Glomus Tumours of Temporal Bone Origin. Study of 17 Cases

Ángel Ramos Macías[a], Silvia Borkoski Barreiros[a], Daniel Pérez Plasencia[a], Isidoro Lisner Contreras[a], Aser Armesto Fernández[a], Carlos Cenjor Espanol[b], and Elisabeth Masgoret[a]
[a]Servicio de Otorrinolaringología y Patología Cérvico-Facial. Complejo Hospitalario Universitario Insular Materno Infantil de Gran Canaria. Universidad de Las Palmas. Las Palmas de Gran Canaria. Las Palmas. Spain.
[b]Fundación Jiménez-Caprio. Madrid. Spain.

Objective: The objective of this paper is to make a retrospective analysis in patients with glomus tumours of temporal bone origin. We present the results according to the surgical approach applied in each case.
Materials and method: This retrospective study presents the findings in 17 patients with diagnosis of glomus jugulare of the temporal bone, who were observed and treated in our department over a 5-year period (1999 to 2004). We performed a general otolaryngology exam, systemic evaluation and radiological exam. Surgical treatment was performed in 16 cases out of 17. In 1 case treatment with stereotaxic surgery was performed.
Results: The surgical approaches were: retroauricular transcanal approach, radical or modified mastoidectomy through facial recess, and infratemporal fossa approach. Pre-operative embolization was used in 11 of our cases. In all cases the diagnosis of glomus tumour was confirmed. The most frequent post-operative complications found were: transitory paralysis of the facial nerve, sensorineural hearing loss, imbalance, paralysis of the IXth and XIth cranial nerves, and salivary fistula. No recurrences were found after 8 years of follow-up. One case of persistence was found in the case treated with radio surgery.
Conclusions: In our series surgery was found as the elective therapy for patients with glomus tumour of the temporal bone with no recurrences after 8 years of follow-up. Pre-operative embolization diminishes surgery time and intraoperative bleeding. Stereotaxic therapy cannot provide tumour growth control. Complications are discussed and compared with the bibliography.

Key words: Tumours of the temporal bone. Glomus jugulare. Glomus tumours.

Tumores glómicos del hueso temporal. Estudio de 17 casos

Objetivo: El objetivo de este trabajo es realizar un estudio retrospectivo en pacientes con diagnóstico de paraganglioma de localización en hueso temporal, analizando los resultados según la conducta terapéutica aplicada para cada caso.
Pacientes y método: Se incluyó a 17 pacientes con diagnóstico de paraganglioma localizado en hueso temporal tratados en nuestro servicio entre 1999 y 2004. A todos se les realizó un examen otorrinolaringológico, sistémico y de imagen. En 16 casos se realizó cirugía para su exéresis y en 1 caso se realizó tratamiento con cirugía estereotáxica.
Resultados: Los abordajes quirúrgicos fueron: resección transcanal (tras abordaje retroauricular), mastoidectomía radical y modificada con abordaje de receso facial y, por último, abordaje infratemporal. Se realizó embolización preoperatoria en 11 de los casos. El diagnóstico de glomus fue confirmado en todos los casos. Las complicaciones postoperatorias más frecuentes fueron: paresia transitoria del VII par, hipoacusia neurosensorial, desequilibrio, parálisis del VII, parálisis del IX y el XI par y fístula salival. No se observó recidiva de enfermedad con seguimiento entre 1 y 8 años. En un caso tratado con radiocirugía persiste el tumor, con control.
Conclusiones: En nuestra serie la cirugía constituye la terapia con la que se logró la resección tumoral total en todos los casos. El uso de embolización preoperatoria reduce el tiempo quirúrgico y el sangrado peroperatorio. La radiocirugía estereotáxica no permite el control tumoral, y persiste la enfermedad.

Palabras clave: Tumores del hueso temporal. Glomus yugular. Paraganglioma.

The authors have not indicated any conflict of interest.

Correspondence: Dr. A. Ramos Macías.
Venegas, 12, 3.º A.
35003 Las Palmas de Gran Canaria. Las Palmas. Spain.
E-mail: ramosorl@idecnet.com

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INTRODUCTION

Glomus tumours or paragangliomas originate in the baroreceptor structures arising from the neural crest, located in vascular regions and, in our territory, predominately in the bulbus of the jugular vein.

