

TITLE: Acoustic Neuroma and Hearing Loss

SOURCE: Grand Rounds Presentation, UTMB, Dept. of Otolaryngology

DATE: December 6, 2006

RESIDENT PHYSICIAN: K. Kevin HO, M.D.

FACULTY PHYSICIAN: Vicente A. Resto, M.D., Ph.D.

SERIES EDITORS: Francis B. Quinn, Jr., MD and Matthew W. Ryan, MD

"This material was prepared by resident physicians in partial fulfillment of educational requirements established for the Postgraduate Training Program of the UTMB Department of Otolaryngology/Head and Neck Surgery and was not intended for clinical use in its present form. It was prepared for the purpose of stimulating group discussion in a conference setting. No warranties, either express or implied, are made with respect to its accuracy, completeness, or timeliness. The material does not necessarily reflect the current or past opinions of members of the UTMB faculty and should not be used for purposes of diagnosis or treatment without consulting appropriate literature sources and informed professional opinion."

Introduction:

This manuscript serves to review the evidence-based literature on hearing loss in acoustic neuroma (AN) patients. The focus will be on the natural history of acoustic neuroma and how hearing progression may be influenced by factors such as tumor growth.

Historical Perspectives:

The first documented case of AN was recorded in 1777 by Dr. Sandifort. The mortality rate associated with surgery was alarmingly high at that time up to 80% due to delayed diagnosis and primitive instrumentation. Major progress in the surgical management of AN was made during the early twentieth century owing to the contribution of three surgeons. Dr. Harvey Cushing employed meticulous dissection and hemostasis, successfully lowering surgical mortality to 4%. His student, Dr. Walter Dandy, further refined technique with use of vessel clips and ligatures. Dr. Dandy was also the first surgeon to perform a complete resection of AN. Dr. William House introduced operating microscope and surgical drills to revitalize the once-condemned translabyrinthine approach in the 1960s. In addition, Dr. House introduced the concept of combining the expertise of neurosurgeons and otologists in the management of AN patients. The current state-of-the-art micro-instrumentation and intraoperative neural monitoring have enabled surgeons to achieve complete tumor removal with a very low mortality rate (<2%).

Anatomy of the Cerebellopontine Angle (CPA):

The cerebellopontine angle is a rather small area located in the posterior fossa near the origins of several vital cranial nerves. The medial and lateral borders of this space are the brainstem and the petrous portion of the temporal bone, respectively. Superiorly, it is bounded by the middle cerebellar peduncle, and inferiorly by the arachnoid tissue of the lower cranial nerves. Posteriorly, the cerebellar tonsil limits this space, as does the clivus anteriorly. Within this space lie several cranial nerves, including portions of VII – XII, as well as the CSF of the quadrimenial cistern, the arachnoid tissue of the above cranial nerves, and several blood vessels, most notably the anterior inferior cerebellar artery.

Acoustic Neuroma:

Epidemiology:

AN represents about 6% of all intracranial tumors and 80-90% of all CPA masses. Its incidence in the States is about 10 cases per million every year. It predominantly presents in adulthood. There are two forms of AN: 1) the sporadic form represents about 95% of all ANs and it presents unilaterally; 2) Neurofibromatosis type 2 (NF-2) has a strong genetic predisposition on chromosome 22, presenting as bilateral ANs. There is currently no known gender and race predilection in the development of AN. AN is a benign tumor arising from the vestibular nerve (hence it is also known as vestibular schwannoma). Majority of ANs arise from the internal auditory canal (IAC).

Hearing in AN patients:

Hearing loss is the most common symptoms of AN and can be found in about 95% of AN patients. Classically, AN patients present with asymmetric sensorineural hearing loss (SNHL) at high frequencies (down-sloping). Speech discrimination scores (SDS) typically decrease out of proportion of pure tone thresholds (PTT).

According to the American Academy of Otolaryngology-Head and Neck Surgery classification published in 1995, patients falling into categories of either class A (>70 % SDS and < 30 dB PTT loss) or B (>50% SDS and < 50 dB PTT) have serviceable hearing. Hearing of Class C (> 50% PTT and >50% SDS) and class D (< 50% SDS) patients are considered non-serviceable.

The etiology of hearing loss in AN is not known. Some proposed mechanisms include compression on the cochlear nerve, vascular disruption of the internal auditory artery, and biochemical changes in inner ear fluids.

The importance of following hearing progression in AN patients cannot be overstated. Preoperative hearing status has been shown to be the most important predictor for the success of hearing preservation surgery (middle fossa or retrosigmoid approach). A number of studies addressing the relationship between hearing progression, tumor size, and tumor growth are described in the following sections.

Tumor size:

AN can be classified by size according to the Jackler's system: 1) Intracanalicular; 2) small < 10 mm; 3) medium 11-25 mm; 4) large 25-40 mm; 5) giant > 40 mm.

It was once thought that tumor size might be correlated with hearing progression in AN. Lustig identified a higher proportion of smaller tumors in the normal hearing group compared to his overall AN group. However, he found that hearing could be normal even in larger tumors > 3 cm. A study by Stipkovits also failed to identify any relationship between hearing and tumor size. To date, there exists no conclusive correlation between tumor size and change in hearing threshold in AN patients.

