CASE REPORT

Malignant Vestibular Schwannoma

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Abstract—A 61-year-old woman underwent a translabyrinthine resection of a right intracanulicular acoustic neuroma, which had been detected in the work-up of sudden hearing loss. At the time of surgery, the tumor was roughly twice as large as indicated by the magnetic resonance scan taken only 2 months previously. The tumor eroded the vertical and transverse crests and extended well into the cerebellopontine angle. It was impossible to distinguish the facial nerve proximal to the geniculate ganglion. All visible tumor was resected, along with the facial nerve. Histological evaluation showed a highly cellular tumor, with many mitoses and areas of necrosis, meeting the criteria for malignant schwannoma. The patient has no stigmata of neurofibromatosis, and has no known relatives with that condition. This case is only the fourth reported of a malignant vestibular schwannoma. The relationships between vestibular schwannoma, neurofibromatosis, and malignancy are discussed. (*Skull Base Surgery, 4(4): 227–231, 1994*)

Malignant schwannomas are uncommon neoplasms, occurring most frequently in patients with neurofibromatosis type 1 (von Recklinghausen disease). Thirteen percent to 29% of neurofibromatosis type 1 patients develop a malignant nerve sheath tumor during their lifetime.[1] Only 10% of these malignant schwannomas are located in the head and neck.[2] Even more uncommon is the occurrence of a malignant schwannoma arising from a cranial nerve.[3,4] Roughly one half of these rare tumors arise from the extracranial branches of the trigeminal nerve.

In an exhaustive search of the English literature, only seven reports of previous *intracranial* malignant nerve sheath tumors were located. Three of these lesions were malignant vestibular schwannomas.[5–7]

Vestibular schwannomas (acoustic neuromas) are generally felt to be benign tumors, and under certain circumstances, some authors have suggested a conservative nonoperative approach.[8–10] We present the fourth case of a *malignant* vestibular schwannoma, in a patient where the initial presentation was sudden unilateral hearing loss. Previous cases of malignant acoustic neuroma are summarized, and an attempt is made to incorporate consideration of these rare tumors into the general management of sudden hearing loss and of acoustic neuromas.

CASE REPORT

Patient Presentation

A 61-year-old woman with previously known symmetric sensorineural hearing loss presented with a right-sided sudden hearing loss, associated with imbalance. An audiogram revealed a severe-profound loss in the right ear (Fig. 1). Electronystagmography revealed a right vestibular weakness. Smooth pursuit, saccades, and optokinetic responses were normal. There was no nystagmus.

After nonresolution of her imbalance, a gadolinium-enhanced magnetic resonance imaging study was obtained. This demonstrated an enhancing lesion filling the right internal auditory canal, extending medially into the cerebellopontine angle cistern without brain stem compression (Fig. 2). Based on her presentation and magnetic

