Early and long-term results of the treatment of jugular paragangliomas using different ranges of surgical approach

Bezpośrednie i odległe wyniki leczenia przyzwojaków otworu szyjnego z wykorzystaniem różnych zakresów dostępu operacyjnego

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Abstract

Background and purpose: The applied approach to the jugular foramen is a combination of the juxtacondylar approach with the subtemporal fossa approach type A. The purpose of this study is to present our results of treatment of jugular paragangliomas using the aforementioned approach.

Material and methods: Twenty-one patients (15 women, 6 men) with jugular paragangliomas were included in the study. The neurological status of the patients was assessed before and after surgery as well as at the conclusion of treatment. The approximate volume of the tumour, its relation to large blood vessels, cranial nerves and brainstem, as well as consistency and vascularity were also assessed.

Results: The duration of symptoms ranged from 3 to 74 months. In 86% of patients hearing loss was the predominant symptom. The less frequent symptoms included pulsatile tinnitus in the head, dysphagia and dizziness. Approximate volume of the tumours ranged from 2 to 109 cm^3. A gross total resection was achieved in 71.5% of patients. The postoperative performance status improved in 38% of patients, did not change in 38% and deteriorated in 24% of patients.

Conclusions: A proper selection of the range of the approach to jugular foramen paragangliomas based on their topography and volume reduces perioperative injury without negative consequences for the radicality of the resection.

Key words: jugular foramen, paraganglioma, microsurgery, skull base surgery.

Streszczenie

Wstęp i cel pracy: Stosowany dostęp do otworu szyjnego to połączenie dostępu okołokłykciowego i dostępu podskroniowego typu A. Celem niniejszej pracy jest przedstawienie własnych wyników leczenia przyzwojaków otworu szyjnego z wykorzystaniem powyższego dostępu.

Materiał i metody: Analizie poddano grupę 21 chorych z przyzwojakami otworu szyjnego (15 kobiet i 6 mężczyzn). U 10 chorych wykorzystano pierwszy, u 7 – drugi, a u 4 – trzeci zakres dostępu do otworu szyjnego. Ocenie podlegały stan neurologiczny chorych przed rozpoczęciem leczenia, po operacji i po zakończeniu leczenia oraz zmiany ich aktywności życiowej. Określano przybliżoną objętość operowanych guzów, ich stosunek do dużych naczyń, nerwów czaszkowych i pnia mózgu.

Wyniki: Długość wywiadu wahała się od 3 do 74 miesięcy (mediana wyniosła 6 miesięcy). W 86% przypadków wiodącym objawem był niedosłuch. Rzadziej występowały pulsujący szum w głowie oraz zburzenia połykania i równowagi. Przybliżona objętość usuniętych guzów wahała się od 2 do 109 cm^3. W 71,5% przypadków przeprowadzone resekcje były doszczętne. Aktywność życiowa u 38% pacjentów poprawiła się, u 38% nie zmieniła się, a u 24% uległa pogorszeniu.

Wnioski: Dobór zakresu dostępu do otworu szyjnego, podyktowany topografią i rozmiarami przyzwojaka rozwijającego się w tym otworze i jego otoczeniu, pozwala na ograniczenie urazu operacyjnego bez negatywnego wpływu na radykalność resekcji.

Słowa kluczowe: otwór szyjny, przyzwojak, mikrochirurgia, chirurgia podstawy czaszki.

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Introduction

A dilemma that arises from the proclivity toward highly radical resection on one hand while maintaining relatively low disability on the other is an unavoidable part of surgical treatment of tumours of the jugular foramen and its vicinity, particularly paragangliomas. The very quintessence of this dilemma arises from the fact that the tendency toward a radical resection of highly vascular tumours such as paragangliomas that present multidirectional growth and encase important anatomical structures forces the extension of the surgical approach.

Anatomical boundaries of the vicinity of the jugular foramen, however, render surgical approach extension in this region prone to permanent neurological sequelae. The authors see the solution of this dilemma in the proper choice of the surgical approach that is flexible and allows intraoperative modification. Our cohort of patients treated for jugular foramen paragangliomas underwent surgery with a single surgical approach that was implemented in three various ranges dependent on the size and direction of tumour growth.

Material and methods

The jugular foramen approach has been used in the Department of Neurosurgery in Sosnowiec since 2000. Thirty-one cases have been treated with this approach; they include 21 patients treated for paragangliomas. A retrospective analysis of this particular group that includes 15 women and 6 men aged 24 to 70 years was performed. Ten cases treated via the jugular foramen approach for tumours other than paragangliomas included: meningioma, neurinoma, haemangiopericytoma, chondrofibroma, chondrosarcoma and undifferentiated malignant tumour. These patients, due to the histopathology of their tumours, were excluded from the cohort that was analysed in this study.

The authors perform the surgical approach to the jugular foramen and its vicinity in three ranges depending on the tumour topography within the petrous part of the temporal bone assessed on preoperative magnetic resonance imaging (MRI). The first range that opens an approach to the posterior aspects of the foramen is implemented in order to remove tumours that encompass the jugular foramen and hypotympanum. The second range approach in addition to the aforementioned visualizes the superior margin of the foramen, thus facilitating the removal of tumours that invade the tympanic cavity. Further extension of the approach to the third

range makes it possible to access the anterior boundaries of the foramen, which consequently allows the dissection of tumours that encase the carotid artery and penetrate the petrous apex.

Neurological status of the patients at the beginning of the treatment, immediately after surgery and at the end of the treatment (Tables 1 and 2), along with the changes of everyday performance assessed with the Karnofsky scale (Table 3), were analysed. The volumes of the tumours were approximated with the formula for a rotational ellipsoid [$volume = \Pi/6\ (x \times y \times z)$], based on MRI. The extent of resection was assessed based on pre- and postoperative MRI examinations. The relationship of tumours to large vessels, cranial nerves, brain stem, as well as their consistency and vascularity, were also analysed.

The distribution of variables, i.e. average times of catamnesis and postoperative follow-up and tumour volume, were analysed with the Shapiro-Wilk test. Based on the fact that all the variables were normally distributed, they were presented as means with standard deviations (SD).

Results

Ten patients out of 21 treated for paragangliomas of the jugular foramen were operated using the first range approach. These tumours on top of the jugular foramen encompass the hypotympanum and upper parts of the jugular neurovascular bundle (Fig. 1). Even though some of these tumours penetrated the posterior fossa, they did not cross the dura. Their volumes varied from 2.3 to 6.3 cm³ (mean 4.1 cm³, SD 1.1 cm³). The second range approach was used in the treatment of 7 patients. Their tumours, aside from the jugular foramen, hypotympanum and the jugular neurovascular bundle, infiltrated the tympanic cavity, the petrous apex and the mastoid process (Fig. 2). Some of these tumours infiltrated the intradural space in the posterior fossa. Tumour volumes varied from 13.4 to 53.6 cm³ (mean 26.0 cm³; SD 14.1 cm³). Four cases forced the surgeon to extend the approach to the third range. Above and beyond the anatomical areas shared by the tumours from the first two groups, the latter invaded the subtemporal fossa and extensively encased the internal carotid artery (Fig. 3). Their volumes varied from 82.4 to 109.3 cm³ (mean 98.1 cm³, SD 12.1 cm³).

