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Surgical treatment of jugular foramen schwannomas

Arkadiusz Nowak *, *Tomasz Dziedzic*, *Tomasz Czernicki*, *Przemysław Kunert*, *Andrzej Marchel*

Department of Neurosurgery, Medical University of Warsaw, Warsaw, Poland

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ABSTRACT

Objective: We present our experience with surgery of jugular foramen schwannomas with special consideration of clinical presentation, surgical technique, complications, and outcomes.
Methods: This retrospective study includes ten patients with jugular foramen schwannomas treated by the senior author between January 2007 and December 2012. Three patients had undergone partial tumour resection elsewhere. The initial symptom for which they sought medical help was hearing loss, dysphagia, hoarseness, and shoulder weakness. Preoperative glossopharyngeal and vagal nerve deficits were the most common signs. In our series, tumour extension was classified according to Kaye-Pellet grading system. In two cases the tumours were classified into type A and 8 patients presented with type D tumours. A retromastoid suboccipital craniotomy was performed for type A tumours and modifications of cranio-cervical approach were suitable for type D.
Results: No death occurred in this series. Four patients deteriorated after surgery: in two patients preoperative cranial nerve deficits deteriorated after surgery while new cranial nerve palsy occurred in 2 other patients. In four patients, the cranial nerve dysfunction had improved at the last follow-up examination. In all other patients, the cranial nerve dysfunction remained the same. One patient experienced tumour recurrence over a follow-up period of 40 months. This patient underwent a successful second surgery without further evidence of tumour growth.
Conclusions: Jugular foramen schwannomas can be radically managed with the use of skull base surgery techniques. However, the surgical treatment of jugular foramen schwannomas carries a significant risk of the lower CN deficits.

1. Introduction

Intracranial schwannomas comprise approximately 8% of all primary brain tumours [1]. Schwannomas arising from sites other than vestibular nerves are rare, constituting 2.9–4% of such lesions [2,3]. Among these, the most common site is the trigeminal nerve (40%), followed by the facial (23%) and then the lower cranial nerves (20%) [2,4]. Fewer than 200 cases of schwannomas arising from the jugular foramen have been

* *Corresponding author at*: Department of Neurosurgery, Banacha 1a, Warsaw, Poland. Tel.: +48 606 787 433; fax: +48 225991574.
E-mail address: arkady.n@wp.pl (A. Nowak).
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reported in the literature [5]. Schwannomas located at the jugular foramen may arise from the glossopharyngeal, vagus or accessory nerve. Some authors classify hypoglossal schwannomas with jugular foramen schwannomas because of clinical similarity as well as difficulty in locating the nerve of origin. The position of the tumour depends on its point of origin from the nerves as they pass through the jugular foramen [6]. Based on this characteristic Kaye and Pellet [7,8] proposed grading system that classifies jugular foramen schwannomas into four groups: Type A tumours are primarily intracranial with only a small bony extension, Type B tumours are primarily within the bone with or without an intracranial component, Type C are primarily extracranial with only minor extension into bone or into the posterior fossa, and Type D are dumbbell-shaped tumours with both an extra- and an intracranial components. Surgical management of jugular foramen schwannomas is complex and difficult due to the proximity and involvement of critical neurovascular structures.

We present our experience with surgery of jugular foramen schwannomas in regard to clinical and radiological features, details of the surgical methods used, complications and surgical outcomes.

2. Materials and methods

2.1. Patient population

We retrospectively reviewed 10 cases of jugular foramen schwannomas treated by the senior author (A.M.) in our centre between 2007 and 2012. Patients who were diagnosed as having neurofibromatosis type 2 were excluded from this study. Schwannoma diagnosis was established based on tumour histopathology. Our cohort included 4 women and 6 men. Patient's age varied from 25 to 54 years (mean 38.5 years). The initial symptom for which they sought medical help was hearing loss, dysphagia, hoarseness, and shoulder weakness. In one case (No. 7) there had been no neurological symptoms before surgery: the large jugular foramen schwannoma had been diagnosed in the course of rare epileptic seizures. On examination glossopharyngeal and vagal deficits were the most common signs. None of the patients had hydrocephalus diagnosed. The most important demographic and clinical features in our series are outlined in Table 1. Three patients had undergone previous non-radical surgery elsewhere: one patient (No. 5) had had deafness, facial palsy, and facial desaesthesia and two other patients (No. 9 and No. 10) were admitted to the department with cranial nerve VII, IX, X, and XI deficits and deafness present after the first surgery. Another patient (No. 8) had undergone a few years earlier unsuccessful exploration surgery of tumour located in the neck without creating additional neurological deficits. One patient (No. 6) had undergone complete surgery of Astrocytoma protoplasmaticum of lateral ventricle with subsequent conformal radiotherapy 19 years before diagnosis of jugular foramen schwannoma.

