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# EAR AND SKULL BASE PARAGANGLIOMA: REVIEW OF LITERATURE

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**Key words:** *glomangioma, neurosurgery, tympanoplasty, embolization, rare tumors*
**Ключові слова:** *гломангіома, нейрохірургія, тимпанопластика, емболізація, рідкісні пухлини*

**Abstract. Ear and skull base paraganglioma: review of literature. Abdrakhmanova D., Niemczyk K., Mukhamadieva G., Mustafin A., Papulova N.** *The actuality of the research is determined by the lack of systemized information on the ear and skull base paraganglioma. Currently, from 500 to 1000 cases of paraganglioma or pheochromocyte are diagnosed in the United States of America annually. The combined morbidity of benign tumor forms is evaluated to be between 0.7 to 1.0 for 100000 people per year. Even though the results in understanding the nature of paraganglioma, as well as treatment thereof, are successful, the amount of literature on this topic only increases. The research aims to analyze study of the terminology, classification, diagnostics, and treatment of paraganglioma. Special attention was brought to the preoperative preparation and embolization of vessels, which supply blood to the tumor as well as to the problem of repeated bouts of glomangioma. The objective of the research is the selection of relative sources, argumentation analysis, and material systematization. For the analysis, 53 research articles were included. During the compilation of this study, such methods, as analysis, synthesis, information collection and systematization, and generalization, were used. In this article, various paraganglioma classifications, endoscopic treatment methods, other aspects of paraganglioma therapy, and diagnosis and treatment thereof, were presented. More and more researchers recommend individual and adapted approaches for each case with the inclusion of factors, related to the patient, including age, treatment goals, other infections, gene status, anatomical issues, caused by the tumor growth, its sizes, and other factors. This literature review focuses on ear and skull base paragangliomas, rare, usually benign, hyper-vascularized neuroendocrine tumors often treated surgically. Alternative treatments like cyber-knife, gamma-knife, and endoscopic approaches are also discussed, highlighting their efficacy for specific cases. The study emphasizes individualized treatment plans considering patient-specific factors, with stereotactic radiosurgery being a safe and effective option for many patients. Materials of the research can be used by doctors of various specializations to enrich their knowledge and for further study of paragangliomas of various localizations.*

**Реферат. Парагангліома вуха та основи черепа: огляд літератури. Абдрахманова Д., Немчик К., Мухамадієва Г., Мустафін А., Папулова Н.** *Актуальність дослідження визначається відсутністю систематизованої інформації про парагангліому вуха та основи черепа. На цей час у Сполучених Штатах Америки щорічно діагностується від 500 до 1000 випадків парагангліоми або феохромоцитоми. Сукупна захворюваність на доброякісні форми пухлини оцінюється в межах 0,7-1,0 на 100 тис. осіб на рік. Незважаючи на те, що результати щодо розуміння природи парагангліоми, а також її лікування успішні, кількість літератури з цієї теми тільки збільшується. Метою дослідження є аналіз вивчення термінології, класифікації, діагностики та лікування парагангліом. Особливу увагу приділено передопераційній підготовці та емболізації судин, які кровопостачають пухлину, а також проблемі повторних випадків гломангіоми. Метою дослідження є відбір відповідних джерел, аналіз аргументації та систематизація матеріалу. До аналізу було включено 53 наукові статті. Під час виконання цього дослідження були використані такі методи, як аналіз, синтез, збір і систематизація інформації, узагальнення. У статті представлені різні класифікації парагангліом, ендоскопічні методи лікування, інші аспекти терапії парагангліом, їх діагностика та лікування. Усе більше дослідників рекомендують індивідуальні та адаптовані підходи для кожного випадку з урахуванням факторів, пов'язаних з*

