Temporal Paragangliomas. A 12-Year Experience

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Abstract

Objective: Our objective was to perform a retrospective analysis of patients with jugulotympanic paragangliomas. We present the results according to the surgical approach applied in each case.

Materials and methods: This retrospective study presents the findings in 21 patients with jugulotympanic paragangliomas who were observed and treated in our department over a 12-year period (1999-2011). We performed a general otolaryngology exam, systemic evaluation, and radiological exam. Surgical treatment was performed in 20 cases out of 21. In 1 case, treatment with stereotactic radiosurgery was carried out.

Results: The surgical approaches were: endaural, retroauricular transcanal, radical or modified mastoidectomy through facial recess, and infratemporal fossa approach. Preoperative embolisation was used in 12 cases. In all cases the diagnosis of paraganglioma was confirmed. The most frequent postoperative complications found were transitory palsy of the facial nerve, sensorineural hearing loss, imbalance, paralysis of the cranial nerves IX and XI, and salivary fistula. No recurrences were found after 12 years of follow-up. One case of persistence was found in the case treated with radiosurgery.

Conclusions: In our series surgery was found to be the elective therapy for patients with paraganglioma, with no recurrences after 12 years of follow-up. Preoperative embolisation decreases surgery time and intraoperative bleeding. Stereotactic radiotherapy cannot eliminate the tumour.

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Paragangliomas temporales. Experiencia de 12 años

Resumen
Objetivo: El objetivo de este trabajo es realizar un estudio retrospectivo en pacientes con diagnóstico de paraganglioma temporal, analizando los resultados según la conducta terapéutica aplicada para cada caso.

Material y métodos: Se incluyó a 21 pacientes con diagnóstico de paraganglioma temporal tratados en nuestro servicio entre enero de 1999 y abril de 2011. A todos se les realizó un examen otorrinolaringológico, sistémico y de imagen. En 20 casos se realizó cirugía para su exéresis y en un caso se realizó tratamiento con cirugía estereotáctica.

Resultados: Se realizó embolización preoperatoria en 12 casos. Los abordajes quirúrgicos fueron: extirpación endaural, resección transcanal tras abordaje retroauricular, mastoidectomía radial ampliada con abordaje de receso facial y abordaje infratemporal. El diagnóstico de paraganglioma fue confirmado en todos los casos. Las complicaciones postoperatorias más frecuentes fueron: paresia transitoria del VII, parálisis del VIII, parálisis del IX y X par y fistula salival. No se observó recidiva de enfermedad con seguimiento entre 1 y 12 años. En un caso tratado con radiocirugía persiste el tumor, estabilizado.

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 intraoperatorio. La radiocirugía estereotáctica no permite la desaparición tumoral y persiste la enfermedad.

Introduction

Paragangliomas are structures formed by paraganglionic cells derived from the neural crest and from autonomic ganglion cells that relate to the neuroendocrine system. The term paraganglioma is the most appropriate to refer to the extra-adrenal paraganglionic tumours, although they have also been called glomus tumours or chemodectomas. In the cervicocephalic region, paragangliomas are related to the embryonic development of the branchial or pharyngeal arches, so they are known as branchiogenic paragangliomas.

The classification of paragangliomas varies depending on the parameter selected: patient age, secretory capacity, whether tumours are sporadic or familial syndromes, isolated or multifocal, benign or malignant, etc. In addition, there are classifications based on aspects such as location, extension, surgical approaches, immunohistochemical characteristics, and so on. No classification has been universally accepted so far. As for location, cervicocephalic paragangliomas are divided into 2 groups: temporal (which in turn are subdivided into jugular and tympanic, orbital, carotid, subclavian, and laryngeal) and vagal paragangliomas. With respect to extension of temporal paragangliomas, the most commonly used classifications are those by Fisch–Valavanis (a review of Fisch–Jenkins, from 1981) and by Glassock–Jackson.

