CASE REPORT

Vestibular Schwannoma: Unusual Recurrence Presenting as an External Auditory Canal Mass

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ABSTRACT—Vestibular schwannomas (VS) commonly are limited to the internal auditory canal (IAC) and cerebellopontine angle. Extension to labyrinth is less frequent, and involvement of the middle ear or external acoustic canal (EAC) is very rare. In this report we present the case of a 41-year-old woman with a VS, which recurred after a previous surgical removal 4 years before. The tumor involved the IAC, cochlea, vestibule, semicircular canals, cavum tympani, mastoid cells, and EAC. Total removal of the tumor was achieved by a transotic approach, without neurological sequela.

Vestibular schwannomas (VS) (or acoustic neuromas) are the most frequent benign tumors at the cerebellopontine angle. They usually arise at the internal acoustic canal (IAC), although there have been reported cases originated at the labyrinth. Few of these cases had been described as masses presenting beyond the labyrinth, in the middle or external ear. In this paper the authors report a case of VS that presented as a mass in the external acoustic canal 4 years after a previous surgical removal.

CASE REPORT

P.G.L., a 41-years-old woman, was referred by her physician to ENT for consultation because of deafness (June 1993). Initial symptoms included progressive hearing loss and tinnitus in the left ear, recurrent episodes of vertigo and dysequilibrium, facial numbness, and occipital headache for 3 years. Head and neck examination revealed anacusia of the left ear, grade I left beating nystagmus, and corneal hypesthesia of the left eye. Lower cranial nerves were intact. Otoscopy was normal. The patient had some difficulty in walking.

Tympanogram revealed normal compliance and absence of the acoustic reflex on the left ear. A computed tomography (CT) scan with contrast revealed a mass in the left cerebellopontine angle (Fig. 1). With the preoperative diagnosis of VS, the patient was referred to the neurosurgery department of a tertiary center (August 1993). She was surgically treated by an occipital approach, with apparently complete removal of an acoustic neuroma invading the cerebellopontine angle. As a result of the treatment, she developed a left facial paresia grade VI (House–Brackmann).¹ To ameliorate it, a spinofacial anastomosis was performed, allowing partial improvement (grade IV paresia).

For 4 years the patient was asymptomatic and was lost to follow-up. In March 1997 she consulted for persistent discharge of the left ear for 3 months, without any additional symptoms. Otoscopy revealed a pinkish polypoid tissue filling the EAC. It was treated with topical eardrops without any improvement. A CT scan revealed a soft tissue mass affecting the middle ear and the EAC, with bone destruction, but without invasion of the cerebellopontine angle (Fig. 2). Biopsy of the mass revealed to be neurogenous in origin. MRI revealed occupation of the IAC, cochlea, vestibule, middle, and ex-

Figure 1. Preoperative CT scan (June 1993) shows a large mass in the left cerebellopontine angle. Mastoid is free of tumor.

ternal ear with a mass enhanced by gadolinium, without any extension to the cerebellopontine angle (Fig. 3).

With the suspicious diagnosis of a schwannoma recurrence, the patient was submitted to our institution for surgical treatment. For an extensive exposure of the tumor, a transotic approach² (performed by C. Suárez) was performed on May 1997. The tumor filled the IAC (without affecting the cerebellopontine angle), cochlea, vestibule, and semicircular canals. The otic capsule was disrupted and tumor extended into cavum tympani, from mastoid cells to the level of the carotid artery and Eustachian tube. Tumor was removed totally, and the surgical cavity was obliterated with a pedicled temporalis muscle graft. The patient had an uneventful con-

Figure 2. CT scan 4 years after first surgical procedure (March 1997). Recurrence of the tumor with occupation of the vestibule, semicircular canals, attic, cavum tympani, and external ear canal by a soft-tissue mass.

Figure 3. T1 (gadolinium-enhanced) axial MRI (April 1997). Observe the tumor at the lateral portion of the internal acoustic canal, with extension to the labyrinth. Tympanic cavity is filled by a soft-tissue mass without enhancing.

lescence and was discharged from the hospital without any complications. She has been followed for 1 year and has been symptom-free. A CT scan conducted 8 months postoperatively was reported without any evidence of recurrence (Fig. 4).

DISCUSSION

VS are common benign neoplasias and constitute 8–10% of all intracranial tumors.³ Incidence has been estimated about 1 new case per 100,000 population per

Figure 4. CT scan after the transotic approach (January 1998). Surgical defect has been closed with a temporalis muscle pedicled graft.

year,⁴ although histologic studies from autopsies had reported that microtumors are present in 2% of individuals.⁵

VS arise from schwann cells of the vestibular nerves, usually at the IAC. Tumor growth is slow, and usually they extend to the cerebellopontine angle. Lateral extension to the labyrinth is relatively common, but progression to the middle ear is rare. Some authors have reported vestibular schwannomas extended to the middle ear, but most of them were possibly originated into the labyrinth.⁶⁻⁹ To our knowledge, there are only two previously published cases in the English literature of neurinomas presented as a mass filling the EAC, reported by Tran Ba Huy et al¹⁰ and Sekiya et al.¹¹ It is difficult to assess the origin of these large tumors, as the present one. Taking into account that reported isolated intralabyrinthine schwannomas do not extend to the cerebellopontine angle,⁶⁻⁹ they probably arise at the fundus of the IAC.

According to the large volume of the tumor, perhaps removal was incomplete at the initial procedure due to residual tumor at the end of the IAC or because ignored extension to the labyrinth. Tumor progressed into the labyrinth and extended through the oval window to middle ear, mastoid, and EAC. Such a progression in 4 years indicates a high rate of growth, far from the 1–2 mm per year commonly accepted.¹²

There are several surgical approaches to schwannomas removal. By far the most used are the translabyrinthine and the suboccipital/retrosigmoid approaches.¹³ In cases of large tumors with extension to the petrous bone, the transotic approach gives a convenient exposure. The major disadvantage of this approach is the increased risk of facial lesion after the nerve translocation.¹⁴ Due to the previous facial paresia in our patient, this was overlooked and the transotic approach allowed complete removal of a large tumor involving the inner, middle, and external ear.

The present case points the possibility of schwannoma recurrence. A careful preoperative study of extension is mandatory, particularly when a nontranslabyrinthine approach is used (due to ignored labyrinthine involvement). Of note is the possibility of unusual patterns of tumor growth in the petrous bone and the limitations of the different approaches. Although the rate of recurrence is low (about 8%, with the exclusion of the neurofibromatosis-2),¹³ the suboccipital approach seems to be the approach with more recurrences, maybe

due to its limitations in removal of tumors extended into the fundus of the IAC.^{15,16} Thus, despite the approach used, an adequate follow-up is necessary to detect and treat asymptomatic recurrences.¹⁶

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