FIVE-YEAR EXPERIENCE IN SURGICAL TREATMENT OF TEMPORAL BONE PARAGANGLIOMAS

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ABSTRACT

The article describes our five-year experience in the surgical treatment of temporal bone paragangliomas. Considering our experience, the surgical treatment of patients with different types of paragangliomas based on a detailed study of the CT scan and MRI data using the preoperative selective embolization of tumor-feeding vessels and the navigation system intraoperatively as well as the endoscopy assistance allows total removing of the tumor with minimal damaging of the vital structures of the lateral skull base.

Keywords: *temporal bone paragangliomas, ear surgery, surgical treatment, CT, MRI, endoscopy, navigation system, vascular embolization*

INTRODUCTION

A paraganglioma is a rare tumor that arises from cells of nonchromaffin paraganglia, also called glomus bodies. In consideration of that it has many synonyms: glomus tumor, jugular and tympanic glomus tumor, chemodectoma, receptoma, nonchromaffin paraganglioma, etc. Paragangliomas are observed in various organs and tissues. They are located most often in the area of the carotid artery bifurcation (carotid artery tumor), along the vagus nerve (vagal paragangliomas), in the jugular fossa and tympanic cavity (jugulotympanic paragangliomas). Paragangliomas take the second place among tumors of the temporal bone after schwannomas of the vestibulocochlear nerve (M.A.S. Garcia 2007). The degree of incidence of temporal bone paraganglioma is 1:1300000 (1). Most often, paragangliomas originate from the jugular vein bulb over the promontory and along the Jacobson (tympanic branch IX) or Arnold (auricular branch X) nerve. This is a benign tumor, but according to the literature, it can sometimes metastasize, even a few years after a successful surgery (2). The paraganglioma is characterized as a tumor with slow growth, it tends to invade the bone structures and the dura mater and is highly vascularized, which leads to a frequent recurrence (3,4). The temporal bone paraganglioma is more common in people aged 40 to 50, although it can occur at any age. According to the literature, women suffer more often than men (5,6,7,8).

The patients complain most often of pulsating tinnitus and hearing loss. Damaging of the cranial nerves can be manifested by tongue paralysis, swallowing problem, and glottic incompetence (9). A less frequent symptom is facial nerve dysfunction. Further growth of the mass in the posterior cranial fossa can lead to compression of the cerebellum, brainstem, and even to occlusal hydrocephalus (10). According to the literature, the time interval between the first symptoms and the diagnosis of paragangliomas is from 4 to 6 years (11). At the stage of diagnosing and planning the further treatment, CT scanning is useful for detecting the temporal bone damage and the degree of spread at an early stage of the disease. MRI assessment is the method of choice in assessing the involvement of the middle ear, the skull base or the posterior fossa and monitoring the growth of the paraganglioma (12). Morphologically there are different types of paraganglioma differentiated based on investigation of Diab Kh.M. and authors (13). According to the Sanna's modified classification of U. Fisch and D. Mattox dated 2013, depending on the degree of spread, they are divided into four main types: type A, type B, type C, and type D (14,15).

All these qualities make glomus tumors one of the most difficult pathologies in otosurgery and emphasizes the importance of early detection of these tu-

mors and timely complete surgical removal with the possibility of preserving the function of vital anatomical structures (16). There are various strategies of treatment of patients with common paragangliomas. With the improvement of microsurgical techniques, the advent of intraoperative monitoring of cranial nerves, the ability to use navigation systems and video-endoscopic assistance, the surgical removal of the tumor is preferred. The exception is provided by the patients with the tumor invasion into the internal carotid artery and with cranial nerves involved in the pathological process, to which a radiosurgery is indicated (17).

The purpose of the study is to increase the effectiveness of treatment by using additional equipment in the surgical treatment of patients with various forms of temporal bone paragangliomas, depending on the prevalence of the process.

MATERIALS AND METHODS

During the period from February 2015 to March 2020, 103 patients with temporal bone paragangliomas (36 men and 67 women) aged 2 years to 82 years were examined and operated in the National Medical Research Center of Otorhinolaryngology Federal State Budgetary Institution. To determine the degree of spread of the tumor process, the involvement of important anatomical structures, and the choice of the optimal tactics of surgical treatment, all patients underwent computed tomography of the temporal bones and contrast-enhanced MRI of the head at the presurgical stage.

