Atypical Invasion of the Temporal Bone in Vestibular Schwannoma

Abstract—Vestibular schwannomas typically present with erosion of the temporal bone that is limited to the boundaries of the internal auditory canal. Four patients with extensive erosion, excavations, and pseudosatellite defects in the petrous apex are presented. These bony changes extended anteriorly up to the level of the carotid canal. The patients did not have stigmata of neurofibromatosis type 2 and they did not have systemic bony disorders. The tumors did not show any unusual histopathologic changes. Such invasive tumors present an unusual surgical challenge compared to vestibular schwannomas with a more typical pattern of bony erosion. Radiological features and surgical considerations relevant to vestibular schwannomas with this rare presentation are discussed. (*Skull Base Surgery*, 5(1):33–36, 1995)

Vestibular schwannomas cause erosion of the internal auditory canal (IAC) in varying degrees. The factors that determine the extent of erosion of the IAC typically depend on the size of the tumor as well as its location within the IAC. Tumors that arise medially along the course of the eighth nerve could grow to a large size within the cerebellopontine angle (CPA) without causing pressure or erosion of the IAC,¹ while more lateral tumors cause varying degrees of bony erosion before they extend into the CPA. In 1980, Linthicum and Brackmann² reported that bilateral vestibular schwannomas were more aggressive than unilateral tumors and that, in one case, the tumor eroded the tegmen, beyond the confines of the bone of the IAC. To our knowledge, such aggressive biologic behavior has not been described in cases of unilateral vestibular schwannomas.

At the New York Center for Cranial Base Surgery, we have included computed tomographic (CT) scanning in the preoperative work-up of all patients with vestibular schwannoma. While magnetic resonance imaging (MRI) is more useful for the diagnosis of these tumors, we have found that CT scans are helpful in surgical planning. The CT scans led us to identify four cases of unilateral vestibular schwannoma that presented with unusually extensive invasion of the IAC and the temporal bone. The findings in these four cases and their significance are discussed below.

CASE PRESENTATION

Case 1

A 57-year-old female presented with a progressive hearing loss. She was diagnosed as having a 3.4-cm vestibular schwannoma on MRI. On CT scans it was noted that the pattern of erosion of the left IAC was unusual. The anterior wall of the IAC was completely effaced, and the tumor had grown into the petrous apex. The petrous bony invasion was lobular, giving rise to pockets of tumor that extended around the cochlea and within the apex. The medial cortex of the temporal bone was not involved (Fig. 1). The patient did not have any known systemic disease of bone. Plain radiographs of the spine did not show unusual signs of osteoporosis. Her blood profile, including her serum calcium, phosphorus, and alkaline phosphates, was normal.

The transcranial translabyrinthine approach was used for the resection of the tumor, as recently described by the authors.³ It consists of a suboccipital/retrosigmoid exposure combined with a planned dissection of the labyrinth that is performed from within the intracranial cavity, from medially to laterally. It allows the exposure of the lateral extent of the IAC and the facial nerve at Bill's bar. It is interesting that when the temporal bone was being drilled

Figure 1. Case 1. A: The temporal bone erosion (arrow) reaches the level of the cochlea. B: The cavitation in the temporal bone gives the impression of satellite defects (arrows).

and dissected, it was felt that the bone itself was softer than normal. The consistency and color of the tumor were typical of vestibular schwannoma. Laterally in the IAC the tumor was seen to originate in the vestibular nerves. As the dissection was carried out medially, the dissection of the facial nerve presented a particular challenge. Because of the irregular invasion of the temporal bone, the nerve was elongated by the convoluted and lobular shape of the tumor and was stretched over the crests of bone within the petrous apex. The nerve was markedly thin in multiple areas but was kept intact. A small remnant of tumor extended in a bony defect anterior to the facial nerve. It could not be removed without jeopardizing the facial nerve, so the facial nerve was spared. The tumor was examined histologically. Both a frozen-section examination and definitive pathologic examinations had results that were typical of vestibular schwannoma. In particular, there were no abnormal vascularity, mitotic figures, or other malignant features. A pathologic examination of the cortex of the petrous bone was unremarkable. The patient had postoperative weakness of the facial nerve that started to recover slowly at about 3 months postoperatively. At one year postoperatively she had achieved a grade III House-Brackmann facial nerve function.⁴

