SPONTANEOUS SHRINKAGE OF VESTIBULAR SCHWANNOMA. CASE REPORT AND REVIEW OF THE LITERATURE

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SPONTANEOUS SHRINKAGE OF VESTIBULAR SCHWANNOMA. CASE REPORT AND REVIEW OF THE LITERATURE (Abstract): Vestibular schwannoma (VS), commonly known as acoustic neuroma, is a slow-growing, benign tumor that originates from the transition zone between the central and peripheral myelin of the vestibular branches of cranial nerve VIII. After making the diagnosis, a conservative approach is an option depending on the tumor size and involves the “watch, wait and rescan” (WWR) policy. We present a rare case of a 59 years old woman, who presented with bilateral hearing loss. Routine imaging showed both otosclerosis and a vestibular schwannoma. She was followed-up with annual MRI between 2006 and 2019 showing a complete shrinkage of the vestibular tumor. Studies describe the natural history of a vestibular schwannoma to be slow growing or in small cases spontaneous shrinkage. However, there are still no pathological theories for the tumor regression process. An internationally method of strategies based on size, radiological aspect and symptoms is needed for a standardized care by analyzing a larger series of patients.

Keywords: ACOUTIC NEUROMA, BENIGN, SPONTANEOUS SHRINKAGE, VESTIBULAR SCHWANNOMA.

Vestibular schwannoma (VS), also known as acoustic neuroma, a tumor with the origin from the transition zone between the central and peripheral myelin of the vestibular branches of the VIIIth cranial nerve, is a slow-growing, benign tumor. It is considered to be the most frequent tumor of the cerebellopontine angle (CPA) (1).

The natural history of these tumors consists in most of the cases of continuous slow growth, followed by stagnation or even shrinkage. Upon diagnosis, the most common attitude involves its treatment through either microsurgery or radiosurgery. However, in some cases, a conservative approach is an option depending on the tumor size and involves the “watch, wait and rescan” (WWR) policy (2).

A review of the literature shows that more than a half of the tumors that are diagnosed do not grow (69%), 16% regress and from those who grow, 70% have a 2 mm/year growth (3). The mean growth rate of VS varies from 0.4 to 2.9 mm/year, and spontaneous shrinkage is observed in 3.8 percent of tumors during observation.
Spontaneous shrinkage of vestibular schwannoma. Case report and review of the literature

We present a rare case, which presented two independent causes for hearing loss, otosclerosis and vestibular schwannoma on the same side and which showed for the first time in our experience spontaneous regression on the long-term follow-up.

CASE REPORT
A 59 years old woman, after a visit at the ENT doctor because of progressive hearing loss, was recommended a head CT scan that showed that she suffered from otosclerosis. The CT imaging also revealed in the cerebellopontine angle a mass occupying lesion that was further investigated through MRI imaging.

The patient came to us with the head MRI and we concluded that she had a vestibular schwannoma in the left cerebellopontine angle.

Clinical assessment showed a good neurological condition, the cause of the bilateral hearing loss being caused by the otosclerosis.

After discussing the various types of management of this tumor, the patient decided for the “watch, wait and rescan” protocol (WWR). She was instructed to rescan the tumor once in two years or when new symptoms appeared.

The first MRI was performed in 2006, but we only have the written description. The next scan was performed in 2007 and it showed no signs of growth of the tumor with a size stationary at 6.73 mm x 11.33 mm.

Spontaneous shrinkage was confirmed in the next scan, done in 2013, as the tumor size decreased by more than 2 mm. The tumor continued to shrink. The next scan performed in 2016 and 2019, showing a tumor measuring 4.22 mm x 5.48 mm.

The tumor size was measured in coronary, sagittal and axial planes (fig. 1). No additional neurologic symptoms were present during this period. The hearing loss followed the common pattern associated with the ear pathology.

Fig. 1. Vestibular schwannoma in series of MRI with measurements showing the tumor shrinkage in axial view.

DISCUSSION
Spontaneous tumor shrinkage is a rare phenomenon but marked in the literature in some studies with follow-ups as long as 27 years (1, 2).

A review of the studies that mention tumor shrinkage in vestibular schwannoma are presented in the first table, including the current study.

In 27 reviewed studies, between 1998 and 2019, tumor shrinkage has been reported in 229 out of 4,002 patients (5.69%) with a mean follow up of 4.37 years (1-28).

As described by Huang et al., 2013 a
complete regression of a VS from a 14.1 mm tumor was seen during a follow-up period of 12 years (10).

A growth-regression process was described by Yasumoto and Ito (28), in a 75-year-old patient with a right small VS that initial grew from 5.2 to 16.7 mm over 7 years, then spontaneously shrank to 8.2 mm with improvement of the tinnitus, dizziness and the other initial symptoms.