The time of catamnesis for all the patients analysed varied from 3 to 74 months (mean 32 months; SD 22 months). Hearing deficits present at admission in

Table 1. Changes in neurological abnormalities that occurred before surgical treatment of jugular foramen paraganglioma (21 cases)

Neurological abnormality	Examination on admission	Examination on discharge	Follow-up examination
Hypoacusis	18	18	17
Disequilibrium	10	8	5
Seventh nerve palsy or paresis	7	5	5
Dysphagia	12	10	4
Dysarthria	5	4	4
Eleventh nerve palsy or paresis	6	6	6
Ninth nerve palsy or paresis	8	8	8
Pulsatile hum in the head	17	0	0
Hemiparesis	2	0	0
Wasting	5	4	0

Table 2. Neurological abnormalities that occurred as side effects of surgical treatment of jugular foramen paraganglioma (21 cases)

Neurological abnormality	Examination on the 1st day post op	Examination on discharge	Follow-up examination
Hypoacusis or deafness	3	3	1
Disequilibrium	not assessed	4	0
Seventh nerve palsy or paresis	6	6	5
Dysphagia	9	8	4
Dysarthria	7	6	2
Eleventh nerve palsy or paresis	5	5	4
Ninth nerve palsy or paresis	4	4	4
Hemiparesis	1	0	0
Wasting	3	3	1

Table 3. An assessment of the daily activities of patients treated surgically because of jugular foramen paragangliomas (Karnofsky scale)

Patient no. and sex	Examination on admission	Examination on discharge	Follow-up examination
1 (F)	70	70	80
2 (F)	50	70	80
3 (F)	70	70	80
4 (M)	80	60	70
5 (F)	80	50	60
6 (F)	90	70	90
7 (F)	80	70	90
8 (M)	90	80	80
9 (F)	90	90	90
10 (F)	60	70	80
11 (F)	80	80	80
12 (F)	80	80	80
13 (M)	60	50	80
14 (F)	90	70	90
15 (M)	80	60	70
16 (F)	60	70	80
17 (F)	80	70	80
18 (M)	80	70	80
19 (M)	90	80	90
20 (F)	80	70	90
21 (F)	80	70	80

F – female; M – male

88% of cases at the end of the treatment either remained level or deteriorated. Conversely, swallowing disturbances, present in 57% of patients, improved with rehabilitation in the majority of cases. A significant improvement of disequilibrium that had been present in 48% of cases was noted as well. Facial nerve paresis was present at admission in 33% of patients and varied from grade II to V on the House-Brackmann scale. Following treatment, it was still present in the majority of these patients. However, in some of them it regressed while in the others it intensified.

Hemiatrophy of the tongue subsequent to hypoglossal nerve injury was present in 38% of cases, with no significant improvement following treatment. Conversely, all the patients who reported a pulsatile hum in the head noticed that the symptom disappeared after surgery. Complete symptomatology at admission and its changes following treatment are summarized in Table 1. Some of the patients whose tumours required the first or second range approach to the jugular foramen presented scarcely any while others showed a plethora of symptoms. All of the patients who required the third range approach presented the full clinical syndrome of jugular paraganglioma.