Case 2

A 38-year-old female presented with a history of headaches of several years and ataxia of a few months duration. She was transferred from another hospital following the placement of a ventriculoperitoneal shunt for hydrocephalus. A neuroradiological work-up demonstrated an aneurysm of the left supraclinoid carotid artery at the level of the ophthalmic artery and a 6-cm right vestibular schwannoma. Bony invasion of the petrous apex similar to case 1 was also noted on CT scans. The erosion involved the pneumatized parts of the temporal bone around the cochlea. The patient did not have any systemic disease of bone. She had no family history or stigmata of neurofibromatosis type 2. Her surgery was performed in two stages. In the first stage, the carotid artery aneurysm was clipped. In the second stage, the vestibular schwannoma was resected via the transcranial

translabyrinthine approach.³ At the lateral end of the IAC the tumor could be separated from the facial nerve and was seen to originate from the vestibular nerves. The gross appearance of the tumor was typical of vestibular schwannoma. The consistency of the temporal bone was felt to be normal. The dissection of the facial nerve within the temporal bone was tedious. The difficulty, as in case 1, was that the facial nerve was compressed against the irregular and lobular anterior wall of the petrous bone. The anterior stretching thinned the nerve and made it transparent. The nerve was kept in continuity despite the large CPA mass. The patient's face was severely weakened and never improved beyond a House-Brackmann grade IV⁴ at 10 months postoperatively. The patient's vestibular schwannoma had normal histopathological features as shown by histopathologic examination.

Case 3

A 77-year-old female presented to our center with progressive gait difficulty of 6 months duration. She had started to use a walker. Ten years prior to her presentation she had undergone a partial removal of a right vestibular schwannoma at another institution. Her facial nerve had been scarified and her face had been rehabilitated with muscle slings. Four years prior to her presentation, MRI showed residual/recurrent vestibular schwannoma with a mild compression of the brainstem. She was advised not to have surgery because of her age. When she finally presented to us, her tumor had grown and caused further compression of the brain stem. A CT scan showed that the temporal bone component of the tumor had excavated the petrous apex, grown around the cochlea, and reached the carotid artery in its petrous portion (Fig. 2). The areas that were eroded by tumor corresponded closely to the pneumatized areas of the opposite temporal bone, as if the tumor had followed the pneumatized tract of the bone leaving the cortices intact. The tumor was resected via a classical transmastoid translabyrinthine approach. At surgery the consistency of the temporal bone was normal. The tumor itself was atypical in that it was divided into two separate components, one within the temporal bone and the other in the CPA. The two components were separated

Figure 2. Case 3. A: The temporal bone is eroded (lower arrow) past the cochlea to the level of the carotid canal (upper arrow). B: Temporal bone erosion anterior and posterior to the internal auditory canal. C: The magnetic resonance imaging appearance of the tumor.

by tenuous fibrous membrane that, we suspect, was scarred from her previous surgery and was continuous with the posterior fossa dura. A total removal was achieved with resolution of the patient's preoperative symptoms. As in the previous cases, the vestibular schwannoma had normal histopathologic features.

Case 4

A 43-year-old male was referred from South America with a 2-year history of hearing loss and numbness of the left face. Magnetic resonance imaging revealed a left vestibular schwannoma that measured 3.5 cm in its medial to lateral dimension. The extension of the tumor within the petrous apex also measured 3.5 cm. The area of the petrous bone that was eroded was symmetrical to the pneumatized bone in the contralateral petrous apex. The posterior lip of the porus acousticus was also eroded (Fig. 3).

The transcranial translabyrinthine approach³ was used for the resection of the tumor. At surgery the consistency of the temporal bone was felt to be normal. The vestibular schwannoma was typical in its color and consistency. The tumor originated very laterally in the IAC from the vestibular nerves, but in this case, the plane between the tumor and the facial nerve could not be followed. With high magnification the tumor could be seen to splay the nerve fibers before completely engulfing and invading it. The tumor was excised completely but the facial nerve

could not be saved, despite the use of intraoperative monitoring. One week later a hypoglossal-facial nerve cross-over was performed. Long-term follow-up could not be done with this patient because he went back to his country postoperatively. On pathologic examination, as in the other three cases, the tumor did not show any unusual features that could explain its aggressive behavior.