*пацієнтом, включаючи вік, цілі лікування, інші інфекції, генний статус, анатомічні проблеми, викликані ростом пухлини, її розміри та інші фактори. Цей огляд літератури присвячений парагангліомам вуха та основи черепа, рідкісним, зазвичай доброякісним, гіперваскуляризованим нейроендокринним пухлинам, які часто лікують хірургічним шляхом. Також обговорюються альтернативні методи лікування, такі як кібер-ніж, гамма-ніж та ендоскопічні підходи, підкреслюючи їхню ефективність у конкретних випадках. Дослідження наголошує на необхідності індивідуальних планів лікування з урахуванням конкретних факторів пацієнта, при цьому стереотаксична радіохірургія є безпечним й ефективним варіантом для багатьох пацієнтів. Матеріали дослідження можуть бути використані лікарями різних спеціальностей для збагачення своїх знань і подальшого вивчення парагангліом різної локалізації.*

More than 200 years ago, scientist G. Valentin [1] spotted a small cellular formation, located near the tympanic nerve, which he first identified as a ganglion, and called it “gangliolum tympanicum”. In the year 1878, House demonstrated, how vascularized this organ is, as well as its histologic similarity to the so-called sleeping gland, and proposed to rename it into the tympanal gland, or the glandula tympanicum [2]. Currently, in the literature of the 21st century, paraganglioma is also denoted with the following terms: chemodectoma [3]; glomangioma [4, 5, 6] and angiomyoneuroma [7]. Pheochromocytomas and paragangliomas are formed from the neural crest tissue. The latter serves as a basis for sympatic and parasympatic paragangliomas [8]. Sympatic paragangliomas are considered: adrenal medulla; paraotric body (chromaffnic bodies), located near the aortic bifurcation; other paragangliomas of the sympatic neurological system of the human. The following are considered parasympatic paraganglioma: carotid body; other paragangliomas of the X and IX branch couples of the skull nerves [8].

Paragangliomas are rare neuroendocrine tumors that secrete catecholamines (norepinephrine). They are usually localized in the preaortic and paravertebral sympathetic plexus or at the base of the skull. Paragangliomas of the head and neck in the jugular foramen, ear, or body of the carotid artery are less differentiated tumors. They secrete norepinephrine, while more differentiated intra-abdominal tumors of the adrenal medulla, such as neuroblastoma and pheochromocytoma, mainly secrete adrenaline [9]. There are numerous case reports in which paragangliomas and pheochromocytomas develop synchronously in the same patient [10]. The name paraganglioma usually coincides with the localization of the tumor: the carotid body lesion occurs in the region of the bifurcation of the carotid artery, glomus tympanicum occurs in the middle ear, but is not found in the jugular foramen, while the jugular-tympanic paraganglioma (formerly called glomus jugulotympanicum) is localized in the middle cavity, ear and upper lateral region of the jugular foramen, glomus vagale is located in the posterolateral part of the pharynx, by analogy, paraganglioma of the larynx is diagnosed in the larynx. However, there is an exception to the rule: chemodectoma. Its location is not so obvious due to the function of chemoreceptors. It should be noted that parasympatic paragangliomas are also called non-chromaffin, while sympatic paragangliomas are chromaffin secreting [11].

Glomus tympanicum arises from middle ear paraganglia cells and differs from glomus jugulare in the absence of bone erosions around the jugular bulb. Most tumors are benign and non-secretory, but new genetic data suggest that some patients are at increased risk for more aggressive, functional, and/or multifocal diseases [12]. Annually, from 500 to 1000 cases of paraganglioma or pheochromocyte are diagnosed in the United States of America. Combined morbidity of benign paraganglioma or pheochromocyte tumors forms is evaluated to be between 0.7 to 1.0 for 100000 people per year. Malignant forms of paragangliomas occur with a frequency of 90 to 95 cases per 400 million people per year. Sporadic paraganglioma is usually diagnosed between the ages of 30 and 50 and is three times more common among women. The hereditary type is usually diagnosed earlier, at the age of 30, and it is equally often detected among men and women [13, 14, 15]. There are still unresolved questions regarding determining radiation treatment effects on patients with head or neck paragangliomas [16], the value of routine disease indicators employment [17], preoperative embolization [18], functional visualization [19], genetic level diagnostic [20], resolution of which are being discussed by many authors. As such, the information, regarding the ear and skull base paragangliomas, is being collected, even though there are still gaps in the systematization and generalization of data.