Tumours were adjacent to the facial nerve within the petrous part of the temporal bone in 43% of cases. This contact varied from adhesion of the tumour to the nerve to the complete encasement of the nerve within
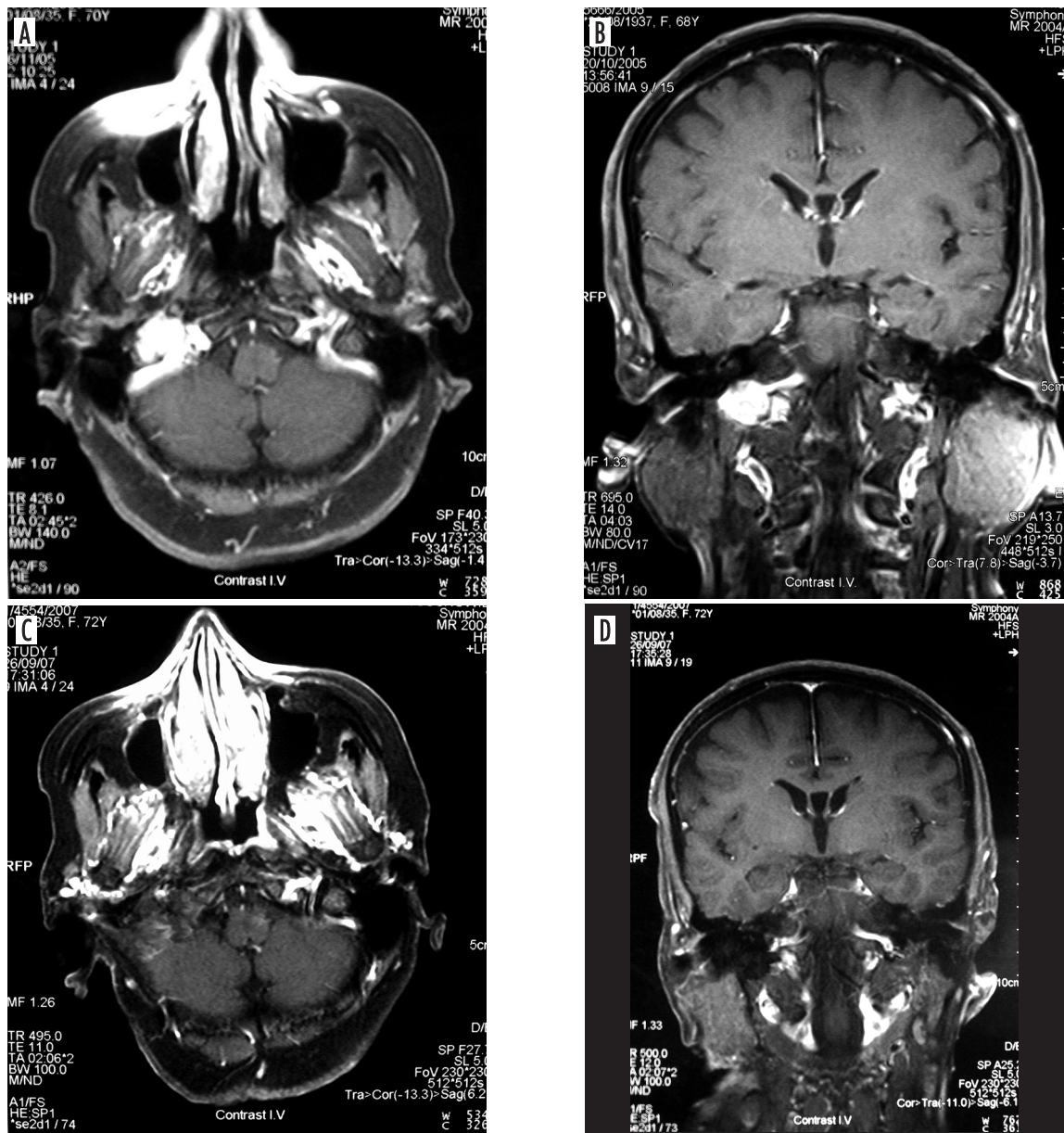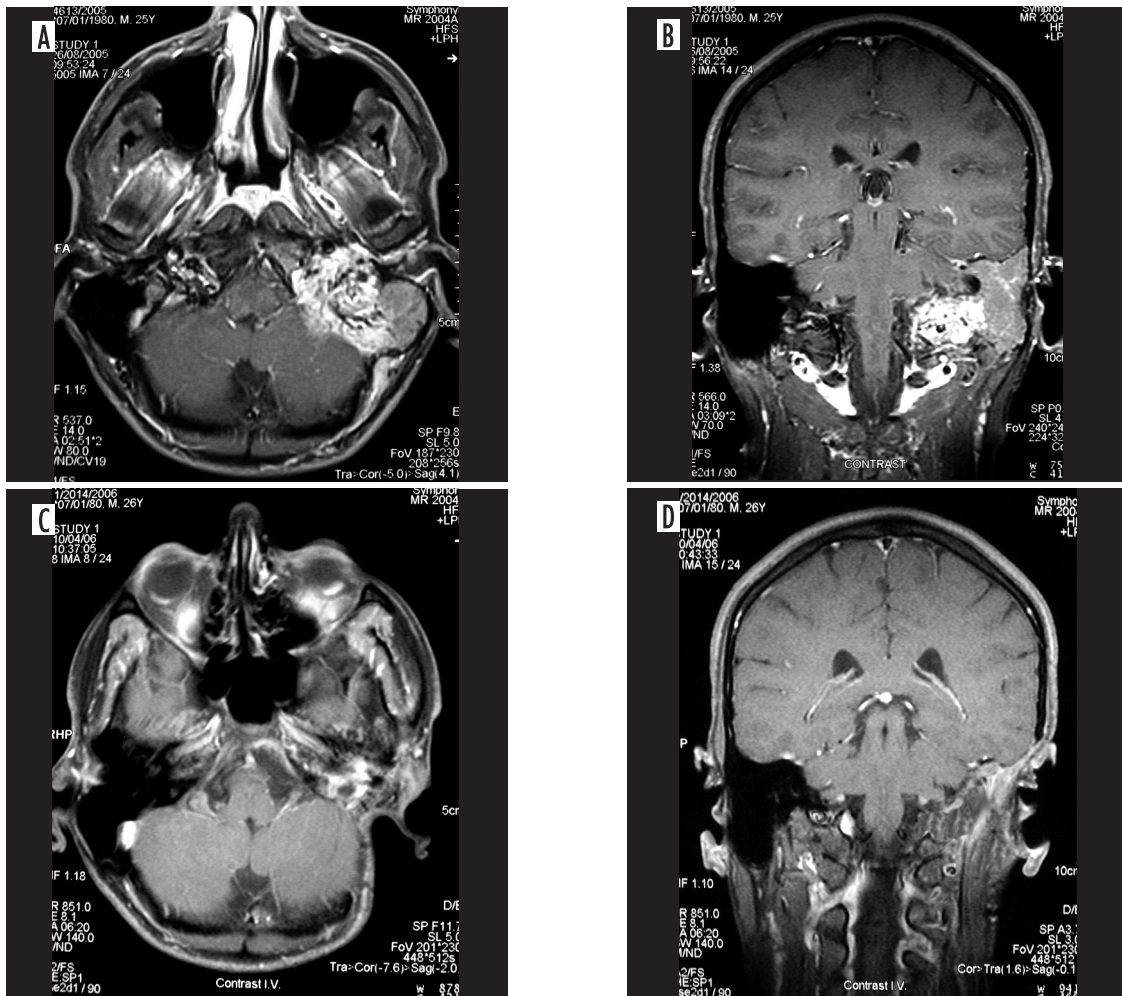
Fig. 1. Jugular foramen paraganglioma spreading into hypotympanum on the right side (patient no. 7, 70-year-old female). The patient developed dysphagia and pulsatile hum in the head within 6 months. Tumour volume was assessed as 3 cm³. She underwent surgical treatment with the first range approach implemented. The tumour did not reach the neural compartment of the foramen. Dysphagia diminished following surgery.
A) preoperative axial MRI, B) preoperative coronal MRI, C) postoperative axial MRI, D) postoperative coronal MRI

the tumour. In two cases, the tumour connected with the facial nerve simultaneously within the petrous part of the temporal bone as well as within the cerebellopontine angle. All patients with preoperative facial paresis presented with the aforementioned contact. Two patients, however, did not show any deficits despite the presence of adhesions between the tumour and the nerve. In seven cases, anterior mobilization of the facial nerve without

the need for reconstruction was performed. Three cases in which preoperative facial nerve palsy had been present and the nerve required dissection from the tumour postoperatively developed permanent paralysis. On the other hand, four other cases with facial nerve palsy, following the period of full paralysis, finally stabilized between the second and the third grade on the House-Brackmann scale. Four cases required reconstruction of

