The majority of patients in whom VS is diagnosed present with symptoms including hearing loss, tinnitus, disequilibrium, or vertigo; rarely they may also present with symptoms of fifth or seventh cranial nerve dysfunction, or with symptoms referable to hydrocephalus such as headache. With the increasing availability and decreasing cost of MRI scans, the “incidental” discovery of VS may become more common. We use the term “incidental diagnosis” of VS here to refer to the diagnosis of VS in asymptomatic patients who undergo radiological imaging for reasons other than a suspected VS. By definition this excludes patients undergoing MRI for asymmetrical hearing loss or dizziness, but does not exclude patients with subjectively “normal” hearing who nevertheless have audiometric evidence of mild hearing loss.

Although a relatively large body of literature exists concerning the natural history of patients with VS with regard to hearing and tumor progression (including those with good hearing), few studies look specifically at tumors that are diagnosed incidentally. These patients therefore present a challenge to the surgeon, in that recommendations made to the patient are at best extrapolations from data describing mixed populations of both asymptomatic and symptomatic patients with “good hearing.” At the present time, however, there is insufficient evidence to suggest that incidentally discovered tumors behave differently from those in patients with minimal symptoms and/or minimal hearing loss.

The goal of the present paper is to assist the surgeon in counseling the asymptomatic patient with VS. For purposes of patient counseling we summarize the evidence available about the natural history of VS (with regard to growth and hearing loss) and the implications of tumor size, hearing status, and other prognostic factors for predicting tumor behavior and hearing preservation of the incidentally diagnosed VS. We conclude with an algorithm for the management of incidental VS based on these considerations.

Methods

Literature Review

A systematic search was performed using the PubMed and MEDLINE databases to identify papers dealing with an incidentally found acoustic neuroma or VS. We first searched the databases by going to the main site and performing a search using the key words “Acoustic Neuroma Incidental,” which yielded 23 results. We then limited these results to English-language articles, and this narrowed it down to 21 papers. To ascertain that all papers on the subject were found, we alternatively searched using the key words “Vestibular Schwannoma Incidental,” which resulted in 26 articles. After limiting this search to English-language articles, it yielded 23 papers. To make our search as comprehensive as possible and to ensure that we would not to miss any relevant articles, we therefore alternately used the key words “Vestibular Schwannoma Asymptomatic” and “Acoustic Neuroma Asymptomatic,” which returned 57 and 54 articles, respectively. After transferring all of these articles to an EndNote library, we came to a total of 78 articles.
To further ensure that we found all articles on the topic, a search of MEDLINE for articles published between 1948 and April 2012 was performed. The key words “Acoustic Neuromas” and “Incidental” (7 results), “Vestibular Schwannoma” and “Incidental” (0 results), “Acoustic Neuroma” and “Asymptomatic” (16 results), and “Vestibular Schwannoma” and “Asymptomatic” (5 results) were used for the query. Four additional articles were identified in these results but yielded papers not written in English, so they were excluded from our analysis. Next, we systematically reviewed the remaining 78 articles to determine which papers met our criteria for discussing incidental VSs. We excluded case reports and papers that analyzed patients who presented with symptomatic VSs. We eventually narrowed these 78 articles to the 9 papers ultimately used in our analysis.

The quality of evidence in the selected articles was categorized according to the US Preventive Services Task Force criteria for ranking evidence (Table 1). Articles were reviewed for data on methodology (retrospective vs prospective), number of patients, tumor size on discovery, and presenting symptoms, if applicable.

**Results**

As stated above, we use the term “incidental” VS to refer to lesions that are identified in asymptomatic patients who undergo radiological imaging for reasons other than suspected VS. Some have suggested that this group of patients is more appropriately referred to as “asymptomatic at presentation,” because these patients may acknowledge a symptom that they have ignored but that is attributable to the VS. A total of 9 studies were identified that specifically included incidental tumors. Case reports were excluded; thus all the studies were retrospective case series. Table 2 shows the data for the retrospective case series on incidental VS. The studies did not consistently describe how the workup was conducted, but of the ones that did, there seemed to be a trend toward a higher likelihood of MRI being the definitive modality. Most of the patients were older than 50 years of age. The average tumor size was 13 mm at the time of detection.

