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REVIEW ARTICLE - TUMOR - SCHWANNOMA

Hearing preservation in small acoustic neuroma: observation or active therapy? Literature review and institutional experience

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Abstract

Background Current imaging modalities enable early diagnosis of a large number of small acoustic neuromas in patients with well-preserved hearing, whose best management is still a debatable matter.

Methods Comparing the hearing outcome of the various therapeutical approaches, including observation, radiosurgery, or resective surgery, is not easy because of the numerous classifications measuring hearing in different ways. In this review, a literature review was performed and papers selected dealing with small tumors, short- and long-term hearing, tumor control or radical resection, and effect of pre-treatment hearing and size on outcome. Two different surgical institutional series of sporadic vestibular schwannomas provided us comparative data on the outcome of observation alone.

Results and conclusions Our experience suggests that active treatment with hearing preservation surgery, unlike observation alone, offers a better chance of hearing preservation, also enabling a more effective treatment of the tumor and an appropriate rehabilitation with hearing aids or cochlear implants.

Keywords Acoustic neuroma · Hearing preservation · Observation · Radiosurgery · Hearing preservation surgery

Introduction

Imaging enables today the early diagnosis of a large number of small acoustic neuroma in patients with well-preserved hearing, whose best management is still a debatable matter. Different therapies are equally feasible, providing they all offer similarly good results in terms of control of disease and overall neurological and facial nerve losses. The problem is therefore how to prevent hearing loss or rehabilitate any loss caused by the tumor or its treatment. The importance of preventing and/or rehabilitating unilateral deafness is motivated by the fact that it is associated with a severe loss of quality of life, especially in adults, because of a consequently impaired speech recognition in noise, lack of directionality of sound, and daily fatigue.

This article is part of the Topical Collection on *Tumor—Schwannoma*

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Comparing the hearing outcome of the various therapies is not easy because the current classifications measure hearing in different ways. The Gardner-Robertson classification [5], which is the most often used in the fields of neurosurgery and radiotherapy, measures hearing as the average of the three thresholds at frequencies of 500, 1000, and 2000 Hz. The AAO-HNS American Academy of Otolaryngology, Head and Neck Surgery [1] (AAO-HNS) considers the average of the thresholds at 500, 1000, 2000, and 3000 Hz in the American otological community, and at 4000 Hz in Europe. PTA stands for pure tone average, and this value is coupled with the ability to discriminate speech, or SDS (speech discrimination score) to classify hearing as: class A, good hearing = PTA $\leq$ 30 dB, SDS $\geq$ 70%; class B, serviceable hearing = PTA $\geq$ 50 dB, SDS $\geq$ 50%; class C, measurable hearing = any PTA, SDS $\geq$ 50; class D, poor hearing = any PTA, SDS < 50.

The Tokyo [10] classification is a further evolution: the PTA was obtained from 500, 1000, 2000, and 4000 Hz, and more strict classes of PTA were considered (A to F) combining with the corresponding speech discrimination score (SDS). For class C and lower classes, an upgrading to the immediately preceding class is done when the SDS belongs to a better class than the PTA. The AAO-HNS and Tokyo classifications are considered and used in the present work.

Methods

The PubMed ResearchGate was investigated about hearing preservation surgery in vestibular schwannoma, in the years between 2005 and 2018. Only the retrosigmoid procedures were considered. Key words were used: "Retrosigmoid approach," "vestibular schwannoma," "hearing preservation surgery," "hearing preservation and vestibular schwannoma." Over the mentioned period, papers were filtered and selected on the bases of the availability of data about (1) number of patients (only series with more than 20 patients were considered); (2) preoperative A-B hearing class according to the AAO-HNS classification; (3) overall rates of postoperative class A-B hearing preservation (series with hearing preserved in worse classes than A-B were excluded); and (4) when available, postoperative outcome of class A-B as it related to small tumors (these data were not present but in two authors [4, 7, 9, 17, 19, 22, 23].

