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The Changing Landscape of Vestibular Schwannoma Diagnosis and Management: A Cross-Sectional Study

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Objectives: To assess the current state of the diagnosis and management of vestibular schwannoma (VS) as well as treatment trends, and to evaluate the role of treatment setting and various specialists in treatment plan.

Methods: Patients diagnosed with VS completed a voluntary and anonymous survey. The questionnaires were distributed through Acoustic Neuroma Association website, Facebook page, and e-mail newsletters from January to March 2017.

Results: In total, 789 VS patients completed the survey. Of those, 414 (52%) underwent surgery; 224 (28%) underwent radiotherapy; and 121 (15%) were observed. General otolaryngologists diagnosed 62% of responders, followed by primary care (11%) and neurotologists (10%). Patients who underwent surgery were significantly younger and had larger tumors compared to those treated with radiation or observation. The ratio of patients having nonsurgical versus surgical resection changed from 1:2 to 1:1 for the periods of 1979 through 2006 versus 2007 through 2017, respectively. Neurosurgeons (40%) and neurotologists (38%) were the most influential in treatment discussion. Neurotologists ($P < 0.001$) and general otolaryngologists ($P = 0.04$) were more influential than neurosurgeons for the decision process in patients with smaller tumors. Patients treated at academic versus nonacademic private institutions reported similar tumor sizes ($P = 0.27$), treatment decisions ($P = 0.09$), and decision satisfaction ($P = 0.78$).

Conclusion: There is a continuing trend toward nonsurgical management, with approximately half of the patients opting for nonsurgical management. In this cohort, the patients commonly presented with otologic symptoms and otolaryngologists made the most diagnoses. Neurotologists and neurosurgeons were the most influential in treatment discussion.

Key Words: Vestibular schwannoma, acoustic neuroma, treatment trend, influential specialist, decision satisfaction.

Level of Evidence: NA

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INTRODUCTION

Vestibular schwannomas (VS) are benign tumors arising from the eighth cranial nerve and comprising 6% to 8% of all intracranial tumors.[1,2] Whereas the annual incidence of 1.1 to 1.7 per 100 thousand people has remained steady, tumors have been diagnosed at smaller sizes over time.[3–6] After diagnosis, VS patients are faced with a multifaceted decision-making process to manage their tumor via watchful observation, radiotherapy, or surgical resection. Retrospective studies of large cohorts demonstrated that 48% to 59% of patients underwent microsurgery, and 21% to 24% underwent radiotherapy, with surgical resection correlating with younger age and larger tumor size.[3,5,6] With innovations in imaging and radiotherapy, some authors have begun to advocate for the benefits of observation and radiotherapy over surgical treatment.[3,7–10] However, a review of VS treatment demonstrated that selection bias and confounding factors, such as age and pretreatment status, limit comparing efficacy and safety across treatment modalities.[11]

It is yet unclear whether the rise in advocacy for conservative management is independent from the earlier detection of these tumors at smaller sizes. Both diagnosis and treatment of VS are continuously evolving toward greater collaboration between neurotologists, neurosurgeons, and radiation oncologists, among others.[12–14] However, the specific roles and influence of these providers in diagnosis and treatment discussion have yet to be compared. This patient-centered study aims to assess the current state of diagnosis and management of VS, as well as to evaluate the role of treatment setting and various specialists' influence in treatment plan and satisfaction.

MATERIALS AND METHODS

In collaboration with the Acoustic Neuroma Association (ANA) and after institutional board approval from the University of California, Irvine, a comprehensive and anonymous survey was distributed to all ANA members from January to March 2017 using a secure and confidential REDCap interface (Vanderbilt University, Nashville, TN).[15] The link to the survey was available on the ANA website https://www.anausa.org/ and Facebook page https://www.facebook.com/ANAssociation/; in addition, notification was sent via e-mail to listed ANA members. A VS diagnosis was required to participate in the study. Survey

From the Department of Otolaryngology–Head and Neck Surgery (K.G., M.A., O.M., R.S., A.S., H.W.L., H.R.D.); and the Department of Biomedical Engineering (R.S., H.R.D.), University of California, Irvine, Irvine, California, U.S.A.

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questions were mostly in the following formats: free response, choosing the best answer, or checking all that apply. The survey evaluated patients' demographics, diagnosis information including involved physicians and VS tumor characteristics, treatment modality with the associated course and outcome, treatment centers, and influential specialists contributing to treatment.

Patients' eventual treatments were categorized into surgery, radiation, watchful observation, surgery–radiation combination, and undecided. The latter two treatments were excluded from some analyses due to a low response rate. Surgery cohort consisted of patients undergoing complete or incomplete resection; radiation cohort consisted of patients receiving stereotactic radiosurgery or radiotherapy; and watchful observation patients underwent serial magnetic resonance imaging (MRI) scans. In this article, we define conservative (nonsurgical) management as either radiation or watchful observation. Because most questions were optional to answer, not all number breakdowns in tables add up to 100% of the respective cohort.