Investigators have attempted to quantify the frequency of incidental VS. Studies based on autopsy reports have noted an incidence as high as 1%. Recently, Lin and colleagues attempted to estimate the prevalence of incidental VS from an intracranial MRI database of 46,414 patients and noted a 0.02% estimated prevalence of incidental VS. The authors suggest that, whereas incidental VS may be less prevalent than autopsy studies suggest, incidental VS may actually be more prevalent than suggested by epidemiological studies. Similarly, recent studies in Denmark estimated the incidence of VS to be 19.4 VSs per million per year as of 2008, whereas another recent study estimated the prevalence of VS in asymptomatic patients at 0.2%. Stangerup and Caye-Thomasen reported on 180 patients with VSs, noting that 65% experienced growth of their tumor, although some that did, there seemed to be a trend toward a higher likelihood of MRI being the definitive modality. Most of the patients were older than 50 years of age. The average tumor size was 13 mm at the time of detection.

**Review of the Literature**

**Evidence-Supported Guide for Discussion With Patients With Asymptomatic VS**

The statements below are supported by our review of the literature and are intended as evidence-based “talking points” for discussion with patients receiving a diagnosis of asymptomatic VS. These statements are summarized in Table 3.

**Approximately Two-Thirds of Tumors Do Not Grow During an Observation Period of Approximately 5 Years (Class III).** An understanding of the natural history of VS aids management decision making for incidental VS. Nikolopoulos and colleagues have recently reviewed the literature for VS growth and found that as much as 75% of tumors (range 6%–75%) exhibited no growth during the follow-up period (the mean growth rate in millimeters/year ranged from 0 to 10.3 mm/year, and mean follow-up ranged from 19 months to 5.5 years). Fucci and colleagues, have recently reviewed the literature and are intended as evidence-based “talking points” for discussion with patients receiving a diagnosis of asymptomatic VS. These statements are summarized in Table 3.

<table>
<thead>
<tr>
<th>Level of Evidence</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Evidence obtained from at least 1 properly randomized controlled trial</td>
</tr>
<tr>
<td>II-1</td>
<td>Evidence obtained from well-designed controlled trials w/o randomization</td>
</tr>
<tr>
<td>II-2</td>
<td>Evidence obtained from well-designed cohort or case control analytic studies, preferably from &gt;1 center or research group</td>
</tr>
<tr>
<td>II-3</td>
<td>Evidence obtained from multiple time series w/ or w/o the intervention—dramatic results in uncontrolled experiments (such as the results of the introduction of penicillin treatment in the 1940s) could also be regarded as this type of evidence</td>
</tr>
<tr>
<td>III</td>
<td>Opinions of respected authorities, based on clinical experience, descriptive studies &amp; case reports, or reports of expert committees</td>
</tr>
</tbody>
</table>