According to the results of radiological research methods, depending on the prevalence of the process according to the international classification of temporal bone paragangliomas (Table 1), the patients were divided into 3 groups according to tumor types.

The first group included 36 patients with type A paraganglioma among them 13 patients with a type A1 tumor, and 23 patients with a type A2 tumor.

The patients of this group complained with pulsating tinnitus and hearing loss; facial nerve dysfunction was not noted. The otomicroscopy showed, a red mass behind the eardrum. The CT scan of the temporal bones showed a soft-tissue substrate, partially filling the tympanic cavity. (Fig. 1 type A CT).

The contrast-enhanced MRI images of the brain showed an active contrast accumulating tumor in the tympanic cavity.

The second group consisted of 44 patients with paraganglioma type. Depending on the spreading of the pathology process, type B1 tumor was detected in 14 cases, type B2 tumor was detected in 13 patients, and type B3 was detected in 17 cases. The patients complained of pulsating tinnitus and hearing loss on the affected side, and temporary facial numbness. During otoscopy, a red pulsating mass involving the eardrum was determined. According to the CT scan of the temporal bone a soft-tissue mass filled the tympanic cavity with spreading to the antrum and mastoid process cells. The tegmen tympany was thinned. The bone wall of the canal of the internal carotid artery eroded in the vertical and horizontal parts in patients with type B3 glomus tumor. The tumor was spread into the infralabyrinthine tract cell with bone destruction in this area (Fig. 2 type B CT).

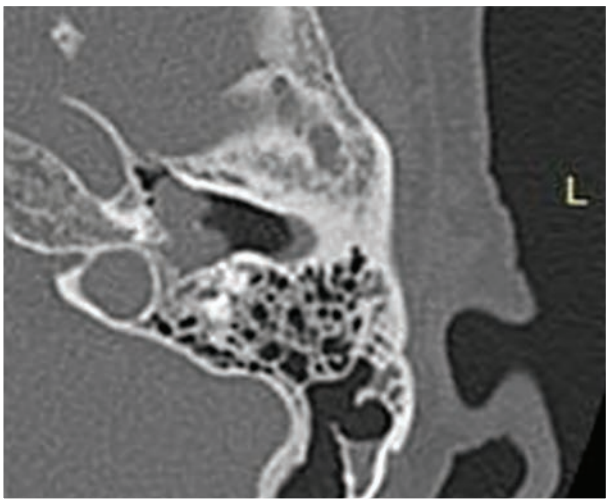

Fig. 1. CT scan of the left temporal bone, axial projection. Type A paraganglioma: the tympanic cavity is filled with a soft-tissue substrate, the destruction of the jugular vein bulb is not observed

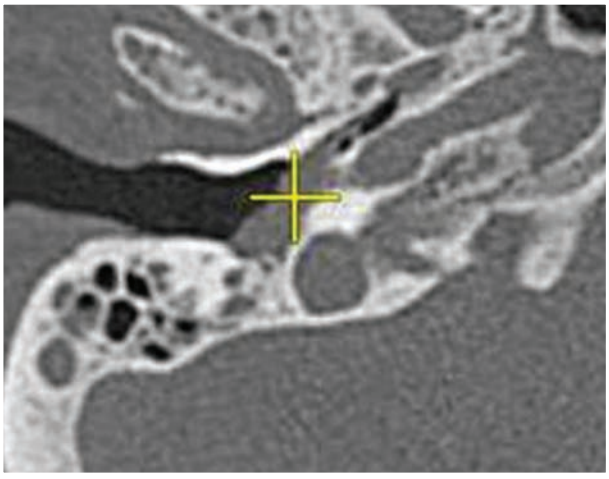

Fig. 2. CT scan of the right temporal bone, axial projection. Type B paraganlioma: the tympanic cavity is filled with a soft-tissue substrate, thinning of the bone wall of the jugular bulb is observed

The contrast-enhanced MRI of the brain and soft tissues of the neck showed the spreading of the tumor into the cells of the mastoid process, without invasion of the jugular vein bulb and internal carotid

artery). A fluid filling the cells of the mastoid process was detected (Fig. 3 type B MRI).