PASW Statistics 18.0 software (SPSS Inc., Chicago, IL) was used for statistical analysis, with a 0.05 alpha considered significant. Independent sample *t* test and chi-square test were used for numerical and categorical variables, respectively. Univariate analysis of variance (ANOVA) was used to compare numerical variables among different cohorts; those that demonstrated a significant *P* value were further analyzed for subset comparison via Bonferroni's correction. Lastly, when comparing the treatments of patients diagnosed since 2007 versus those prior to that, the potential confounding effect of their tumor size at diagnosis was determined via a bivariate analysis using binary (surgery or conservative) or multinomial (surgery, radiation, or observation) logistic regression.

RESULTS

Demographics and Diagnosis

In total, 789 individuals diagnosed with VS completed the survey, with an average age at diagnosis of 52.0 ± 11.8 years. Patients' demographics and different treatment modalities are summarized in Table I. The tumor size diagnosed by different specialists and eventual treatments are compared in Table II. One-way ANOVA showed a significant difference between specialists in diagnosed tumor size ($P = 0.003$). Multiple comparison analysis revealed that tumors diagnosed by general otolaryngologists ($P = 0.012$) and neurotologists ($P = 0.004$) were smaller than those diagnosed by neurosurgeons. There was no difference in treatment decision (i.e., surgery or nonsurgical management) when comparing patients diagnosed by different specialists ($P = 0.607$).

Whereas 60 (7.6%) patients had their tumors found incidentally, 726 (92.0%) patients obtained MRI due to concerning symptoms on presentation. The most common symptoms were hearing loss (n = 559, 70.8%), imbalance/dizziness (n = 304, 38.5%), tinnitus (n = 295, 37.4%), aural fullness (n = 232, 29.4%), and vertigo (n = 152, 19.3%). Overall, 704 (89.2%) patients experienced at least one of these symptoms.

TABLE I.
Vestibular Schwannoma Patients' Demographics.

Treatment Modality	N (%)	Tumor Size (SD) in cm	Age at Diagnosis (SD)	Female Sex (%)	Years Since Diagnosis (SD)
Surgery	414 (52.5)	2.38 (1.33)	48.6 (11.3)	285 (68.8)	8.4 (8.4)
Radiation	224 (28.4)	1.72 (1.10)	55.6 (11.1)	139 (62.1)	6.3 (5.1)
Observation	121 (15.3)	1.12 (0.88)	57.2 (10.3)	73 (60.3)	5.4 (5.1)
Surgery + radiation	14 (1.8)	2.98 (1.31)	42.4 (13.0)	9 (64.3)	5.2 (2.9)
Undecided	16 (2.0)	1.70 (1.18)	56.9 (7.5)	11 (73.3)	3.5 (5.6)
Total	789 (100)	2.02 (1.28)	52.0 (11.8)	517 (65.5)	7.2 (7.1)

SD = standard deviation.

Treatment Trend

One-way ANOVA showed that the surgically treated patients were younger and had larger tumor sizes compared to those treated with radiation (both $P < 0.001$) or observation (both $P < 0.001$). In Table III, participants diagnosed between 2007 and 2017 (n = 580) were compared to those diagnosed prior to 2007 (n = 209) regarding tumor size at diagnosis and eventual treatment. Tumor size at diagnosis was smaller in the 2007 through 2017 cohort compared to the 1979 through 2006 period (1.87 vs. 2.33 cm, $P = 0.013$). The former cohort underwent surgery in 47.1% (n = 273) of cases versus nonsurgical management in 48.1% (n = 279) of cases. This was significantly different than those diagnosed between 1979 and 2006, with 66.0% (n = 138) surgical and 31.1% (n = 65) nonsurgical patients ($P < 0.001$). When we adjusted for tumor size as a possible confounder, there still was a significant difference between nonsurgical versus surgical management of the two time periods ($P = 0.002$).

Specialists and Treatment Centers

During the course of VS management, 700 (87.8%) patients had seen a neurosurgeon, 549 (68.9%) a neurotologist, 307 (38.5%) a radiation oncologist, and 39 (4.9%) a general otolaryngologist. Patients were asked which specific specialist played the most influential role in the discussion and decision of treatment. Of the 759 responders, 301 (39.7%) designated neurosurgeons, 287 (37.8%) neurotologists, 77 (10.1%) general otolaryngologists, and 50 (6.6%) radiation oncologists. Average tumor sizes were 2.34 ± 1.39, 1.79 ± 1.13, 1.88 ± 1.22, and 1.87 ± 1.26 cm, respectively ($P < 0.001$). Post hoc analysis showed that neurotologists ($P < 0.001$) and general otolaryngologists ($P = 0.04$) were most influential in smaller tumor sizes compared to neurosurgeons.

