Objective: To describe the stage-related clinical features of 8th cranial nerve schwannoma.

Material and method: Descriptive study of 71 patients whose diagnosis and/or treatment have been carried out at our centre between 1997-2003. Gender, age, and symptoms were considered, with special attention to gender. Determination of tumoral stage following Tos and Thomsen image criteria.

Results: The mean age was 64.6 (range, 20-87) with a marked incidence between 52 and 70 years (62 % of the whole), slightly higher in females and in the left ear. The main symptoms were those derived from involvement of the 7th and 8th cranial nerves, with other cranial nerves and cerebello-pontine structures being involved in larger tumours, although a high variability was noted in clinical patterns of same-stage cases and in the first symptom. Our study also found a high variability in hearing conservation and a marked frequency of vestibular or facial (motor and sensory) symptoms. There were also very infrequent forms of presentation that are highlighted.

Conclusions: There is no typical clinical pattern and no typical first symptom in 8th cranial nerve schwannomas. Any audiovestibular or facial symptom, even the slightest, may be the first expression of 8th cranial nerve schwannoma. There is no stage-specific symptom except for those with involvement of the cerebello-pontine or cerebral structures.

Key words: Neurinoma. Schwannoma. 8th cranial nerve. Clinical picture. Symptoms. Acoustic neuroma.

Objective: Determinar la semiología característica del schwannoma del VIII par craneal en función de su estadio por imagen.

Material y método: Estudio descriptivo a partir de 71 sujetos diagnosticados en nuestra consulta o remitidos a nuestro centro para tratamiento entre 1997 y 2003. Se recoge el sexo y la edad, así como los síntomas que presentaba el paciente, diferenciando el síntoma de inicio. Determinación del estadío del tumor según criterios de imagen de la clasificación de Tos y Thomsen.

Resultados: La media de edad es de 64,6 (intervalo, 20-87) años. llama la atención que el tramo de edad entre los 52 y 70 años acumula el 62 % de los casos. La incidencia es levemente mayor en mujeres y en el oído izquierdo. Los síntomas predominantes son los derivados de la afeción del VIII y el VII pares craneales, asociándose síntomas por afeción de otros pares craneales y del ángulo pontocerebeloso en tumores mayores, aunque existe una enorme variabilidad tanto entre casos del mismo estadío como en la clínica de inicio (first symptom). Destacamos en nuestro estudio la variabilidad de la preservación de la audición entre los diferentes pacientes y el alto número de casos afectos de clínica vestibular o facial, tanto motora como sensitiva. Asimismo, hay formas de inicio sorprendentes por su infrecuencia.

Conclusiones: No existe patrón clínico típico de comienzo del schwannoma del VIII par. Ningún síntoma es patrimonio de ningún estadío, con la excepción de los propios de la afeción de la fosa posterior.


INTRODUCTION

The VIIIth cranial nerve schwannoma is by far the most common tumour of the cerebellopontine angle, as it accounts for approximately 80% of all tumours involving this
anatomical region. It mainly affects middle-aged females and their symptoms result directly from the compression of the vascular nerve of the inner auditory canal and the structures of the posterior fossa. It is generally sporadic and unilateral, but it is bilateral in 4% of cases, usually associated with type 2 neurofibromatosis.

The clinical manifestations they provoke depend on the structures affected by their growth, generally cochleovestibular initially and over time, endocranial symptoms and symptoms due to the involvement of other nerve pairs. Nevertheless, schwannomas do not follow a fixed semiological pattern in terms of clinical manifestations, symptom-intensity or chronology of appearance, which can cause us to delay a diagnosis that should be made early in the course of the tumour and will determine the possibility of function-conserving treatments.

Bearing this circumstance in mind, we undertake this study with the purpose of analyzing the many forms of presentation an VIIIth cranial nerve schwannoma can have by studying our centre’s case series in order to answer the question in the title of our text: can we assert that VIIIth cranial nerve schwannomas have typical symptoms?