* System proposed by Harris et al.
### TABLE 2: Review of the literature for incidentally diagnosed VS*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Study Design</th>
<th>No. of Cases</th>
<th>No. of VSs</th>
<th>Mean Age</th>
<th>Avg Tumor Size at Detection</th>
<th>Presenting Sx/Reason for Imaging Study†</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lustig et al., 1998</td>
<td>retro</td>
<td>546</td>
<td>4</td>
<td>NA</td>
<td>NA</td>
<td>1) eval for breast cancer mets; 2) MS; 3) slurred speech; 4) weakness</td>
</tr>
<tr>
<td>Tos et al., 1999</td>
<td>retro</td>
<td>NA</td>
<td>970</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Anderson et al., 2000</td>
<td>retro</td>
<td>24,246</td>
<td>17</td>
<td>56 yrs</td>
<td>8 pts &lt;1 cm; 6 pts 1–2 cm; 3 pts &gt;2 cm</td>
<td>NA</td>
</tr>
<tr>
<td>Nutik &amp; Babb, 2001</td>
<td>retro</td>
<td>433</td>
<td>NA</td>
<td>54.0 yrs (M), 54.9 yrs (F)</td>
<td>13.4 mm (M), 16.2 mm (F)</td>
<td>NA</td>
</tr>
<tr>
<td>Lin et al., 2005</td>
<td>retro</td>
<td>46,414</td>
<td>9</td>
<td>58 yrs</td>
<td>13.8 mm</td>
<td>1) seizures; 2) contralat HFS; 3) juvenile angiofibroma; 4) breast cancer mets; 5) Horner syndrome; 6) SDH; 7) dizziness; 8) TIA</td>
</tr>
<tr>
<td>Smouha et al., 2005</td>
<td>meta-analysis</td>
<td>1,345</td>
<td>7</td>
<td>62 yrs</td>
<td>11.8 mm (900 of 1,345 pts)</td>
<td>NA</td>
</tr>
<tr>
<td>Stangerup et al., 2010</td>
<td>retro</td>
<td>2,283</td>
<td>NA</td>
<td>55.4 yrs</td>
<td>16.4 mm</td>
<td>NA</td>
</tr>
<tr>
<td>Varughese et al., 2012</td>
<td>retro</td>
<td>4.8 mil</td>
<td>178</td>
<td>NA</td>
<td>0.71 cm³</td>
<td>1) balance problems (58 of 178); 2) tinnitus (129 of 178); 3) vertigo (77 of 178); 4) hearing: Gr A (52), Gr B (50), Gr C (20), Gr D (54). 2 missing data</td>
</tr>
<tr>
<td>Jeyakumar et al., 2007</td>
<td>retro</td>
<td>121</td>
<td>15</td>
<td>55.7 yrs (symptomatic grp), 52.8 yrs (incidental grp)</td>
<td>1.5 cm (symptomatic grp), 1.1 cm (asymptomatic grp)</td>
<td>1) neck &amp; ear pain; 2) blurry vision; 3) exotropia; 4) family Hx of brain aneurysms; 5) head trauma; 6) contralat ear sudden hearing loss; 7) migraine HA; 8) visual changes, ataxia, contralat ear hearing loss, ipsilat deafness since birth; 9) contralat hearing loss, HAs, bilat ear pressure; 10) parkinsonism; 11) peri orbital numbness, HAs; 12) Lt facial twitching &amp; numbness, fatigue, HAs, Lt hand tremor, ataxia, visual disturbances, tinnitus, MRI done because demyelinating disease suspected; 13) migraines; 14) sinusitis, long-term ipsilat hearing loss thought to be due to noise damage (gunfire); 15) tremor, unsteady gait</td>
</tr>
</tbody>
</table>

* The evidence level in all studies was Grade III according to the US Preventive Services Task Force system (see Table 1). Abbreviations: avg = average; eval = evaluation; Gr = Grade; grp = group; HA = headache; HFS = hemifacial spasm; Hx = history; mets = metastasis; mil = million; MS = multiple sclerosis; NA = not available; pts = patients; retro = retrospective; SDH = subdural hematoma; Sx = symptoms; TIA = transient ischemic attack.
† Numbers followed by parenthesis represent the cases of VS.
Approximately 50% of patients may maintain their hearing during an observation period of approximately 5 yrs.

Tumors exceeding 1.5–2 cm in maximal dimension have a higher probability for growth.

Initial hearing loss, even small degrees of loss, may predict a greater chance of loss of good hearing over time.

The presence or absence of a normal RVR & ABR, presence of fundal fluid, & tumor size may help one to counsel patients on prognosis.

Approximately 2/3 of patients will retain serviceable hearing following hearing preservation surgery.

Approximately 50% of patients will retain serviceable hearing following radiosurgery.

*The evidence level in all studies was Grade III according to the US Preventive Services Task Force system (see Table 1).
preservation, or at least heighten suspicion for an unusual anatomical configuration. Goddard and colleagues noted that the presence of fundal fluid and tumor origin from the superior vestibular nerve were predictive of better hearing outcomes, with both factors having a higher correlation with preservation of the patient’s American Association of Otolaryngology–Head and Neck Surgery hearing class and with preservation of serviceable hearing. These prognostic factors aid in counseling patients about surgical treatment and may aid in convincing an appropriately selected patient to proceed with microsurgical resection.