Of those who specified their treatment setting (n = 684), 411 (60.1%) patients received treatment at academic institutions, 259 (37.9%) at private nonacademic medical centers, and 14 (2.0%) at the Veteran Administration hospitals. There were no significant differences in treatment decision ($P = 0.09$), tumor size at diagnosis ($P = 0.27$), days of hospitalization ($P = 0.05$), or discussion of most influential specialist in treatment ($P = 0.36$) between academic and private nonacademic centers' patients. On a scale of 1 to 5, there was no difference ($P = 0.78$) in satisfaction, with treatment decision between receiving treatment at academic (3.87 ± 1.1) versus private nonacademic (3.85 ± 1.1) institutions.

TABLE II.
Initial Specialist Diagnosing Vestibular Schwannomas and Respective Frequency, Average Tumor Size, and Eventual Treatment Modality.

	Diagnosing Physician					
N (% of 749*)	Otolaryngologist 486 (61.6%)	PCP 85 (10.8%)	Neurotologist 81 (10.2%)	Neurosurgeon 69 (8.7%)	Neurologist 28 (3.5%)	*P* Values
Tumor size (SD)	1.94 (1.28)	2.15 (1.41)	1.72 (1.06)	2.50 (1.28)	2.21 (1.16)	0.003
Treatment modality						0.372
Surgery	253 (52.1%)	39 (45.9%)	43 (53.1%)	33 (47.8%)	18 (64.3%)	
Radiation	131 (27.0%)	24 (28.2%)	20 (24.7%)	25 (36.2%)	5 (17.9%)	
Observation	77 (15.8%)	13 (15.3%)	15 (18.6%)	5 (7.2%)	3 (10.7%)	

*749 is the number of responders that participated in this question. Undecided and surgery-plus-radiation patients were excluded from treatment modality's analyses.

PCP = primary care physician; SD = standard deviation.

TABLE III.
Comparison of Patients Diagnosed Within Last 10 Years Versus Those Diagnosed Previously Regarding Tumor Size and Eventual Treatment.

	Diagnosing Window		
	2007–2017	1979–2006	*P* Value
N (% total)	580 (73.3%)	209 (26.2%)	
Tumor size (SD)	1.87 (1.24)	2.33 (1.41)	0.013
Treatment modality			<0.001*
Surgery	273 (47.1%)	138 (66.0%)	
Radiation	179 (30.9%)	45 (21.5%)	
Observation	100 (17.2%)	20 (9.6%)	

Undecided and surgery-plus-radiation patients were excluded from treatment modality's analyses.

**P* value remained significant after adjusting for size as a potential confounder via multinomial binary logistic regression ($P < 0.001$).

SD = standard deviation.

DISCUSSION

This patient-centered study of VS patients demonstrates that there is a trend toward nonsurgical management even when adjusted for the decreasing tumor size at diagnosis. The patients mostly presented with otologic symptoms and were diagnosed by otolaryngologists. Neurosurgeons and neurotologists were perceived as the most influential in discussing treatment, and patients reported no meaningful differences between the treatment institutions.

Diagnosis

The common clinical symptoms experienced in VS patients are reported as hearing loss (57%–95%), tinnitus (12%–83%), vertigo/dizziness (14%–75%), and trigeminal neuropathy (4%–9%).[16–20] These wide ranges are partly due to the heterogeneity of tumor size and location (intracanalicular vs. mostly in cerebellopontine angle). Hearing loss was seen in most of our cohort (71%) who underwent initial diagnostic MRI, but tinnitus and imbalance/dizziness (both 38%) also represented a significant number of complaints leading to imaging. It is important to note that our five most common presenting symptoms were all otologic-related. This is in agreement with our breakdown of diagnosing physicians, in which otolaryngologists and neurotologists were identified as having diagnosed more than 70% of the VS cases.

We also found that tumors diagnosed by neurosurgeons were larger than those diagnosed by neurotologists and general otolaryngologists. It may be because VS starts manifesting itself with otologic symptoms and thus naturally draws the patient to otolaryngologists, as opposed to neurosurgeons who may receive referrals after a large enough tumor has led to enough mass effect for neurologic symptoms or an emergency department setting. Although not significant, we observed that patients diagnosed by neurosurgeons received slightly higher rates of radiosurgery and lower rates of surgery despite the fact they were shown to have larger average tumor size. This might be due to some neurosurgeons' higher level of comfort with radiosurgery versus surgical resection or the setting of their practice.