Fig. 2. Jugular foramen paraganglioma spreading into hypotympanum and tympanic cavity on the left side (patient no. 4, 24-year-old man). The patient developed hypoacusis, dysphagia and pulsatile hum in the head within 5 months. Tumour volume was assessed as 23 cm^3. He underwent surgical treatment with the second range approach implemented. Prior to tumour resection, anterior reposition of the facial nerve was performed. Still the patient developed permanent facial palsy. Dysphagia and hypoacusis did not change following surgery.
A) preoperative axial MRI, B) preoperative coronal MRI, C) postoperative axial MRI, D) postoperative coronal MRI
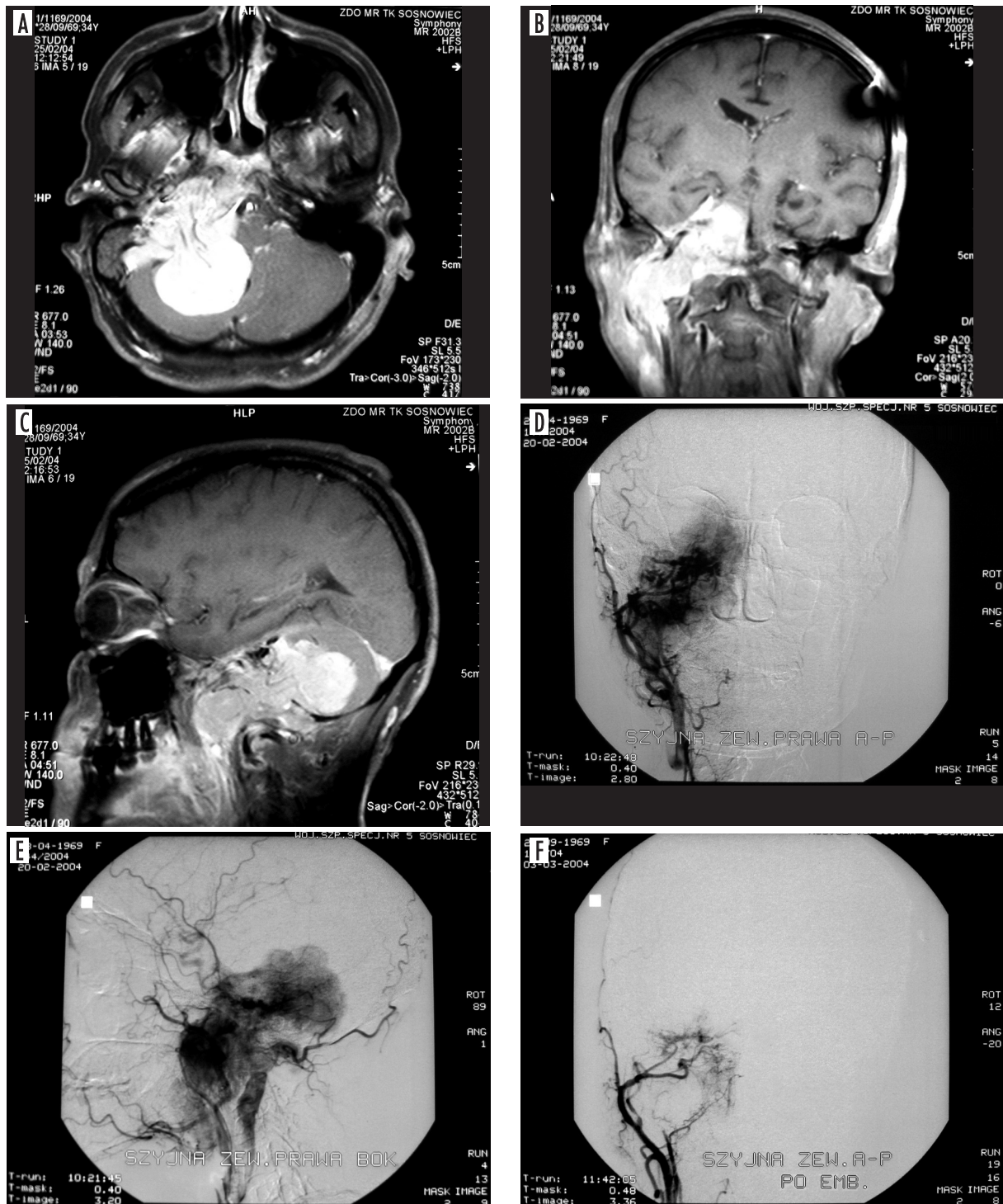
the infiltrated facial nerve with either the great auricular nerve or the sural nerve.

In 85% of cases, tumours were adherent to the nerves that transverse the jugular foramen, i.e. the glossopharyngeal, vagus and accessory nerve. Interestingly, at admission a third of the patients had no neurological deficits that could be related to it. Nonetheless, penetration of the tumour into the neural compartment in those cases where the tumour was adherent to the aforesaid nerves within the jugular foramen, even in patients without preoperative deficits, resulted in full paralysis after surgery. A more favourable outcome, especially after rehabilitation, was achieved in

patients who had similar adhesions in the cerebellopontine angle or upper parts of the jugular neurovascular bundle.

In 76% of cases, tumours were adherent to the hypoglossal nerve, usually in the upper parts of the jugular neurovascular bundle. Still, only half of the patients presented with hemiatrophy of the tongue. In 19% of cases, this sign appeared as a new, permanent deficit.

Contact or adhesion to the brain stem was found in 38% of cases. Fortunately it caused no intraoperative complications and had no effect on the final outcome. Table 2 contains a full description of neurological deficits rendered as side effects of the surgery.