2.2. Neuroimaging studies

Radiographic evaluation included brain Magnetic Resonance Imaging (MRI) scans as well as temporal bone Computed Tomography (CT). The nature of the tumour, extensions and its relationship to neighbouring structures were studied using MR images. CT scans with the aid of bone algorithm were obtained for analysis of bone involvement and position of jugular bulb. Digital Subtraction Angiography (DSA) and Magnetic Resonance Angiography (MRA) were performed in order to evaluate the position and size of the jugular bulb, tumour vascularization and the size, dominance and tributaries (superior petrosal, inferior petrosal, and vein of Labbe) of the transverse and sigmoid sinuses (DSA in 7 patients, MRA in 3 patients). Extensive audiological evaluations were done before and after surgery.

2.3. Surgical technique

The choice of surgical approach was determined by the type of tumour extension identified. The retrosigmoid approach was suitable for Type A jugular foramen schwannomas (Fig. 1). In one case (No. 2) the dorsal part of the jugular foramen was opened extradurally and the dural incision was extended to the jugular foramen. For Type D tumours (Figs. 2 and 3), depends on tumour extension and patency of the jugular bulb various modifications of a cervical-transmastoid approach were used. The general approach consists of:

Table 1 – Clinical presentation of 10 patients with jugulare foramen schwannomas.

Case no.	Age (years)	Sex	Initial symptoms	Cranial nerve deficit					
				V	VII	VIII	IX–X	XI	XII
1	38	M	Decreased hearing		+	+	+		
2	50	M	Decreased hearing			+			
3	25	F	Shoulder weakness				+	+	
4	26	M	Shoulder weakness					+	
5	35	M	Decreased hearing	+	+	+		+	
6	31	F	Dysphagia				+		
7	38	F	Seizures						
8	54	M	Hoarseness				+	+	+
9	41	M	Hoarseness		+	+	+	+	
10	47	F	Dysphagia		+	+	+		