Approximately Two-Thirds of Patients Will Retain Serviceable Hearing Following Hearing Preservation Surgery (Class III). Friedman and colleagues reported long-term (5 years) preservation of serviceable hearing in 16 (70%) of 23 patients who underwent middle fossa resections for intracanalicular tumors (1.1 ± 0.4 cm; mean ± SD) who exhibited serviceable hearing in the immediate postoperative period. Woodson and colleagues reported 26 (88%) of 26 patients with VS who underwent middle fossa resection and who maintained a word recognition score of > 70% at > 5 years of follow-up. Woodson and colleagues concluded that initial postoperative findings are predictive of long-term hearing results. Kutz and colleagues recently reported serviceable hearing preservation in 24 (63.2%) of 38 patients with preoperative serviceable hearing. These authors noted that patients with VSs ≤ 10 mm compared with patients with VSs > 10 mm were more likely to exhibit serviceable hearing preservation (73.3% vs 25%). Although these surgical results represent the outcomes from some experienced groups and cannot necessarily be generalized to all surgeons, these results do give a sense of what is possible and achievable in appropriately selected patients.

Approximately 50% of Patients Will Retain Serviceable Hearing Following Radiosurgery (Class III). In a recent meta-analysis, preservation of serviceable hearing, defined as a speech reception threshold < 50 dB and an SDS > 50% occurs in approximately 51% of patients following stereotactic radiosurgery, regardless of radiation dose, tumor size, or patient age. This meta-analysis of 4234 patients identified 1322 who had serviceable hearing prior to stereotactic radiosurgery for VS. This rate is similar to the natural history of hearing loss in sporadic VS. Stereotactic radiosurgery in this meta-analysis consisted of single-shot treatment with a mean margin dose of 14.2 ± 2.4 Gy (range 11.5–21.5 Gy). The mean tumor size, for those studies in which it was available, was 3.9 cm. The mean follow-up was 44.4 ± 35 months (the mean margin dose and follow-up are expressed ± SD). In this meta-analysis, 542 patients received an average radiation dose of ≤ 13 Gy, and 671 patients received an average radiation dose of > 13 Gy. The lower-dose (≤ 13 Gy) and higher-dose (> 13 Gy) groups had hearing preservation rates of 60.5% and 50.4%, respectively (p = 0.0005).

Discussion

Management Considerations

We propose an algorithm for management of the incidentally diagnosed VS (Fig. 1).

Given the natural history of growth and the greater likelihood of smaller tumors in patients with incidentally discovered VS, an initial period of observation to assess for tumor growth is a reasonable initial treatment option. Consideration should be given to the patient’s preferences, including but not limited to hearing preservation. If observation is the elected initial approach after a full discussion with the patient, assessment with serial MRI studies is recommended. Despite the fact that some studies indicate that growth usually occurs within the first 5 years of observation, monitoring MRIs beyond this time period is necessary because these tumors can continue to grow slowly and/or unpredictably over time.

Subsequent decision making depends on a combination of factors, including but not limited to patient symptomatology, significant tumor growth (> 2 mm/year), and patient preferences. Initial workup in addition to imaging should include an audiogram and may include an ABR and videonystagmography study for prognostication and counseling purposes. Whereas the mean tumor growth rate varies 1 and 2 mm/year for all tumors and 2–4 mm/year for those tumors that grow, some tumors may exhibit exceptional growth that exceeds 18 mm/year. Furthermore, Mick and colleagues have noted that the majority of tumors identified as growing continue to grow on subsequent observation. Demonstrated significant growth (> 2 mm/year) is the rationale for intervention, either by a microsurgical or radiosurgical approach, according to many studies.

After follow-up imaging, tumors are divided into subgroups of stable and growing tumors. Tumors that presented incidentally may also of course become symptomatic over time. We believe that treatment is indicated for incidentally discovered tumors that become symptomatic or exhibit significant growth. However, the timing of intervention depends not only on initial tumor size and demonstrated growth or symptoms, but is also influenced by the patient’s age and general medical condition as well as the patient’s and surgeons’ preferences.

Parameters for Intervention

Stable Tumors. Incidental tumors with relative stability (≤ 2 mm/year) on serial imaging that remain asymptomatic may safely continue to be observed; microsurgery may be offered in patients who begin to experience progressive onset of symptoms, including progressive hearing loss or imbalance for example.