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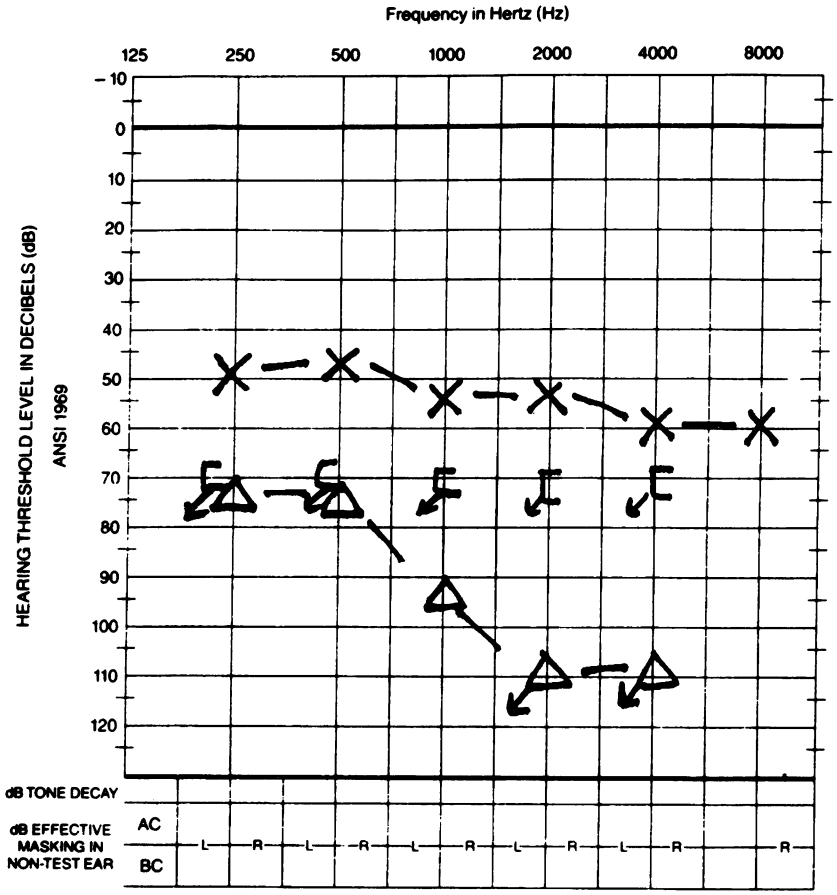
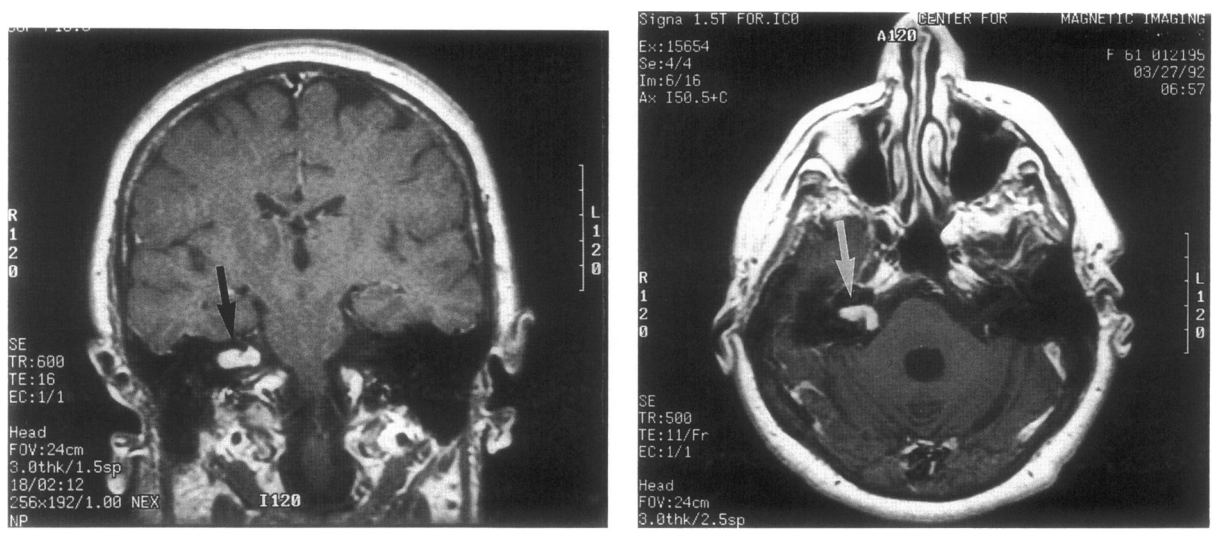


Figure 1. Audiogram obtained following right sudden hearing loss (presenting complaint) in a patient with a malignant vestibular schwannoma.



A B

Figure 2. T1-weighted magnetic resonance imaging with gadolinium contrast showing mostly intracanalicular tumor (arrows). A: Coronal. B: Axial.

resonance imaging findings, the working diagnosis of a solitary vestibular schwannoma was given.

Surgical Procedure

Two months later a translabyrinthine approach for excision of the tumor was performed. At surgery the lesion was much larger than anticipated from the preoperative magnetic resonance imaging. The transverse and horizontal crests were effaced. The tumor was found to *invade* the facial nerve, and there was significant brain stem compression. All visible tumor was resected, along with the facial nerve. Intraoperatively, a frozen section was returned as benign spindle cell neoplasm consistent with vestibular schwannoma.

Final histopathologic evaluation demonstrated a cellular, spindle cell neoplasm with nuclear pleomorphism, frequent mitotic figures, and numerous areas of individual cell necrosis (Figs. 3–6). Reticulin stain demonstrated abundant reticulin fibers, particularly around the periphery of the lesion. No positive fibers were seen coursing through the tumor. The Vimentin and S-100 stains were positive. The glial fibrillary acidic protein stains were negative. Basing our conclusions on this evaluation, we characterized the lesion as a low-grade malignant vestibular schwannoma.

Clinical Course

Additional family history revealed no known relatives with neurofibromatosis. The patient had an uneventful postoperative recovery, and a 2-year follow-up magnetic resonance imaging revealed no evidence of recurrent or persistent tumor. The facial paralysis was treated with a gold weight implanted into the upper eye lid and facial slings.

DISCUSSION

Woodruff et al[11] described the characteristics of a malignant nerve sheath tumor and gave criteria needed for this diagnosis: (1) more than a rare or isolated mitosis, usually accompanied by nuclear pleomorphism and increased cellularity, and (2) invasion by the tumor into a previously nonoperated field. Only one of these criteria was necessary to separate the malignant tumors from the more common "cellular schwannoma." The tumor described in this report fulfills both criteria.