Results

The hearing preservation outcomes of the chosen representative papers were reported in Table 1. All the cases, as defined in the inclusion criteria, were preoperative class AB. In our series [17, 19], data referred to preoperative class A.

The reported hearing preservation rates were defined as "overall" when referring to any preoperative size of tumor. When available, data on "small size tumor" were reported. Overall rates ranged from 21.4 to 85% within the preoperative conditions of class A-B hearing, disregarding tumor size. Only few [17, 19, 22] papers considered class A-B hearing preservation rates in small tumor, which ranged from 39 to 85%. A small tumor was < 1.5 cm [6] or < 1 cm [17, 19] in cerebellopontine angle (CPA) according to the different series.

Discussion

Observation, or abstaining from any active therapy, is believed nowadays to offer the best chance of preserving long-term hearing. It can also be advantageously combined with the fact that the majority of small tumors spontaneously stop growing. But such a wait-and-see policy is clearly associated with two issues: hearing inexorably declines and, even when it is partially preserved, it is of poor quality. As emerged in a representative paper [13], observation for intrameatal tumors is associated with a progressive hearing decline over the years, with a long-term good hearing preservation rate of 17% at 10 years. In the most favorable condition of patients with intrameatal tumors and a SDS of 100%, hearing was reportedly preserved in 73% of cases at 10 years, with a SDS > 70%, but there is no mention of the sound level at which this speech discrimination was achieved. According to the AAO-HNS classification, the reported [11] PTA of 46 should coincide with a class B, which means a serviceable hearing.

Whether an early diagnosis can avoid the patient, any further hearing loss with currently available therapies remains to be seen. The fate of hearing function has been the object of inconclusive debate in recent papers. "*Early* risk of deafness is likely to be greatest with microsurgery, followed by radiosurgery and observation, but the greatest number of serviceable hearing years comes with observation compared with proactive treatment" [2, 3]. HPS has "predictable and satisfactory results" [19]. "The postoperative preserved hearing can be considered *durable* hearing preservation as it is unlikely to change markedly" [6]. Small tumors in patients with good hearing are "preferably observed" and only submitted to radiotherapy in the event of growth [19] (comments to 18). The spontaneous arrest in the tumor's growth and the long-term hearing preservation enabled by observation alone "makes it the benchmark with which every other therapy should be compared" [13]. The present report moves from this comparison recommended by Kirchmann [13].

Table 1 Selected papers (see "Methods" for "inclusion criteria")

Author	Year	Patients (only RS approach)	Overall postop A-B class (AAO-HNS) (%)	Small T post op A-B class (AAO-HNS) (%)	Comments
Hilman et al.	2010	26	38.5	ND	
Sameshima et al.	2010	43	76.7	ND	
Mohr et al.	2005	128	24.2	39% (< 1.5 cm)	
Han et al.	2010	18	61	ND	
Di Maio et al.	2011	28	21.4	ND	Only large tumors
Mazzoni et al.	2011	322 (96)[a]	63	83% (< 1-cm extrameatal size)	
Mazzoni et al.	2018	100[b]	67	85% (< 1-cm extrameatal size)	

[a] 96 refers only to preop class A cases

[b] 100 refers only to preop class A cases

The Danish study [13] on observation reported that good hearing (class A) was preserved at 10 years in 17% of cases (5/29), and class AB hearing in 34% (25/75 cases). The hearing loss in these two groups amounted to 83% and 67%, respectively. The subgroup of 27 cases with SDS 100% at diagnosis retained a remarkably better long-term hearing, with an average PTA of 46 dB and a SDS of 73%, which coincides with AAO-HNS class B.

Radiotherapy appears to be effective in stopping tumor growth [8], with a still unknown rate of success in growing vs non-growing tumors [14, 21]. Hearing is preserved in the short term, but declines inexorably in the longer term, with only a 23% rate of preservation of good hearing at 10 years [2, 3] (24 and 12% at 10 and 15 years, respectively [24]) according to recent reports. Hearing preservation surgery (HPS) is attributed to such different success rates [11] that compare it with other therapies; it is possible if the subcategory of small tumors is considered [12, 16, 20].