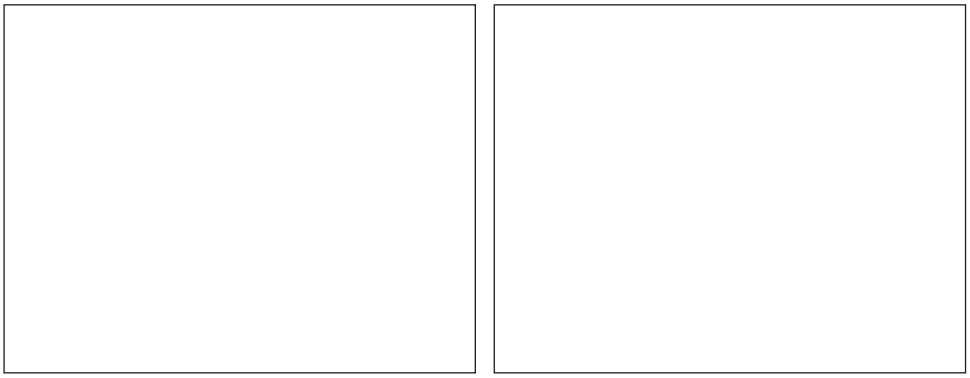

Figure 1. Computerized tomography of a space-occupying lesion, bone destruction secondary to a type C jugulotympanic glomus.

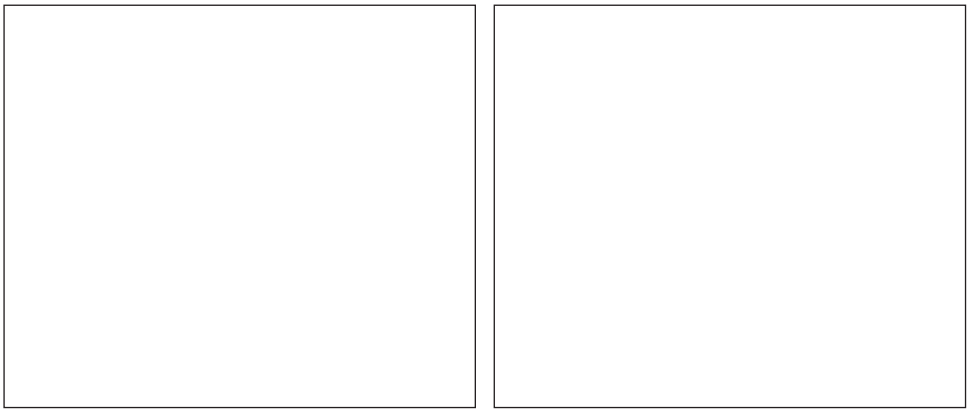

Figure 2. Imaging of embolization of a glomus tumour following administration of sclerosing particles by interventionist vascular radiology. A: Tumour vascularization prior to embolization. B: Appearance post-embolization.

A first study on paragangliomas was conducted by Guild in 1941. In his study, he reported "epithelioid cells in capillaries and pre capillaries close to or at the level of the wall of the bulbus venae jugularis". However in 1945, Rosenwasser was the first to identify the possible relation between Guild's glomus jugularis and paragangliomas located on the temporal bone[1].

They are highly vascularized and histologically benign tumours. Growth, though slow, can cause compression, displacement and invasion of nearby structures, bone, blood vessels, dura mater and cranial nerves[2,3]. These tumours are more common in women, with ratio of 1:3; it is the most common benign neoplasm of the middle ear and the second most common in the temporal bone[4]. It may debut as a bilateral tumour in 1%-2% of the cases; only 1% is capable of secreting catecholamines. They can metastasize in exceptional cases (1%-3%)[5].

Ten percent of the paragangliomas originating in the head and neck are familial with a modified autosomal dominant pattern of heritance[6-8].

Despite the fact that to date there is no standard nomenclature for this type of tumour, the Fisch and Glassock-Jackson classifications[9] are accepted and used without distinction.

