Hydrocephalies and Vestibular Schwannoma of Great Size: Therapeutic Strategies: About a Series of 32 Cases

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Abstract

Introduction: The association hydrocephalus and vestibular schwannoma (VS) has been known for a long time. However, there is no therapeutic consensus, especially the place of drainage of cerebrospinal fluid (CSF). We report the result of our experience on the management of this pathology from a group of patients with a high volume VS (Koos IV) and operated consecutively. After reflections based on current literature data, we propose a therapeutic decisional algorithm. Materials and Methods: This is an analytical, retrospective study of 171 patients operated on KOOS IV vestibular schwannoma from January 2003 to December 2016 at the Marseille University Hospital Center. Of these, 32 patients with hydrocephalus and stage IV vestibular schwannoma were included. Radio-diagnostic criteria for hydrocephalus were based on Evans’ index, cortical furrow status, and the presence of trans-ependymal resorption. Our sample was divided into 2 groups. The first consisted of patients first operated on their hydrocephalus and secondarily treated with schwannoma surgery (group I); patients who underwent surgical resection of their first-line tumor were group II. Epidemiological, clinical, radiological, therapeutic and monitoring data were analyzed. The comparison of the quantitative variables was made by Fisher’s test. Results: During our study period (13 years), 171 cases of stage IV SV had been operated. The association between hydrocephalus and SV stage IV of Koos accounted for 18.7%. The average age of our patients was 53 years with a sex ratio of 0.7. The clinical picture was primarily composed of otological signs (90.6%), headache (56.3%) and cerebellar involvement (43.8%). The average diameter of VS in ponto-cerebellar angle (PCA) was 31.5 mm. The treatment consisted of placing a first shunt of the CSF in 34.4% (group I). The ventriculoperitoneal shunt (VPS) was performed in 90.9% of cases. The first surgical removal of the tumor (group II)
involved 65.6% of the patients. The postoperative tumor residue averaged 0.76 cc. The Evans index was evaluated on average at 0.33 in each of the 2 groups postoperatively. The average follow-up time for patients was 51 months. Eight cases of complications were recorded during the study. Secondly, in group II, VPS was performed in 9.5% (2 cases). **Conclusion:** Hydrocephalus is a condition commonly associated with stage IV vestibular schwannoma. The first optimal surgical excision of the tumor seems to be the treatment of choice for this pathological association. The success of the surgery is very often related to the management of hydrocephalus pre, per and post operative.

**Keywords**
Hydrocephalus, Ventriculoperitoneal Shunt, Vestibular Schwannoma, Exeresis

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**1. Introduction**

Vestibular schwannoma (VS) is a benign, extra-axial tumor developed in the ponto-cerebellar angle (PCA) at the expense of the Schwann sheath of the vestibular nerve, which is a divisional branch of the 8th cranial pair called the vestibulo-cochlear nerve [1]. The association between vestibular schwannoma and hydrocephalus is not a recent discovery. Cases have long been reported in the otolaryngology and neurosurgical literature, although for the most part based on anecdotal reports [2] [3]. In the literature, this pathological association accounts for 3.7% to 18% of all VS cases. Many controversies persist around the therapeutic management of the association between hydrocephalus and stage IV vestibular schwannoma; especially the place of treatment of hydrocephaly. We report the result of our work on this pathological association through which we will try to extricate a strategy of therapeutic approach by analyzing also the current data of the literature.

The main objective was to determine the rationale for schwannoma primary surgery in patients with hydrocephalus. The secondary objective was to propose an algorithm for the management of hydrocephalus on stage IV of vestibular schwannoma.

**2. Materials and Methods**

This was an analytical, retrospective study of 171 patients operated on KOOS IV vestibular schwannoma between January 2003 and December 2016 at the Marseille University Hospital Center. Of these, 32 patients with hydrocephalus and stage IV vestibular schwannoma were included. The criteria for radiological diagnosis of hydrocephalus were based on the presence of dilatation of the cerebral ventricles including VL, V3 and sometimes V4 with an Evans index greater than 0.33; significant narrowing or obliteration of cortical furrows.
Our study population was divided into 2 groups. The first consisted of patients who received an external ventricular derivation (EVD) or ventriculoatrial derivation (VAD) from the LCS before tumor surgery (group I). The second (group II) consisted of patients who had undergone first-line surgical resection of their tumor and who had secondarily undergone surgery for hydrocephalus. Epidemiological, clinical, radiological, therapeutic and follow-up parameters were studied. These parameters were:

- age, sex; history of NF2, failure of Gamma Knife treatment
- clinical presentation: signs of HTIC, otological signs, cerebellar lesions, long pathway involvement
- Radiological data: Size and volume of schwannoma, volume of postoperative balance, state of ventricles.