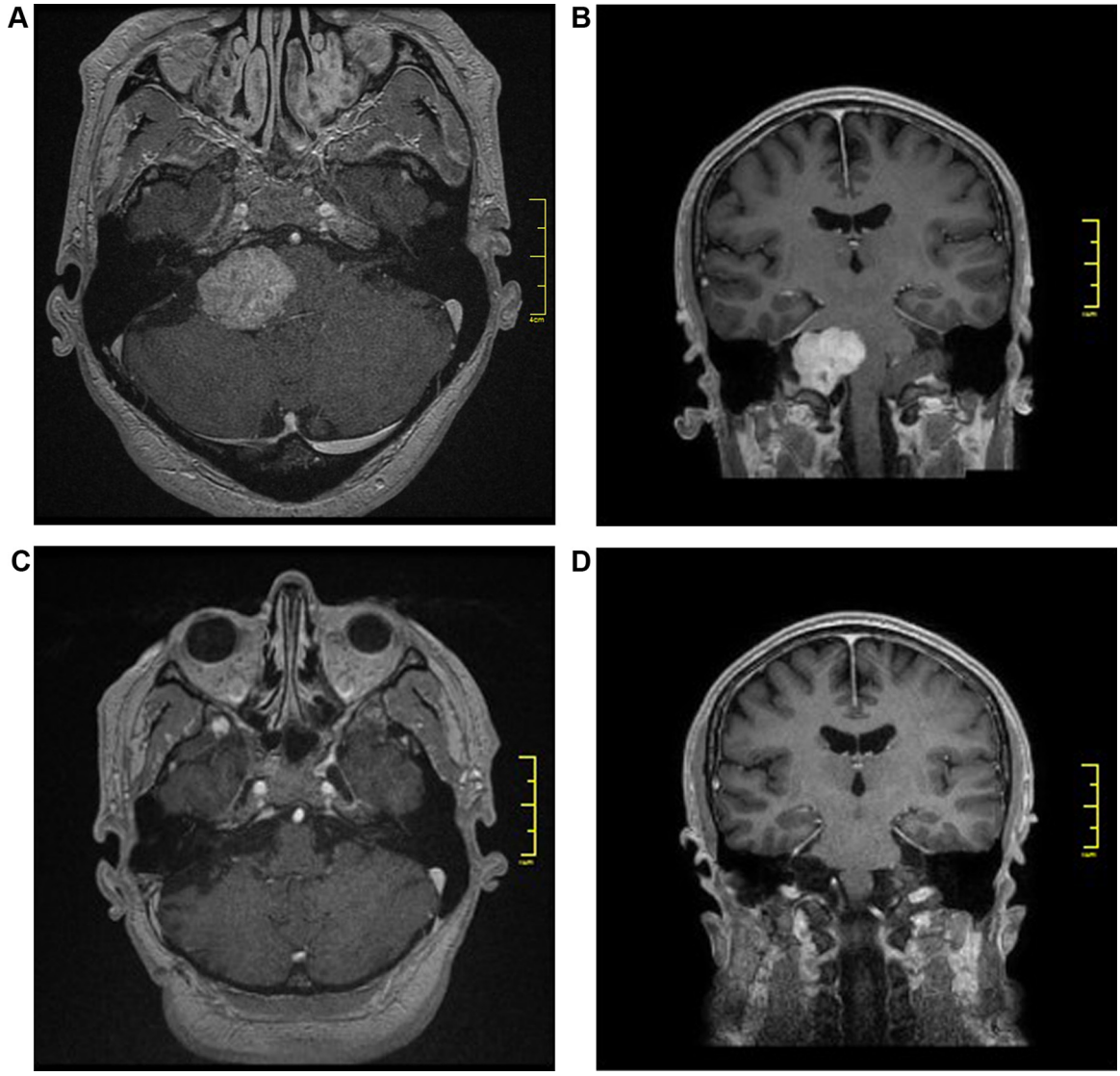

Fig. 1 – Preoperative axial (A) and coronal (B) T1 contrast enhanced MR image demonstrating a Type A jugular foramen schwannoma. Postoperative axial (C) and coronal (D) T1 contrast enhanced MR image obtained after complete resection of the tumour.

- identification of the internal carotid artery, external carotid artery, internal jugular vein, and the 9th, 10th, 11th and 12th cranial nerves in the cervical region
- the sternocleidomastoid muscle is dissected, mobilized and reflected inferiorly, C1 transverse process and vertebral artery are exposed, the facial nerve is identified at the entrance to the parotid gland
- a mastoidectomy is followed by posterior petrosectomy (retrolabyrinthine or translabyrinthine in case of preoperative deafness)
- a small craniectomy is performed and to further expand the exposure, the posterior part of the occipital condyle and the jugular tubercle are removed, thus opening the jugular foramen dorsolaterally
- the dura is opened anterior (extending the incision to the jugular foramen) and posterior to the sigmoid sinus for tumour removal in the two corridors; if tumour involves both the jugular bulb and the internal jugular vein a transjugular approach with ligation of internal jugular vein is necessary
- the extracranial portion of the tumour extending in the neck is resected in the last step.

Cranial nerves 7–12th function was monitored intraoperatively in 5 patients. To avoid cerebrospinal fluid leakage, lumbar drainage was placed for 5 days. A nasogastric tube was kept in place for a week in every patient. If swallowing function was preserved oral nutrition was started over 1 week after surgery. The extent of tumour resection was evaluated based on the intraoperative findings and postoperative brain MRI (made 3 months after the operation). The patient's condition was assessed at discharge from the department along with a long-term follow-up based on neurological examination and