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Fig. 3. Jugular foramen paraganglioma spreading within the skull base into the middle and posterior fossa on the right side (patient no. 2, 35-year-old female). Within 5 years of follow-up the patient developed hypoacusis, disequilibrium and dysarthria with facial nerve palsy, left-sided hemiparesis and pulsatile hum in the head. Severe dysphagia resulted in wasting. Tumour volume was assessed as 95 cm^3. She underwent surgical treatment with the third range approach implemented in combination with an extended subtemporal approach. After surgery and rehabilitation, the only permanent deficits are right-sided deafness and dysarthria. Since cable anastomosis of the facial nerve was futile, it was followed by an anastomosis with the hypoglossal nerve that resulted in improvement from full palsy to facial nerve paresis. Three years after surgery the patient delivered a healthy child and returned to her previous profession. Two years later a small recurrence in the subtemporal fossa was found; the patient underwent radiotherapy. The follow-up is ongoing.
A) preoperative axial MRI, B) preoperative coronal MRI, C) preoperative sagittal MRI, D) external carotid artery in antero-posterior projection, E) external carotid artery in lateral projection, F) external carotid artery following embolisation in antero-posterior projection, G) external carotid artery following embolisation in lateral projection, H) postoperative axial MRI, I) postoperative coronal MRI, J) postoperative sagittal MRI
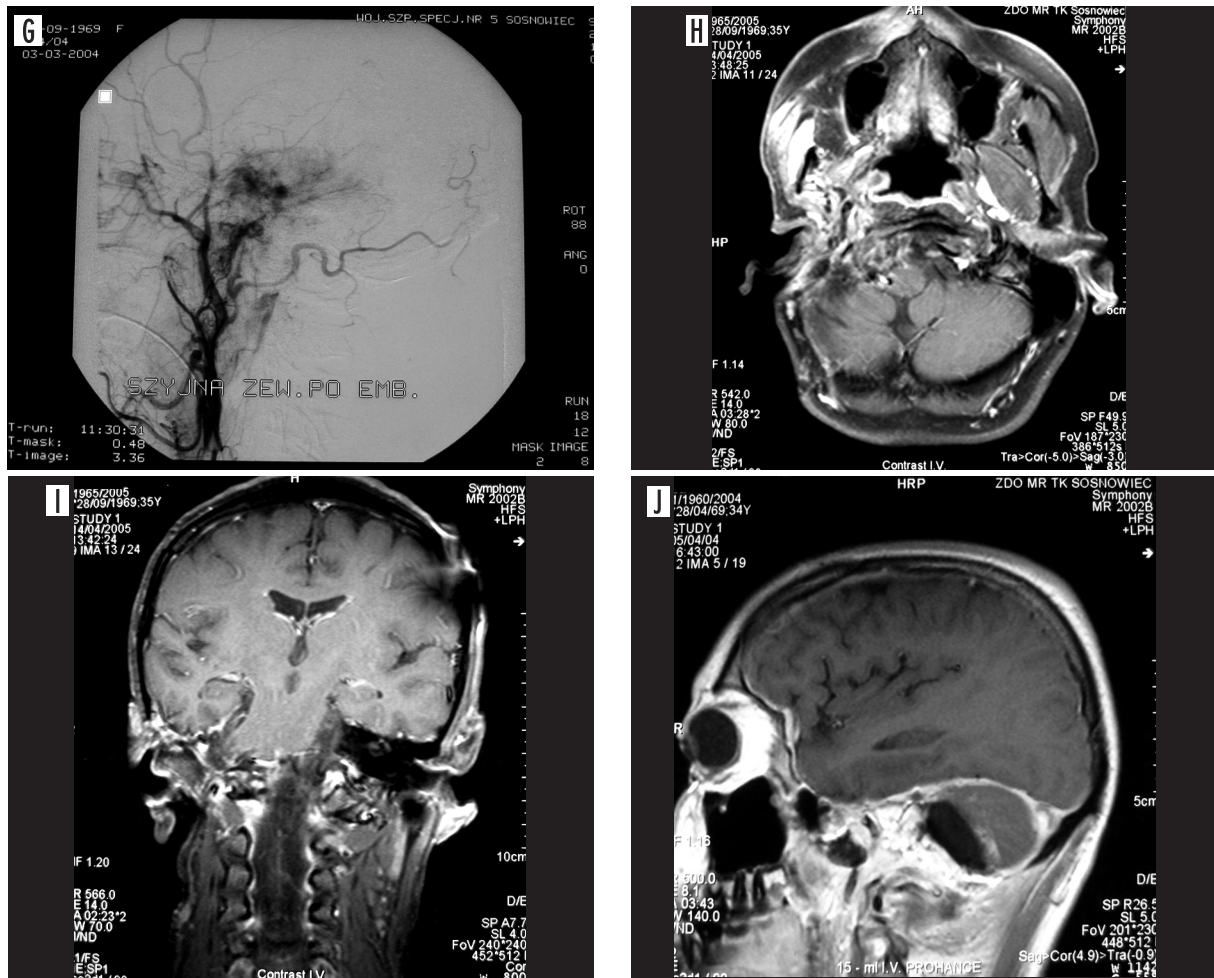
Fig. 3. Cont.

Since all of the tumours infiltrated the venous compartment of the jugular foramen, it required opening during surgery. Each patient had the sigmoid sinus and internal jugular vein obliterated. In three cases, the flow throughout these vessels prior to their closure was preserved. In one of these cases the residual flow was so high that prior to permanent closure of the sigmoid sinus it was closed temporarily in order to assess a possible significant increase of the pressure in the proximal part of the sinus. In one case, the tumour grew into the sigmoid sinus to its full extent, thus forming a loose cast of it. Conversely, excision of this part of the tumour preceded obliteration of the sinus.

In three cases (14%), tumours grew around the internal carotid artery, one of them (Fig. 4) 4 cm lengthwise. In another four cases (19%), tumours were adherent to the artery. Still, identification of the proper layer enabled safe dissection of the vessel from the tumour.

Radical resection was achieved in 71.5% of cases. Lack of radicality usually arose from the tendency to avoid deficits from the lower group of the cranial nerves in the cerebellopontine angle. In four patients who underwent non-radical resection, a local recurrence was found. These patients were subsequently treated with stereotactic radiosurgery. None of the irradiated tumours have shown any signs of progression so far.

The follow-up period for the entire group varied from 9 to 106 months (mean 48 months; SD 22 months). It was long enough to observe either a gradual reduction of postoperative neurological deficits in some of the cases or adaptation of the patients to the permanent disabilities. Comparison of the functional capacities of the patients on admission and during the follow-up as measured with the Karnofsky performance scale revealed improvement in 38%, unchanged condition in 38% and deterioration in 24% of patients, with a clear discrepancy between the

assessments immediately after surgery and subsequent to rehabilitation and convalescence. None of the patients in our cohort died during follow-up.

Discussion

Paraganglioma, neurinoma and meningioma represent the three most common histopathological subtypes of tumours related to the jugular foramen [1-3]. Due to their abundant vascularity and the capacity for multidirectional growth, paragangliomas are the most challenging ones. They originate from parasympathetic glomus bodies, which localize within the walls of the jugular vein (glomus jugulare) [4]. Rarely tumours that originate from glomus bodies within the tympanic cavity (glomus tympanicum) or adjacent to the lower ganglion of the vagal nerve (glomus vagale) spread toward the jugular foramen and require treatment modalities similar to paragangliomas of the jugular foramen [5,6]. Along with the Arnold nerve, i.e. a branch from the upper ganglion of the vagal nerve that anastomoses to the mastoid part of the facial nerve, parasympathetic glomus bodies reach the facial canal. These glomus bodies may be the origin of the tumour that expands toward the jugular foramen as well [7]. Facial canal paragangliomas deform this canal, penetrate toward the parotid gland and secondarily reach the tympanic cavity along with the jugular bulb, which relatively early evokes either facial paresis or paralysis [7].

Paragangliomas might occur as part of von Hippel-Lindau syndrome, type 1 neurofibromatosis (NF1) or of multiple endocrine neoplasia type 2 (MEN2) [8-10]. Familial occurrence of paragangliomas has been observed in 10 to 50% of cases [11]. Their inheritance has an autosomal dominant pattern and is related, among others, to the mutation of a gene for a D subunit of succinate dehydrogenase (SDHD). This gene is located on the short arm of chromosome 11 [10]. An affected male has a 50% chance of having a sick child while a female does not. Nonetheless, she transmits the inactive, mutated gene to her progeny. A male child who inherits such a gene has a 50% chance of having an affected offspring [11]. The aforementioned information has a measurable practical value for it may prompt a diagnostic screening of persons at risk in order to diagnose small, clinically silent tumours that are relatively easy to treat.