Tumor Growth:

Four phases of tumor growth have been described: intracanalicular, cisternal, compressive, and hydrocephalic. Generally, patients in intracanalicular phase display unilateral loss of vestibular or cochlear nerve function, such as hearing loss, vertigo, tinnitus, and disequilibrium. These symptoms progress as tumor grows and expands into the CPA cistern. Cranial neuropathies especially CN V and VII typically occur late in the compressive phase. Lastly, obstruction of the fourth ventricle results in hydrocephalus and can be seen in patients with visual changes and altered mentation.

Numerous papers have addressed the natural history of AN by following tumor growth in patients in the observation, or wait-and-scan group. Despite the retrospective nature and intrinsic selection bias in most of these studies, they illustrate similar results that roughly 50% of ANs grow, 40-50% remain in same size, and less than 10% involute.

The relationship between tumor growth rate and hearing was explored by Massick et al., who found a positive correlation between changes in tumor volume, pure tone average as well as speech discrimination score.

An understanding of tumor growth rate is instrumental for the clinical management of AN. Walsh demonstrated that the risk of loss of serviceable hearing in the observation period is much higher in the tumor growth group (80%) compared to the non-tumor growth group (14%). Undoubtedly, the current management of AN – observation, surgery, and radiation therapy – all hinges upon the concept of tumor growth rate. A non-growing AN that is associated with stable or minimal change in hearing is likely to be observed, while the fast growing ones with progressive deterioration of hearing would be resected or irradiated. Most integral to the success of radiation as a treatment of AN is to arrest tumor growth, which occurs in no more than 10% after radiation. Various studies have attempted to identify predictors for growth of AN without much success. There was no correlation between growth rate and patient age, gender, initial volume, and side of tumor. While potential biomarkers such as fibroblast growth factor receptor may provide an answer to tumor proliferation in the near future, the only reliable means to estimate tumor growth at present is serial MRI scanning as described below.

Diagnosis and assessment of AN:

Timely diagnosis of AN is critical because treating smaller tumors either by surgery or radiation carry a smaller risk of morbidity and higher chance of hearing preservation compared to larger tumors. While the frequencies and patterns of symptoms in AN have been thoroughly described in the literature, we must also be aware of the variations and often insidious disease progression. Patients often ignore the early and non-specific symptoms of hearing loss (~ 94%), vertigo (~39%), disequilibrium (~ 56%), or tinnitus (~64%), resulting in the delay of diagnosis. A complete otolaryngologic and neurologic exam must be performed. A high index of suspicion should be given to those with progressive unilateral SNHL, unilateral audiovestibular symptoms, facial or trigeminal nerve dysfunction, and sudden SNHL.

As described above, AN patients typically present with asymmetric SNHL with decreased SDS. Therefore, a thorough audiometric assessment is a crucial first-step in

diagnosing AN. Audiogram may also demonstrate features of loss of acoustic reflex and acoustic reflex decay that are associated with retrocochlear pathology. The diagnostic efficiency of Auditory Brainstem Response (ABR) has been studied extensively. The sensitivity of ABR has been reported to be > 95% in large tumors > 3 cm. However, the false positive rate has also been described to be as high as 33 % in intracanalicular AN and false positive rate > 80%. More importantly, the response of interaural latency difference for wave V (IT5) is absent almost half the time in large tumors > 2 cm. Because of these limitations, studies on a new technique called Stacked ABR are currently underway to improve its diagnostic efficiency. Otoacoustic emissions (OAE) and vestibular testing (eg. Electronystagmography, rotatory testing, posturography) have little role in the diagnosis of AN because of low sensitivities and specificities.

Gadolinium-enhanced MRI remains the gold standard in the diagnosing AN. It can detect tumors as small as 1 mm and differentiate AN from many CPA lesions based on its density and enhancement on T1 and T2 images. Imaging on temporal bone lesions has been previously described by Cowan et al. in our Grand Rounds Series and will not be discussed here. For those patients unsuitable for MRI scanning (eg. Metallic implant), a contrast-enhanced CT can be obtained, though it cannot reliably detect smaller tumors < 1.5 cm.

References:

1. Bailey BJ et al. Otolaryngology-Head and Neck Surgery 2006
2. Jackler RK et al. Tumors of the ear and temporal bone. 2000
3. Jackler RK et al. Neurotology. 2005
4. Myrseth: Neurosurgery, Volume 59(1).July 2006.67-76
5. Lustig LR. Am J Otol 1998: 19; 212-8
6. Stipkovits EM et al. Am. J. Otol 1998: 19; 834-9
7. Battaglia et al. Otol Neurotol. 2006 Aug;27(5):705-712
8. Massick DD. Laryngoscope 2000: 110; 1843-9
9. Herwadker A. Otol and Neurotology 2005: 26; 86-92
10. O' Reilly BF et al. Otol Neurotol 2000: 25; 791-6
11. Fraysse B et al. First International Conf. on Acoustic Neuroma. 1992
12. Gordon ML. American Journal of Otol 1995; 16: 136-9
13. Chandrasekhar SS et al. Am J Otol 1995;16:63-7
14. Don M et al. Am J. Otol 1997: 21; 148-151
15. Kim AH. Otol Neurotol. 2006 Apr;27(3):372-9
16. Raut V et al.: Clin Otolaryngol 29:505-514, 2004.
17. Rohit MS et al. Ann. Oto. Rhino. Laryng. 2006: 115 (1); 41-6
18. Walsh RM et al. Laryngoscope 2000: 110; 250-5