Clinical and Radiological Evolution of a Group of Untreated Acoustic Neuromas

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Abstract
Introduction: The acoustic neuroma is a benign tumour that originates in the vestibular branch of the eighth cranial nerve. The main treatment is surgery, but many authors suggest that with elderly patients or in small neuromas we can opt for watchful waiting.

Methods: This was a retrospective study from 2007 to 2013 that included 27 patients diagnosed of acoustic neuroma that had not been treated due to the size of the tumour, age and comorbidities, or by patient choice. We evaluated overall condition, hearing thresholds, degree of canal paresis and central disorders.

Results: After 6 years of follow up, clinical manifestations of 18 patients remained unchanged, 5 patients underwent hearing loss and developed tinnitus, 2 cases had more intense tinnitus and 2 cases had dizziness. The radiological controls by magnetic resonance imaging showed that the initial maximum diameters (5–16 mm) increased by 1.7 mm on average, with annual growth rates below 0.5 mm.

Conclusion: In selected cases, such as for small neuromas and in elderly patients, the conservative option of close monitoring with magnetic resonance imaging is an important alternative given that, in our cases, clinical features and radiological image did not suffer major changes. If there were any such changes, therapeutic options could be proposed.

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KEYWORDS
Acoustic neuroma; Vestibular schwannoma; Sensorineural hearing loss; Unilateral hearing loss; Tinnitus

PALABRAS CLAVE
Neurinoma del acústico;
Schwannoma vestibular;  
Hipoacusia neurosensorial;  
Hipoacusia unilateral;  
Acúfenos

Introduction

Cerebellopontine angle tumours account for around 10% of intracranial tumours. Vestibular schwannomas or acoustic neuromas are the most frequent of these, representing 80%–90%. Vestibular schwannoma is prevalent throughout the world, regardless of race. Incidence is similar in the different communities at 0.01 per 1000 inhabitants/year (1 case per 100 000 inhabitants/year); in Spain, incidence may be extrapolated, with 400 new cases being diagnosed each year. Incidence of neurofibromatosis type 2 is lower, with one new case per million inhabitants per year. The ratio of acoustic neuroma cases associated with neurofibromatosis type 2 compared with sporadic unilateral neuroma is 1:10. The incidence of neurofibromatosis type 1 or von Recklinghausen disease is higher (one new case per 40 000 inhabitants/year), although the neurofibromatosis type 1 associated with acoustic neuroma is very infrequent, with few cases diagnosed worldwide.

Acoustic neuroma is a tumour which grows from Schwann cells, from the vestibular nerve myelin sheath; it grows in the area adjacent to the Scarpa’s ganglion, on the inside of the internal auditory canal. The proliferation of these cells lead to the formation of the tumour, which compresses the vestibular nerve axons and also spreads to the adjacent nerves (cochlear and facial).

The tumour is the same in sporadic unilateral form or bilateral hereditary form (neurofibromatosis type 2); however, this hypothesis does not explain why, according to histological studies, the hereditary form is more aggressive, with greater capacity for infiltration of the adjacent facial and cochlear nerves; clinical evidence confirms this, showing that there is higher growth capacity of the form associated with neurofibromatosis type 2 and cases undergoing radiosurgery, in which results in hereditary forms are markedly worse than in lateral acoustic neuroma forms. In either of the 2 forms, the acoustic neuroma tends to dilate the internal auditory canal, albeit not constantly, and when it spreads to the cerebellopontine angle, it occupies the cistern, and comes into contact with the brainstem and cerebella, compressing the fourth ventricle and leading to hydrocephalus and intracranial hypertension symptoms. Hearing and vestibular function are usually affected in early stages, but facial nerve function is preserved intact, even in large tumours, except in exceptional cases.

Vestibular schwannoma is a rare case with regards to development and growth. Therapeutic implications are very clear, yet it leads to one of the greatest controversies over decision-making in diagnosed cases. Tumour growth is generally slow. Extensive acoustic neuroma studies with patients who underwent surgery show a mean increase in diameter of 1–2 mm per year. However, no correlation exists between tumour growth and age, gender, tumour size at diagnosis and clinical manifestations of the patient; correlation only exists between the growth rate and the proportion of cells in the replication stage. This information will only be useful in tumours which have been operated on where residual tumour has remained or been left.