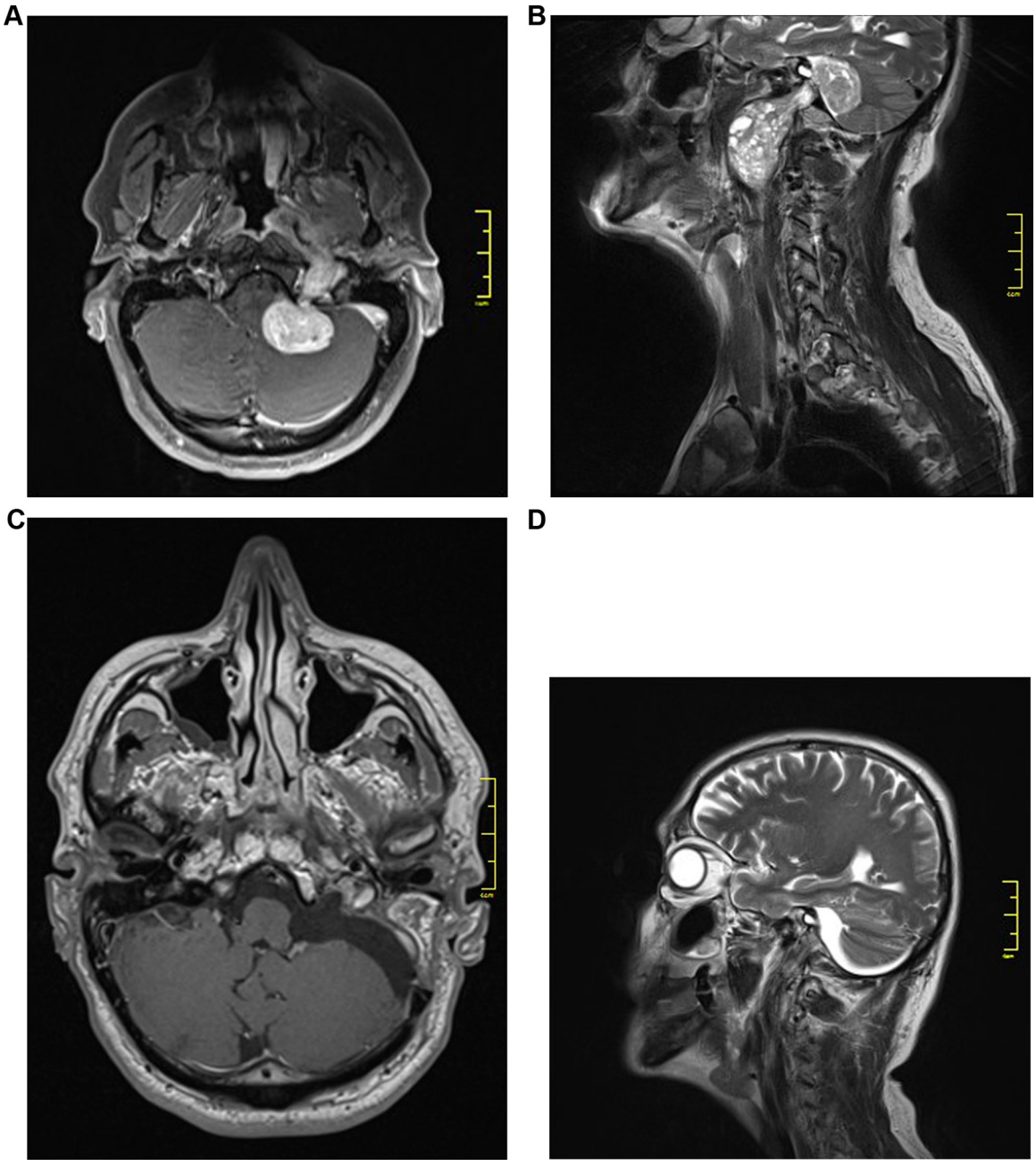

Fig. 2 – Preoperative axial T1 contrast enhanced (A) and sagittal T2 (B) MR image demonstrating a dumbbell-shaped jugular foramen schwannoma. Postoperative axial T1 contrast enhanced (C) and sagittal T2 (D) MR image obtained after complete resection of the tumour.

brain MRI in all of the patients. The postoperative follow-up period ranged from 6 to 71 months (mean 28 months).

3. Results

The surgical findings and operative results are summarized in Table 2 and Table 3. Two tumours belonged to Type A and eight tumours presented with an intra- and extracranial extension were classified as Type D according to the Kay-Pellet classification. In two cases, the tumours presented with cystic degeneration. All lesions were hypovascular on cerebral angiography; in 8 cases (Type D tumours), the jugular bulb was not patent on angiographic studies. Intraoperatively the jugular bulb was completely occluded in 2 patients. In half of cases it was not possible to determine the nerve of origin of tumour: in our series the glossopharyngeal nerve was identified as the nerve of origin in three patients, CN X in one patient and CN XI in one patient.

