GLOMUS JUGULARE TUMORS. A SERIES OF 24 CASES

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ABSTRACT

Objective: describe the surgical technique and experience of a neurotology surgical team working is Lisbon, Portugal, in the surgical treatment of glomus jugulare tumors.

Methods: the retrospective study included 24 patients operated between 1988 and 2007. Data was obtained from the medical records and from a recent reevaluation of the patients. Data evaluation included: clinical manifestations, preoperative embolization, surgical technique, tumor removal, morbidity, mortality and evolution of residual lesions.

Results: symptoms included hearing loss (33%), pulsatile tinnitus (21%) and paralysis of the cranial nerves X (29%), VII (25%) and XII (21%). Preoperative embolization was performed in 92% of the cases. In all cases, an infratemporal fossa approach type A was used for glomus tumor removal. Total removal of the tumor was obtained in 11 (45%) cases. Only 21% of the patients had a grade III-IV House-Brackmann or worse after 12 months. Two patients died from pneumonia. Five (21%) of the patients required a second surgery but only one patient required radiotherapy after incomplete removal of the tumor.

Conclusions: glomus jugulare tumors and their treatment have significant morbidity and mortality. Neurotology surgical teams should be created to develop surgical expertise and competence in the treatment of the disease, in order to offer the best options and outcome to the patients.

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INTRODUCTION

Glomus jugulare tumors are neuroendocrine tumors, originating in the paraganglionic cells, which are elements of the neural crest[1]. They are histologically benign, but behaves aggressively, eroding the jugular foramen and invading the temporal bone, the skull base, the neck and, less often, the cranial cavity.

First considered irresecable, the surgical treatment of these lesions was stimulated by the Swiss otolaryngologist Ugo Fisch[2], who designed the novel technique named Infratemporal Fossa Approach type A.

Currently there is consensus that these tumors should be treated by multidisciplinary teams that include otolaryngologists, neurosurgeons and interventional radiologists. These teams should combine competencies to perform advanced surgical techniques designed to explore the ear and the temporal bone, the neck, the skull base, the related vessels, the intracranial structures, and to monitor Intraoperatively the involved cranial nerves[3].

Presently, the Egas Moniz University Hospital is the major referral center in the surgical treatment of the jugulare tumors in Portugal. A first generation of surgeons pioneered this operations in 1988 (JPV) and presently this surgical activity is developed by a surgical team, comprising an Otolaryngologist (PE) and a Neurosurgeon (GNA).

The purpose of this paper is to describe the surgical experience of the Departments of Otolaryngology and Neurosurgery, Hospital de Egas Moniz, in the surgical treatment of jugulare glomus tumors, between 1988 and 2007.

METHODS AND MATERIALS

STUDY DESIGN

Retrospective study based on medical record review and in a recent reevaluation of the patients. .

PARTICIPANTS

The study included all the 24 patients who underwent surgical treatment for jugulare glomus tumors at the Hospital de Egas Moniz, in a 20-year period from 1988 to 2007.

A total of 24 patients aged between 28 and 74 were enrolled, 17 (71%) females and 7 (29%) males.

Patients with glomus tumors limited to the middle ear without involvement of the jugular foramen were excluded, because these tumors were treated by classical tympanotomy or tympanomastoidectomy approaches.

Recent observation of 8 of the patients completed the data of the medical records.

PARAMETERS EVALUATED

Study variables are shown in Table 1

Table 1: Study variables

Clinical manifestations
Preoperative embolization
Surgical technique
Tumor removal
Morbidity
Mortality
Evolution of residual lesions

RESULTS

The most common manifestations of the disease were hearing loss, pulsatile tinnitus and deficits in cranial nerves VII and IX-X. A list of the symptoms and signs at presentation is presented in Table 2.

Table 2: Clinical manifestations

Symptom or sign	*n*	*%*
Hearing loss	8	33%
Paralysis of the cranial nerve X	7	29%
Facial nerve paralysis	6	25%
Pulsatile tinnitus	5	21%
Paralysis of the cranial nerve IX	5	21%
Paralysis of the cranial nerve XII	5	21%
Vascular lesion of the external auditory canal	4	17%
Otorrhagia	3	13%
Paralysis of the cranial nerve IX	2	8%
Otalgia	1	4%
Epistaxis	1	4%

Preoperative embolization was performed in 22 (92%) of the patients. Vascular supply was seen to come from the ascending pharyngeal, occipital or ascending palatine arteries.

