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# Evolution of a cochlear schwannoma on clinical and neuroimaging studies

## Case report

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The authors report on a patient with a rare schwannoma that arose from the cochlear division of the vestibulocochlear nerve. Distinctively, the lesion appeared to arise from the cochlea itself and was monitored with clinical and neuroimaging studies for 12 years before it was diagnosed and treated. The atypical occurrence of schwannomas of the vestibulocochlear nerve originating in the inner ear structures underscores the high level of clinical suspicion required for the diagnosis of these lesions in patients presenting with persistent auditory and vestibular symptoms.

**KEY WORDS • acoustic neuroma • vestibular schwannoma • vestibulocochlear nerve • Obersteiner–Redlich zone**

SCHWANNOMAS of the eighth cranial (vestibulocochlear) nerve, misnamed acoustic neuromas, account for approximately 8% of all intracranial tumors and have an annual incidence of approximately 10 per million population.[3,6,8] Most of these tumors grow at a rate of 0.2 to 2 mm/year; 95% arise sporadically and 5% occur in association with neurofibromatosis.[3,6,10] Almost all schwannomas of the vestibulocochlear nerve arise from the vestibular rather than the cochlear division of this nerve. Current opinion favors an equal origin from either the superior or inferior subdivisions of the vestibular nerve in the IAC in close proximity to the vestibular ganglion (Fig. 1).[6,7,10] We report on a patient with progressive tinnitus, hearing loss, vertigo, and hemifacial spasm in whom medical diagnosis was delayed, despite her persistent symptoms and repeated evaluations. Distinctively in this case, the origin and evolution of the tumor, a rare schwannoma of the cochlear division of the vestibulocochlear nerve, were inadvertently recorded in clinical and neuroimaging studies performed over a period of 12 years before eventual diagnosis and surgical treatment.

### Case Report

*History.* This 48-year-old woman was referred to our institution for treatment of an intracranial neoplasm. Twelve years before her referral, the patient had experienced insidious onset of high-pitched tinnitus and concurrent hearing loss in her left ear in the course of 1 year. An MR image of the head, which was obtained at that time (Fig. 2), was reported to be negative for intracranial pathological findings and a provisional diagnosis of Ménière disease was made. Three years later, the patient noted near-complete left-sided deafness and a sensation of fullness or pressure in her left ear. Hydrochlorothiazide therapy was initiated, which she thought relieved the sensation of fullness. Two years later the patient experienced episodes of nausea and unsteadiness, and 1 year after this she suffered her first episode of acute-onset, transient, severe vertigo. Results of repeated MR imaging performed at that time (Fig. 3) demonstrated normal findings. The patient stated that during the subsequent 6 years her left-sided deafness remained stable and that she experienced less severe, intermittent episodes of vertigo. A few months before her referral—that is, 12 years after the onset of her first symptoms—she experienced sudden onset of left hemifacial spasm while running on a trail; the episode lasted for approximately 1 minute. At that time, after her third MR image (Fig. 4), the patient was referred to our institution for treatment of a lesion provisionally diagnosed as a left-sided acoustic neuroma.

*Clinical and Neuroimaging Evaluation.* At the time of our evaluation, the patient was neurologically intact except for complete sensorineural hearing loss on the left side, which was confirmed on audiometry. Intermittent contractions of her left orbicularis oculi muscle were noted during the examination. The patient displayed no cutaneous stigmata of neurofibromatosis. On reviewing her most recent MR image (Fig. 4), a relatively uniformly enhancing mass was seen filling and expanding the left IAC, extending into the left cerebellopontine cistern without compression of the brainstem. In addition, an abnormal signal in the vestibule,

*Abbreviations used in this paper:* IAC = internal auditory canal; MR = magnetic resonance.