**TABLE I**

<table>
<thead>
<tr>
<th>Author, year</th>
<th>No. of patients with shrinkage/total patients studied (%)</th>
<th>Age of patients (mean)</th>
<th>Mean size of tumor (mm)</th>
<th>mm or % of shrinkage</th>
<th>Follow up years (mean)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Luetjie et al., 1988 (13)</td>
<td>1/115 (9%)</td>
<td>71</td>
<td>13</td>
<td>5.38%</td>
<td>5</td>
</tr>
<tr>
<td>Bederson et al., 1991 (5)</td>
<td>4/70 (8%)</td>
<td>N/A</td>
<td>N/A</td>
<td>5.70%</td>
<td>2.7</td>
</tr>
<tr>
<td>Tschudi et al., 2000 (26)</td>
<td>8/74 (11%)</td>
<td>N/A</td>
<td>N/A</td>
<td>16.00%</td>
<td>3</td>
</tr>
<tr>
<td>Luetjie, 2000 (14)</td>
<td>6/47 (13%)</td>
<td>66.83</td>
<td>18.75</td>
<td>6.24%</td>
<td>8.46</td>
</tr>
<tr>
<td>Shin et al., 2000 (22)</td>
<td>7/60 (12%)</td>
<td>63</td>
<td>N/A</td>
<td>N/A</td>
<td>2.6</td>
</tr>
<tr>
<td>Hoistad et al., 2001 (9)</td>
<td>3/102 (3%)</td>
<td>64</td>
<td>N/A</td>
<td>N/A</td>
<td>2.4</td>
</tr>
<tr>
<td>Stipkovits et al., 2001 (24)</td>
<td>3/44 (7%)</td>
<td>58.3</td>
<td>19</td>
<td>12.53%</td>
<td>7.6</td>
</tr>
<tr>
<td>Mohyuddin et al., 2003 (17)</td>
<td>10/50 (20%)</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>1.65</td>
</tr>
<tr>
<td>Slatteri WH III et al., 2004 (23)</td>
<td>16/56 NF2 (29%)</td>
<td>N/A</td>
<td>N/A</td>
<td>1-7 mm</td>
<td>4.3</td>
</tr>
<tr>
<td>Bozorg Grayeli et al., 2005 (6)</td>
<td>7/111 (6%)</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>2.75</td>
</tr>
<tr>
<td>Al Sanosi, 2006 (1)</td>
<td>6/205 (3%)</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>3.2</td>
</tr>
<tr>
<td>Battaglia et al., 2006 (3)</td>
<td>6/111 (5%)</td>
<td>N/A</td>
<td>11-25</td>
<td>0.7 mm</td>
<td>3.1</td>
</tr>
<tr>
<td>Yasumoto and Ito, 2006 (28)</td>
<td>1</td>
<td>75</td>
<td>16.7</td>
<td>5.51%</td>
<td>4</td>
</tr>
<tr>
<td>Penido et al., 2007 (2)</td>
<td>2</td>
<td>54</td>
<td>8.5</td>
<td>N/A</td>
<td>3</td>
</tr>
<tr>
<td>Mick et al., 2008 (16)</td>
<td>2</td>
<td>60.1</td>
<td>N/A</td>
<td>N/A</td>
<td>2.1</td>
</tr>
<tr>
<td>Ferri et al., 2008 (7)</td>
<td>6/123 (5%)</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>4.8</td>
</tr>
<tr>
<td>Hajioff et al., 2008 (4)</td>
<td>16/72 (22%)</td>
<td>61</td>
<td>N/A</td>
<td>N/A</td>
<td>10</td>
</tr>
<tr>
<td>von Eckardstein et al., 2010 (27)</td>
<td>2 (NF2)</td>
<td>55</td>
<td>28.55</td>
<td>8.36%</td>
<td>6</td>
</tr>
<tr>
<td>Pennings et al., 2011 (19)</td>
<td>4/47 (9%)</td>
<td>N/A</td>
<td>Intracanalicular</td>
<td>N/A</td>
<td>5.1</td>
</tr>
<tr>
<td>Huang et al., 2013 (10)</td>
<td>48/1261 (4%)</td>
<td>56.7</td>
<td>12.04</td>
<td>6.25</td>
<td>9.5</td>
</tr>
<tr>
<td>Huang et al., 2013 (11)</td>
<td>1</td>
<td>67</td>
<td>14</td>
<td>14.10%</td>
<td>12</td>
</tr>
<tr>
<td>Romani et al., 2016 (20)</td>
<td>2/223 (1%)</td>
<td>28.5</td>
<td>27</td>
<td>6.22%</td>
<td>6</td>
</tr>
<tr>
<td>Kontorinis et al., 2016 (8)</td>
<td>23/540 (4.3%)</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>0.97</td>
</tr>
<tr>
<td>Sebök et al., 2018 (21)</td>
<td>1</td>
<td>61</td>
<td>2.6</td>
<td>0.11 mm</td>
<td>10</td>
</tr>
<tr>
<td>Lahlou et al., 2018 (12)</td>
<td>14/196 (7%)</td>
<td>60</td>
<td>1.6</td>
<td>0.2 mm/year</td>
<td>5</td>
</tr>
<tr>
<td>Tikka T et al., 2018 (25)</td>
<td>28/540 (5.2%)</td>
<td>N/A</td>
<td>N/A</td>
<td>3.9 mm</td>
<td>2.54</td>
</tr>
<tr>
<td>Amoo et al., 2019 (15)</td>
<td>1</td>
<td>41</td>
<td>3.96</td>
<td>3.52 mm</td>
<td>10</td>
</tr>
<tr>
<td>Present report</td>
<td>1</td>
<td>59</td>
<td>8.15</td>
<td>4.85 mm</td>
<td>13</td>
</tr>
<tr>
<td>TOTAL</td>
<td>229/4002</td>
<td>58.9</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Spontaneous shrinkage of vestibular schwannoma. Case report and review of the literature

