on the high-resolution, bone algorithm temporal bone CT (Fig. 1A) and the Gd-enhanced 3D SPGR sequence (Fig. 1D, E). Under the impression of transotic schwannoma, she underwent a surgical resection of the mass via the transotic approach, and was found to have a tumor involving the CPA, IAC, whole membranous labyrinth, and MEC. The mass extended from the labyrinth through the round and oval windows to the MEC. The histological and immunohistochemical findings of the resected specimen from the CPA, IAC, membranous labyrinth, and MEC were consistent with schwannoma (Fig. 1F, G). The postoperative course was unremarkable with intact facial nerve functions. She was followed-up clinically and with MR imaging without any evidence of recurrence until 22 months after the operation.

DISCUSSION

The management of acoustic schwannoma involves a surgical resection with a goal of preserving the hearing and facial nerve functions. When the schwannoma involves the inner ear, a hearing-preservation surgery is not an option, as removing the tumor from the labyrinth is expected to result in a profound sensorineural hearing loss (SNHL) (1). As such, the accurate preoperative delineation of the tumor extent is prerequisite. Kennedy et al. (1) and Salzman et al. (2) have classified intralabyrinthine schwannoma into six categories: intracochlear (a tumor confined to the cochlea), intravestibular (a tumor centered in the vestibule with or without extension into the semicircular canals), vestibulocochlear (a tumor within the vestibule and cochlea), transmodiolar (a tumor centered in the cochlea with extension through the modiolus into the IAC), transmacular (a tumor centered in the vestibule with extension into the IAC via the macula cribrosa), and transotic (a tumor within the labyrinth with extension into the IAC and MEC). To make the accurate diagnosis of transotic schwannoma, the radiologist needs to carefully interrogate not just the CPA and IAC, but the labyrinth

and MEC in all patients referred for SNHL. Salzman et al. (2) have revealed intralabyrinthine schwannoma by using the high-resolution, thin-section fast spin-echo (FSE) T2-weighted imaging and the Gd-enhanced spin-echo (SE) T1-weighted imaging. The lesions of intralabyrinthine schwannoma were identified as the filling defects with a replacement of the normal high-signal intensity fluid of the membranous labyrinth on the high-resolution, thin-section FSE T2-weighted images, and the focal homogeneously enhancing mass on the Gd-enhanced SE T1-weighted images (2). With the development of the high field magnets and the refined MR imaging sequences, more cases of intralabyrinthine schwannoma may be detected with the MRI. The FIESTA sequence can provide a strong T2 contrast, which emphasizes the cerebrospinal fluid signals. In addition, this sequence has a high signal-to-noise ratio and the inherent flow compensation, and is suitable for the direct 3D imaging (6). Therefore, this sequence can depict small structures surrounded by the cerebrospinal fluid (7) and are thus used to evaluate the posterior fossa lesions, such as acoustic schwannomas (8). We have used the 3D thin-slice, high-resolution FIESTA and the Gd-enhanced SPGR sequences, which may have enhanced the delineation of the extent of transotic schwannoma. In our case, transotic schwannoma was excellently demonstrated as a mass in the CPA-IAC and MEC and a hypointense replacement of the normal high-signal intensity fluid within the membranous labyrinth. These findings corresponded to the intensely enhancing masses in the CPA-IAC and MEC, and an intense enhancement of the membranous labyrinth. Notably, the extension of the schwannoma from the inner ear to the MEC occurred via the round and oval windows, which was excellently demonstrated by the high-resolution, bone algorithm temporal bone CT and the Gd-enhanced 3D SPGR sequence, and was confirmed by the surgical findings.

Intralabyrinthine schwannoma is usually managed by observation with the serial MR imaging. Surgery is indicated for intractable vertigo, extension of tumor to the CPA, or evidence of the tumor growth in patients who are medically fit for surgery. In our case, the surgical resection was performed, because of the extension of the tumor into the IAC, CPA, and MEC. In summary, transotic schwannoma, an extremely rare entity, represents a tumor within the labyrinth with the extension into the IAC and MEC. Extension of the schwannoma from the inner ear into MEC may occur via the round and oval windows. The

thin-section, high-resolution, 3D FIESTA and the Gd-enhanced SPGR sequences under a 3.0-T unit may enhance the assessment of the extent of the transotic schwannoma.

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경이 신경초종의 전산화단층촬영 및 자기공명영상 소견: 증례 보고[1]

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경이 신경초종은 극히 드물다. 저자들은 38세 여자에서 발생한 경이 신경초종 1예의 CT, MRI 및 병리 소견을 보고하고자 한다. 병변은 고해상도 골 알고리즘 측두골 CT에서 내이도의 팽창성의 종괴와 중이의 종괴로 관찰되었고, 3차원 fast imaging employing the steady-state acquisition (FIESTA) 영상에서 소뇌교각-내이도 및 중이의 저신호강도의 종괴와 달팽이관, 전정기관, 세반고리관의 정상적인 물에 의한 고신호강도가 저신호강도로 대치되는 소견으로 나타났으며, 3차원 조영증강 후 spoiled gradient-recalled echo (SPGR) 영상에서 소뇌교각-내이도 및 중이에 강한 조영증강을 보이는 종괴와 달팽이관, 전정기관, 세반고리관의 강한 조영증강으로 관찰되었다. 종괴는 경이적 접근에 의해 제거되었으며, 병리 소견은 신경초종에 합당하였다. 경이 신경초종의 범위를 평가하는데 있어서 3차원 FIESTA 영상과 조영증강 후 SPGR 영상은 그 정확도를 높여줄 것으로 생각된다.

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