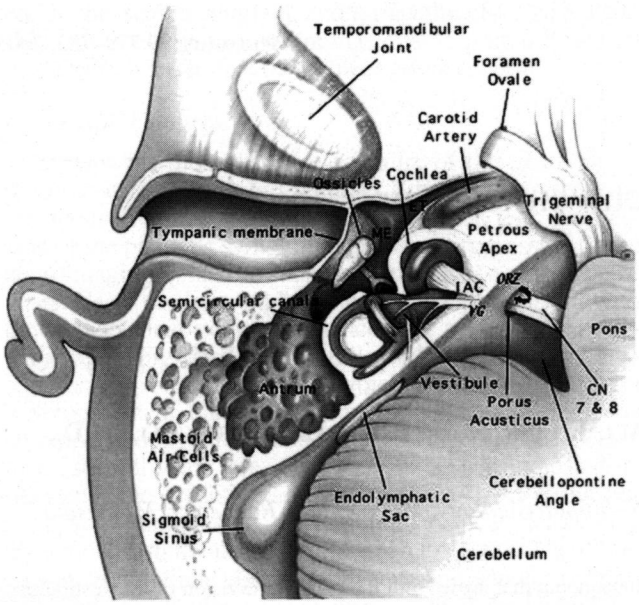


FIG. 1. Drawing showing the anatomy of the vestibulocochlear nerve and the inner ear (axial section). Most acoustic neuromas (vestibular schwannomas) arise from the vestibular division of the vestibulocochlear nerve within the IAC. They are thought to originate from the region of the vestibular (Scarpa) ganglion (VG), in the vicinity of the Obersteiner–Redlich zone ([ORZ], *curved arrow;* see text). Schwannomas of the vestibulocochlear nerve that originate in the labyrinthine vestibule or cochlea are exceedingly rare. CN = cranial nerve; ET = eustachian tube; ME = middle ear. Reproduced with permission from Jackler RK, Driscoll CLW (eds): **Tumors of the Ear and Temporal Bone.** Philadelphia: Lippincott Williams & Wilkins, 2000.

semicircular canals, and cochlea was also noted. On reviewing her original MR image obtained 12 years previously (Fig. 2), a small enhancing lesion was noted in her left cochlea. By the time of her second MR image 6 years later (Fig. 3), the lesion had increased in size to involve her vestibule, semicircular canals, and distal IAC also.

*Treatment and Histopathological Findings.* After placement of appropriate electromyography electrodes for monitoring the fifth, seventh, and 11th cranial nerves, a left postauricular, full-thickness scalp flap was elevated. A wide corticomastoidectomy was then performed in a typical fashion for a translabyrinthine approach. Tumorous tissue was found in and resected from the middle ear, basal turn of the cochlea, semicircular canals, and vestibule. The IAC was surgically unroofed, and after electromyographic confirmation of the location of the intracanalicular portion of the facial nerve, the tumor was debulked from the IAC. The left cerebellopontine angle was exposed and the remainder of the tumor in this region was resected. The origin of the vestibulocochlear nerve was identified at the brainstem and sharply divided; the remaining tumor in the porus acusticus was then removed. Ultimately, complete tumor resection was thought to have been achieved, along with anatomical and physiological preservation of the seventh cranial nerve. The pathological diagnosis in all of the specimens was schwannoma.

*Postoperative Course.* The patient made an excellent recovery and was discharged from our hospital on postoperative Day 5 with no new neurological deficit except for mild (House–Brackmann[5] Grade 2/6) left facial weakness. This resolved within a few weeks of the operation, and at the time of her 3-month clinical follow-up visit, she was asymptomatic and neurologically intact except for the expected left-sided deafness, which was unchanged from before her operation.

## Discussion

The diagnosis of a cochlear schwannoma is unique in our experience. Despite a small number of published cases of this tumor,[2,4,9,11] we were unable to find any similar report