Growing Tumors. Intervention is indicated in those tumors that exhibit significant growth on follow-up imaging (> 2 mm/year). These patients may be treated with either microsurgery or radiosurgery, depending on such factors as tumor size, patient age, and hearing status. Patients without hearing who have a growing VS and who elect microsurgical resection should be offered a resection via the translabyrinthine approach. Patients with serviceable hearing can be offered a hearing preservation approach. For small tumors ≤ 1.5 cm, in general we favor the middle fossa approach for microsurgical resection. With respect to the retrosigmoid approach, we reserve this approach for smaller tumors with minimal extension into the IAC (less...
than one-half of the proximal IAC) with a predominantly cerebellopontine angle component, in which the patient desires an attempt at hearing preservation. A discussion of the relative advantages and disadvantages of the middle fossa approach versus the retrosigmoid approach for hearing preservation is beyond the scope of this paper.

Microsurgery Versus Radiosurgery. A discussion of the extensive literature on microsurgery and radiosurgery for VS is beyond the scope of this review. Although we favor observation for most incidentally discovered tumors, treatment at the time of initial diagnosis may be reasonable for tumors that at the time of diagnosis are sufficiently large to have mass effect on the cerebellar peduncle and/or brainstem. Most patients with incidental VS will, however, have smaller tumors and desire hearing preservation. Therefore, if treatment is contemplated, consideration for the relative odds of hearing preservation with treatment (that is, microsurgery or radiosurgery) versus observation is advised.

Summary Remarks

Our review of the literature provides evidence on which to inform our recommendations for management of incidentally diagnosed VS. Admittedly, there is a dearth of high-quality evidence (that is, Class I or II) to guide clinical decision making in the treatment of incidental VS; however, we do believe that careful evaluation of the available Class III data can aid in patient counseling. Clearly, there will never be Class I evidence (that is, randomized, blinded data) on which to base these clinical decisions. This review is not intended to be an exhaustive or comprehensive review of the literature—no attempt at meta-analysis has been made, and the data presented should not be interpreted as such. Rather, this is a highly selected review of the literature consisting of high-quality retrospective studies and expert opinion.

Approximately two-thirds of VS tumors do not grow, and approximately 50% of patients maintain their hearing during an observation period of approximately 5 years. Incidentally discovered VSs tend to be smaller, so observation is a reasonable option if a patient follows up with an initial Gd-enhanced MRI study at 6 months and then, as long as no growth occurs initially, MRI studies yearly. With significant growth with or without symptoms, microsurgical resection may be offered as a treatment option. This position is strengthened by a recent report from Sughrue and colleagues in which it was noted that patients who had tumors with growth rates > 2.5 mm per year tended to have worse hearing results versus those who had tumors with growth rates ≤ 2.5 mm per year.

Initial hearing loss predicts a greater chance of loss of good hearing over time and provides support for microsurgical resection via a hearing preservation approach without initial observation, because hearing loss may continue to progress despite lack of tumor growth. Large initial tumor size may be a predictor of future growth, so observation for tumors exceeding 1.5–2 cm is not recommended given the higher probability for growth.

We believe that only significantly growing tumors (> 2 mm/year) should be treated with radiosurgery. Stereotactic radiosurgical treatment carries a small risk of malignant transformation, which must be discussed with patients. Microsurgical resection is generally more difficult after radiation, due to the formation of significant adhesions between the facial nerve and tumor, and it presents increased risk of facial nerve injury.

Conclusions

An understanding of the natural history of VS growth and changes in hearing associated with these tumors over time is essential to the management of the incidentally diagnosed VS. Furthermore, an understanding of the con-
Discourse

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conceptualization and design: Lekovic, Hoa, Schwartz. Acquisition of data: Lekovic, Hoa, Drazin, Hanna. Analysis and interpretation of data: all authors. Drafting the article: Lekovic, Hoa, Drazin, Hanna. Critically revising the article: Lekovic, Hoa, Drazin, Schwartz. Reviewed submitted version of manuscript: Lekovic, Hoa, Drazin. Approved the final version of the manuscript on behalf of all authors: Lekovic. Study supervision: Lekovic.

References


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