Clinical and Radiological Evolution of a Group of Untreated Acoustic Neuromas

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

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

defienden que en pacientes de edad avanzada o en neurinomas de pequeño tamaño se puede optar por simple vigilancia.

**Métodos:** Estudio retrospectivo, del 2007 al 2013, que incluye a 27 pacientes diagnosticados de neurinoma del acústico que no fueron tratados, bien por el tamaño del tumor, por la edad y las comorbididades asociadas, o por decisión del paciente. Se evaluaron el estado general, los umbrales auditivos, el grado de paresia canalicular y las posibles alteraciones a nivel central.

**Resultados:** A los 6 años de seguimiento, la clínica –hipoacusia (70%), acúfeno (15%) y vértigo (4%)– de 18 pacientes se mantuvo sin variaciones; 5 casos experimentaron calda del umbral auditivo y desarrollaron acúfeno, 2 casos intensificaron su acúfeno y 2 casos presentaron vértigo. Los controles radiológicos por resonancia magnética demostraron que los diámetros máximos iniciales (5-16 mm) se incrementaron en 1.7 mm como media, con tasas de crecimiento anual inferiores a 0.5 mm.

**Conclusión:** En casos seleccionados, neurinomas de pequeño tamaño y pacientes de edad avanzada, la opción conservadora mediante vigilancia estrecha con resonancia magnética es una alternativa importante, puesto que, según nuestra muestra, ni la clínica ni la imagen radiológica sufren cambios significativos y, en caso de que lo hagan, siempre estamos a tiempo de una actuación terapéutica.

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**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 compress 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 auditory 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
vestibular schwannoma in 6 years, who had not been treated with stereotactic radiosurgery or traditional cerebellopontine angle surgery.

**Materials and Methods**

We present a review of 27 patients diagnosed with sporadic 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 reasons: 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 second 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 audiometry to control auditory thresholds; a bithermal caloric test to detect the existence or variations in canaliculic presiia levels, and a neurological examination with particular attention 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 seneoneural 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 sensation of objects spinning around and autonomic symptoms which lasted a few minutes. The final group included 3 asymptomatic patients for whom the discovery of acoustic 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 loss, tinnitus, instability, or asymptomatic conditions presented initially. Only 9 patients presented variations in their symptoms: in 5 patients who initially presented with sensorial hearing loss, minor losses in the auditive 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 sensorial 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 nausea and vomiting. This was self-limiting and isolated, with no recurrences to date. It should be noted that during 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 development 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>
<thead>
<tr>
<th>Table 1</th>
<th>Evolution of the Clinical Manifestations in the 6-year Follow-up.</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>Number of Patients (2007)</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>19</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>4</td>
</tr>
<tr>
<td>Vertigo</td>
<td>1</td>
</tr>
<tr>
<td>Other atypical symptoms</td>
<td>0</td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>3</td>
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</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.

**Discussion**

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.4,5 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.3 Trigeminal symptoms may appear less frequently, as may facial, cerebellar, cerebral or other cranial nerve symptoms.4,5 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.6-9 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.14-16 Here between 60% and 70% of cases presented growth, with only between 10% and 20% showing growth over 2 mm per year.17-24 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

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

<table>
<thead>
<tr>
<th>Level</th>
<th>Discrimination (dB)</th>
<th>Number of Patients (2007)</th>
<th>Number of Patients (2013)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Level I</td>
<td>0-30</td>
<td>8</td>
<td>6</td>
</tr>
<tr>
<td>Level II</td>
<td>31-50</td>
<td>14</td>
<td>11</td>
</tr>
<tr>
<td>Level III</td>
<td>51-90</td>
<td>5</td>
<td>10</td>
</tr>
<tr>
<td>Level IV</td>
<td>91-100</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Level V</td>
<td>No response</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

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**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.
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.\textsuperscript{25} 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.\textsuperscript{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.\textsuperscript{20}

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 reuptake of surgery or radiotherapy.

Conflict of Interest

The authors have no conflict of interest to declare.

References