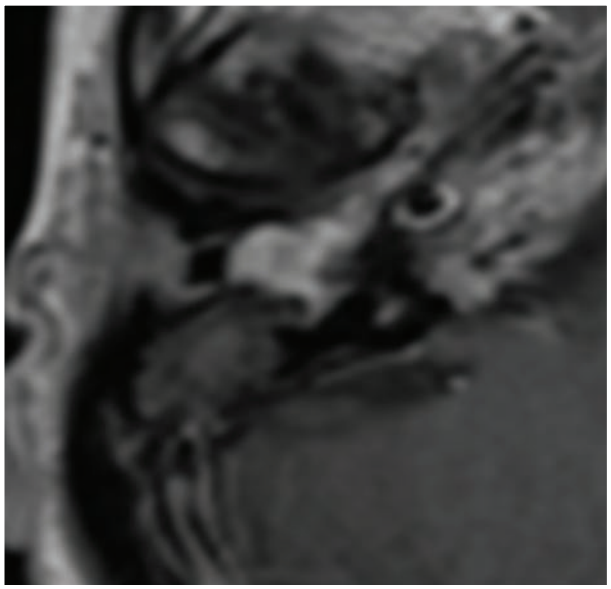

Fig. 3. MRI of the brain and soft tissues of the neck, T2-VI with signal suppression from adipose tissue, in the coronal plane. Type B paraganglioma

The third group consisted of patients (n=23) with widespread type C temporal bone paraganglioma. According to the degree of tumor spreading, they were divided into 3 types: type C1 - 9 patients, type C2 - 7 patients, type C3 - 7 patients.

The patients with paraganglioma type C complained of pulsating tinnitus and hearing loss on the affected side, periodic facial numbness, and periodic bleeding from the acoustic meatus. Six patients who had undergone surgical interventions repeatedly before entering our clinic had a facial nerve paresis stages 4-6 (House-Brackmann score) on the affected side. The video endoscopy showed a pulsating red mass protruding into the lumen of the external acoustic meatus. The CT of the temporal bones showed a soft-tissue substrate filling the tympanic cavity. The tegmen tympany was thinned, and eroded (1). The bone wall of the internal carotid artery canal in the vertical and horizontal parts eroded. The bone wall of the jugular bulb was destroyed by the tumor (2) (Fig. 4 type C CT).

The MRI of the brain and soft tissues of the neck showed the spread of the mass to the cochlea capsule and to the area of the right cisterna magna (Fig. 5,6). The corresponding parts of the brain were not involved. The tumor involved the internal jugular vein (1). The lumen of the internal jugular vein was not observed in this area (2) (Fig. 6)

The patients received computer angiography 24 hours before surgical treatment and endovascular

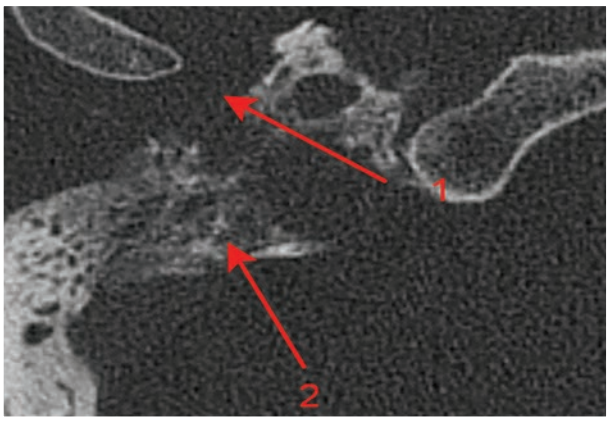

Fig. 4. CT scan of the right temporal bone, axial projection. Type C paraganglioma: the posterior wall of the mandibular fossa is not observed in some places (1). The soft-tissue content that destroys the bone walls of the hypothympanum is observed at the level of the jugular fossa (2)

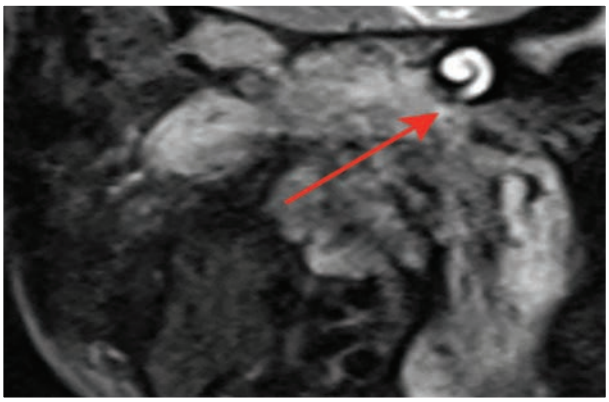

Fig. 5. MRI of the brain and soft tissues of the neck, T2-VI with signal suppression from adipose tissue, in the coronal plane. Type C paraganglioma: the spread of the mass to the cochlea capsule is observed

