Intradural Jugular Paragangliomas: Complications and Sequelae

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ABSTRACT

Objectives: To present the long-term results of a group of patients who underwent surgery for intradural jugular paragangliomas. We discuss the complications, sequelae, and evolution of recurrences and behavior of residual tumors. **Methods:** From 1989 to 2002, 11 patients with intradural jugular paragangliomas underwent surgery using different approaches. The paragangliomas were grouped according to the classification of Fisch. **Results:** Total removal of the paraganglioma was possible in eight patients, while a subtotal resection was achieved in the other three cases. The tumor remnant remained stable in two patients but continued to grow in one. Recurrence was observed in one patient. There were preoperative deficits in cranial nerves IX to XI in four patients and of XII in two patients. Deficits of cranial nerves IX to XI were acquired as a result of surgery in three cases and of XII in another. Six patients had a pure-tone average of 45 to 75 dB while the others had dead ears. Persistent dysphagia and dysphonia were managed in two patients by injection of the paralyzed cord with fat and subsequent medialization of the vocal cord by thyroplasty. Two patients had a cerebrospinal fluid leak. **Conclusions:** Surgical results in patients with extensive jugular paragangliomas are consistent and offer an acceptable quality of life even in advanced cases.

KEYWORDS: Jugular paraganglioma, skull base, intradural extent, classification, complications

Although there have been many improvements in imaging, anesthesia, intensive care, surgical techniques, and radiotherapy over the past few decades, the treatment of jugular paraganglioma is still very controversial. Every treatment strategy risks the functional integrity of cranial nerves and

damage to major vascular structures, each of which has a negative impact on quality of life.¹⁻¹⁵

There is no consensus on how to treat paragangliomas that have an intradural extension. Debate rages over best management of these patients, especially with regard to the preparation

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Table 1 Distribution of the Patients

Patient	Sex	Side	Age	Class	Approach	Stage	Removal
1	F	Right	24	C1De1Di1	STP; RS	1	Total
2	F	Right	52	C2De2Di1	IT; TC	1	Total
3	F	Left	55	C2De1Di1	IT; RS	1	Total
4	M	Left	31	C2De2Di2	IT; RS	2	Subtotal
5	F	Left	38	C3De1Di1	IT; RS	2	Total
6	M	Left	56	C3De2Di1	IT; RS	1	Total
7	M	Right	61	C2De1Di1	IT; RS	2	Total
8	F	Right	34	C3De2Di2	IT; RS	2	Subtotal
9	F	Left	48	C2De1Di1	IT; RS	1	Subtotal
10	M	Right	51	C2De2Di2	IT; RS	2	Total
11	F	Left	32	C3De2Di1	IT; RS	1	Total

STP, subtotal petrosectomy; RS, retrosigmoid; IT, infratemporal; TC, transcochlear.

for tumor resection from the infratemporal (internal carotid) or intradural (vertebral/basilar) artery systems, the treatment of recurrences, and the role of radiotherapy.¹⁻¹⁵ The few papers on this topic are sometimes difficult to compare because of different strategies adopted and different methods of reporting the results.

This study presents the long-term outcomes of a group of patients who underwent surgery for intradural jugular paragangliomas. We discuss the complications, sequelae, and evolution of recurrences and the behavior of residual tumor.

MATERIALS AND METHODS

This study documents 11 patients with jugular paragangliomas who underwent surgery between 1989 and 2002. Demographic data on these patients in relation to sex, age, and side of tumor are summarized in Table 1. All of the patients presented with intradural extension into the cerebello-pontine angle. The temporal bone was involved to varying degrees. The paragangliomas were grouped according to the classification of Fisch³ (Table 1, Fig. 1).

Different surgical techniques were used. In nine patients the infratemporal and retrosigmoid approaches were combined to access the cerebello-

pontine angle. In one of the remaining patients, retrosigmoid surgery was combined with a subtotal petrosectomy, and the last patient was managed by a combined infratemporal and transcochlear approach^{1,2,4,8,14}. The operation was performed in one stage in six patients and in two stages in five.

All of the patients underwent yearly magnetic resonance imaging (MRI) for the first 5 years after surgery. In the absence of recurrences or residual tumors, examination with imaging techniques was subsequently performed at 2-year intervals along with an evaluation of cranial nerve function.

Figure 1 Preoperative magnetic resonance image of an intradural jugular Di2 paraganglioma.

Figure 2 Sealing the jugular foramen with abdominal fat after removing a jugular paraganglioma and sacrificing cranial nerves IX to XI.

Figure 3 Postoperative magnetic resonance image shows total removal of the paraganglioma.

RESULTS

Total tumor removal was achieved in eight patients (Figs. 2 and 3). In the other three cases a subtotal resection was performed. The tumor was excised as completely as possible and what was left was coagulated (Table 1). At the most recent follow-up, the tumor had remained stable in two of these patients after 8 and 6 years, respectively. In the remaining patient, the tumor remnants were found to have increased in size over a follow-up period of 4 years.