Institutional experience

Two different series of sporadic vestibular schwannomas provided us with the data for comparing with the outcome of observation alone [17–19]. Our latest series of 100 consecutive cases of HPS [19], with good hearing at the diagnosis, preserved postop class A hearing (AAO-HNS) in 53% of cases, and a good or serviceable hearing (postop class AB, AAO-HNS) in 85% of cases, both groups in patients with *favorable* parameters at diagnosis. These *favorable* parameters were tumors less than 10 mm in CPA and a hearing function of at least 30 PTA and 70 SDS (class A AAO-HNS). This outcome has a minimum follow-up of 2 years. The long-term outcome (5–10 years) is unavailable as yet. In a previous study on the long-term outcome of HPS in a group of 117 cases followed up for 6 to 21 years (mean 14, median 9 years), the loss of class A-B hearing [1] occurred in 15% of cases, and the loss of class A in 8%.

The recent series of our ongoing experience [25] included all the currently available therapies, i.e., observation (O), radiotherapy (RT), HPS, and conventional surgery, which were used according to the guidelines in Table 1. HPS replaced O when it had a high probability of success, e.g., in patients with the mentioned good hearing at diagnosis. As the choice of therapy was based both on our institutional guidelines and on the patient's preference, homogeneous O and HPS groups were available for comparison. Hearing was measured according to the Tokyo classification [10], which we consider representative of the quality of hearing function. There were 86 patients with tumor size up to 10 mm in the CPA. O was adopted in 75% of cases, HPS in 20%, excision via a translabyrinthine approach in 5%. Radiotherapy was never indicated as a first choice at diagnosis, but only during the follow-up. Four outcomes are meaningful for purposes of the comparison considered here: the outcome of O, including failures leading to active therapy; the results of HPS; the outcome of secondary RT; and the time course of hearing function in O vs HPS. The O approach failed due to tumor growth in 28% of cases, over a mean period of 22 months (range 9–48 months): 25% were switched to active treatment (surgery in 20%, radiotherapy in 5%), while O continued for 3%. The tumors submitted to RT showed no further growth, but the only case with good hearing at diagnosis was no longer in class A after 27 months. In the group under O with class AB hearing (24 cases), a decline in hearing was evident in 42% of cases (10 cases). No correlation emerged between hearing loss and tumor growth, but in the first 2 years of follow-up; the cumulative hazard of hearing impairment related to tumor growth was 4.8% for tumors remaining the same size, and 12.5% for growing tumors in the first year, and 11.9% and 22.2%, respectively, in the second. The group of cases treated with HPS, as a first choice or after a period of O, had two different outcomes depending on tumor size and hearing function. The favorable factors were a tumor less than 10 mm in size in the CPA, and a hearing function of at least PTA 30/SDS 70, with a normal or slightly altered ABR (auditory brain response). The factors coincided with the preservation of good hearing (class AB) in 77% of cases, and an overall 85% of cases in which a good or serviceable hearing (class ABC) was preserved. The failure rate was 15%, with poor hearing or deafness. The subgroup whose hearing function and tumor size were outside the abovementioned favorable limits had a success rate of 33%.

As for the time course of the decline in hearing function in the O vs HPS groups:

- the patients with good hearing submitted to O showed a hearing deterioration rate of 42% after a mean follow-up of 25 months (range 9 to 48 months); in other words, class AB hearing was maintained in 58% of cases.
- among the cases in the HPS group with favorable factors (tumor size and hearing function), failures (class C-F) occurred in 23% of cases, while 77% preserved a good hearing function in the short term (postoperatively). After a mean 2 years of follow-up (range 9 to 78 months), one patient's hearing had deteriorated (from class A to C), and the rate of preserved class AB hearing was 69%, and the loss of class A was 10%.