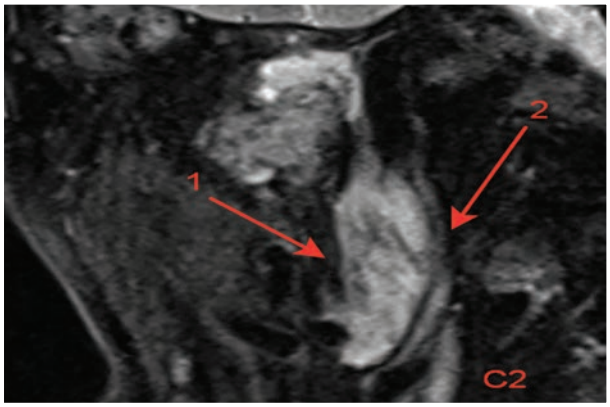

Fig. 6. MRI of the brain and soft tissues of the neck, T2-VI with signal suppression from adipose tissue, in the coronal plane. Type C paraganglioma: the signs of the process spreading along the internal jugular vein caudally to level C2 are observed, with the size of the extracranial component being 50x18 mm (1). The lumen of the internal jugular vein in this area is not visible (2), the sub-stenotic sections are not significantly expanded

Table 1. Surgical approaches depending on the prevalence of the process n=103

	Type	Patients	Surgical Access
Group I n=36	A1	13	retroauricular transmeatal
	A2	23	
Group II n=44	B1	14	transmastoid with dilated posterior tympanotomy
	B2	13	
	B3	17	transmastoid without preserving the posterior wall of the external acoustic meatus
Group III n=23	C1	9	retrofacial access
	C2	7	
	C3	7	infratemporal access

selective embolization of tumor-feeding vessels was performed.

All surgeries were performed using neuromonitoring of the facial, glossopharyngeal and recurrent nerves. To facilitate the orientation in patients of the third group with type C tumors, when removing the tumor tissue along the vertical portion of the canal of the internal carotid artery and from the infralabyrinthine cell tract, an electromagnetic navigation system and video-endoscopic assistance were used.

The retroauricular transmeatal approach was used in cases of paraganglioma type A (1 group-n=36). After canaloplasty and tympanotomy, the tympanic membrane was separated totally from the malleus and the tumor was totally removed from the tympanic cavity as well as from the oval, round window niches and the Eustachian tube. The ossicles remained intact in some cases we cut a part of the long process of the malleus. At the end of the procedure an underlay tympanoplasty using temporal fascia graft was performed (Fig. 7). The use of photoangiolytic laser in the surgical treatment of temporal bone paraganglioma type A, B can be useful to re-