Pre-operative suspicion must be high regarding any audiological or vestibular manifestations of a unilateral or asymmetrical nature (hearing loss, sensorineural hearing loss, vertigo or instability) and investigation should take place. The use of brainstem auditory evoked potentials is still a useful detection method, particularly in cases of lower clinical significance, such as tinnitus or minor asymmetries in the audiogram, and in elderly patients. However, immediate use of MRI does tend to be made, which is invaluable to diagnosis. Furthermore, systematic use of MRI for headaches and other neurological conditions has led to the diagnosis of asymptomatic vestibular schwannomas, even when there is no alteration of auditory brainstem evoked potentials.

The treatment of choice is surgery, immediately following diagnosis if possible, since, according to several authors, the smaller the tumour, the better the results obtained from surgery. On the contrary, many other authors defend watchful waiting for elderly patients or when neuromas are small.

The aim of our study was thus to comment upon the clinical and radiological evolution of 27 patients diagnosed with
vesticular schwannoma in 6 years, who had not been treated with stereotactic radiosurgery or traditional cerebellopon-
tine angle surgery.

Materials and Methods

We present a review of 27 patients diagnosed with spo-
radic acoustic neuroma, who were not treated surgically or
with radiosurgery, but were placed under observation. The
patient ages of our sample ranged between 35 and 72 at
diagnosis, with a mean age of 59. There was a predominance
of women with a female to male ratio of 2:1. Neuromas
affected both ears almost equally, with 48% of schwannomas
located in the right ear and 52% in the left ear.

As previously indicated, surgical approach for the
patients included in the study was rejected for several rea-
sons: tumour size, which in the majority of cases was not
above 1 cm, advanced age and associated comorbidities,
or express decision of the patient. There were 9 patients
in the first group, in which the maximum diameter of the
tumour ranged between 5 and 9 mm, with a mean of 6 mm.
There were 13 patients over 65 years of age in the sec-
ond group who presented multiple concomitant disorders
and too high a risk to undergo surgery. Tumour size in
these patients ranged initially between 5 mm and 16 mm,
with a mean size of 11 mm. The last group included those
patients who rejected any treatment due to the possible
risks and complications, and who opted for watchful waiting.
The maximum tumour diameter for these patients ranged
between 8 and 14 mm. These patients received annual
check-ups for almost 6 years, specifically from 2007 until
2013. In all cases diagnosis of neuroma was made the year
prior to the beginning of follow-up. In all cases and in each
annual check-up the following was performed: a thorough
anamnesis on symptom development at the beginning or
appearance of other new symptoms; a complete assessment
of the general state of health; a luminal tonal audiome-
try to control auditory thresholds; a bithermal caloric test
to detect the existence or variations in canalicular presia
levels, and a neurological examination with particular atten-
dion paid to the cranial nerves. MRIs were also performed
annually for radiological control, to identify changes in the
maximum tumour diameter which was the chosen parameter
for the study of size in these neuromas.

Results

Initial clinical manifestations were, in order of frequency:
unilateral sensoneural hearing loss presented by 19 of the
27 patients as an initial symptom; 4 patients presented
at the clinic for the first time with unilateral hearing
loss; only one patient presented with vertigo and a sensa-
tion of objects spinning around and autonomic symptoms
which lasted a few minutes. The final group included 3
asymptomatic patients for whom the discovery of acous-
tic neuroma was coincidental to radiological tests from
MRIs requested for other conditions (Fig. 1). Symptoms
were assessed at each annual visit. During the observed
period, the clinical findings presented on initial assessment
remained unchanged in 18 patients, where no subjective
or objective fluctuations were detected in the hearing

Figure 1 Initial clinical manifestations presented by the
patients included in the study and the frequency of appearance
of each one of them in our sample.

loss, tinnitus, instability, or asymptomatic conditions pre-
presented initially. Only 9 patients presented variations in their
symptoms: in 5 patients who initially presented with sensoneural
hearing loss, minor losses in the auditory threshold
were identified after 6 years of study and they developed
tinnitus, and in only 2 patients who began with tinnitus
was the appearance of a minor sensoneural hearing loss
detected and a worsening in the tinnitus intensity during
follow-up. Another 2 patients presented with short-lived
episodes of vertigo (spinning sensation), associated with
nosea and vomiting. This was self-limiting and isolated,
with no recurrences to date. It should be noted that dur-
ing this follow-up no atypical clinical manifestations were
detected, such as facial paresis or sudden deafness, in any
cases (Table 1). Table 2 shows the audiometric develop-
ment of these patients according to the Gardner-Robertson levels,
comparing the thresholds at the initial moment of diagnosis
and at 6 years.