The amount of regrowth was limited and a wait-and-see strategy was adopted. Recurrence was observed in one patient (originally a C2De1Di1) who was thought to have undergone a total removal. This residual tumor was subsequently removed.

Anterior transposition of the facial nerve was performed in nine patients; two had House-Brackmann⁷ class III facial function 1 year after surgery (Table 2). One patient with a complete facial palsy preoperatively was managed with a sural nerve interposition graft. This facial nerve function

Table 2 Pre- and Postoperative Function of Cranial Nerves VII to XII

Patient	VII* Preop	VII* Postop	IX-X-XI Preop	IX-X-XI Postop	XII Preop	XII Postop	VIII Preop	VIII Postop
1	I	III	Normal	Normal	Normal	Normal	Normal hearing	PTA 55 dB
2	I	II	Normal	Normal	Normal	Normal	Dead ear	Dead ear
3	I	I	Normal	Paretic	Normal	Normal	Normal hearing	PTA 45 dB
4	I	II	Normal	Paretic	Normal	Paretic	Normal hearing	PTA 53 dB
5	I	III	Paretic	Paretic	Normal	Normal	Normal hearing	Dead ear
6	I	II	Paretic	Paretic	Normal	Normal	Dead ear	Dead ear
7	I	II	Normal	Paretic	Normal	Normal	PTA 35 dB	PTA 65 dB
8	VI	IV	Normal	Normal	Normal	Normal	Normal hearing	PTA 50 dB
9	I	II	Normal	Normal	Normal	Normal	Normal hearing	Dead ear
10	II	III	Paretic	Paretic	Paretic	Paretic	PTA 48 dB	Dead ear
11	I	II	Paretic	Paretic	Paretic	Paretic	PTA 55 dB	PTA 75 dB

*Cranial nerve VII grades follow the House-Brackmann scale.
PTA, pure tone average; dB, decibels.

recovered to House-Brackmann class IV. Deficits of cranial nerves IX to XI and XII were acquired preoperatively in four and two patients, respectively. Postoperatively, the damage involved nerves IX to XI in another three cases and nerve XII in one other (Table 2).

Postoperative hearing function ranged from a pure-tone average (PTA) of 45 to 75 dB in six patients; the other five patients had dead ears. The postoperative course in one patient was complicated by bronchopneumonia which was treated medically. This patient required temporary tube feeding. Persistent dysphagia and dysphonia in two patients were first treated by infiltrating fat into the paralyzed cord and then by thyroplasty with medialization of the vocal cord.

Two patients developed cerebrospinal fluid (CSF) leaks which settled spontaneously in one patient, while the other required surgical closure. The internal carotid artery was preserved in all patients and no vascular accidents (stroke) occurred.

DISCUSSION

Jugular paragangliomas have always presented a challenge because of their behavior, the complexity of the anatomical site involved, and the importance of the structures that they affect. The severity of potential complications can have a very negative impact on the patient's quality of life. Moreover, the risk of recurrence is high even after prolonged follow-up.¹⁻¹⁵ The choice of treatment is certainly not straightforward. It is further complicated by the relative rarity of the disease. Some of the few series published in the literature lack complete data and this makes comparisons difficult. As a result of the pioneering work undertaken by Fisch in the late 1970s,² clear indications and innovative surgical techniques were developed to replace traditional radiotherapy as the first line of treatment. Nonetheless, the incidence of postoperative complications and sequelae remains

controversial.⁵ The situation has been further complicated by developments in radiosurgery, but these are viewed with suspicion and many questions remain without satisfactory answers. For example, should the lack of tumor growth after radiotherapy be considered a success, or is it just a feature of natural history of the tumor, which is known to grow slowly and insidiously? Does the fibrosis arising from radiation therapy increase the risk of damage to the nerves should surgery become necessary? Is there a risk of damage to the underlying brain? Is the patient at significant risk of developing malignancies in the radiation field? These uncertainties make it important to present surgical results to obtain valid data for realistic and useful comparisons.

In this study, we examined patients with intradural extensions, which are recognized as the most difficult to treat by surgery. Jackson and colleagues⁸ and Briner and associates³ devised different classification systems for jugular paragangliomas. Although the latter classification has proved to be the more popular, only a few studies have grouped cases using the most recent version. The difficulties in comparison are immediately clear when one tries to evaluate the incidence of total resection of the tumor and the frequency of recurrences according to the area of tumor growth. Careful analysis of the English literature reveals that 70% and 96% of the surgically treated tumors were reported as total resection.⁵ Subtotal resection is more frequent in tumors with intracranial extent, class D tumors according to Fisch.³ The most recent version of the classification groups both extradural (classes De1 and De2) and intradural (classes Di1 and Di2) tumors with intracranial spread above or below 2 cm in size, respectively. When the percentage of subtotal resections is considered, some reports have to be excluded from any comparison. Although Jackson and coworkers⁸ and Pareschi and others¹² reported 10% and 16% subtotal resection rates in patients with intracranial tumors, respectively, they did not specify the incidence of extra- and intradural lesions. Tran Ba Hui et al¹⁵ classified the jugular paragangliomas that they

treated surgically using the Fisch classification and reported that 30% were subtotal resections. However, the authors did not indicate the distribution of those paragangliomas by class. Oghalai et al¹¹ described a series of six class Di1 and Di2 patients with subtotal resection in five cases, but did not give more precise detail.