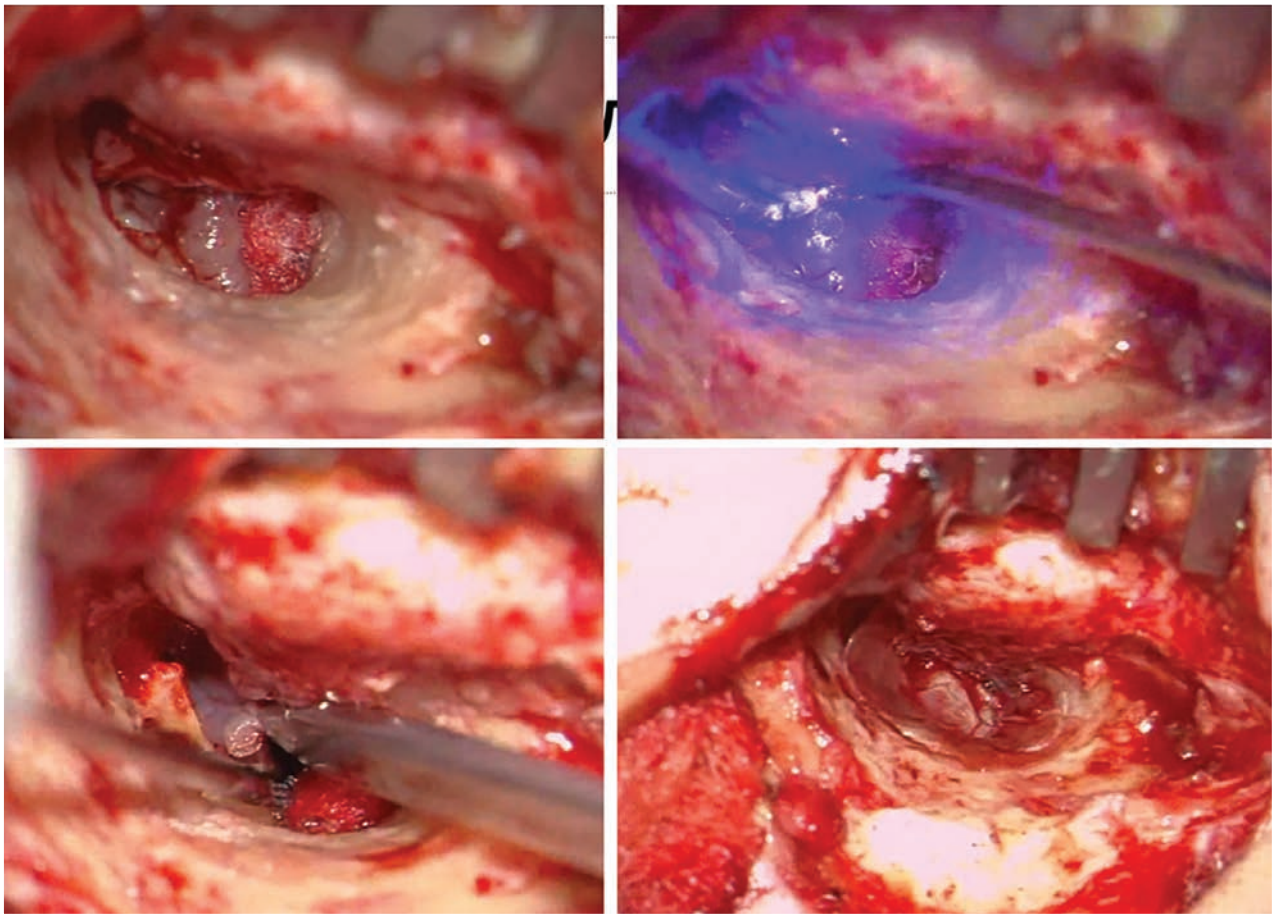

Fig. 7. Type A2 paraganglioma. Stages of retroauricular transmeatal approach

duce the bleeding during separation and removal of the tumors (18).

In the second group 44 patients the transmastoid access has been used for tumor removing. During the intervention, the soft tissues and skin of the walls of the external acoustic meatus were separated in patients with type B1 paraganglioma. Canaloplasty and tympanotomy were also performed. A wide antro-attico-mastoidotomy with posterior tympanotomy was done then the retrofacial access was used to the hypotympanum. The feed vessels were coagulated and the tumor was removed under videoendoscopic control with the preservation of the ossicular chain and anatomical structures of the middle ear.

The patients with type B2 and B3 tumors underwent an extended radical mastoidectomy, the tumor was completely removed from the tympanic cavity, the mastoid process, Eustachian tube and area of the internal carotid artery canal were reviewed under endoscopic control and electromagnetic navigation. Tympanoplasty with using temporal fascia graft was performed (Fig. 8).

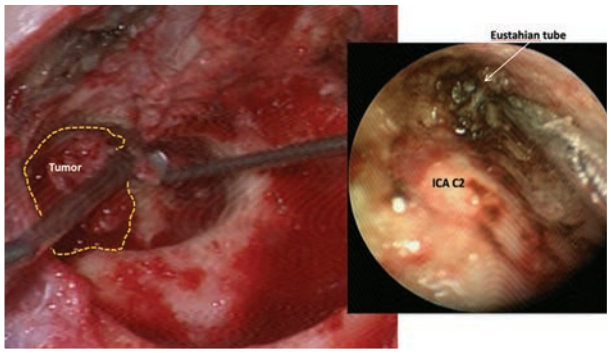

Fig. 8. Type B3 paraganglioma – deep anterior extension to the Eustachian tube with destruction of the petrous part of ICA canal (C2). Intraoperative photos

The patients of the third group (n=23) with type C1-C2 tumor underwent surgery using retrofacial access since the complete removal of the tumor required good visualization of the internal jugular vein bulb, internal carotid artery and VII, IX-XII pairs of cranial nerves (Fig. 9).