This analysis aims to highlight the issues of terminology, classification, diagnosis of paragangliomas (glomus tumors), and modern advances in their treatment (both in surgical and radiation and chemotherapeutic methods). The issues of transcanal surgical techniques and the restoration of sound conduction during the removal of a glomus tumor and its impact on the patient's quality of life will also be considered. Attention was brought to the preoperative preparation and embolization of vessels, which supply blood to the tumor as well as to the problem of

glomangioma repeated bouts. The objective of the research is the selection of relative sources, argumentation analysis, and material systematization.

**MATERIALS AND METHODS OF RESEARCH**

This article is mostly theoretical. As such, these methods were used: analysis (to determine research mode, data analysis mode, statistical corpus employment, highlighting specific parts and data of publications), synthesis (combining separate elements, parts, and attributes into a single unit), information systematization and collection (a division of research results into a classification of paragangliomas, diagnostics, treatment), generalization. Publications, dated as years 1840 to 2022, were used. PubMed publication base and Cochrane library were used. All literature was in the English language. To determine potential articles, the following medical terms were used during the search: glomus tympanicum, paraganglioma of the ear, paragangliomas of the head and neck, paragangliomas treatment, paragangliomas diagnosis, middle ear paragangliomas.

This strategy detected 825 possible articles, which can be used in the research. Articles were excluded based on one or more of the following criteria: only an abstract and no access to the full article, no abstract or access to the full text of the article, the original language of the article is not English, the use of alternative medicine methods in the treatment or diagnosis. Amongst them, 647 articles were excluded, while 203 articles were employed for a complete review. Then, the full text of the 178 remaining articles was reviewed. As a result, 53 articles were included in the final article. Investigators examined general descriptive data from the article (e.g., country of origin, number of patients, mean age, sex and age structure of the study subjects), procedures (method of diagnosis, treatment, and control of bleeding, study design, duration of follow-up), and observed treatment complications (their number, nature, the need for blood transfusion, the need for interventions to stop bleeding and death) and other data.

As such, during the literature review, the authors, have concluded: general evaluation; analytical evaluation; format evaluation. The confidential data of each patient was kept secret during the research. All personal information, such as medical record numbers, was not included in the dataset of either the individual studies or the review as a whole, and the patients described by the data remain anonymous. All procedures performed in the study were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki Declaration and its later amendments. Ethical consent was given by the Astana Medical University Ethics Board, approval number JF-138001. Data were entered into a computer and then structured and systemized with the help of a standard Microsoft Office Excel package (2016).

**RESULTS AND DISCUSSION**

**Tumor classification**

Great progress has been made in the understanding of these tumor nature, as well as their treatment, during the last decade. The proper classification of this tumor variety, however, is still not compiled. As such, additional work should be focused on that [21]. Below the main classifications, described in the literature, are the classifications, employed in the medical practice. Glomus tumor classification on the initial growth location is presented in Table 1. This classification does not have an author and was created during practice.

*Table 1*

**Glomus tumor classification on the initial growth location**

| | |
|---|---|
| **Tympanal glomus (Glomus tympanicum and hypotympanicum)** | **Initial growth on the Tympanic cavity, medially located** |
| **Glomus jugulare** | **Glomus tumor on a bulb of jugular vein** |
| **Glomus vagale** | **Tumor in the tenth cranial nerve** |
| **Carotid body tumor** | **Initial growth in the bifucation zone of carotid artery** |

U. Fisch and D.E. Mattox [22] classification of paragangliomas of the lateral skull base is presented in the Table 2 and was employed during the patient treatment. Literature also describes the usage of C.G. Jackson et al. [23] paragangliomas of the lateral skull base classification (Table 3) and U. Fisch and D.E. Mattox [22] crotaphic paragangliomas classification (Table 4).