The most common signs and symptoms of jugular foramen paragangliomas include hearing disturbances or loss, dysphagia, disequilibrium, facial paresis or paralysis, hemiatrophy of the tongue and pulsatile hum in the head [9,12-14]. Symptomatology of paragangliomas in our cohort was similar. What is worth mentioning, however, is wasting seen in 5 patients with dysphagia.

Diagnostic imaging of jugular foramen paragangliomas includes computed tomography (CT), MRI and angiography. CT in the bone window is of particular importance for it can visualize the destruction of bony margins of the jugular foramen along with loss of airiness of the petrous part of the temporal bone. It was clearly visible in 8 cases in our cohort. On the other hand, MRI provides information on the progress of
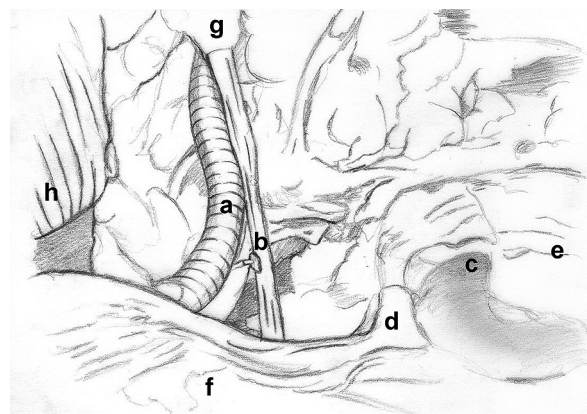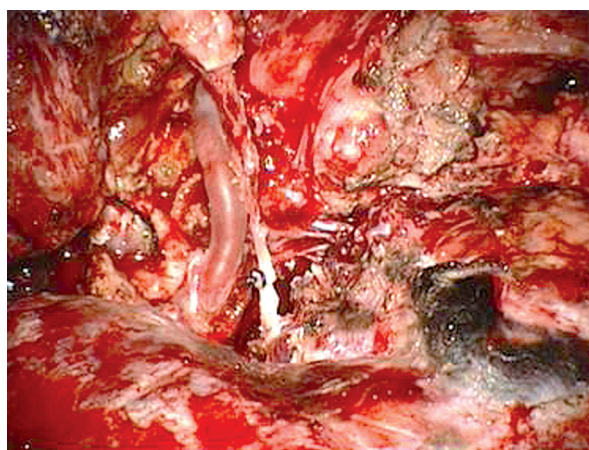
Fig. 4. Intraoperative view of jugular foramen paraganglioma (patient no. 2, 35-year-old female). Extradural part of the tumour was removed and complete amputation of the petrous part of the temporal bone was already performed (right side); a — petrous part of the internal carotid artery, b — facial nerve fully mobilized, infiltrated by tumour, c — obliterated sigmoid sinus, d — dura infiltrated by tumour between superior and inferior petrous sinus, e — dura in the suboccipital region, f — dura in the temporal region, g — parotid gland, h — temporal muscle

the tumour. Paragangliomas significantly enhance after gadolinium injection in T1-weighted sequences [4,9,13]. Notably, angiography of internal and external carotid arteries along with vertebral arteries is necessary in order to achieve exhaustive assessment of vascular disturbances related to jugular foramen paragangliomas. Pathological vascularity might originate from any of the aforesaid arteries. It is of particular importance to notice a displacement and narrowing of the internal carotid artery encased in the tumour. Occasionally, a vertebral artery might be similarly involved as well. Obliteration of the sigmoid sinus and internal jugular vein along with the presence of arteriovenous fistulas can also be found [12]. Additionally, angiography aids in the appraisal of indications as well as feasibility of tumour embolisation. Virtually all of the jugular foramen paragangliomas have indications for embolisation. It drastically reduces intraoperative blood loss and facilitates reduction of the time of the procedure [14]. In most cases, tumour feeders that originate from the external carotid artery, such as the ascending pharyngeal artery, occipital artery or posterior auricular artery, are feasible for selective catheterization [4]. Cases from our cohort fully confirmed this. Even though elective embolisation of vessels that can be easily and with impunity litigated is problematic, every attempt to reduce intraoperative blood loss is advisable [15]. Abolition of vascularity that originates from the external carotid artery results in increased circulation in other vessels that feed the tumour, which originate from the internal carotid artery or vertebral artery [16]. This explains the lack of justification for embolisation as a separate therapeutic modality on one hand and supports the limitation of time between embolisation and surgery on the other [12]. The authors performed surgery no later than four days after embolisation. Importantly, recent advances have enabled intraoperative embolisation of paragangliomas via direct injections of their tissue.

An increased level of catecholamines was reported in 3 to 4% of patients with paragangliomas [11,16,17]. Levels of their metabolites, i.e. vanillylmandelic acid, hydroxyindoleacetic acid and metanephrine, should be routinely assessed then [4,16,17]. None of the patients in our group proved positive when evaluated accordingly. One- or twofold increase of metabolite levels in urine has no clinical manifestation. Four- or fivefold increase might present as increased blood pressure and dangerous blood pressure variability during surgery [13]. Patients with the aforesaid disturbances require two weeks of preparation with alpha- and beta-blockers

[13,16]. Patients with paragangliomas with increased excretion of catecholamines are prone to complications following embolisation and radiotherapy as well, which are explained by the release of these substances from necrotic tumour tissues into the blood stream [17,18].

Paragangliomas are considered benign tumours that double their volume within 4.2 years [19]. The clinical basis necessary to confirm their malignant transformation, which occurs in 4 to 6% of cases, if distinct histopathological features are lacking, consists of the presence of metastases to organs that are endocrinologically silent, mainly the lymph nodes [11].