Treatment

Participants who underwent surgical resection were younger and had larger tumor sizes, which is consistent with other studies.[3,5,6] Our mean age at diagnosis (52.0 years) was comparable to a large epidemiological study (53.1 years),[5] but our average tumor size (1.12 cm) was consistent with that reported from a large meta-analysis (1.18 cm).[8] When separating the cohort into recently diagnosed (2007–2017) patients and those diagnosed prior to 2007, we observed a different treatment breakdown. Namely, nonsurgical management has become more prevalent from a previous 1:2 (nonsurgical to surgical) ratio to the 1:1 of the recent decade. This is consistent with the current body of literature suggesting the consideration of observation and radiation therapy for the appropriate tumors, which is due to earlier detection, more diagnoses in elderly patients with indolent tumors, better monitoring, limited number of regrowth in VS tumors, and avoiding surgical-related complications.[3,9,10,21–24]

The cohort diagnosed in 2007 through 2017 had a smaller tumor size at diagnosis, but we showed that there was a persisting trend toward nonsurgical management even after adjusting for tumor size. It has been suggested that tumors ≤3 cm in the cerebellopontine angle are potential candidates for radiosurgery and that observation can show success for slow-growing and smaller VS

tumors, especially <2 cm total length.[3,9,22,25–27] A portion of the surgical patients in this study, especially those diagnosed prior to 2007, fall within these ranges. This suggests that some of these patients could have benefitted from a discussion regarding conservative management as a viable option. Alternatively, it is also important to point out recent emerging evidence suggesting a possible association of poor long-term hearing outcomes with conservative management. Evidence from modern, conformal, low-dose radiation techniques demonstrated poor long-term hearing preservation rates, from 80% to 23% in 2-year and 10-year posttreatment hearing preservation rates, respectively.[28] Likewise, a 2018 study not included in this review showed a similar trend in 466 conservatively managed patients, demonstrating a drop of 94% to 44% in 1-year and 10-year serviceable hearing maintenance rates (defined as pure tone average ≤50 dB hearing loss and word recognition score ≥50%).[29] A 2019 literature review and institutional experience of hearing preservation surgery on small vestibular schwannomas concluded that, although observation may have better short-term hearing function, active surgical treatment can offer a better chance of long-term hearing preservation.[30] With the trend toward smaller tumor size at diagnosis, all treatment options—including risks and benefits associated with each—should be discussed with patients whose tumor characteristics renders them candidates for such approaches.

Influential Specialists and Treatment Centers

Our data showed that neurosurgeons were closely followed by neurotologists as the most influential specialists in treatment discussion. Otolaryngologists or neurotologists who were most influential in almost half of the patients had patients with smaller tumor sizes compared to those diagnosed by neurosurgeons. Of those who specified their treatment setting, 60% were treated at academic institutions compared to 38% receiving treatment at private non-academic institutions. A geographical analysis of access to cancer care centers suggested that 50% and 75% of the U.S. population live within less than 30 minutes of academic and private facilities providing cancer care, respectively.[31] This implies that, although more people are likely to live close to a private institution, most of our cohort sought treatment at an academic center. Studies suggest that many baseline confounders may be present, including age, socioeconomic status, ethnicity, baseline morbidity, or hospital caseloads, rendering simple comparison challenging.[32–34] Regardless, we demonstrated that this study's patients receiving care at academic and nonacademic private institutions had similar eventual treatment distribution and satisfaction with this decision.

Limitations

Although great effort was taken to ensure the validity of our findings, this study relied on retrospective self-reported data and lacked a control group. Moreover, diagnosis and management may be influenced by factors and confounders beyond the scope of this study, such as patients or physicians' subjective preferences, access to care, and outside influencers like online resources or family and support groups. The patient–physician relationship and the perception of bedside manners, knowledge, and time spent can vary from case to case as well.[35] Participation bias may also play a role because ANA members active in the online community were more likely to access and spend time completing the survey. Also, it is plausible to consider that patients with either poor or excellent outcomes may be more inclined to participate. The ANA members may also be socioeconomically different than the general population of VS patients. Lastly, some of the participants completed the survey a long time after their initial diagnosis and treatment, which can lead to recall bias. In addition to recall bias, this difference of range of time from diagnosis/treatment to survey participation can influence the diagnosis and treatment regimens because the standard-of-care guidelines may be determined or changed by the time period. Regardless of these limitations, this study's large cohort and extensive questions can offer additional insight, especially from the patients' perspective.

CONCLUSION

We found that patients diagnosed with VS from 2007 through 2017 had smaller tumor sizes, along with an increase in nonsurgical management. Otologic symptoms were the most common presenting symptoms in VS patients; accordingly, general otolaryngologists and neurotologists diagnosed the majority of patients. Patient treatment decision was influenced the most by neurosurgeons when tumor size was larger or by otolaryngologists and neurotologists when the tumor size was smaller. Although more patients sought care at an academic center, treatment decision or satisfaction was similar to that at private institutions.

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