Abstract

Background/Objective: Hemorrhagic vestibular schwannomas (HVS) are a rare clinical entity, with less than 50 reported cases in the English literature. The pathogenesis of acute intratumoral hemorrhage remains unknown since the majority of published cases have occurred in patients without precipitating trauma or underlying risk factors such as anticoagulation. Presentation is variable; however, most patients experience acute decline in symptoms presumably caused by abrupt tumor enlargement from intratumoral bleeding. The objective of the current study is to review our experience managing 6 patients with HVS that presented over the last 15 years in order to further define risk factors, patient course, and outcome.

Study Design: Retrospective chart review of all patients diagnosed with HVS, evaluated at the authors’ center between 2000-2015.

Setting: Single tertiary academic referral center.

Main Outcome Measures: Disease presentation, radiologic features, management strategy, and clinical outcome.

Results: Six HVS were evaluated between 2000 and 2015, representing approximately 0.3% of all vestibular schwannoma evaluated at the authors’ center during this time. The median age at time of diagnosis was 67 years (range 39-93). 5 of 6 cases occurred in men, and all had sporadic unilateral tumors. No patients reported prior head trauma, however 3 of 6 were receiving warfarin for indications including atrial fibrillation and pulmonary embolism; the median INR in this subgroup was 2.3 (1.7-2.8).

The most common presenting symptoms were hearing loss (AOO-HNS Class D), disequilibrium, and headache (all cases). Four patients reported recent vertigo, 3 had trigeminal hypesthesia, 2 acute facial paresis (HB 4), and 2 hydrocephalus. The median tumor size at diagnosis was 2.6cm (range 0.4-3.5) and 2 demonstrated macrocystic features. Four of 6 (67%) HVS had focal areas of pre-contrast T1-weighted hypointensity within the tumor, and 2 of 6 exhibited subarachnoid blood products.

Five of 6 patients underwent microsurgical resection while one subject with an intracanalicular tumor was observed. The latter patient presented with sudden sensorineural hearing loss and acute vestibulopathy; however shortly after evaluation his dizziness and hearing loss spontaneously improved (Class D to B) and there has been no evidence of growth for 68 months. Of the 5 subjects that underwent surgical treatment, 3 received gross total resection while 2 were managed with aggressive subtotal removal. The median HB score for patients greater than 12 months of postoperative follow-up was 2. One of the two patients with preoperative HB grade 4 paresis has recovered good function (HB 2) over the course of 25 months following surgery, the second recently underwent surgery and follow-up data is not yet available. There were no perioperative deaths, and at a median follow-up of 19 months (6-9 months) no patient has experienced recurrence.

Conclusions: HVS are rare and commonly present with acute worsening of headache, hearing loss, imbalance, and cranial neuropathy. Contrasting previous reports, we found a high percentage (100%) of HVS associated with anticoagulation use. In surgically fit patients with large tumors, we advocate urgent microsurgical resection. Observation with serial radiography may risk repeated hemorrhage with further neurologic decline. The role of radiosurgery is not yet defined.

Background

Historically, it has been estimated that approximately 1% of all vestibular schwannomas have a hemorrhagic component. Several theories for the development of hemorrhage exist, such as the formation of dilated vasculature, hypervascularity, recent trauma, anticoagulation use, or rapid tumor growth. Recent advances in imaging suggest that historical incidence estimates may underestimate the true frequency of this phenomenon, leaving the question of whether or not this is simply part of the natural evolution of tumor growth over time. Previous reports have demonstrated that HVS are associated with a more aggressive disease course in terms of symptoms, growth rate, and morbidity. We aim to provide our recent experience with this relatively rare tumor subtype.

Patient #4: T1 pre-contrast (left), Contrast-enhanced (right), T2-weighted (bottom left and right)

Table 1: Patient Demographics

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)</th>
<th>Size (cm)</th>
<th>Symptoms</th>
<th>Follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>63</td>
<td>0.4</td>
<td>Ataxia, Vertigo, Tinnitus</td>
<td>68</td>
</tr>
<tr>
<td>2</td>
<td>39</td>
<td>3.5</td>
<td>Ataxia, Hearing Loss</td>
<td>12</td>
</tr>
<tr>
<td>3</td>
<td>66</td>
<td>2.0</td>
<td>Ataxia, Vertigo, Headache, Trigeminal Weakness</td>
<td>69</td>
</tr>
<tr>
<td>4</td>
<td>68</td>
<td>3.5</td>
<td>Facial Weakness, Vertigo, Headache, Diplopia</td>
<td>25</td>
</tr>
<tr>
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<td>72</td>
<td>3.1</td>
<td>Ataxia, Headache, Nausea, Vomiting, Hydrocephalus</td>
<td>4</td>
</tr>
<tr>
<td>6</td>
<td>61</td>
<td>2.4</td>
<td>Facial Weakness, Vertigo, Hearing Loss, Tinnitus</td>
<td>3</td>
</tr>
</tbody>
</table>

Discussion and Conclusions

We report our experience managing 6 HVS since the year 2000. Similar to previous reports, this subtype of tumor is quite rare, representing only 0.3% of all vestibular schwannoma evaluated during the 15-year study period. In agreement with other reports, we found that patients with HVS more commonly present with cranial neuropathy and rapid neurologic decline. In fact, a third of our patients presented with advanced facial weakness.

Currently there is no universally agreed upon theory of pathogenesis for the development of intratumoral hemorrhage, as the process is likely multifactorial and patient dependent. Contrasting previous reports, we found a high percentage (90%; patients 1, 2, and 4) of HVS associated with anticoagulation use, 66% of which were initiated in close proximity to the development of symptoms. It is possible that the use of anticoagulation in an at-risk patient (large tumor, rapidly growing tumor) resulted in tumoral hemorrhage and symptom progression.

The presence of cystic change in conjunction with intratumoral hemorrhage is worth mentioning. It is possible that tumors with large intratumoral cysts, which may rapidly expand, may experience subsequent intracystic hemorrhage. It is also possible that instead of resulting from rapid cyst growth, it may also be that intratumoral hemorrhage leads to cyst formation. The latter theory is supported by the sarcomatous appearance of cyst fluid seen during surgery.

Our study demonstrates that surgical resection results in improvement in symptomatology and tumor control in the majority of cases. Due to the often rapid development of symptoms and neurologic decline in this cohort, we advocate for microsurgical resection in surgically fit patients. The role of radiosurgery is not yet defined.

References