There was no operative mortality. There was no cerebrospinal fluid leakage. In the immediate postoperative period neurological deterioration occurred in 4 patients. Preoperative

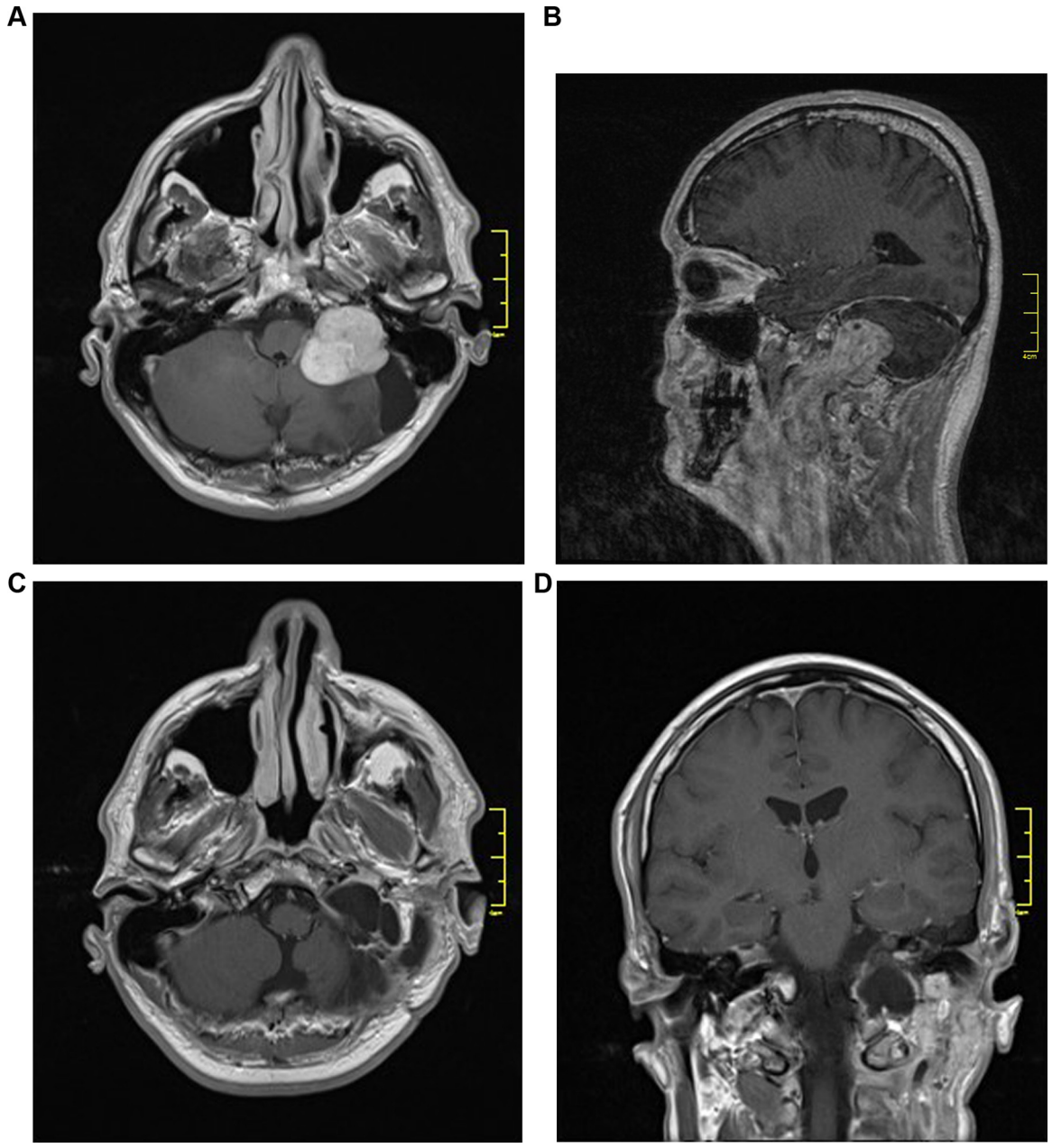

Fig. 3 – Preoperative axial (A) and sagittal (B) T1 contrast enhanced MR image demonstrating regrowth of a dumbbell-shaped jugular foramen schwannoma after previous partial resection at another institution. Postoperative axial (C) and coronal (D) T1 contrast enhanced MR image obtained after complete resection of the tumour.