*Table 2*

## Paraganglioma of the lateral skull base classification

| | |
|---|---|
| **Class A** | **Tumor is restricted by the middle ear cavity** |
| **Class B** | **Tumor is restricted by the tympan-mastoid cavity, without the infralabirintial compartment.** |
| **Class C** | **Tumor, located in the infralabirintial compartment of a temporal bone and grown on a pyramide apex** |
| **Class C1** | **Limited infection of carotid artery vertical element** |
| **Class C2** | **Inner carotid artery vertical element invasion** |
| **Class C3** | **Inner carotid artery hotizontal element invasion** |
| **Class C4** | **Sphenotic foramen and cavernous sinus invasion** |
| **Class D** | **Tumor with inner-skull spread** |
| **Class De** | **Extradural expansion** |
| **Class Di** | **Intradural expansion** |
| **Class D1** | **Tumor with a size no more than 2 cm** |
| **Class D2** | **Tumor with a size no bigger than 2 cm** |

**Note.** Source – U. Fisch and D.E. Mattox [22].

*Table 3*

## Paraganglioma of the lateral skull base classification

| | |
|---|---|
| **Type 1** | **Tumor edges are fully visible during otoscopy** |
| **Type 2** | **Tumor is in the middle of ear cavity** |
| **Type 3** | **Tumor is spreading on a middle ear and tympan-mastoid appendage** |
| **Type 4** | **Tumor on the eardrums and the outer external auditory canal** |

**Note.** Source – A.D. Sweeney et al. [12].

*Table 4*

## Crotaphic paragangliomas classification

| | |
|---|---|
| **Type** | **Paraganglioma location** |
| **Type A** | **Across the tympanic plexus on the cape** |
| **Type B** | **Hypotympanon spreading, the tubular bone near jugular bulb is intact** |
| **Type C1** | **Paraganglioma with an erosion of carotid hole** |
| **Type C2** | **Carotid canal infection** |
| **Type C3** | **Carotid canal infection, sphenotic foramen is intact** |
| **Type C4** | **Tumor grows into sphenotic foramen and cavernous sinus invasion** |
| **Type De1/2** | **Intercranial distribution, without arachnoid cavity infiltration. De1-De2 on the shift of the pachymeninx** |
| **Type Di1/2/3** | **Intradural spreading. Di1-Di3, depending on the rear skull cavity depth invasion** |

**Note.** Source – U. Fisch and D.E. Mattox [22].

**Diagnostics**

Most paragangliomas are diagnosed during the examination of unknown tumors or the checkup due to the abnormal catecholamine level, and during the catecholamine screening in patients with the diagnosed family syndrome (Von Hippel-Lindau disease, 1st type neuromatosis and patients with hereditary tumors due to mutation of succinate dehydrogenase gene resistance) [8, 24]. Magnetic resonance tomography (MRT) and Computer Tomography (CT) are the main tools of glomus tumor diagnosis and complete each other [25, 26, 27, 28]. In the research, in which a selection of 26 patient's disease history was analyzed and was divided into two groups: with neck paraganglioma and with temporal bone paraganglioma, MRT sensitivity, in these tenth cranial nerve tumor diagnostics, were up to 75%, while specifics was at around 90%. In the same cohort, temporal bone paragangliomas appeared as pulsatile masses behind the tympanic membrane, causing varying degrees of hearing loss in 91% of cases. The facial nerve was the most infected cranial nerve (was spotted in more than 2/3 of cases) [29].

Suffice it to note, that any patient with hearing loss, dizziness, and tinnitus, is subject to a close otologic and otomicroscopic examination during the clinical checkup. In addition, the role of imaging and interventional radiology in improving diagnosis and therapeutic management cannot be overstated [30]. Genetic testing and assessment of tumor neurosecretory function should be performed based on the patient's medical history and family history [12]. Determining the metastatic potential of a tumor is difficult, yet it is important for an individual assessment and adjuvant treatment of each patient after the initial surgery. Primary biochemical tests are metanephrine plasma, 3-Methoxytyramine as well as chromogranin-A. Genetic tests are important for consultation, and prognosis values. Aside from CT and MRT, molecular visualization through positron emission tomography (PET) should be employed [24].