In the annual radiological studies, the following results
were obtained: maximum initial diameters of our 27 patients
ranged between 5 and 16 mm, with a mean of 8 mm. The
final mean increase in this maximum diameter after 6 years
was only 1.7 mm. Furthermore, we can state that 18 of the
27 tumours showed some type of growth, with an annual
growth rate under 0.5 mm; 9 neuromas showed no growth
and remained completely stable with regards to maximum

| Table 1 Evolution of the Clinical Manifestations in the 6-
<table>
<thead>
<tr>
<th>year Follow-up.</th>
<th>Number of Patients (2007)</th>
<th>Number of Patients (2013)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing loss</td>
<td>19</td>
<td>21</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>4</td>
<td>9</td>
</tr>
<tr>
<td>Vertigo</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Other atypical symptoms</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>3</td>
<td>3</td>
</tr>
</tbody>
</table>
diameter and in the remaining 2 patients initial tumour size decreased by 1 and 2 mm respectively (Fig. 2).

We would indicate that we were unable to correlate tumour size with the clinical history of our patients, as tumour size was similar in the asymptomatic patients and the patients with sensoneural hearing loss or tinnitus and there was no relationship between the appearance of new symptoms in the development of certain patients with tumour growth.

No significant changes in the follow-up of these patients showed up in caloric test results. Initially, only 2 patients were diagnosed using this test for a unilateral vestibular hypofunction with a canalicular paresis of 45% and 38%, respectively. Curiously, only one of them had initially presented with peripheral instability and vertigo. The other individual belonged to the asymptomatic group. In the other patients, the caloric test showed normal values, with lower canalicular percentages of paresis under 22% and a directional preponderance under 28%. Over the 6 year check-up period, these values hardly changed, with caloric tests showing normal results in the last check-up in 24 of the 25 patients for whom results had initially been normal, and pathological in the other 3, with canalicular paresia values of 28% in the new case and without any apparent clinical results and hardly any change in the 2 known cases (48% and 39%, respectively).

We noted that neurological examination in all patients during the 6-year check-up period was completely normal, with no detection of deficiency or loss of strength or sensitivity in the cranial nerves.

**Table 2** Audiometric Evolution (According to Gardner-Robertson Classification Over the 6-year Follow up).

<table>
<thead>
<tr>
<th>Level I (discrimination 0–30 dB)</th>
<th>Number of Patients (2007)</th>
<th>Number of Patients (2013)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Level II (discrimination 31–50 dB)</td>
<td>14</td>
<td>11</td>
</tr>
<tr>
<td>Level III (discrimination 51–90 dB)</td>
<td>5</td>
<td>10</td>
</tr>
<tr>
<td>Level IV (discrimination 91–100 dB)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Level V (no response)</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

**Figure 2** Recording of the maximum diameters (in millimetres) of the vestibular schwannomas of our patients, measured using MRI at the beginning (2007) and end (2013) of the study.

Vestibular schwannoma is a benign tumour which forms part of the cerebellopontine angle tumour group. As previously commented upon, 2 forms are distinguished: sporadic, which is the most frequent, with one case per 100,000 inhabitants/year and hereditary, linked to neurofibromatosis type 2 and which is uncommon, with one case in every million inhabitants/year. In our study, and in accordance with the statistics described, we had no cases of bilateral vestibular schwannoma associated with neurofibromatosis type 2. All were isolated forms, some of which were even discovered incidentally.