MATERIAL AND METHODS

A retrospective, descriptive study was conducted involving a population of 71 diagnosed subjects from our centre or referred to our centre for treatment between the years 1997 and 2003. We collected a series of basic statistical data such as gender and age, as well as the symptoms they presented, distinguishing between the initial symptom and the time of evolution until diagnosis in months.

We used the taxonomy published by Tos and Thomsen for radiological stage classification, taking into account the considerations put forth by Magnan and Zini regarding this classification with respect to tumour compression of the brainstem:

- Stage I: intracanalicular tumour
- Stage II: tumour <2 cm. The tumour presents a slight extrameatal extension and does not come into contact with any brain structures
- Stage III: tumour between 2 and 4 cm. The tumour may touch brain structures, but there is no compression of the fourth ventricle
- Stage IV: tumour >4 cm. The tumour compresses or displaces the fourth ventricle

Having collected these data, we proceeded to the classification and analysis by sub-groups of stages on the basis of imaging studies.

RESULTS

The following epidemiological data are revealed by the analysis of the study population. The population distribution by age is shown in Figure 1, with a mean age of 64.6 years (range, 20-87). It is striking to note that the 52-70 year age group accounts for 62% of the cases. By gender, the distribution was 44 females (61% of the cases) and 28 males (39%). The populational distribution by imaging-based tumour stage is presented in Figure 2; it is particularly striking that 42 cases (59%) were diagnosed in the early stages of illness (stages I and II). The distribution based on the location of the neurinoma reveals that 55% (39 cases) were on the left, whereas 45% (32 cases) were on the right, which is very much in line with the data reflected in the literature we consulted.

The following results were derived from the analysis of our patient population according to the initial symptoms they presented:

- Hypoacusia, 93% (66 cases)
- Tinnitus, 83% (59 cases)
- Painful hearing, 14% (10 cases)
- Vertigo, 28% (20 cases)
- Facial neuralgia, 4% (3 cases)
- Problems swallowing, 5.63% (4 cases)
- Symptoms of essential HBP or of compression of the brainstem, 7% (5 cases)

There was a remarkably high number of cases with vertigo. Figure 3 presents the results of the analysis of these data according to a distribution of tumour stages.

Figure 1. Distribution of the population by age.

Figure 2. Distribution of the population by imaging-based tumour stage.
The initial clinical manifestations the patients in our series presented can be found in Table I.

Finally, if we select cases of VIIIth cranial nerve schwannoma without hypoacusia, we can find 7 cases that corresponded to stage I in 3 cases, stage II in 2 cases, stage III in 1 case, and stage IV in 1 case.

DISCUSSION

The analysis of the data extracted from our population reveals that, consistent with what is reflected in the literature, the highest incidence of VIIIth cranial nerve schwannomas is found during middle age (41-60 years), which accounts for more than 60% of the cases, with a predominance of females and in the left ear.4-6

In all the stages, the most common clinical manifestations and initial symptoms are hypoacusia, often discovered by chance by the patient (this typically takes place when speaking on the phone) as evidence of its insidious nature, regardless of the underlying functional impairment, that can nevertheless exist and be proven by means of auditory evoked potentials, and tinnitus generally accompanies hearing loss. It presents from the very early stages and oftentimes, it is this tinnitus that the patient is most bothered and alarmed by and what causes the patient to seek care.5-11

Occasionally, the hearing loss is sudden; that is to say that the patient’s hearing is acutely and significantly affected. According to some series, this can occur in close to 15% of all schwannomas, and up to 2% of the cases of sudden hypoacusia are due to VIIIth cranial nerve schwannoma.1,4,6,11-17

In our series, this was the initial clinical manifestation in 11% of the cases, above all in small tumours and probably as the result of temporary ischaemia due to tumour...
as occurred in our study. There is a lower percentage of instability, imbalance, dizziness and less exact feelings, present.