These two groups were homogeneous in terms of hearing parameters, but the choice of O or HPS was made by the patient (not randomly).

In principle, O afforded a better hearing preservation rate than HPS in the short term because of the early hearing loss experienced in the cases where surgery failed. The progressive decline in hearing function under O gradually led to much the same rate of hearing loss for both O and HPS over time: after 2 years of follow-up, the rate of preservation of a good hearing function in the cases with favorable factors was 58% in the O

Table 2 (modified from Martini et al. [15]). Sporadic small acoustic neuroma: hearing-focused management

Acoustic neuroma size (mm in the CPA angle)	Decision factors	Treatment
< 10 mm	Good hearing (< 30 dB, > 70% SDS, normal or slightly modified ABR)	Hearing preservation surgery or observation[a]
	Good hearing (< 30 dB, > 70% SDS, normal or slightly modified ABR) + surgical risk /unwillingness to undergo surgery	Observation[a]
	Poor hearing (> 30 dB, < 70% SDS)	Observation[a] or surgery (+ hearing rehabilitation with cochlear implant)

[a] Active treatment (surgery or RT) in the event of tumor growth to > 15 mm, or vertigo, or VII cranial nerve impairment

group and 69% in the HPS group. The surgically preserved class AB cases maintained their hearing function class in 90% of cases at 2 years. A longer follow-up would be needed to assess the degree of further hearing decline in the O and HPS groups, but is not yet available.

Comparison of O and HPS was here carried out between a universal therapy like O and a variable one from center to center like HPS. The retrosigmoid approach with retrolabyrinthine meatotomy (RLM) [19] was used by us. The rationale of the RLM is both the favorable control of the full internal auditory canal and wide, multiangled exposure for instrument handling as obtained with two steps. First, the occipital craniotomy of the RS approach is extended posteriorly as to afford an oblique view which bypasses the bulge of the labyrinth and controls the canal up to its fundus. It also allows an improved view of the nerve-tumor interface for dissection. Second, the petrous bone is widely removed around the canal allowing room for multiangled view and handling of instruments and tissues.

Conclusions

Imaging offers the chance of an early diagnosis of usually small acoustic neuromas, and their management challenges the general assumption that an early diagnosis means a better prognosis. There is a general consensus that the currently available therapies offer similarly good results concerning neurological losses, facial nerve preservation, and cure or control of the tumor, but hearing function generally suffers a more or less severe deterioration. Hearing loss, or deafness, is therefore the residual morbidity confronting the therapy.

Within the limitations of a clinical study weakened by referral and selection biases, the comparison between the O and HPS groups shows that hearing function takes a different course under these two treatment options.

- Observation offers a better hearing function in the short term (because HPS is burdened with immediate postoperative hearing loss in the event of failure).
- The course of hearing deterioration is inexorable under observation as it reflects the natural history of the disease. The outcome of HPS is more durable, despite some decline in hearing function over time. The hearing/time diagram for observation vs HPS shows a convergence after around 2 years, then the decline in hearing function continues at a faster rate under observation. The difference between the two options becomes considerable at 5 and 10 years.
- Patients with a SDS of 100% at diagnosis and submitted to observation maintain high levels of speech recognition [14], although their mean PTA of 46 designates an AAO-HNS class B, i.e., a serviceable rather than a good hearing.
- Good hearing in the long term appears to be due to a good hearing at the postoperative baseline and a modest time loss in HPS; this is an uncommon event in O since hearing shows considerable loss with time. Hearing remains serviceable only in patients with an intrameatal tumor and a normal (100% SDS) speech discrimination ability.

By now, our experience has reached shared indications (Table 2) [25] which suggest that active treatment with HPS, unlike observation alone, can offer a better chance of hearing preservation. HPS also enables a more effective treatment of the tumor and an appropriate rehabilitation with hearing aids or cochlear implants.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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