Initial, most frequent symptoms of acoustic neuroma are audiological or vestibular. Trigeminal symptoms may appear less frequently, as may facial, cerebellar, cerebral or other cranial nerve symptoms. In the House clinic, in 180 consecutive vestibular schwannoma cases, between 1992 and 1993, initial symptoms were, by order of frequency: hearing loss (75%), tinnitus (9%), dizziness (9%), headache (1%), facial paresia (1%) and others (5%). Symptoms found during disease progression were: hearing loss (90%), tinnitus (86%), dizziness (57%), headache (40%), facial paresthesia (15%), facial weakness or spasm (2%) and others (10%). A progressive character has always been attributed to these symptoms. Similar results were obtained in our study with regards to initial symptoms, with unilateral sensorineural hearing loss being the primary symptom (70%), followed by tinnitus (15%) and vertigo (4%). We found greater discrepancies regarding the House study in symptom progression. Percentages at 6 years were as follows: hearing loss (77%), tinnitus (33%), and vertigo (11%). According to the results obtained, clinical manifestations suffered by the patients included in our study were slow to progress and there were no significant changes.

There are currently series of vestibular schwannomas with no surgical intervention, that are radiologically controlled using computerized tomography and magnetic resonance over long periods. Here between 60% and 70% of cases presented growth, with only between 10% and 20% showing growth over 2 mm per year. Our study obtained more promising results, with mean tumour growth over 6 years of 1.7 mm, and an annual growth rate under 0.5 mm. These results logically give rise to the belief that a conservative approach should become an alternative to consider for this type of tumour under certain conditions where the success of surgical intervention of radiotherapy could be overshadowed. Rosenberg, in his study on the natural history of the vestibular schwannoma in a group of 130 patients, concluded that measuring the maximum tumour diameter with MRI is a reliable method for growth follow-up of acoustic neuromas. He adds that it is not necessary to perform rigorous analysis of the tumour to determine whether it is growing significantly and that the great majority of patients over
65 with acoustic neuromas do not require therapeutic intervention of any type. According to this study, indications for therapeutic intervention should be based on a combination of rapid tumour growth with development of symptom.  

We agree with these conclusions. Out of our patients, practically 70% experienced no significant change in 6 years in the clinical manifestations with which they began, whilst the remaining 30% presented with small decreases in auditory thresholds, a slight increase in hearing loss, or isolated episodes of vertigo which did not reoccur throughout the study and which could have been due to benign paroxysmal positional vertigo. The latter greatly affects the general population and there is an obvious lack of argument in favour of an ethiopathogeny connecting both processes. Radiologically, 30% of these tumours experienced no change in maximum diameters after 6 years of examinations. 60% increased in size during this period, but only by 1.7 mm on average, with annual growth rate under 0.5 mm. On the contrary, in 10% of the sample a reduction in tumour size was even detected. We may thus deduce that the behaviour of an acoustic neuroma is unpredictable; its pattern may vary and that the same tumour does not always present the same uniform growth pattern throughout its evolution, as was previously believed. There is therefore a need to consider all therapeutic options within a wide range for vestibular schwannoma, including watchful waiting with close monitoring, that is highly recommended in certain situations. The patient must always be told that there is no therapeutic approach, merely observation, and that clinical and/or radiological changes could occur.17–19 An informed patient consent to verify agreement with this watchful observation would be useful to include in the patient’s clinical record.

Another aspect to be considered is the problem of assessing the growth rate of the vestibular schwannoma. Apparently insignificant changes in the increased diameter of the tumour could lead to considerable changes in its volume; the trend towards introducing computerized volume measurement systems is inevitable and would lead to alterations in the classification of different cerebellopontine angle tumours.26

We should note that this was a retrospective study with well defined inclusion criteria and therefore subject to their corresponding bias. It only includes patients who did not undergo surgical intervention or radiotherapy and those for whom an annual 6-year follow up was maintained.

Conclusions

Our study results and relevant published literature lead us to conclude that, in selected cases (small neuromas, elderly patients and/or patients with conditions which could be contraindicative to conventional surgery or radiosurgery, and patients who expressly rejected any therapeutic approach), the conservative option based on observation and periodic monitoring using MRI is an alternative to be considered, given that, in general, neither the typical audiovestibular clinical manifestation of this tumour nor radiological imaging showed any important changes in our patients. Furthermore, given the promising results of our observations, with consideration of the different bias of our study, we should consider this as another ‘therapeutic’ option for any type of patient diagnosed with acoustic neuroma. If watchful waiting is the chosen option, any clinical or radiological change detected in successive check-ups will always allow room for the reappraisal of surgery or radiotherapy.

Conflict of Interest

The authors have no conflict of interest to declare.

References