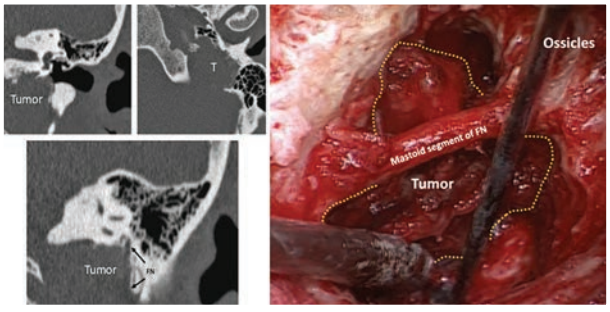

Fig. 9. Type C1 paraganglioma. Retrofacial approach

The infratemporal access (5,17,18,20) was used in 7 patients with a type C3 tumor due to the spread of the tumor into the jugular vein lumen, the jugular vein was ligated at the level of the C1 vertebra behind the posterior bell of the digastric muscle in the area of its insertion (Fig. 10).

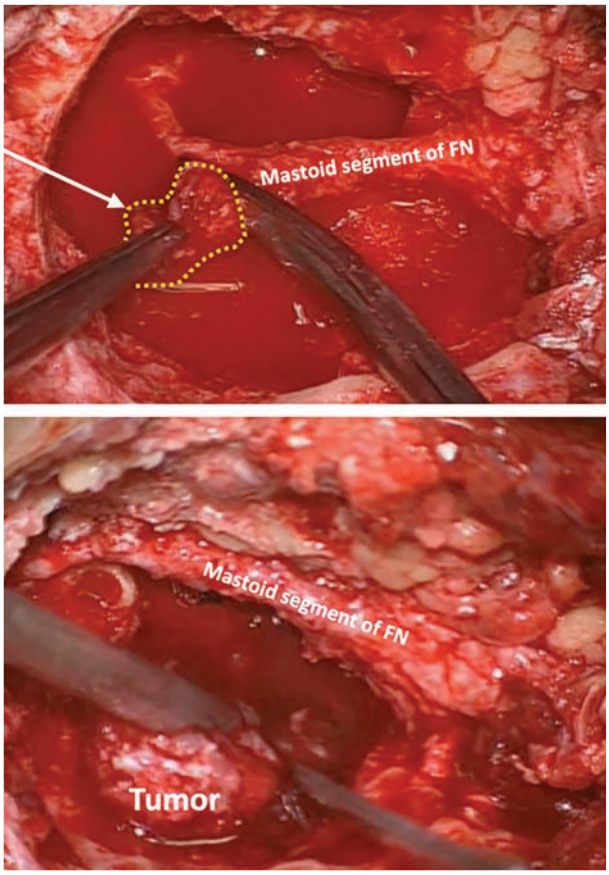

Fig. 10. Type C2 paraganglioma. Infratemporal approach is used

During the surgery, the facial nerve was mobilized from the stylomastoid foramen to the geniculate ganglion and transposed anteriorly. In all cases, the medial and lower surfaces of the mastoidal segment of the facial nerve were adherent to the tumor tissue. Then the sigmoid sinus was skeletonized, opened and obliterated; the tumor was removed together with the jugular vein bulb. It was possible to compare and contrast the MRI and CT scans of the process spread intraoperatively with the help of an electromagnetic navigation system (Fig. 5,6,7,8,9). The tumor fragments were removed from the walls of the internal carotid artery under the control of navigation system. After removal of the tumor and hemostasis, the facial nerve was covered by auto-fascia graft; the postoperative cavity was obliterated with abdominal fat. The acoustic meatus was blindly closed (cull de sac).

RESULTS AND DISCUSSION

The results were evaluated in the immediate and long-term postoperative period, the quality of tumor removal and the saving of facial nerve function were assessed. To assess the functional results, the degree of paresis was determined in all patients. An improvement in hearing in the postsurgical period was observed in group I patients with type A paragangliomas; the facial paresis was not observed. Twenty-four hours before surgery, 6 patients with a type A2 tumor underwent selective embolization of the tumor-feeding vessels; the remaining 30 patients were operated without preliminary embolization. Intraoperatively, a decrease in bleeding from the tumor was observed in patients after embolization, but no significant differences in the total volume of blood loss between the group I patients with and without embolization were detected. During re-examination after 6 to 12 months, according to MRI, no tumor recurrence was observed in any of the patients with type A tumors.