Classifications of paragangliomas are based on their multidirectional growth along with related difficulty to dissect them. Glasscock and Jackson created the first classification [20]. They separately analysed paragangliomas of the tympanic cavity and jugular foramen. The latter ones were classified as follows: grade I – tumour involving the jugular bulb, middle ear and mastoid process; grade II – tumour extending under the internal auditory canal that may have intracranial extension; grade III – tumour extending into the petrous apex that may have intracranial extension; grade IV – tumour extending beyond the petrous apex into the clivus or infratemporal fossa that may have intracranial extension. Fisch described the most renowned classification of paragangliomas of temporal bone [21]: class A – tumours limited to the tympanic cavity; class B – tumours limited to the middle ear or mastoid without involvement of the jugular foramen and carotid canal; class C – tumours that originate from the jugular foramen with variable extension toward the middle ear, inner ear and cranial cavity along the sigmoid sinus, internal jugular vein, lower complex of the cranial nerves in the cerebellopontine angle and internal carotid artery. Within class C, the degree of extension along the internal carotid artery is defined with the following numbers: 1 – tumour with limited involvement of the vertical portion of the carotid canal – it destroys the wall of the carotid canal but does not invade it; 2 – tumour invading the vertical portion of the carotid canal; 3 – tumour encasing the internal carotid artery in its vertical and horizontal portion but not reaching the foramen lacerum, i.e. not encasing the paratrigeminal part of the internal carotid artery; 4 – tumours encasing the internal carotid artery beyond the foramen lacerum toward the cavernous sinus. The last class, D, has a complementary role for the whole classification and defines the range of intracranial extension of these tumours. Class De tumours show an intracranial, extradural penetration. Number 1 signifies

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intracranial extension of less than 2 cm in diameter. Number 2 pertains to intracranial extension greater than 2 cm in diameter. Class Di encompasses tumours with intradural cranial cavity penetration. Within this class number 1 indicates penetration of less than 2 cm in diameter, number 2 indicates penetration greater than 2 cm in diameter, while number 3 describes extensive intracranial penetration that renders these tumours inoperable. Patel *et al.* [4], on the other hand, proposed a descriptive classification of jugular foramen paragangliomas that takes into account the range of tumour penetration into structures and regions of the skull base, its relation to the large arteries, brain stem and the presence of hydrocephalus. The Fisch classification has a considerable practical value due to its explicitness. Every attempt to frame a biological phenomenon that is based on variability is made at the cost of accuracy of the phenomenon's description. This is relevant to the Fisch classification as well. A certain lack of information that it provides is complemented by the descriptive system of Patel *et al.* A description of Di3 class in the Fisch classification that is inoperable due to the extent of its intracranial penetration is particularly controversial. The discussed classification as cited above was published in 1998. Significant progress that resulted from the introduction of extended surgical techniques in skull base surgery has been made since. A tumour considered inoperable by one surgical team might not be inoperable for another one. The objective inoperableness of benign skull base tumours that include the vast majority of paragangliomas, if it exists at all, arises from the combination of the burdens that are related to the neoplasm and its treatment on one hand and general condition of the patient on the other. Extensive paragangliomas require a combination of one of the transpetrosal approaches with an extended subtemporal approach as well as a transcondylar approach. It seems to the authors of this paper that the Di3 class should be reserved for paragangliomas whose dissection requires approaches to the skull base different from the aforementioned ones.

Surgical treatment of jugular foramen paragangliomas has its own distinct specificity. The standing rule is that en route to the tumour it is obligatory to reveal as much of the tumour as possible before one even starts the resection. While visualizing a tumour one should identify as much as possible of the anatomical structures that either enter or exit it. These include cranial nerves, the sigmoid sinus, the internal jugular vein and, most importantly, the internal carotid artery. Based on

paraganglioma size and topography, various approaches should be implemented in order to carry out the aforesaid assumptions. It is worth mentioning however that, omitting detailed solutions proposed by various authors, two concepts of surgical approach to the jugular foramen exist. Fisch is the author of the first one [21]. It is called a subtemporal approach type A and involves getting control over the neurovascular bundle, amputation of the external auditory canal and temporal cavity content, anterior reposition of the facial nerve and exposure of the petrous part of the internal carotid artery. George has propagated the other concept [1]. It is named a juxtacondylar approach and similarly to the previous one it initially encompasses identification of the neurovascular jugular bundle and on top of that identification of the distal part of the V_3 section of the vertebral artery. Subsequently, the posteroinferior part of the mastoid is resected along with the jugular process of the occipital bone and the posterolateral part of the occipital condyle. Both concepts are used alternatively in the surgical treatment of jugular foramen paragangliomas [8,22-24].

In our opinion, a starting point for the treatment of jugular foramen paragangliomas should be an approach that is the least harmful and brings the lowest possible risk of complications. The juxtacondylar approach fulfils most of these conditions. It preserves the external auditory canal, facial nerve canal and tympanic cavity. In our understanding this approach meets the requirements of the first range approach to the jugular foramen and its vicinity. It opens the access to the posterior limitations of the foramen. If an obtained access is not sufficient for radical tumour removal, which can be determined during surgery, it is possible to modify it to the second range approach that is an equivalent to the subtemporal type A approach in most of the stages. An access to the posterior as well as superior edges of the jugular foramen is open upon completion of the first and second range approach. Provided that the obtained entry is still insufficient, it is possible to modify it to the third range approach that opens a route toward the anterior edge of the jugular foramen. The third range approach is an equivalent to the final stages of the subtemporal type A approach and involves exposure of the petrous part of the internal carotid artery. However, in selected cases, despite the fact that a full approach (i.e. in all three ranges) to the jugular foramen has been performed a total resection of the paraganglioma is still not feasible. These circumstances usually are related to extensive, intradural tumour penetration, which requires a com-

bination of the jugular foramen approach with either a partial, transcondylar approach or an extended, subtemporal approach [25]. Ten patients out of 21 treated for paragangliomas of the jugular foramen in our cohort were operated using the first range approach, seven with the second range approach and four with the third range approach. In two cases, additional surgical approaches were implemented. In one case we used an extended, subtemporal approach and partial transcondylar in the other.

The selected paraganglioma cases, even though they require all three approach ranges to the jugular foramen and its vicinity, are amenable to gross total resection without infringement of the anterior boundaries of the venous compartment of the jugular foramen. Consequently, there is no need to breach the neural compartment, thus avoiding a conflict with cranial nerves within the jugular foramen [8,16].

Resection of a jugular foramen paraganglioma usually forces the surgeon to sacrifice the sigmoid sinus before and the internal jugular vein after the tumour. As the sigmoid sinus might be obliterated relatively early during surgery, an internal jugular vein should remain patent up to the final stages of resection in order to drain blood from the tumour [16]. It is important to advise the anaesthetist to seek central venous access, for example a subclavian vein catheterization on the tumour side. The contralateral side upon obliteration of the sigmoid sinus and internal jugular vein has to carry the full burden of the venous drainage and should not have any obstacles en route.

Paragangliomas rarely force the surgeon to dissect and reconstruct the internal carotid artery, even though they often encase it [12,16]. It is feasible with the microscope to find a layer that enables safe preparation of the vessel from the tumour. It is of particular importance to systematically coagulate and cut internal carotid artery branches in the petrous part where they feed the tumour [16].

It is still a matter of discussion whether the resection of large jugular foramen paragangliomas should take place in two stages: an extradural and intradural one. This concept has its supporters [21] as well as opponents [16]. The authors staged paraganglioma resection into two stages once with a favourable outcome.

