■ BRIEF REPORT

Unusual Clinical Presentations of Vestibular Schwannomas

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The aim of this study is evaluate the unusual ways of initial presentation of the vestibular schwannomas. We performed a retrospective study of the patients who underwent resection of acoustic neuromas in our service, including for analysis only the cases which initial symptom was not the hearing loss. Tumour size, localization, clinical presentation, and age of the patients were considered. Nine patients present with atypical symptoms. The most common complain in this group were facial paresthesias (22.2%). None of them complained about other otological symptoms. A significant group of patients did not present with the otological symptoms classically associated with vestibular schwannoma. Clinical knowledge of these kinds of symptoms may lead to earlier detection of these lesions.

Key words: Atypical symptoms. Vestibular schwannomas. Acoustic neuroma. Hearing loss. Cerebellopontine angle.

Formas clínicas inusuales de presentación de los neurinomas del acústico
El objetivo de este estudio es mostrar las formas inusuales de presentación con que pueden manifestarse inicialmente los neurinomas del acústico. Se realiza un estudio retrospectivo de los pacientes intervenidos de neurinomas del acústico en nuestro servicio; se incluye en el análisis sólo los casos cuyo síntoma inicial no se encuadraba en la hipoacusia unilateral. Se recogen datos de tamaño tumoral, localización, forma de presentación y edad del paciente. Encontramos 9 pacientes con síntomas atípicos y sin hipoacusia, entre los que la queja más común (22,2 %) fueron las parestesias faciales. Ninguno de ellos refirió otra clínica otológica acompañante. Un grupo significativo de los pacientes de nuestro estudio no se presentó con los síntomas clásicos de los neurinomas del acústico. Que los clínicos conozcan este tipo de síntomas puede llevar al diagnóstico precoz de esta afección.

Palabras clave: Síntomas atípicos. Schwannoma vestibular. Neurinoma del acústico. Hipoacusia. Ángulo pontocerebeloso.

INTRODUCTION

The most common form of presentation for acoustic neurinomas is gradual unilateral loss of hearing, together with tinnitus.[1] In such patients, audiometry reveals unilateral sensorineural hypoacusia, for which reason an imaging test is requested in order diagnose the condition. However, there are some patients with vestibular schwannomas who do not present with these typical symptoms. This small percentage of patients is able to demonstrate normal hearing or even symmetrical sensorineural hearing loss, thus hindering the diagnostic suspicion of acoustic neurinomas.[2] For this reason, it is important to understand the more atypical forms of presentation of VIII pair neurinomas, so as to ensure specialists remain alert and to allow the early diagnosis of this tumour.

With our analysis, we have attempted to highlight the unusual symptoms with which a neurinoma may come to the fore and test the diagnostic skill of even the most experienced specialists. This study includes such data as tumour size and the age of the patients at diagnosis, so as to show scant symptoms that might arise with a tumour of considerable size, as well as the fact that age cannot be a factor for excluding this disorder from our diagnostic options.

METHODS

We have revised the case studies of the patients treated for acoustic neurinomas at our otorhinolaryngological department between November 1994 and September 2006.

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Data for Patients With Unusual Presenting Symptoms of Acoustic Neurinomas[a]

Patient	*Age*	*Gender*	*Presenting Symptom*	*Size, cm*	*Audiometry, dB, Threshold*	*Vestibular Tests*
1	49	F	Hemifacial numbness	3.2	20	Left vestibular dysfunction
2	24	M	Cephalea	2	15	Normal
3	36	F	Hemifacial numbness	4.3	20	Right vestibular arreflexia
4	36	M	Sensation of blocked ear	1	15	Left vestibular hyporeflexia
5	46	M	Right lingual hypoaesthesia	3	20	Right vestibular dysfunction
6	32	F	Tinnitus	1.8	20	Normal
7	50	F	Facial pain	2.2	15-20	Right vestibular arreflexia
8	51	F	Otalgia	3	20	Left vestibular arreflexia
9	32	F	Vertigo	4	15	Left vestibular arreflexia

[a]F indicates female; M, male.

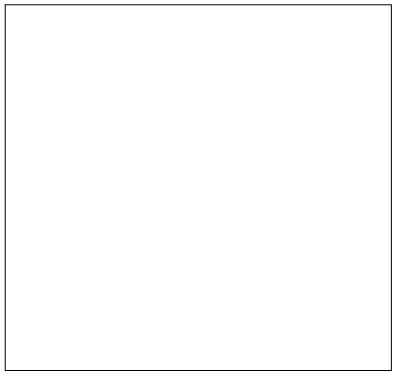

Figure. T1 sequence gadolinium-enhanced magnetic resonance image of patient number 3, showing a neurinoma measuring 4.3 cm on its longest diameter.

Of the 120 patients identified, we chose for the present study all those who had not attended the consultation in connection with hearing loss and who did not report subjective hypoacusia during anamnesis; ie, those presenting normal results in the audiometric study.