any information as to tumour volume should there be one guarantee the absence of tumour nor does it provide us with therefore affirm that the conservation of hearing does not particularly remarkable that 2 cases of advanced stage overt vertigo varies widely among authors. Although patients who develop vertigo, although the percentage of involvement of Wrisberg’s intermediate nerve, although it may be that, depending on the tumour stage, these clinical symptoms should be related to the findings from neuromuscular evoked potential and facial sensitivity studies, which is beyond the scope of this study. As regards facial mobility, we would like to emphasize that the VIIIth cranial nerve is highly resistant to slowly progressive compression before giving rise to clinical symptoms. It is generally affected by bulky tumours, which is a priori a warning sign. Our study reveals a small percentage of cases with this type of symptom at all stages, even in cases of small tumours (Figure 3). It may be more the result of ischaemia than pressure, hence, this clinical indicator also fails to constitute a foolproof sign of a large tumour, although it may be orientative.

On the other hand, it may be worth wondering what percentage of cases display electrical involvement (in other words, electrophysiological or functional involvement) of the facial nerve without any evidence of symptoms, a circumstance that is taken very much into account when it comes to treatment. There has not been a single case of hemifacial spasm in our series, something that has been observed in various conditions affecting the cerebellopontine angle. Nor have we found any case in which the initial symptom was clinically evident facial paralysis or parasthesia.

Signs of severity, such as gait disturbances, loss of strength in a limb, swallowing problems or impaired consciousness are indicative of a bulky tumour and appear in cases of such tumour formation. In our study, they appeared only in stage IV tumours thanks to the tremendous resistance of the nervous system to slowly increasing pressure. This is due to the fact that tumour presence and growth are tolerated for a long time until they give rise to manifestations. The clinical picture at onset varies widely. We would only point out the high percentage of vestibular symptoms (24%) as the initial symptom and of isolated tinnitus (23%), particularly in the low stages of illness with respect to other studies in the literature.

We therefore believe that any case of tinnitus should be thoroughly investigated, since it could be the first symptom of an acoustic schwannoma, with the peculiarity that this is very likely to be small in size, thus making hearing-sparing treatment feasible.

<table>
<thead>
<tr>
<th>Stage I</th>
<th>Stage II</th>
<th>Stage III</th>
<th>Stage IV</th>
<th>Total (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sudden hearing loss</td>
<td>2</td>
<td>4</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Gradual hearing loss</td>
<td>11</td>
<td>5</td>
<td>5</td>
<td>10</td>
</tr>
<tr>
<td>Tinnitus (in general)</td>
<td>12</td>
<td>5</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Tinnitus without hearing loss</td>
<td>9</td>
<td>3</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Instability</td>
<td>3</td>
<td>4</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Vertigo</td>
<td>2</td>
<td>2</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Facial pain-hypoesthesia</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Brainstem</td>
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<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Painful hearing</td>
<td>1</td>
<td>0</td>
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<td>0</td>
</tr>
</tbody>
</table>
As an interesting case in point, we have recorded an VIIIth cranial nerve schwannoma that began with painful hearing and another 2 cases in which the initial symptom was alterations of facial sensitivity (one suffered from hypoesthesia and the other one, neuralgia).

Finally, we would like to underscore the fact that unfortunately there is still a high percentage of patients who are diagnosed in advanced stages of disease (40%); these figures should motivate us to make the clinical criteria for referral better known, in our opinion preferably in primary care, and to persevere in demanding the best of ourselves as specialists in investigating the key symptoms of this illness.

CONCLUSIONS

There is no clinical pattern for onset of VIIIth cranial nerve schwannoma that can be considered typical. Any audiovestibular symptom and even facial symptoms, no matter how subtle, may be the first clinical manifestation of an VIIIth cranial nerve schwannoma and should be duly investigated. No symptom is exclusive to any stage, with the exception of the classical symptoms of involvement of the fossa posterior.

REFERENCES