Treatment planning for jugular foramen paragangliomas has to take into account age, general condition of the patient, size and location of the tumour, signs and symptoms with hormonal activity assessment included, as well as possible complications [11]. The risk of com-

plications increases with the size of the tumour. This fact emphasizes the importance of early diagnosis. Treatment of small tumours with a clear tendency for growth should be considered as a prevention of potential deficits [8]. Contemporary surgical treatment of jugular foramen paragangliomas aims at curing the patient of the disease with simultaneous minimization of permanent deficits that arise from injuries to the cranial nerves, brain stem and large vessels [4]. Importantly, the main problems arise from cranial nerve injuries. Jackson *et al.* [26] reported, based on their series of 152 cases, that complete resection of jugular foramen paragangliomas without cranial nerve injuries is feasible in 31% of cases. The authors achieved similar results in 24% of cases, which pertained to tumours removed via the first range of approach to the jugular foramen. The frequency of preoperative deficits of the lower group of cranial nerves is as high as 40%; postoperative deficits may reach 40% as well [26,27]. Preoperative deficits of cranial nerves are closely related to the intracranial expansion of the tumour [16]. We are inclined to relate them to penetration of tumour into the neural compartment of the jugular foramen. Paraganglioma cases that qualify for the first or second range of approach to the jugular foramen might present with or without cranial nerve deficits. Tumours that qualify for the third range of approach always present with such deficits. Preoperative dysphagia, dysarthria and disequilibrium have a tendency to subside after surgery despite the fact that they might temporarily intensify in the early postoperative period [12,16]. The improvement of swallowing and phonation seen in our patients following the rehabilitation period resulted mainly from adaptation to the use of the contralateral structures. Facial palsy and hearing loss, on the other hand, are usually permanent [12,16]. In our opinion, this pertains to hypoglossal nerve injuries as well. Another significant problem relates to facial palsies following surgery. According to the literature, no less than 70% of patients who undergo anterior reposition of the facial nerve within a year after surgery are in the first three grades of the House-Brackmann scale, and only occasionally in the fourth one [4,8,11,28,29]. We performed anterior mobilization of the facial nerve seven times, without the need for its reconstruction. The final outcome seemed to be closely related to the preoperative status of the nerve; however, the small number of cases rules out any definite conclusions. All the cases with initial facial paresis after reposition developed full palsy with no subsequent recovery. All the other cases who had had no facial nerve deficits preoperati-

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vely after surgery developed full palsy that within six months after surgery recovered to paresis equivalent to grade two or three on the House-Brackmann scale. Out of four cases that required cable anastomosis of the facial nerve, one has not regained any function. In this particular case, a very severe preoperative facial paresis had been present, which could have affected the unfavourable outcome of the reconstruction. The remaining three cases eventually improved to grade four on the House-Brackmann scale, which can hardly be recognized as a favourable outcome.

Radicality of the resection of jugular foramen paragangliomas varies from 70 to 90% [8,11,28,29], while the recurrence rate was reported as 5 to 25% [8,11,28]. Our radicality reached 71%, while the recurrence rate was 19%.

Routine use of tracheotomy in patients treated for jugular foramen tumours, including paragangliomas, is rarely advocated [30]. Nonetheless, it is being advised to perform an elective tracheotomy postoperatively in cases with severe dysphagia [4,20]. These patients usually have gastrostomy performed as well. Patients with impaired articulation subsequently might qualify for surgical vocal cord reposition [11,28,31].

According to Al-Mefty and Teixeira [16] it is prudent to distinguish a subgroup of "complicated jugular foramen paragangliomas". In order to assign a given case to this subgroup it ought to fulfil at least one of the following criteria: large size, multiple localizations unior bilaterally, malignant transformation, increased levels of catecholamines, coincidence with other pathologies such as arteriovenous malformations, adrenal gland tumours and previous, unsuccessful treatment. In their opinion, treatment of complicated jugular foramen paragangliomas poses a real challenge for the surgeon and these tumours should be carefully distinguished from the others, i.e. typical jugular foramen paragangliomas.

One should not neglect stereotactic radiosurgery as a therapeutic modality in the treatment of paragangliomas. Indications for its use are: tumour size less than 3 cm in diameter, infiltration of internal carotid artery with unfavourable results of balloon obturation test, presence of contralateral tumour with cranial nerve dysfunctions and venous circulation impairment, poor general condition of the patient or denial of surgical treatment consent, nonserviceable hearing contralateral to the tumour, or lack of clinical signs of the tumour that would validate surgical intervention [8,12,15,28,32]. Radiotherapy results in tumour necrosis and loss of its proliferative abilities. It

should not be identified as a cure. Neither the range of control achieved nor its duration can be predicted [4]. Radiotherapy is more beneficial in older people. Moreover, it is not free of complications. They appear late after irradiation and usually include necrosis and demyelization [33]. In patients with jugular foramen paragangliomas, these changes might involve the brain stem, cranial nerves and temporal lobe [34]. In the context of the aforementioned indications for radiotherapy of jugular foramen paragangliomas, it is necessary to specify indications for surgical treatment of these tumours. They include: age below 45 years, persistent cranial nerve deficits, tumour with a low risk of postoperative complications, intracranial expansion, paragangliomas in other locations on the same side, malignant transformation signs, internal carotid artery infiltration with favourable results of balloon obturation test, and recurrent tumour following previous radiotherapy [28]. An elective combination of surgery and radiotherapy is also feasible. Initially, a large tumour is surgically reduced to the approximate size of 3 cm without any permanent deficits evoked. Radiotherapy follows as a next step of the therapeutic protocol either obligatorily or implemented in the case of progression of the remaining tumour [28].

The question arises whether to forbear the treatment of jugular foramen paragangliomas in favour of observation with systematically performed neuroimaging studies. In our opinion, the answer is yes, but in those cases where tumours are found accidentally without any clinical signs. Tumours with discrete signs might be followed as well, up to the point where radiological or clinical signs of progression occur. The only group of tumours that requires immediate attention constitutes those that are hormonally active [11]. The decision to wait and see becomes easier with the growing age of the patient.

Conclusions

1. A proper choice of a jugular foramen approach based on topography and size of paraganglioma that develops within the jugular foramen and its vicinity facilitates reduction of perioperative injury without a negative influence on the resection radicality.
2. Lack of radicality in the surgical treatment of paragangliomas of the jugular foramen and its vicinity arises from the effort to avoid postoperative deficits of the lower complex of the cranial nerves within the cerebellopontine angle.
3. The vast complex of disorders resulting from surgical treatment of paragangliomas of the jugular fora-

men and its vicinity that includes both persistent signs and symptoms from the tumour and adverse effects of the treatment is significantly limited and mitigated by rehabilitation and convalescence.

Disclosure

The authors report no conflict of interest.

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