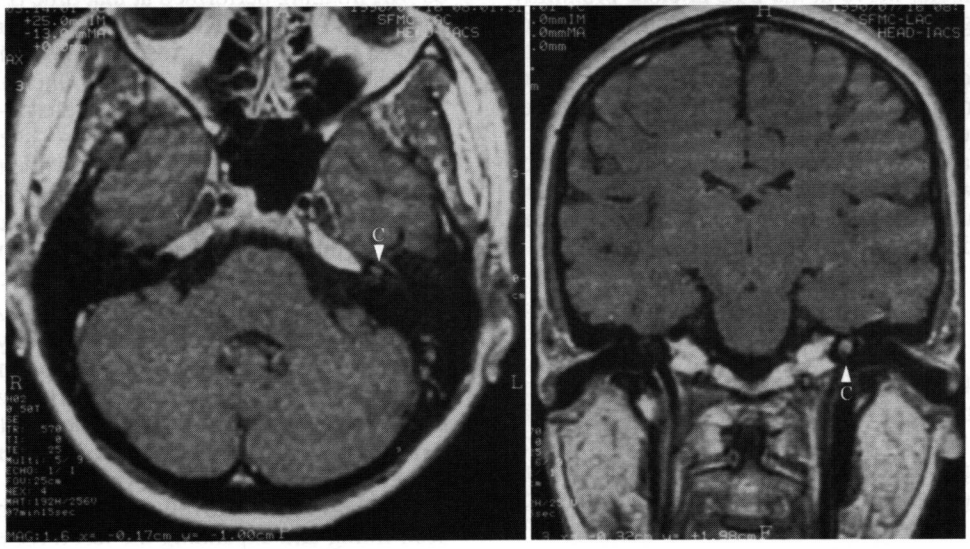


FIG. 2. Magnetic resonance images obtained 12 years before diagnosis. Although the results were reported to be normal, axial *(left)* and coronal *(right)* postcontrast images demonstrate a small enhancing lesion in the left cochlea (C).

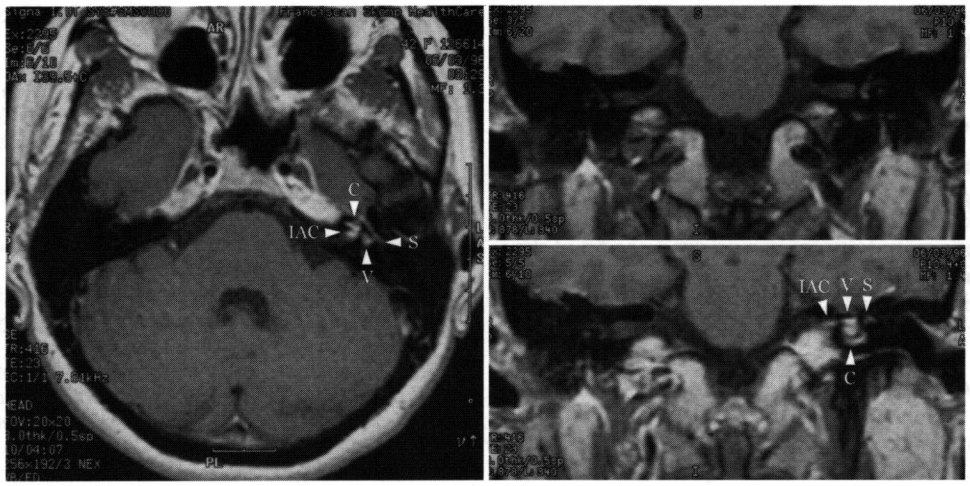


FIG. 3. Magnetic resonance images obtained 6 years before diagnosis. Results of this study were also reported to be normal, although axial postcontrast *(left)* and coronal pre- *(upper right)* and postcontrast *(lower right)* images demonstrate the left-sided lesion involving the labyrinthine vestibule (V) and semicircular canals (S) in addition to the basal turn of the cochlea and distal IAC.

in which the evolution of this rare lesion was documented concurrently on clinical and neuroimaging studies.