Type of treatment proposed as first-line
- the derivation of CSF in first option
- the excision of schwannoma in first intention
- the derivation of the CSF after excision of schwannoma

Evolution
- postoperative complications and the future of patients
- the duration of patient follow-up

The data collected from the archived paper files and from the computer system of the APHM (public assistance of Marseille hospitals) were put on an Excel file and processed by Stata 11.

The comparison of the quantitative variables was made by Fisher’s test. The P-values less than or equal to 0.05 were considered statistically significant.

A total of 32 patients with hydrocephalus and vestibular schwannoma (VS) were enrolled between January 2003 and December 2016.

3. Results

1) Descriptive characteristics of the study population (Table 1)

Our study population was overwhelmingly female (sex ratio 0.7) and had a mean age of 53 (range, 13 to 82 years). As antecedents, three patients in our study sample had neurofibromatosis type 2 (NF2) and two others had Gamma knife radiosurgery. More than half of our patients had otological signs at the time of their treatment. Otologic signs were predominant in our study population 71.9%; 34.4% tinnitus, 28.1% dizziness and 28.1% cophosis. This damage to the ear was more found in patients in group II (95.2%). It consisted of 42.8% tinnitus, 38.1% vertigo and 14.3% cophosis. The difference found in otologic impairment in the 2 groups was not significant. The Group I, were marked by the predominance of signs of intracranial hypertension. Signs of long-track involvement and those of the cerebellar hemisphere were found in both groups. They were unstable at walking 81.8% for group I and 52.4% for group II and disorder 45.4% group I and 42.8% for group II.

The Evans index was 0.36 (range 0.33 - 0.39) in the study population. Initially
Table 1. Descriptive characteristics of the study population.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Total population</th>
<th>Group I</th>
<th>Group II</th>
</tr>
</thead>
<tbody>
<tr>
<td>Effective</td>
<td>32</td>
<td>11 (34.4%)</td>
<td>21 (65.6%)</td>
</tr>
<tr>
<td>M/F</td>
<td>14 (43.75)/18 (56.25)</td>
<td>4 (36.4%)/7 (63.6%)</td>
<td>10 (47.6%)/11 (52.4%)</td>
</tr>
<tr>
<td>Age:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>average</td>
<td>53 ans</td>
<td>58.4 ans</td>
<td>50.19 ans</td>
</tr>
<tr>
<td>extremes</td>
<td>(13 - 82)</td>
<td>(17 - 78)</td>
<td>(13 - 82)</td>
</tr>
<tr>
<td>Antecedents</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>NF2</td>
<td>3 (9.4%)</td>
<td>0</td>
<td>3 (14.3%)</td>
</tr>
<tr>
<td>Gamma knife</td>
<td>2 (6.3%)</td>
<td>2 (18.2%)</td>
<td>0</td>
</tr>
<tr>
<td>Clinical data</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>tinnitus</td>
<td>11 (34.4%)</td>
<td>2 (18.2%)</td>
<td>9 (42.8%)</td>
</tr>
<tr>
<td>cophosis</td>
<td>3 (9.4%)</td>
<td>0</td>
<td>3 (14.3%)</td>
</tr>
<tr>
<td>fear of heights</td>
<td>9 (28.1%)</td>
<td>1 (9.1%)</td>
<td>8 (38.1%)</td>
</tr>
<tr>
<td>instability</td>
<td>20 (62.5%)</td>
<td>9 (81.8%)</td>
<td>11 (52.4%)</td>
</tr>
<tr>
<td>cerebellar syndrome</td>
<td>14 (43.8%)</td>
<td>5 (45.4%)</td>
<td>9 (42.8%)</td>
</tr>
<tr>
<td>headaches</td>
<td>18 (56.3%)</td>
<td>11 (100%)</td>
<td>7 (33.3%)</td>
</tr>
<tr>
<td>Radiological data</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Evans Index</td>
<td>0.36 (0.33 - 0.39)</td>
<td>0.37 (0.35 - 0.38)</td>
<td>0.36 (0.33 - 0.39)</td>
</tr>
<tr>
<td>tumor diameter</td>
<td>3.15 cm (2 - 5.5)</td>
<td>2.86 cm (2 - 3.7)</td>
<td>3.29 cm (2 - 5.5)</td>
</tr>
<tr>
<td>tumor volume</td>
<td>20.3 cc (4 - 87)</td>
<td>12.5 cc (4 - 22)</td>
<td>24.6 cc (4 - 87)</td>
</tr>
<tr>
<td>tumor laterality R/L</td>
<td>16 (50%)</td>
<td>8 (72.7%)/3 (27.3%)</td>
<td>8 (38.1%)/13 (61.9%)</td>
</tr>
</tbody>
</table>