In the largest study on the conservative management of vestibular schwannoma, Huang et al. (10), after observing 1261 patients, noted 48 that showed spontaneous shrinkage (4%). The approach of “watch, wait and rescan” (WWR) was analyzed by Kontorinis et al., 2016 in a retrospective analysis of the medical records of 540 VS over a 15 years period, 23 (4.3%) of which demonstrating tumor shrinkage (8). Tikka T. et al. in 2018 achieved a direct comparison of MRI of the internal auditory meatus (IAM) in a group of patients with sporadic VS demonstrating spontaneous regression compared with a control group of patients with growing VS (25). In our case report the patient showed a linear tumor shrinkage during the 12 year follow up.

The study of Romani R and Pollock J, 2016 made some remarks regarding the mechanism of tumor shrinkage that is not well understood even today. They proposed environmental factors, genetic-molecular and a reduction in vascular supply hypothesis (20).

In a recent study, Lahlou G. et al., 2018 (12) identified two specific radiologic features in a series of cases of vestibular schwannoma shrinkage. First, it’s described a festooned or scalloped appearance, defined by multiple curves in the tumor outline. We found this aspect of scalloped “line” in the first MRI done in 2007, as shown in green (fig. 2).

Second, on T2 weighted imaging of cerebrospinal fluid (CSF) infilling the intrameatal portion of the tumor, associated with asymmetry of the intrameatal (IAM) size. This feature was recognized in T1 weighted imaging with contrast, as displayed by the blue line, marking the CSF infilling of the IAM.

The study concluded that the two radiological features could allow neurosurgeons and neuro-otologists to easily recognize the vestibular schwannoma that will shrink spontaneous, therefore it is important to look for them and opt for a WWR protocol (12).
CONCLUSIONS

Although spontaneous shrinkage of acoustic neuroma is described as a potential natural evolution in a consistent body of literature, there are still no pathological theories for this evolution. As the genetic variations in schwannoma do not indicate a predisposition for involution, we propose a mechanism involving tumor metabolism. The failure to recruit vascularization might be, for example, an individual mechanism that can lead to tumor shrinkage (as “failure to thrive”).

We argue that cases like the one we presented are a strong argument for “watch, wait and rescan” approach (WWR) towards the patients that harbor small sized tumors. The current literature endorses this policy for small or medium sized tumors, with minimal or absent symptoms. The Lahlou G. et al., 2018 study provides evidence for tumor shrinkage even in large tumors (12).

An internationally standardized method of strategies based on size, radiological aspect and symptoms is needed by analyzing a larger series of patients.

REFERENCES

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**EARLY MEASLES VACCINATION AND THE IMMUNE RESPONSE**

It is well known that during measles epidemics World Health Organization (WHO) recommends that the first dose of measles-containing vaccine (MCV 1) should be given at 9 months of age. But, globally, there were reported cases on infants who are too young to receive MCV1, and they remain at high risk of contracting measles. This study is the first systematic review to examine the effect of MCV1 administered to infants younger than 9 months on the immunogenicity and vaccine effectiveness of subsequent MCV doses. It was reviewed evidence of humoral and cellular immunity, including seropositivity, geometric mean titres, avidity, T-cell stimulation, and vaccine effectiveness. Their findings suggest that administering MCV1 to infants younger than 9 months followed by additional MCV doses results in high seropositivity(98%), vaccine effectiveness(95%), and T-cell responses, which are independent of the age at MCV1, supporting the vaccination of very young infants in high-risk settings. However, they also found some evidence that MCV1 administered to infants younger than 9 months resulted in lower antibody titres after one or two subsequent doses of MCV than when measles vaccination is started at age 9 months or older. The clinical and public-health relevance of this immunity blunting effect are uncertain (Lochlainn LM, de Gier B, van der Maas N, van Binnendijk R, Strebel PM, Goodman T, de Melker HE, Moss WJ, Hahné SJ. Effect of measles vaccination in infants younger than 9 months on the immune response to subsequent measles vaccine doses: a systematic review and meta-analysis. The Lancet Infectious Diseases. 2019 Sep 20.).