This study presents a retrospective, descriptive study of patients with a diagnosis of paragangliomas of the temporal bone; the general characteristics and results obtained are analyzed following treatment of a total of 17 patients in our department.

PATIENTS AND METHOD

Seventeen patients are reviewed following diagnosis of paraganglioma of the temporal bone in our department between 1999 and 2004.

The main reasons for consultation were pulsatile tinnitus, the most common symptom, which presented in 14 patients (83.2%). The next most common symptoms are transmission hearing loss (12 patients), otalgia (8), and otorrhea (5). In all cases, a full otorhinolaryngological examination was performed, including case history, physical examination with otomicroscopy and audiological studies.

All patients had a high resolution computerized tomography (HRCT) and magnetic resonance (MR) (Figure 1). Angiography was performed in the interventionist vascular radiology department and the diagnosis in all cases was Fisch type B and C[9]. Collateral circulation was evaluated and the afferent and efferent tumour vessels were studied (Figure 2). Twenty-four hour urinary vanilmandelic acid determinations were also performed. Tumours were staged according to the Fisch and Glassock-Jackson glomus classifications[9].

The surgical approaches used were the transcanal retroauricular approach for tumours classified as Fisch class A, radical tympanomastoidectomy for class B tumours and a type A radical infratemporal approach for those tumours classified as Fisch class B, C and D (Figure 3).

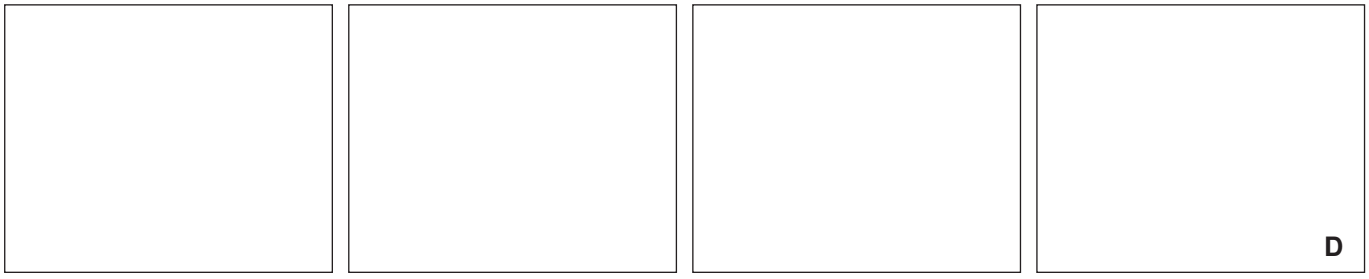

Figure 3. Imaging of the surgical times in the Fisch type A infratemporal approach.

RESULTS

In our series, the age interval was 29-77 years, with a mean age of 39.6 years. There was a 10:17 predominance of females.

During otoscopic examination, the tympanic membrane appeared purple in 12 patients (70.5%); one inflammatory polyp-like formation was observed in the external auditory canal in 4 (23.5%), and only 1 subject (5.8%) presented otorrhea associated with a purplish mass. The serial determination of vanilmandelic acid in 24-hour urine gave normal results in all cases.

According to Fisch's classification, 41% (n=7) presented tumour type A; 29.4% (n=5), type B, and 29.4% (n=5), type C2-C3. According to the Glassock classification, type I was detected in 24.4% (n=5); type II, in 11.7% (n=2); type III in 24.4% (n=5), and type IV in 24.4% (n=5).

In 1 case and in light of the patient's advanced age and poor general status, the decision was made to perform stereotactic radio surgery. In all cases, an arteriographic study of the supra-aortic arterial trunks was conducted and tumour embolization was carried out in 11 patients (68.7%) 72 hours prior to surgery.

The distribution of the surgical approaches used is presented in Table I.

According to Makek's classification[10], infiltration of the facial nerve was observed in 3 cases: degree II in 1 patient (6.25%) and degree III in 2 (12.5%). The diagnosis of glomus was confirmed in all cases by pathology study of the surgical specimen.

The postoperative complications according to each type of approach performed are summarized in Table II.

Surgery was capable of controlling the tumours and no relapse was observed in any of the patients after a follow-up period of between 1 and 8 years. The tumour was seen to persist in the patient who underwent radio surgery; however, it did not grow and it displayed areas of tumoral necrosis without increasing size.