cm = centimeter; cc: cubic centimeter (milliliter); R: right, L: left, M/F: male/female; NF2: neurofibromatosis type 2.

this index was evaluated at 0.37 (0.35 and 0.43) in group I and 0.36 (0.33 and 0.46) in group II.

2) Radiological aspects

The Evans index associated with a reduction or obliteration of the cortical furrows with or without trans-ependymal resorption made it possible to make the radiological diagnosis of hydrocephalus. This index was evaluated on average at $0.36 \pm 0.04$ cm preoperatively and $0.33 \pm 0.01$ cm postoperatively in our study population. This difference was statistically significant $p = 0.03$. Preoperatively, in group I, the Evans index was, on average, 0.37 (0.35 and 0.43) versus 0.36 (0.33 and 0.46) in group II. In each of the two groups postoperatively, it was 0.33 with extremes of 0.28 - 0.35 for group I and 0.32 - 0.36 for the second group. There was a parity of location of the tumor. The latter averaged a diameter of 3.15 cm (2 - 5.5) for a volume of 20.3 cc (4 - 87) (Figure 1(a) and Figure 1(b)).

3) Treatment

It consisted of the laying of a first derivation of the LCS in 34.4% (group I). Of these, a ventriculo-peritoneal shunt (VPS) derivation was performed in 90.9% of cases and a ventriculo-atrial derivation (VAD) in one patient.
Surgical excision was performed in 21 first-line patients (65.6% of cases) constituting group II. A EVD was placed postoperatively in 4 patients in group II (19%); for hematoma PCA 3 cases (14.3%) and for meningitis a case (4.7%).

Macroscopically, the intraoperative appearance of the tumor was typical in half of the patients in our study sample (50%). It was a soft, lipidized, yellowish lesion. The fleshy and indurated schwannoma in 12 patients (37.5%) and mixed, that is to say, fleshy, indurated and cystic in 3 patients (9.4%). The macroscopic aspect was not mentioned in a patient’s chart (3.1%). In group I, the lesion was typical in 63.6% of cases, fleshy-indurated in 18.1% of cases (2 patients) and mixed in the same proportion. The fleshy and indurated tumor accounted for 47.6% (10 cases) of patients in group II, followed by the typical form 42.8% and the mixed lesion 4.7% (1 case). The consistency was not reported in one patient (4.7%).

Surgical excision was subtotal in all our patients with an average tumor residue of 0.76 ± 0.6 cc. This tumor residue was 0.62 cc (0.05 and 1.3) in group I patients and 0.82 cc (0.08 and 2.9) in group II patients. There was no significant difference in tumor size between the two groups (p = 0.09).

4) Evolution and Complication (Table 2)
### Table 2. Patient follow-up characteristics.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Group I</th>
<th>Group II</th>
</tr>
</thead>
<tbody>
<tr>
<td>Follow-up period (month)</td>
<td>48.5 (2 - 106)</td>
<td>52.2 (12 - 106)</td>
</tr>
<tr>
<td><strong>Complications</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CSF rhinorrhea</td>
<td>1 (9.09%)</td>
<td>2 (9.5%)</td>
</tr>
<tr>
<td>keratitis</td>
<td>1 (9.09%)</td>
<td>1 (4.7%)</td>
</tr>
<tr>
<td>meningitis</td>
<td>0</td>
<td>1 (4.7%)</td>
</tr>
<tr>
<td>shunt complication</td>
<td>1 (9.09%)</td>
<td>0</td>
</tr>
<tr>
<td>meningocele</td>
<td>0</td>
<td>1 (4.7%)</td>
</tr>
</tbody>
</table>

The mean follow-up duration of our patients was 51 ± 31 months. It was 48.6 months (2 and 106) for patients in group I and 52.2 months (12 and 106) for those in group II. This difference was not significant (p = 0.31). 53.1% of patients (17 cases) in our study population had benefited from complementary treatment with Gama Knife. They consisted of 15.6% (5 cases) of patients in group I and 37.5% (15 cases) of those in group II. At four years of follow-up, the mean tumor residue volume was 0.78 ± 0.8 cc in the population; 0.51 cc and 0.9 cc for groups I and II, respectively.