In the group II patients with type B paragangliomas an improvement in hearing after surgery was observed in patients with type B1 tumors; the hearing remained unchanged or, in some cases, a moderate hearing decrease was noted in patients with type B2 and B3 tumors. Twenty-four hours before surgery, 13 patients with a type B tumor underwent embolization of the tumor-feeding vessels, the remaining 31 patients were operated without preliminary embolization. Intraoperatively, a decrease in tumor size and a decrease in bleeding from the tumor was observed in patients after embolization; the difference between the volume of blood loss with embolization and without embolization was 100-150 mL. Facial nerve dysfunction in the early and long-term postsurgical period was not observed. According to the radiology examinations, tumor recurrence was not observed after 6-12 months (Fig. 11, 12).

In the third group, 18 patients underwent selective embolization of the tumor-feeding vessel 24-48 hours before surgery; the remaining 5 patients were operated without preliminary embolization (Fig.13, 14). Intraoperatively, a decrease in tumor size and a decrease in bleeding from the tumor after embolization were observed, the difference between the volume of blood loss with embolization and without embolization was 500-700 mL.

It was possible to save the function of the cranial nerves. In the early postsurgical period, all patients with type C1 and C2 tumors had a paresis of 2-3 degrees; facial dysfunction of 4-5 degrees according to House-Brackmann score was observed in patients with a type C3 tumor. In the long-term postsurgical period (6-12 months), the function of the facial nerve

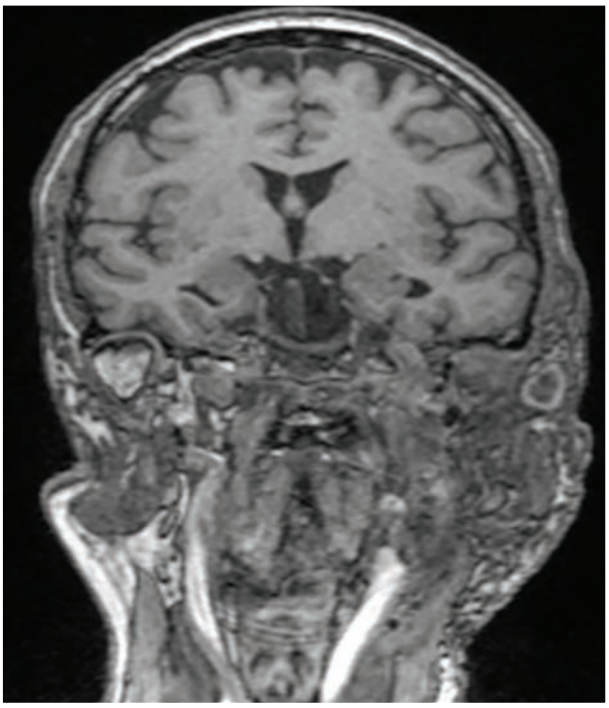

Fig. 11. MRI of the brain and soft tissues of the neck, T2-VI with signal suppression from adipose tissue, in the coronal plane. Type C paraganglioma: postsurgical control 6 months after surgical treatment: no data for tumor recurrence

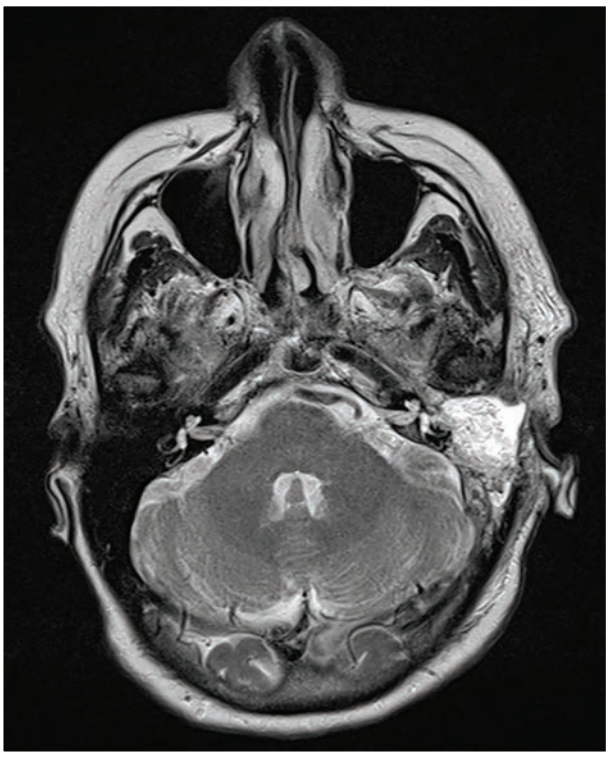

Fig. 12. MRI of the brain and soft tissues of the neck, T2-VI with signal suppression from adipose tissue, in the coronal plane. Type C paraganglioma: postsurgical control 6 months after surgical treatment: no data for tumor recurrence