The clinical characteristics of the three previously reported cases of malignant acoustic neuroma (as well as of the present case) are summarized in Table 1. Only two of these patients were diagnosed as having a malignant vestibular schwannoma after the initial attempt at surgical extirpation. The other two tumors were determined to be malignant from the histology of the recurrence. None of these patients fit the criteria for neurofibromatosis types 1 and 2. One patient was a 2-year-old child, and this tumor should probably be considered separately from the other three. The remaining patients were more than 50 years of age, and all presented with hearing loss as the initial symptom. The two previously reported adults had large tumors, and rapid regrowth occurred following initial attempts at excision. Both patients were dead of their disease within a few months. Our patient presented with a smaller tumor, underwent presumed complete excision (although not en block), and as of this writing shows no evidence of recurrence.

Some authors have advocated conservative nonsurgical management of acoustic neuromas, especially in older patients.[9,10] Slow growth rates[12-14] as well as occasional spontaneous tumor "involution"[15,16] have been reported. Others have argued against the nonoperative management of these tumors.[8,17,18] Our patient and the two other patients with reported malignant vestibular schwannomas

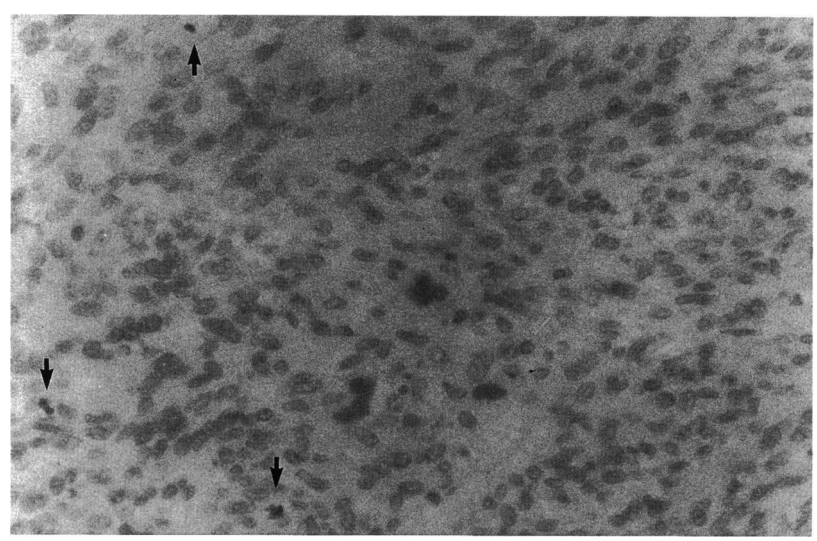


Figure 3. Photomicrograph of malignant vestibular schwannoma demonstrating marked cellularity and numerous mitotic figures (arrows) (hematoxylin and eosin, ×130).

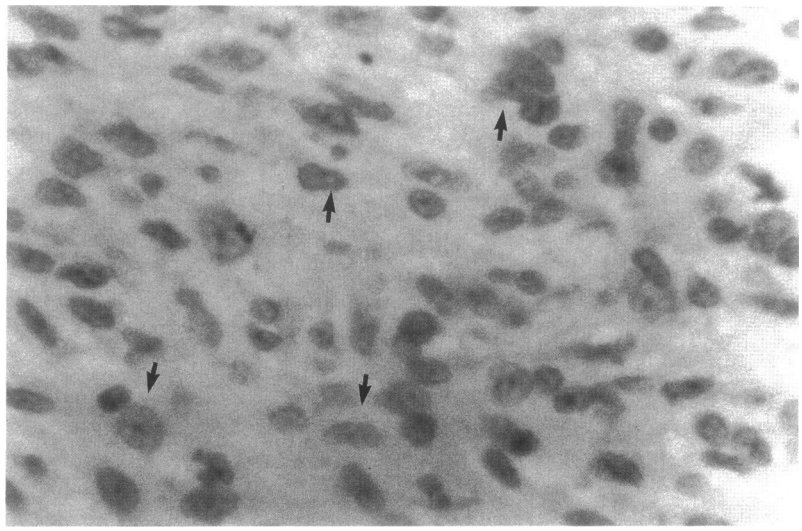


Figure 4. Photomicrograph of malignant vestibular schwannoma demonstrating nuclear polymorphism (arrows) (hematoxylin and eosin, ×260).