After the definition of these criteria, a total of 9 patients (7.5%) were included for analysis; these included 6 females and 3 males, between 24 and 51 years of age (mean, 39.5 years). The largest diameter of the tumour measured on the magnetic resonance images varied between 1 and 4.3 cm (mean, 2.72 cm).

RESULTS

The main symptom leading the 9 patients in the study to consult a physician are given in Table. No-one reported subjective hypoacusia and all of them were shown to be audiometrically normoacusic, ie, they had a pure tone threshold of ≤20 dB with a verbal discrimination of 100%. The most frequent complaint from these patients was facial numbness, in 2 cases (22.2%). One patient (11.1%) complained of cephalea and another of tinnitus, without any other associated symptomatology. One patient reported a sensation of a stuffed ear and another, right lingual hypoesthesia. One patient came to the clinic due to earache and another because of hemifacial lancing pain. As a result, it is noteworthy that the cranial pair which most frequently led to a diagnosis of vestibular schwannoma in this series was the trigeminal nerve, in 5 cases.

We were struck by the mean age of the patients, under 40 years of age, and the fact that they were predominantly female. All of the tumours had attained a considerable size, with a mean diameter of over 2.5 cm, except for a 1×1 cm intracanallicular neurinoma that produced a blocked ear. By way of example, Figure reproduces patient 3's magnetic resonance image. Following the diagnosis, all of the patients were subjected to vestibular tests, in which only 2 patients did not present alterations, although none of them complained of dizziness or instability.

DISCUSSION

The classic description of the symptoms of acoustic neurinomas is based on the development of clinical signs as the tumour grows. Patients begin by complaining of hearing loss, tinnitus, and vertigo, followed by symptoms stemming from the compression of cranial pairs. If this growth continues, there is compression of the cerebellum and brain stem.[3] Already in 1944, Olsen et al[4] noted that typical clinical symptoms, especially hearing loss, were missing in certain patients with neurinomas of the acoustic nerve. Several studies have estimated the incidence of patients with normoacusia at between 2.7% and 12%.[1,5-8]

In 1993, Moffat et al[9] proposed a classification for acoustic neurinomas based on the site of origin and morphology. According to this classification, schwannomas could be categorized as: *a)* lateral (dumbbell shaped), originating in the internal auditory canal (IAC) and leading to early hearing loss; *b)* medial (lollipop shaped), originating in the cerebellopontine angle (CPA) capable of growing without producing significant auditory symptoms; and *c)* intermediate (cone shaped), with a lateral portion spreading into the IAC and a medial portion located in the CPA, producing moderate auditory symptoms.

If we apply these principles to our study (taking intracanallicular neurinomas as lateral, extracanallicular

neurinomas as medial and mixed neurinomas as intermediate), we can see that the 8 mixed lesions in our patients produced the entire range of symptoms explained and the intracanallicular neurinoma produced a blocked ear sensation, from which it is inferred that the vestibular schwannomas in our study did not present the symptoms expected in line with the pattern described.

The incidence of normal hearing in patients with vestibular schwannomas has been estimated at between 3% and 6% in those studies which assessed it specifically.[10-13] The most common presenting symptoms leading to this diagnosis were those related with the trigeminal nerve. In our series, we found 5 patients (55.5%) with trigeminal symptoms. This preponderance of impairment in the V cranial pair in patients with normal hearing has also been encountered by other authors.[6,14] This group of patients would be diagnosed as having a tumour by most clinicians, as they would use a radiological image test to assess any unresolved cranial nerve impairment.

Asymmetric tinnitus and cephalea, observed in our study in 1 patient each, are well-known as possible presenting symptoms for acoustic neurinomas in general and also for presenting in patients with neurinomas and normal hearing.[6,14] This kind of patient would also be diagnosed, as an imaging test would be requested to resolve a persistent cephalea without improvement. More problematic are patients with unilateral tinnitus, as it is unusual for tinnitus to be evaluated in a patient with normal audiometric result. Tinnitus is defined as "sounds in one or both ears, such as buzzing, ringing, or whistling, occurring without any external stimulus." The sounds may be pulsing or continuous (non-pulsing), subjective (perceived only by the patient), or objective (also perceptible by another person). The characteristic tinnitus of neurinomas is non-pulsing and subjective.[14] It is accepted that, in a case of persistent asymmetric tinnitus, the patient should be monitored, even despite a normal audiometric result. However, it is also known that this philosophy may not be favourable in cost-effectiveness terms, as the vast majority of these patients will not have any identifiable retrocochlear condition.

CONCLUSIONS

This study shows that a far from contemptible percentage of patients with vestibular schwannomas may present without any of the typical symptoms attributed to this condition.

This kind of tumour must be considered when dealing with patients presenting continuous unilateral tinnitus, persistent cephalea, and cranial nerve symptoms.

A diagnosis of vestibular schwannoma cannot be discarded in the light of a normal audiometric result.

The awareness by clinicians of this kind of symptomatology may lead to the early diagnosis of this condition.

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