CN deficits worsened in 2 patients and new postoperative paresis in a previously normal CN was seen in 2 patients. All the patients were independent at discharge from department. New postoperative facial palsy and deterioration of pre-existing facial nerve deficit were of a lower degree (grade II or III on the House–Brackmann scale [9]). Two patients with facial palsy showed gradual improvement of the nerve function in the follow-up.

In 4 patients the cranial nerve dysfunction had improved considerably at the last follow-up examination. In all other patients preoperative deficits proved permanent.

One patient has shown signs of recurrence 40 months after treatment. The tumour had been classified as Type A with glossopharyngeal nerve of origin and had been approach via retrosigmoid route. At second surgery mastoidectomy was performed, the sigmoid sinus and jugular bulb were exposed and the posterior part of the occipital condyle and the jugular tubercle were removed. The jugular foramen was found to be enlarged. The tumour was approached posterior to the jugular bulb and completely removed. The postoperative course was uneventful and no further tumour recurrence was found in 3-year follow-up.

Table 2 – Surgical findings in 10 patients with jugular foramen schwannomas.

Case no.	Tumour type[a]	Surgical approach	Tumour origin[b]	Jugular bulb patency (DSA/MRA[c])	Jugular bulb patency (intraoperatively)	Tumour removal
1	A	Retrosigmoid	CN IX	Patent	Patent	Total
2	A	Retrosigmoid	CN IX	Patent	Patent	Total
3	D	Cranio-cervical	CN XI	Not patent	Not patent	Total
4	D	Cranio-cervical	Unknown	Not patent	Patent	Total
5	D	Cranio-cervical	Unknown	Not patent	Not patent	Total
6	D	Cranio-cervical	Unknown	Not patent	Patent	Total
7	D	Cranio-cervical	CN IX	Not patent	Patent	Total
8	D	Cranio-cervical	CN X	Not patent	Patent	Total
9	D	Cranio-cervical	Unknown	Not patent	Patent	Total
10	D	Cranio-cervical	Unknown	Not patent	Patent	Total

[a] According to Kaye and Pellet grading system.
[b] CN, cranial nerve.
[c] DSA, Digital Subtraction Angiography; MRA, Magnetic Resonance Angiography.

Table 3 – Early and late surgical results on cranial nerve status.

Case no.	Early surgical results		Late surgical results		
	New postoperative CN deficit or preoperative CN deficit worsened	Preoperative CN deficit stable	CN deficit improved	CN deficit stable	Outcome (Karnofski scale)
1	VII[b] VIII IX X	–	VII[a] IX X	VIII	90
2	–	VIII	–	VIII	100
3	VII[b] IX X	XI	VII[a] IX X	XI	90
4	–	XI	–	XI	90
5	–	V VII[c] VIII XI	–	V VII[c] VIII XI	80
6	IX X	–	IX X	–	90
7	–	–	–	–	100
8	–	IX X XI XII	–	IX X XI XII	80
9	VII[a] VIII XII	IX X XI	XII	VII[a] VIII IX X XI	80
10	–	VII[c] VIII IX X	–	VII[c] VIII IX X	80

[a] Grade II on the House–Brackmann scale.
[b] Grade III on the House–Brackmann scale.
[c] Grade IV on the House–Brackmann scale.

4. Discussion

Schwannomas located at the jugular foramen may arise from cranial nerves IX–XI. In most patients the lower CN of origin cannot be determined [10] and among cases of identified lower CN the most common origin is glossopharyngeal nerve. There has been 1 report of jugular foramen MPNST (malignant peripheral nerve sheath tumour) [11].

On imaging, enlargement of the jugular foramen is frequently seen. On CT scans of jugular foramen schwannomas enlarged jugular foramen appears as a sharp rounded

shape with sclerotic rims in contrast to hyperostosis and thickness of jugular spine and jugular tubercle (meningioma) or erosion and destruction of bone (glomus jugulare tumours). The jugular foramen schwannomas on MR images show dense enhancement after contrast administration and tend to compress the bulb and the jugular vein rarely showing intraluminal growth.

The clinical presentation of jugular foramen schwannomas depends on the tumour's extension rather than corresponding to the nerve of origin [12]. The most common sign of tumour is compromise of the eighth CN however sometimes the tumour does not manifest clinically until it reaches a large size and presents with increased intracranial pressure. Type A tumours (according to Kay-Pellet classification) usually mimic acoustic neuromas. Based on the position of the tumour and the nerve's point of origin Kaye et al. classified these tumours into Type A (primarily intracranial tumours with only a small extension into the bone), Type B (the main mass of the tumour is within the bone, with or without an intracranial component) and Type C (tumours are primarily extracranial with only a minor extension into bone).