The surgical technique was the infratemporal fossa approach type A, as described by Ugo Fisch, in all the cases.

SURGICAL TECHNIQUE

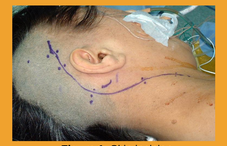

Figure 1. Skin incision

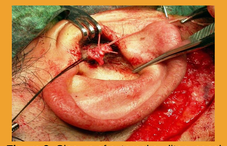

Figure 2. Closure of external auditory canal

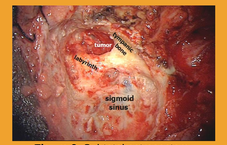

Figure 3. Subtotal petrosectomy

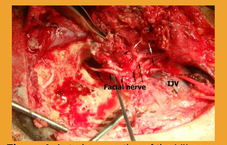

Figure 4. Anterior rerouting of the VII nerve

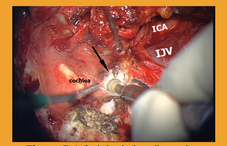

Figure 5. Infralabyrintine dissection

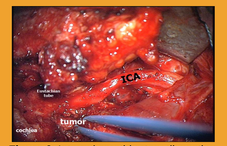

Figure 6. Internal carotid artery dissection

Surgical resection was complete in 11 patients (46%) and incomplete in 13 patients (54%). Incomplete resections were justified by the infiltration of the internal carotid artery, considered irresecable.

Two patients (8%) died from aspiration pneumonia in the immediate postoperative period. These patients were elderly patients operated at the beginning of the series, who developed a paralysis of the cranial nerve X postoperatively.

75% of the patients had a facial paralysis in the immediate postoperative period, but the number of patients who maintained a significant facial paralysis (House Brackmann grade III or worse) beyond 6 months to 1 year was reduced to 5 (21%). Other causes of morbidity were dysphagia, aspiration pneumonia, and CSF fistula, which together occurred in 13 patients (54%).

Of the 24 patients, 5 (21%) underwent a second surgery due to recurrence. Only one patient required radiotherapy after incomplete removal of the tumor.

DISCUSSION

This poster presents the results of a series of 24 patients with glomus jugulare tumors, operated at the Egas Moniz University Hospital in the first 20 years after this surgical treatment was initiated in this institution.

It is a small series that reflects the rarity of the disease. In Portugal, most of the cases are referred to our institution for surgical treatment.

Glomus jugulare tumors are slow growing benign tumors and mortality should not be expected as a result of the surgical treatment. The 2 deaths of our series occurred in elderly patients who had intact cranial nerve X function before the surgical treatment and had a paralysis of this cranial nerve after the surgical procedure. They did not tolerate the aspiration, developed pneumonia and died. These cases made us more cautious in advising surgical treatment for older patients with extensive tumors and intact cranial nerves, and we had no further mortality.

Facial nerve preservation is mandatory in this benign disease. Infratemporal fossa approach type A is a very delicate operation that requires intraoperative monitoring and expertise in the surgery of the intratemporal and extratemporal portions of the facial nerve, that were fully developed by our surgical team with cadaveric training.

Preoperative embolization is critical to reduce intraoperative bleeding. The patients should undergo carotid and cerebral angiography followed by embolization of the tumor vessels 2 to 3 prior to surgery.

Recurrence should be minimized by complete removal of the tumor. However, subtotal removal should be considered when noble structures, like the internal carotid artery, the facial nerve or the cranial nerve X, are firmly infiltrated. In cases of subtotal removal, only a few number of the patients require further surgery.

CONCLUSIONS

When surgical centers have the responsibility to treat patients with uncommon and complex cases, they should periodically reassess, review, analyze and publish their results.

The results of the first 20 years of the surgical treatment of glomus jugulare tumors in our institution allow our new generation of surgeons to strive for better results in the future.

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