Eight cases of complications were recorded during our study (25%). They were distributed as follows: 3 cases of rhinorrhea of the LCS (9.4%) of which two in group II; a case of lymphocytic meningitis (3.1%) in group II; 2 cases of ocular keratitis by peripheral facial palsy (6.2%), one in each group. one case of meningocele in group II (Figure 1(c)). Most of our complications had been treated medically. Cases of rhinorrhea and meningocele were managed by strict decubitus and ACETAZOLAMIDE®-based medication combined with a compressive dressing for meningocele.

Short bi-antibiotic therapy associated with EVD was used to treat meningitis; it was cured without neurological sequelae.

The VPS was implanted in 2 patients (9.5%) of group II for chronic hydrocephalus (Figure 1(d)) 8 and 21 months after the removal of their tumors. Six months after the surgery, a group I patient was taken to the valve malfunction block. Most patients in group II had a good clinical course of their hydrocephalus after excision of their schwannoma (Figure 1(e) and Figure 1(f)).

### 4. Discussion

#### 4.1. Frequency

Vestibular schwannomas account for 80% of adult cerebellar pancreatic tumors [4]. The association between vestibular schwannoma (VS) and hydrocephalus represents 3.7% to 18% of all cases of VS. More frequently, it is a communicating type hydrocephalus [5] [6]. In our study, the prevalence of this pathological as-
4.2. Age, Sex and Location of Schwannoma

In the literature, the mean age of discovery of a vestibular schwannoma is 50 years, with extreme limits ranging from 16 to 85 years according to series [7]; in our work, he was 53 years old with extremes of 13 and 82 years old. This slight difference was due to the fact that our study focused only on Stage IV SV so with a slightly longer evolution time.

A female predominance was encountered in our study with a sex ratio of 0.7. We found parity about laterality in our study population. According to some authors, the sex and the side of the localization of the tumor do not seem to be factors determining the development of hydrocephalus in VS patients. [6] [8] [9].

Studies have shown a high correlation between the presence of hydrocephalus and the size of schwannoma, the concentration of CSF proteins and the duration of disease [6] [9] [10]. They reveal that a prolonged duration of the disease and a high level of protein in the CSF secondary to the presence of the tumor in elderly patients contribute to the formation of communicating hydrocephalus [11]. This notion was not found during our work when lesions of greater volume were found in some younger patients. Jeffrey et al in their work on a cohort of 157 patients found no correlation between the patient’s age and the type of hydrocephalus [12].

4.3. Hydrocephalus

Among the factors involved in the development of communicative hydrocephalus in patients with VS, the high concentration of proteins in the CSF is the most widely discussed [13]. To date, the concentration of CSF proteins necessary for the genesis of hydrocephalus remains unknown. Other factors may play a role in the formation of this hydrocephalus, including the partial obstruction of CSF resorption sites by the secreted proteins by the tumor [14]; it is recognized that CSF protein concentration in patients with SV is a function of tumor size [6] [15]. The presence of schwannoma outside the blood-brain barrier allows blood proteins to pass through the cleft and interendothelial junctions of the tumor to the surrounding CSF [12]. Communicating hydrocephalus is thought to be predominant in older SV carriers [10] [16]. We will simply say that the presence of the neuroma is a predisposing factor to hydrocephalus whatever the age. For cases of obstructive hydrocephalus with schwannoma of large volume, the size of the tumor plays an essential role [8] [17]. In their cohort of 157 patients, Jeffrey et al. found a positive correlation between non-communicating hydrocephalus and tumor volume (P < 0.001) [12].

4.4. Treatment

The treatment of hydrocephalus in patients with stage VI vestibular schwannoma is a function of clinical presentation and age. Despite the evolution of the man-
agement of this pathological association, uncertainties remain around the place of treatment of hydrocephalus. Should a shunt be placed before or after schwannoma resection? Which shunt and for which patient?