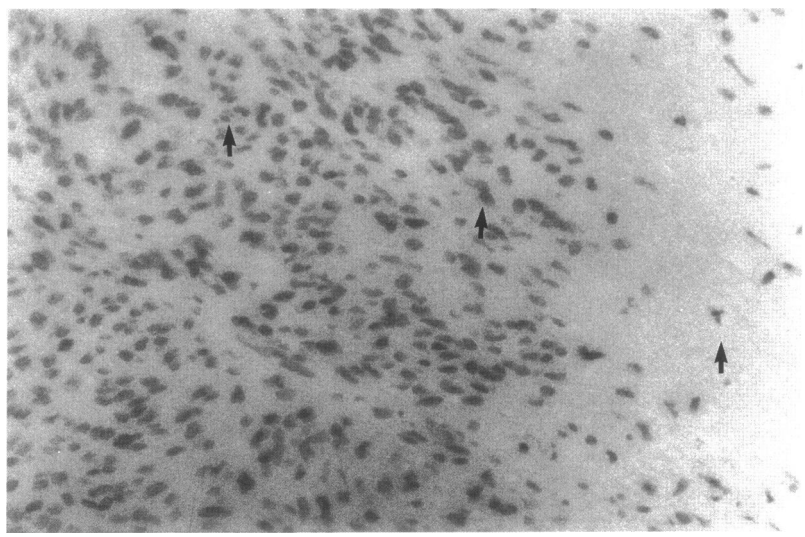


Figure 5. Marked single-cell necrosis is seen in this photomicrograph (arrows) (hematoxylin and eosin, ×130).

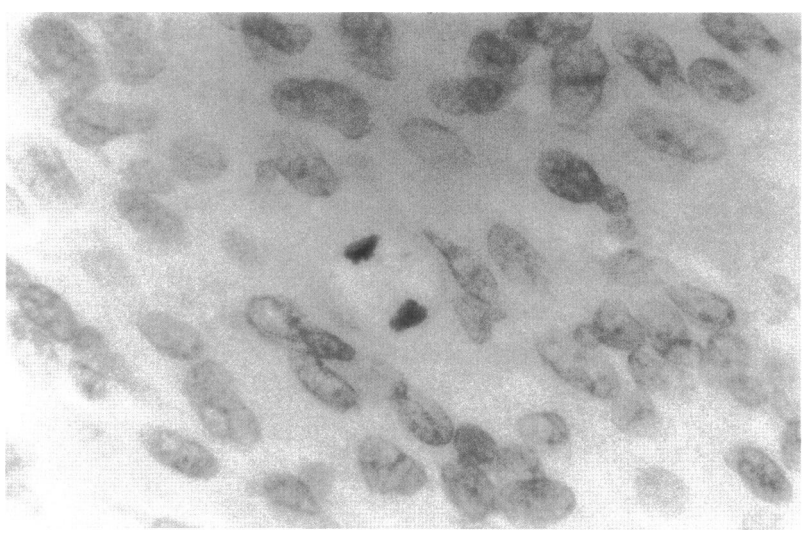


Figure 6. High power image demonstrating a mitotic figure (hematoxylin and eosin, ×650).

Table 1. Summary of Reported Cases of Malignant Vestibular Schwannomas

Reference	*Age*	*Sex*	*Presenting Symptoms*	*Course*
7	22 mo	F	Hearing loss Facial paralysis	2.2-cm tumor Two operations No evidence of disease 1-year follow-up
5	54 y	M	Hearing loss	Massive tumor Rapid regrowth following surgery Dead of disease <1 y
6	61 y	M	Hearing loss Facial hypesthesia	4-cm tumor Massive regrowth following surgery Dead of disease <1 y
Present case	61 y	F	Sudden hearing loss	1.5-cm tumor Translabyrinthine excision No evidence of disease 1 year

were over the age of 50. Conservative management may not be appropriate in all cases.

Although only 1% to 2% of patients with sudden sensorineural hearing loss are subsequently found to have vestibular schwannomas, up to 26% of acoustic neuromas present as sudden hearing loss.[19] The present case emphasizes the importance of a thorough and timely evaluation, as well as a definitive diagnosis, in all patients with sudden hearing loss. If our patient's sudden hearing loss had been treated casually, the tumor might not have been detected until it had grown to substantial size, making the surgical treatment more difficult and possibly resulting in a less favorable outcome.

As malignant vestibular schwannomas are exceedingly rare, any discussion of treatment must extrapolate data from other malignant schwannomas. For those lesions, wide local excision, with or without adjuvant chemotherapy or radiation therapy, is recommended. This surgical approach, however, is not feasible in the cerebellopontine angle. Additionally, it is unlikely that the malignant nature of the lesion will be known preoperatively. The role of adjuvant radiation therapy for malignant vestibular schwannomas is undetermined. It seems reasonable that the use of radiation would be indicated in subtotal resections or recurrent lesions. The morbidity and high mortality rate of these tumors (50% dead of disease within one year) is due to local disease, not distant metastasis.

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