Paragangliomas are highly vascularised but usually benign tumours (although there are malignant cases, especially in the functioning and familial). Their growth, albeit slow, can cause compression, displacement, and invasion of adjacent structures such as bone, blood vessels, dura mater, and cranial nerves.

It is the most common benign neoplasm of the middle ear and the second most common in the temporal bone. These tumours are more common in women, with a ratio of 1:3. Up to 10% of paragangliomas originating in the head and neck are familial and are transmitted in a modified autosomal dominant pattern. They may appear as bilateral tumours in 1%-2% of cases. In rare cases, they may metastasize (1%-3%). If we observe their secretory capacity or biochemical functionality, we note that less than 4% of cervicocephalic paragangliomas are active enough to be considered functional.

This article presents a retrospective and descriptive study of patients with a diagnosis of temporal paraganglioma, and discusses the general characteristics and results obtained after treatment of a total of 21 patients in our service.

Materials and Methods

We carried out a review of 21 patients who were diagnosed with temporal paraganglioma at our department between January 1999 and April 2011.

The main reason for consultation was pulsatile tinnitus, which appeared in 17 patients (81%). Other symptoms leading to consultation were transmission hearing loss (which occurred in 15 patients), otalgia (8), and otorrhea (5).

All patients underwent a complete ENT examination including an otomicroscopy and audiological studies, a high-resolution computed tomography (HRCT), and magnetic resonance imaging (MRI) and a detailed history was taken. In addition we also carried out an angiography at the interventional vascular radiology department for all cases.
Figure 1  Angiography of a Fisch type C2 right jugulotympanic paraganglioma.

diagnosed as Fisch types B and C \(^{11}\) (Fig. 1) to evaluate collateral circulation and study afferent and efferent tumour vessels. In the case of 1 patient with a Fisch–Valavanis type C2 secreting jugulotympanic paraganglioma (Fig. 2A–F) along with two retroperitoneal paragangliomas, we also performed a whole body scan with I\(^{31}\) and I\(^{123}\) metaiodobenzylguanidine (MIBG) on several occasions, as well as I\(^{123}\) MIBG single photon emission computed tomography (SPECT). In all cases, we determined 24-h urine fractionated catecholamines and metanephrines, 24-h urine vanillylmandelic acid levels and plasma free levels of catecholamines and metanephrines.\(^2\)

We used the Fisch–Valavanis classification for jugulotympanic paragangliomas in the staging of tumours.

The surgical approaches used were the endaural approach for 1 case of Fisch type A temporal paraganglioma (Fig. 3A–F), the transcanal retroauricular approach for Fisch type A tumours, radical tympanomastoidectomy for type B, and type A infratemporal approach for Fisch type C tumours (Fig. 4).

Results

In our series, the age interval was 29–77 years, with a mean of 39.6 years. We also found a female predominance (13/21).

Upon otoscopic examination, 16 patients (76.2%) showed a purplish tympanic membrane; a polypoid inflammatory formation in the external auditory canal was also observed in 4 (19%) and only 1 patient (4.8%) presented otorrhea associated with a purplish mass.

Figure 2  (A and B) Coronal CT images of a Fisch type C2 right jugulotympanic paraganglioma. (C–E) Axial CT images of Fisch C2 right jugulotympanic paraganglioma.
The plasma concentrations of free catecholamines and metanephrines, as well as 24-h urine determinations of catecholamines, fractionated metanephrines and vanillylmandelic acid were all within the normal limits, except in 1 case of Fisch–Valavanis type C2 jugulotympanic paranglioma, which turned out to be secreting paranglioma (this patient also presented two retroperitoneal parangliomas of 3.3×2.8 cm and 1×1.1 cm, which were intervened by the Urology Department during a first surgery; she is currently awaiting the results of genetic studies to rule out multiple paranglioma syndrome).

According to the Fisch–Valavanis classification, 43% (n=9) of patients presented type A tumour, 24% (n=5) type B, and 33% (n=7) type C2–C3.