Pellet et al. [8] modified the grading system by adding a Type D which corresponds to the dumbbell-shaped jugular foramen schwannomas with both an extra- and an intracranial extension.

The goal of surgical treatment of jugular foramen schwannomas is to achieve complete tumour removal with cranial nerves preservation [4,12–14]. The lower cranial nerves injury is the source of life-threatening postoperative complications, especially if the patient had no deficits preoperatively. Acute onset of postoperative lower cranial nerves deficits is dangerous because patients have no chance for compensatory mechanisms to develop as we have observed in patients with slowly progressive deficits [4]. Intraoperative neurophysiological monitoring of the lower cranial nerves as well as the facial nerve is important adjuvant for preventing injury to these nerves and we implement it in each operation with an increased risk of cranial nerve injury. Appropriate measures such as the nothing-by-mouth regimen, keeping in place nasogastric tube and use of mechanical diets, are taken in the immediate postoperative period. If the swallowing function remains impaired and prolonged recovery period is anticipated, a percutaneous endoscopic gastrostomy (PEG) or jejunostomy and temporary tracheostomy are performed [5,6]. In the largest reported series of surgically treated jugular foramen schwannomas Sedney et al. [13] noted no improvements in preoperative CN deficits in any of their patients and 44% of new or worsening CN deficits were permanent. However there was a statistically significant decrease in permanent deficits of the CN 9 and 10 with a more conservative technique focused on maximum preservation of neurovascular structures and considering near-total resection acceptable. In a series considering only Type D jugular foramen schwannomas, Kadri and Al-Mefty [12] found no additional postoperative CN deficits and improvement in preoperative lower CN deficits was noted in one-third of patients. Chibbaro et al. [14] reported that in 16 surgical cases of dumbbell-shaped jugular foramen schwannomas all patients showed improvement of pre-operative lower cranial nerve deficits and 56.3% of patients experienced full functional recovery. In their series of 16 patients Samii et al. [15] reported temporary CN morbidity of 38%; in all other patients, the CN dysfunction remained the same as preoperatively. Suri et al. [16] describing a surgical series consisted of 22 Type A tumours and one Type D tumour reported one death (4.3%), permanent morbidity in three patients (13%) in the form of new onset facial nerve paresis, and lower cranial nerve improvement in 13% of patients. The choice of surgical approach is crucial for achieving satisfactory outcome and depends on tumour's location and extension. Type A tumours can be treated via a retrosigmoid approach [12,15,16] but if there is osseous involvement within jugular foramen mastoidectomy should be performed and additional opening the jugular foramen dorsolaterally is desirable [5]. For Types B, C and D tumours modifications of cervical approach combined with mastoidectomy provide adequate exposure. Kadri and Al-Mefty [12] recommend use of the transcondylar suprajugular approach for dumbbell-shaped tumours without opening the wall of the bulb. Bulsara et al. [5] used transjugular approach combined with a high cervical approach and occasional need for the internal jugular vein ligation in cases of dumbbell-shaped tumours with a high cervical extension. Interestingly, there was one case in our cohort of jugular foramen schwannoma found 19 years after radiation treatment for brain glioma. Similar case has been reported of the de novo formation of a jugular schwannoma 20 years after irradiation [17]. The experience with radiosurgery of jugular foramen schwannomas as a primary treatment modality is a topic of debate and it seems reasonable to reserve radiosurgical treatment for cases when difficult venous anatomy presents a considerable surgical risk [12], in elderly patients, in patients who underwent previous subtotal tumour resection [14,18] or as an adjunct treatment for regrowth of residual tumours [13].

5. Conclusions

1. Jugular foramen schwannomas can be radically managed with the use of skull base surgery techniques. The surgical treatment of jugular foramen schwannomas carries a significant risk of the lower CN deficits.
2. Partial jugular foramen schwannomas removal poses considerable surgical risk of CN deficits without patient's cure.
3. Careful lower CN evaluation and appropriate measures taken if patient exhibits CN deficits in the immediate postoperative period allow for life-threatening postoperative complications prevention.

Conflict of interest

The authors declare no conflict of interest.

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None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

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