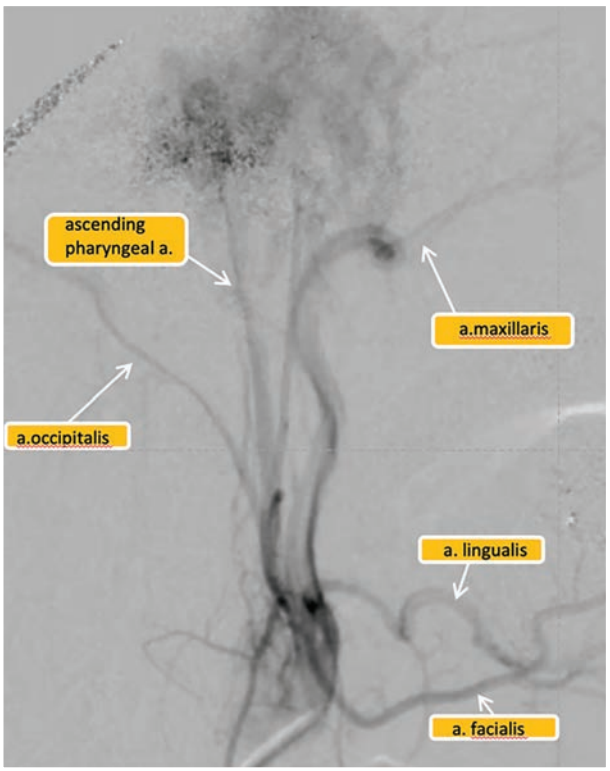

Fig. 13. Angiography before selective embolization of the tumor-feeding vessel

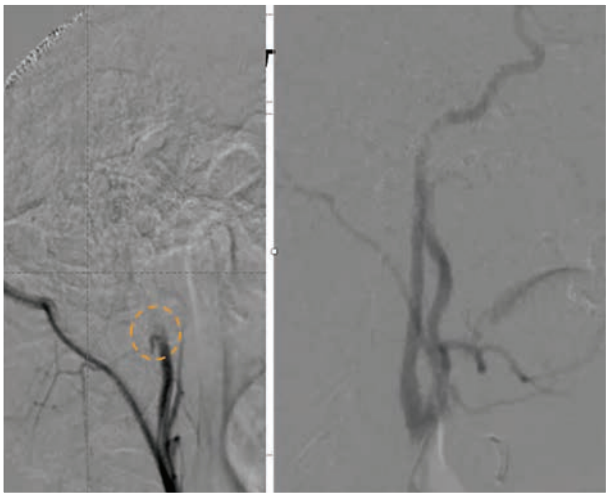

Fig. 14. After selective embolization of the tumor-feeding vessel

in patients with type C1 and C2 paragangliomas was fully restored, and the function of the facial nerve in patients with type C3 was restored to the level of 2-3 degrees according to House-Brackmann score, except for 3 patients in whom paresis was noted prior to our surgical intervention.

According to CT and MRI, tumor recurrence in the postsurgical period was observed in one patient with type C3 paragangliomas; no recurrence was observed in the remaining patients from group III (the maximum follow-up period was 6-12 months).

CONCLUSION

The detailed study of preoperative radiological data in patients with temporal bone paragangliomas allowed to choose an adequate surgical approach to obtain a good visualization of the anatomical structures, which allowed to completely remove the tumor. The use of selective embolization of tumor-feeding vessels reduced intraoperative blood loss in all groups of patients. However, in patients with paragangliomas of type A1, A2, and B1, there were no significant differences between blood loss with and without embolization. The use of an electromagnetic navigation system, video-endoscopic assistance and neuromonitoring of cranial nerves during surgical intervention allowed to prevent serious postsurgical complications and to achieve good functional results: it was possible to save hearing in patients with tumors of type A1, A2, B1 completely, save the functioning of the cranial nerves. In most patients with type C tumors the facial nerve function was completely restored in 6-12 months after the surgery.

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