The only two complete studies that can be compared were published by Briner et al³ and Sanna et al.¹³ The former reported subtotal resections in 17% of surgically treated cases; five were class Di2 and one was class C2. Sanna et al¹³ reported a subtotal resection rate of 9.2% (five cases), with a clear majority of class Di2 (three Di2, one Di1, and one C1). It is notable that subtotal resection was mainly associated with extensive intradural tumors (class Di2). More specifically, 5 of the 14 patients treated by Fisch³ and 3 of the 10 patients treated by Sanna et al¹³ were class Di2. Our experience confirms this trend with a clear predominance of intradural tumors over 2 cm.

An interesting subject of debate is the clinical evolution of patients who have undergone subtotal removal. In Fisch,³ repeated MRI did not reveal any further growth of the tumor residue in three of five patients. The patients in whom growth was seen received radiotherapy after which the recurrent tumor presumably remained unaltered, although the clinical outcome was not actually stated. Sanna et al¹³ described four cases of subtotal resection of tumors with extensive intradural growth and particularly aggressive biological characteristics. Two of these were subsequently treated with radiosurgery and one with embolization; the last patient was not treated. In only two cases did the size of the recurrent tumor increase and then only slightly: one patient had been treated with embolization and another subjected to radiosurgery. In our series of three subtotal resections in Di2 patients, the tumor remnant remained stable in two cases and grew in one case. The variable behavior of residual tumor was also confirmed by Tran Ba Hui and colleagues.¹⁵ Although those authors grouped the remnants without classifying them according to Fisch, they did note that in 73% of the subtotal

resections, tumor residue tended to remain stable or grow at a very slow rate. They concluded that leaving less than a centimeter of residual tumor did not make any substantial difference to the final outcome and modified their therapeutic strategy on this basis. They deliberately opted for a subtotal removal in patients over 60 years of age with intact cranial nerve function. They argued that these patients were at great risk of postoperative complications with total tumor removal, such as damage to the internal carotid artery or cranial nerves, or CSF leak.

In our opinion, the proposed indications for treatment in reported series must be considered carefully because MRI now offers the possibility of better follow-up and because treatment methods are constantly evolving and improving the patient's quality of life.

In analyses of patients undergoing total gross removal, Briner and associates³ reported recurrences in 3% of the cases that were originally class B. Sanna et al¹³ reported recurrences in 10.2% of cases with extracranial or extradural intracranial lesions, two of which originally had a substantial intradural component (Di2 or a combination of extradural and intracranial extensions). Tran Ba Hui et al¹⁵ documented a 6.6% recurrence rate but did not specify the original class of the paraganglioma. In our sample, recurrence developed in one patient with a C3De1Di1 lesion. The limited rate of recurrence is probably related to anterior transposition of the facial nerve which is a fundamental surgical step in the infratemporal approach.^{2,3,8,12,13,15} Tran Ba Hui et al¹⁵ contest this, as according to their data not mobilizing the facial nerve did not compromise the efficacy of tumor removal.

Anterior transposition of the facial nerve increases the incidence of postoperative palsy to ~20%, with an inevitable negative impact on the patient's quality of life. Briner et al³ and Sanna et al¹³ mobilized the facial nerve in all cases. Our experience was similar, which makes it difficult to assess whether surgery without mobilization of the facial nerve is inferior from an oncological point of view. This should be investigated further.

In conclusion, we were able to achieve reproducible surgical results in patients with extensive jugular paragangliomas. In our hands surgery offered a relatively acceptable quality of life, despite the severity and aggressiveness of these tumors.

Other complications and postoperative deficits in cranial nerves VIII and IX to XII did not differ from those reported in the literature. The preservation of cranial nerve IX to XI function in tumors with C3 and D4 spread is problematic due to perineural infiltration. New deficits in the cranial nerves are to be expected postoperatively, varying from 26 to 42% for the glossopharyngeal nerve, 13 to 28% for the vagus, 25 to 26% for the accessory nerve, and 5 to 21% for the hypoglossal nerve.⁵ These outcomes are 25 to 30% worse for intradural paragangliomas, confirming that these are the most complicated to treat surgically.

Patients recover from dysphagia and dysphonia relatively quickly when the lesions are unilateral by natural compensation and their quality of life is acceptable. Surgery is inadvisable when the contralateral side is paretic, or in elderly patients whose capacity for compensation is reduced.

Finally, it is worth noting that the incidence of CSF leakage was not particularly high. This confirms that the procedures used to close the meningeal compartments are effective, even in cases with extensive intradural tumors.¹³

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