*Pathogenesis of the Tumor*

The vestibulocochlear nerve is unusual among cranial nerves in that it possesses a long segment of central myelin. The point of transition between central myelin (derived from oligodendroglial cells) and peripheral myelin (derived from Schwann cells) along the vestibulocochlear nerve is referred to as the Obersteiner–Redlich zone.[6,8,10] This transition zone, which is identifiable histochemically,[12] lies in the IAC approximately 8 to 12 mm from the pial surface of the pons, and most frequently occurs in the vicinity of the vestibular (Scarpa) ganglion (Fig. 1).[7,10,12] Interestingly, the greatest density of Schwann cells along the vestibulocochlear nerve is found in the vestibular ganglion,[12] and it is from this region that most schwannomas of this nerve are thought to arise.[6,8,10] Theoretically, a schwannoma of the vestibulocochlear nerve may originate anywhere along the cranial nerve, from the Obersteiner–Redlich zone to the termination of the Schwann cell sheath at the end organ (vestibular apparatus or cochlea). For the cochlear division of the vestibulocochlear nerve, myelination by Schwann cells begins in the modiolus, just proximal to the spiral ganglion.[1] Thus, intracochlear schwannomas may arise in the region of the modiolus, as was thought to be the case in our patient, based on her earliest symptoms and original MR imaging study (Fig. 2). Note that in the vestibular division of the vestibulocochlear nerve, vestibular fibers are unmyelinated lat-

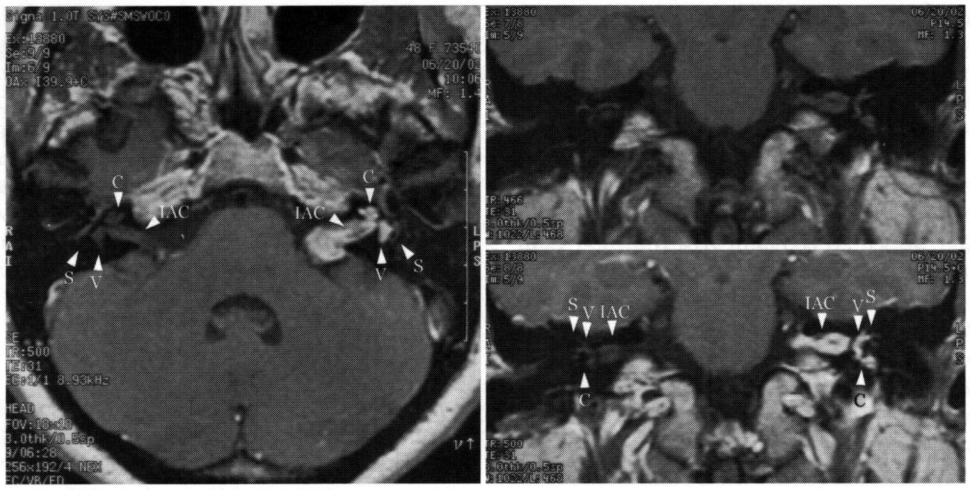


FIG. 4. Magnetic resonance images obtained at the time of diagnosis. Axial postcontrast *(left)* and coronal pre- *(upper right)* and postcontrast *(lower right)* images demonstrate the left-sided lesion filling and expanding the IAC and involving the labyrinthine vestibule and semicircular canals as well as the basal turn of the cochlea. The lesion mushrooms into the cerebellopontine cistern, but does not compress the brainstem. Note the corresponding normal structures labeled on the right side in the *lower right* panel.

eral to the Scarpa ganglion, and it may therefore be reasonable to suspect that schwannomas arising primarily within the labyrinth are in fact of cochlear nerve origin.[1]

*Diagnostic Features*

Perhaps the most distinctive feature of this case, in addition to its being a schwannoma of the cochlear nerve, is the documented evolution of this rare tumor, with progression of the patient's symptoms mirroring progression of the neuroimaging-confirmed abnormalities over a period of 12 years. Early tinnitus and hearing loss were consistent with the intracochlear contrast enhancement seen on the original images (Fig. 2). This was followed by vertigo and dysequilibrium some years later, which were consistent with development of enhancement in the vestibular apparatus demonstrated on subsequent images (Fig. 3). The occurrence several years later of hemifacial spasm indicated irritation of the seventh cranial nerve (Fig. 4). As in our patient, schwannomas of the vestibulocochlear nerve may arise more distally along the IAC than expected. Therefore, cochleolabyrinthine contrast enhancement, which may precede intracanalicular enhancement, should be looked for with MR imaging. We believe that a high level of suspicion for vestibulocochlear schwannoma is required in any patient who has persistent symptoms and signs of vestibular and/or auditory dysfunction.

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