DISCUSSION

Valvassori⁵ described the radiological appearance of the IAC in the vestibular schwannoma. On CT scan the bony changes are usually limited to the IAC and consist mainly of flaring of the porous acousticus. The four patients we described presented with a pattern of invasion of the temporal bone that is more extensive than expected in cases of unilateral vestibular schwannoma. Linthicum and Brackmann² reported the invasive behavior of bilateral vestibular schwannomas in four patients. In one of their cases, the tumor, composed of schwannoma and meningioma, had invaded the tegmen anterior to the internal auditory canal. To our knowledge, aggressive erosion of the temporal bone by unilateral vestibular schwannoma has not been previously reported in the English literature.

We retrospectively examined various factors that could have contributed to the excessive erosion of the petrous apex in the patients we described. In all four patients the tumor exhibited typical color and consistency at surgery. On histological examination none of the four

Figure 3. Case 4. A: The petrous apex is invaded. B: A higher magnetic resonance imaging cut showing the tumor. C: A computed tomography view showing erosion of the internal auditory canal.

tumors exhibited excessive or unusual vascularity or any malignant features. It is interesting to note that three out of the four patients were female. We did not test the tumor in our patients for estrogen or progesterone receptors. None of the patients had signs of advanced osteoporosis and all had normal serum levels of calcium, phosphorus, and alkaline phosphatase. The pattern on invasion of the petrous apex was compared to the pattern of pneumatization of the contralateral temporal bone. These patterns were similar in cases 3 and 4. This suggests that when the cortex of the IAC is eroded, the destruction of bone in the petrous apex can actually follow the pneumatized tracts. However, this does not explain why some tumors erode the anterior wall of the IAC in the first place. In cases 1 and 4, patient-related factors may have played a role. The case 1 patient had a soft consistency to her temporal bone. Of course, this is a subjective finding and a biopsy of the bone itself was normal. In case 4, a thick membranous scar had "trapped" and confined the tumor within the temporal bone and separated it from the recurrent tumor in the CPA. This mechanical trapping could have facilitated bony invasion in this specific patient. The dissection of the facial nerve presented a particular challenge in all cases except for the patient in case 3, who presented with preoperative facial nerve paralysis. In case 4, the tumor invaded the facial nerve in a way that was similar to the invasion of the cochlear nerve in cases of bilateral vestibular schwannomas.² In cases 1 and 2, the challenge was related to the irregular distortion of the facial nerve along the invaded petrous apex. The bony edges and crests complicated the dissection of the facial nerve, even in the absence of obvious facial nerve invasion. Additionally, the pseudosatellite defects, as seen in case 1, made it harder to achieve a complete tumor removal. It is interesting to note that when Linthicum and Brackmann² examined the temporal bones of their patients with bilateral tumors they also found residual tumors, even when the operating surgeon had felt that the tumor had been completely resected.

From a retrospective analysis of our four cases, we cannot determine why some unilateral tumor behave in a fashion that is reminiscent of the typically more invasive bilateral tumors. We feel that, in such cases, achieving the goals of total resection of the tumor and preservation of the facial nerve is more challenging than one expects solely from the size of these tumors. Finally, we feel that the radiological appearance of invasive unilateral invasive vestibular schwannomas should not be confounded with the appearance of other primary or metastatic lesions of the petrous apex.

CONCLUSION

The bony changes in unilateral vestibular schwannomas are typically limited to flaring of the IAC. We showed that, in rare instances, these tumors are aggressive and can invade the petrous apex and the facial nerve. At surgery, the uneven erosion of the temporal bone may make it more difficult to follow the plane that separates the tumor and the facial nerve. This may result in facial nerve preservation results that are poorer than can be expected solely from the size of the tumor.

REFERENCES

1. Lo WWM: Tumors of the temporal bone and cerebello-pontine angle. In Som PM, Bergeron RT, Curtin HD, Reede DL: *Head and Neck Imaging*. St. Louis: Mosby, 1991
2. Linthicum FH Jr, Brackmann DE: Bilateral acoustic tumors: a diagnostic and surgical challenge. *Arch Otolaryngol* 106:729-733, 1980
3. Feghali JG, Kantrowitz AB: The transcranial translabyrinthine approach to acoustic neuromas. *Skull Base Surg* 4:132-135, 1994
4. House JW, Brackmann DE: Facial nerve grading system. *Otolaryngol Head Neck Surg* 93:146-147, 1985
5. Valvassori GE: The abnormal internal auditory canal: the diagnosis of acoustic neuroma. *Radiology* 92:449-459, 1969