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Cochlear Schwannomas

ABSTRACT—Schwannomas arising solely from the cochlear nerve and limited to the internal auditory canal are rare. Only three prior cases have been specifically described in the literature. We report a 38-year-old male with an asymmetric audiogram and poor discrimination. Magnetic resonance imaging revealed a 3-mm mass occupying the inferior portion of the internal auditory canal. Discrimination improved following a course of steroids, but thresholds did not. The patient underwent a translabyrinthine removal of the tumor, which was confirmed to involve only the cochlear nerve. It is important to obtain evidence regarding the possibility that a tumor confined to the internal auditory canal is a cochlear schwannoma. If so, surgery can be deferred until hearing is no longer functional or tumor growth mandates removal.

Schwannomas within the internal auditory canal (IAC) most frequently arise from the superior vestibular nerve. As early as the 1930s, there were reports of tumors arising from the cochlear portion of the eighth nerve. But reports of true cochlear nerve schwannomas within the IAC, confined only to the nerve, are rare.¹⁻³ We describe an intracanalicular schwannoma confirmed at surgery to involve only the cochlear nerve.

His otologic exam was normal. He had a slight facial asymmetry due to hypoplasia of the right maxilla. His facial nerve function was normal and symmetric. An audiogram demonstrated pure-tone averages (PTAs) of 58 dB HL on the right side and 19 dB HL on the left, with the asymmetry limited to frequencies of 3000 Hz and lower. Word discrimination was 24% on the right on initial examination. Gadolinium-enhanced magnetic resonance imaging (MRI) revealed a 3-mm enhancing mass in the fundus of the right IAC, which appeared to be occupying the inferior portion of the canal. Following a course of steroids, discrimination improved to 72%; however, PTA remained unchanged.

A 38-year-old male reported a right-sided hearing loss for 10 years, which was slowly progressive, associated with fullness and tinnitus, but did not fluctuate. His past history was unusual, with a nasopharyngeal angiofibroma removed on two occasions at the age of 12, followed by radiation therapy.

Given the hearing loss, prior radiation therapy, and the presence of an inferior compartment tumor, a middle fossa approach was not recommended, although it was discussed with the patient. He was given options of either deferring surgery, monitoring his hearing level and obtaining annual MRI scans, or removing the tumor. He

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chose to have surgery; therefore, a translabyrinthine approach was performed. At surgery, the inferior and superior vestibular nerves were found to be normal. Tumor involved only the cochlear nerve.

DISCUSSION

It has long been known that most acoustic tumors arise from the vestibular division of the eighth nerve. Many histologic studies and temporal bone dissections have supported Skinner's⁴ original conclusions regarding the embryological predisposition for tumor development. The vestibular nerve is more inclined to have disordered arrangement of sheath cells at the glial-schwann cell junction.⁵

Schwannomas will occasionally arise from the facial nerve, and many reports describe schwannomas originating within the vestibule or the cochlea. Cochlear schwannomas with involvement of the cochlea, labyrinth, or middle ear are more frequently seen than those confined to the cochlear nerve. We found only three prior reported cases of intracanalicular schwannoma originating from and confined only to the cochlear nerve.¹⁻³ Schlitter¹ first reported such a case in 1930. Thomsen and Jorgensen² described a case which originated from the cochlear nerve and was undiagnosed but found on review of temporal bones. Most recently, Ayani and Shea³ histologically documented a 2-mm schwannoma clearly originating from the cochlear nerve. The patient was asymptomatic from the tumor, which was discovered on routine autopsy.

Asymmetric hearing loss may not occur until the tumor enlarges. In the case described by Thomsen and Jorgensen, the patient was 93 years old and was diagnosed with a "senile sensorineural hearing loss." The mixed loss in the case reported by Ayani and Shea was due in part to stapedial otosclerosis and, presumably, presbycusis, with no significant difference between the two ears in bone conduction. The patient we describe is the only reported case of a clinically diagnosed cochlear schwannoma limited to the cochlear nerve in the IAC.

He had primarily a low frequency, asymmetric loss, with an initial PTA of 58 dB and a discrimination score of 24% in the tumor ear.

Management choices for tumors confined to the cochlear nerve and within IAC depend on several factors, including the age and medical status of the patient and the hearing levels in the involved and uninvolved ears. For some patients who are older or who have poor hearing in the other ear, management by observation will allow the patient the opportunity to use the hearing in the involved ear for as long as possible. Even in older patients who have poor hearing in the involved ear, observation may be the appropriate choice, with periodic imaging to evaluate tumor growth. For the younger patient, the option of observation until the hearing is no longer serviceable should be discussed. In the younger patient with poor hearing, the tumor can be observed until demonstrating increased growth or it can be removed by the translabyrinthine approach. The latter option is dependent upon the judgment of the surgeon and the patient's decision to either watch the tumor further or have it removed while it is still confined to the IAC.

It is important to obtain evidence regarding the possibility that a tumor confined to the IAC is a cochlear schwannoma to provide patients with options about treatment—in particular, the reasonable possibility of hearing preservation. If a cochlear schwannoma is present, surgery can be deferred until hearing is no longer functional or tumor growth mandates removal.

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