The recognition of hydrocephalus through its clinical signs is particularly important especially when a surgical procedure is envisaged. In front of a patient with signs of intracranial hypertension marked by intense headache, visual disturbance and drowsiness, a primary derivation of the LCS becomes the priority after radiological confirmation of hydrocephalus. This derivation can be done by a ventriculocisternostomy (VCS) if hydrocephalus is obstructive by compression of V4; in other cases we recommend the establishment of a ventricular shunt. In the event that schwannoma resection surgery is prioritized in a intracranial hypertension patient by hydrocephalus, the risk of intraoperative cerebellar involvement at the opening of the dura mater would expose to an easily understandable surgical risk [3] [12]. Some authors favor the implantation of an EVD in these cases before the excision of SV [18]. The delay between the placement of this EVD and the lesion excision surgery must be reasonable in order to minimize the infectious risks associated with the presence of the drain.

After treatment during our study, no cases of radiological hydrocephalus were recorded in both groups. As a result, we are perfectly in tune with the "current" concept, which proposes tumor resection in the first place without using a shunt for associated hydrocephalus [2] [8] [16] [19]. The optimal resection of the schwannoma could avoid unnecessary placement of a ventricular shunt and thus reduce the possible stresses and complications associated with the presence of this device [11]. In case of post-operative persistence of signs related to CSF hydrodynamic disturbances, a derivation may be considered [20]. In the literature, the incidence of patients requiring shunt placement for post-tumor resection hydrocephalus varies between 3.8% and 29.2% [8] [16] [21] and 9.5% in our study. Nevertheless, it was a selected patient population, presumably with radiological criteria for hydrocephalus, with a compatible clinic, but without intracranial hypertension.

4.5. Evolution and Complication

A rhinorrhea and/or otorrhoea of the CSF translate a communication of the endocranium with the external environment. It is important that they are treated as soon as possible to prevent the occurrence of meningitis, a formidable complication. Measures to reduce this postoperative complication are meticulous obliteration of exposed air cells, including those around the internal auditory canal, adequate closure of the dural envelope, and temporary lowering of intracranial pressure with a ventricular or lumbar drain [8]. Table 3 summarizes recent literature data on the failure of clinical monitoring of hydrocephalus after schwannoma exeresis. The synthesis of current data from the literature and our experience led us to propose a decision algorithm in the treatment of hydrocephalus associated with large vestibular schwannoma (Figure 2).
Table 3. Large volume vestibular schwannomas with hydrocephalus, main publications describing the first excision and analyzing the failure rate on hydrocephalus.

<table>
<thead>
<tr>
<th>Authors/references</th>
<th>Year of publication</th>
<th>Average age (years)</th>
<th>Tumor size (cm)</th>
<th>Effective Failure cases &amp; shunts</th>
<th>Average duration of follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gerganov [16]</td>
<td>2011</td>
<td>47.2</td>
<td>3.6 ± 1.76</td>
<td>48</td>
<td>12.5</td>
</tr>
<tr>
<td>Hussam [22]</td>
<td>2014</td>
<td>41</td>
<td>&gt;3</td>
<td>37</td>
<td>8.1</td>
</tr>
<tr>
<td>Prabhuraj [23]</td>
<td>2017</td>
<td>53.7</td>
<td>4.41</td>
<td>65</td>
<td>15.4</td>
</tr>
<tr>
<td>Xiang [24]</td>
<td>2017</td>
<td>47.54 ± 12</td>
<td>&gt;3</td>
<td>1167</td>
<td>0.6</td>
</tr>
<tr>
<td>Our series</td>
<td>2017</td>
<td>50</td>
<td>3.29</td>
<td>21</td>
<td>9.5</td>
</tr>
</tbody>
</table>

Figure 2. Decision algorithm.

4.6. Limits of the Study

The limitations of this study were: the small size of our study population, lack of randomization (retrospective study), bias in case recruitment as patients treated for this condition during the study period were excluded for incomplete the follow-up file.

5. Conclusion

Hydrocephalus is a relatively common condition in patients with stage IV vestibular schwannoma. Our results and the analysis of the literature indicate that optimal surgical excision of schwannoma would avoid the placement of a shunt, provided that this hydrocephalus is not responsible for acute intracranial hypertension.

References