We carried out an arteriographic study of the supra-aortic trunks in all cases and 12 patients (57.1%) underwent tumour embolisation 72 h before surgery. In 1 case of advanced age and poor general condition, we decided to perform stereotactic radiosurgery.

The distribution of the surgical approaches carried out is shown in Table 1.

According to the Makek\textsuperscript{12} classification, there was infiltration of the facial nerve in 3 cases: in 1 (4.8%) patient in grade II and in 2 (9.5%) patients in grade III.

The diagnosis of paranglioma was confirmed in all cases by anatomopathological examination of the surgical specimen.

The postoperative complications for each type of approach employed are summarised in Table 2.

Surgery achieved control of tumours, and no recurrence was observed in any patient after a follow-up period between 1 and 12 years. In the patient who received radiosurgery, we observed tumour persistence without growth, with areas of tumoral necrosis and with no increase in size.

### Discussion

Paranglioma of the jugular bulb is the most common benign tumour of the middle ear. It is a slow-growing, highly vascular tumour, but with potential for extension and involvement of structures of the skull base, nerves and blood vessels.
Most series report a predominance of females. Pulsatile tinnitus coincident with the pulse and hearing loss are the most frequent forms of clinical presentation of jugular tympanic paragangliomas. Hearing loss is generally conductive, although a sensorineuronal component can also be found, in which case the possibility of labyrinthine invasion should be kept in mind. Other symptoms include occasional otorrhea, otalgia, and vertigo. Up to 15% of these tumours are asymptomatic. Physical examination generally reveals a hypotympanic or mesotympanic mass, which is characteristic but not pathognomonic. It is also possible to observe a formation with polypoid or inflammatory appearance in the external auditory canal.

The existence of symptoms of involvement of low cranial nerves indicates extensive injury by the temporary paraganglioma.

Facial paralysis by involvement of the 7th cranial nerve is the most common form of presentation of neural disease, with this invasion being most frequently located in the mastoid portion of the nerve. Next in order of frequency are involvement of the 10th, 9th, 11th, and 12th cranial nerves, although the degree of nerve dysfunction observed in the preoperative period is not an indicator of neural condition.

Although histochemical and ultrastructural studies have proved that all paragangliomas produce catecholamines, less than 4% of cervicocephalic paragangliomas are active enough to be considered as functional. In those cases where they are functional, preoperative treatment should include alpha and beta blockers.

Preoperative evaluation includes HRCT to obtain a good analysis of the bony structures of the skull base, tumour size, and areas of bone erosion. Gadolinium-enhanced MRI and angiography allow the study of tumour vascularisation, extension and its relationship to surrounding structures. Digital subtraction angiography (this was not used in our cases) enables a diagnosis and acts as a guide for embolisation.

The treatment of glomus includes different therapeutic modalities, such as surgical resection, stereotactic radiosurgery, radiotherapy, chemotherapy, use of intratumoral sclerosing agents (with the aim of necrotizing and reducing the tumour), metabolic therapy with I-MIBG, treatment with somatostatin analogues (octreotide), and expectant attitude with regular controls.

Once a surgical attitude has been adopted, preoperative embolisation of these lesions reduces intraoperative bleeding and surgical time and results in complete tumour resection in all cases.

Complications observed in the postoperative period include CSF leak, imbalance, and tissue ischemia, as well as those resulting from damage to the cranial nerves and major cardiovascular, pulmonary, and cerebral complications.

Glomus is considered as a relatively non-radiosensitive tumour. Fractionated radiotherapy reports 7% of minor complications and 2%-3% of major complications. Benign tumours in patients undergoing radiation therapy may develop secondary tumours; there is an estimated 2.7% chance of developing secondary malignancy within 10 years.

Stereotactic radiosurgery with gamma-knife appears to enable a local control associated with low morbidity and essentially no mortality. It is thus recommended in patients with advanced age, poor general condition, advanced tumour, rejection of surgery, or unresectable tumour.

### Conflict of Interests

The authors have no conflicts of interest to declare.

### References