DISCUSSION

The glomus jugulare or paraganglioma of the bulbus venae jugularis is the most common benign tumour of the middle ear. It is a highly vascularized, slow-growing tumour, although it has the potential to spread and involve structures at the base of skull, the nerves and blood vessels.

A predominance of females is seen in most series[8]. The most common clinical presentation is pulse-synchronous pulsatile tinnitus and hearing loss[8]. Hearing loss is often conductive, although there may be a sensorineural component, in which case the possibility of invasion of the labyrinth must be taken into account[9]. Other symptoms are occasional otorrhea, otalgia, and vertigo.

Table I. Surgical approach in glomus tumours

Types of surgery	*Transcanal resection (retroauricular)*	*Mastoidectomy and facial recess approach*	*Infratemporal approach*	*Not operated*
	6	5	5	1

Tabla II. Postoperative complications observed after a minimum of one year postoperative follow-up

Type of surgery	*Transcanal resection (retroauricular) (N=6)*	*Mastoidectomy and facial recess approach (N=5)*	*Infratemporal approach (N = 5)*	*Not operated (N=1)*
Sensorineural hearing loss	1	2	–	
Imbalance	–	1	2	
Temporary paresis VII	–	2	2	
Paralysis VII G3/4 (House Brackmann)	–	–	2	
Paralysis IX-XI	–	–	2	
Salivary fistula	–	–	1	

The presence of symptoms of low cranial nerve involvement indicates a space-occupying lesion.

The facial paralysis resulting from the involvement of the VII cranial nerve is the most common presentation of neural disease; the tumour spreads preferably in the mastoid portion of the nerve; followed by involvement of the X, IX, XII, and XI cranial nerve in order of frequency, although the degree of nerve dysfunction seen preoperatively is not indicative of the status of neural involvement[3,10]. No symptoms are present in 15% of these tumours.

In spite of the fact that histochemical and ultrastructural studies have proved that all paragangliomas secrete catecholamines, only 1% secrete enough to give rise to clinical effects. Should there be any such effect, alpha and beta blockers constitute the preoperative treatment.

During the physical examination, a hypo- or meso-tympanic mass is characteristic, though not pathognomonic. A polyp-like or inflammatory formation can be seen in the external auditory canal.

The preoperative workup includes HRCT in order to have a good analysis of the bone structures at the base of the skull, tumour size and areas of bone erosion. Gadolinium-enhanced MR and angiography make it possible to evaluate tumour vascularization, spread and its relation with nearby structures. Subtraction digital angiography is diagnostic and serves to guide embolization. It was not used in our cases.

There are 2 classifications elaborated to aid in choosing the best surgical plan, the Fisch and the Glassock-Jackson classifications; both are currently used, although to date there is no uniform nomenclature for this type of tumour[9].

There are several therapeutic modalities for treating the glomus, including surgical resection, radiotherapy and radio surgery[4]. Once surgery has been decided upon, preoperative embolization of these lesions decreases intraoperative bleeding and surgery time and in all cases enables complete tumour excision.

Possible postoperative complications are cerebrospinal fluid fistula, imbalance and tissue ischaemia and those resulting from trauma to the cranial nerves and major cardiovascular, pulmonary and cerebral complications[6,7].

The glomus is considered to be relatively insensitive to radiation[4,11-14]. Fractionated radiotherapy is associated with a 7% rate of minor complications and 2%-3% rate of major complications[12]. Benign tumours in subjects undergoing radiotherapy may develop secondary tumours; it has been estimated that there is a 2.7% possibility of secondary malignancy in 10 years[14,15].

Another treatment option in managing these tumours is stereotactic radio surgery using gamma-knife, which makes it possible to attain local control associated with low morbidity and virtually no mortality. It is recommended in elderly patients or individuals with a poor general status, a diagnosis of advanced tumour, those who refuse surgery or unresectable tumour[4]. Other alternatives include the use of intratumour sclerosing agents in an attempt to necrotize and shrink the tumour, as well as adopting a wait-and-see attitude with regular check-ups.

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