**Treatment**

Paragangliomas of the head and neck are tumors that require good surgical skills from the doctor to ensure complete excision of the pathology. This is important as the results of their therapy in terms of morbidity, recurrence, and rehabilitation are described as "excellent" [21]. Surgical removal of a tumor is a prioritized method of treatment as it has several benefits, such as full removal, low level of complications, low risk of secondary infection, sufficient hearing, and low pain [31]. The literature recommends surgical removal for patients with small tumors due to the high control level and smaller neurological deficit (especially of the cranial nerves) in comparison with larger tumors. At the same time, researchers note, that patients with Fisch C and D temporal bone paragangliomas are treatable via surgical removal, although in most cases, only a subtotal tumor removal is possible. Moreover, the frequent formation of the abovementioned neurological deficit in those patients and the tumor growth progression during a prolonged further examination makes the combination of therapy methods or initial radiation treatment more suitable for larger tumors [32]. Other scientists come to similar conclusions; they note that resection is the only radical method of treating these tumors. However, at the same time, subtotal excision can be used in cases where the tumor affects vital structures to prevent unnecessary complications [12].

In the research [33] authors also described the rich surgical practice of mainly endoscopic treatment glomus tympanicum. The research was concluded as a retrospective review of case series in two installations, in which, medical documentation of 30 patients, gathered over a span of 7 years, who received transcanal tumor of type A1, A2, and B1 (modified classification of Fisch-Mattox) removal, was included. During each surgery, tumor attributes, found during the operation, after-surgery complications, and further functional results were evaluated. Secondary cases or residual diseases were evaluated clinically and with an x-ray on a whole period of patient examination. None of the patients, who had the abovementioned surgery, had an intraoperative complication and required a transition to microscopic intrusion. In 90% of cases, total resection was a result of surgery, while near-total resection was recommended when the remaining pathology was near a carotid artery. The average hospitalization period was 1.6 days, and no after-surgery complications were reported. No secondary cases after the total removal of a tumor after an average monitoring period of 38 months were reported. Researchers have concluded, that middle ear paragangliomas, that did not reach the tympan-mastoid appendage, can be safely treated through transcanal endoscopic intrusion. The low rate of after-surgery complications, short hospitalization period, and high rate of total resection indicates, that transcanal endoscopic intrusion is a safe and effective treatment method.

A similar publication [34] showed the benefits and nuances of transcanal endoscopic intrusion in 12 cases of a single surgeon practice. Amongst the cases, 8 patients with glomus tympanicum ranged from Glasscock-Jackson to stage I-III. None of the patients had after-surgeon dizziness, tinnitus or perceptive hearing loss, facial nerve damage, or tympanic chord damage. Two patients had surgical eardrum perforation due to

the tumor growing on it. However, eardrum integrity was later restored with a transplant. The authors have concluded, that investigated method ensures perfect visualization of a middle ear and is an effective way of middle ear pathology resection. This approach requires less curettage and higher hypotympanum evaluation due to the inability to use both hands in surgery. Publications, that study various transanal endoscopic intrusion attributes, are still being published [35, 36, 37, 38]. Authors of a recent publication [39] note, that endoscopy may contribute valuable information to the usual microscopic approach, although a consensus on a unified best method is still not reached. These conclusions were made after an analysis of 342 surgical cases on a middle ear/petrous bone, performed during years 2005 to 2015. Microscopic or endoscopic methods were employed in all cases. Between them, 5 patients (7.5%) had glomus tympanicum tumors. Researchers [39] note, that doctor has to choose the best strategy for treatment and any complications or secondary diseases avoidance. The same conclusions on the total exsection through transcanal endoscopy have been achieved by another researcher. Scientists note that the procedure was conducted safely, quickly, and effectively thanks to the better visualization of the surgical area by the endoscope camera. In their publication, the problem of bleeding stopping is raised. Hemostasis is achieved through the modified aspiration catheter, which functioned as aspirational diathermy [40].

Blood loss stoppage was also discussed in other publications. As such, hemostatic control was achieved with the use of a gelatin sponge and quantum-molecular-resonance congelation in cases of glomus tympanicum [41]. As stated earlier, surgical removal of the tumor is the main treatment for paragangliomas of the middle ear. However, since the tumor has abundant blood flow, control of bleeding is critical. The patient may undergo preoperative embolization. A recent publication [41] reported the case of a 46-year-old woman who presented to a hospital with complaints of pulsatile tinnitus on the right side. A red pulsating neoplasm was found in the right cavity of the middle ear. A class B1 paraganglioma was diagnosed, which was then surgically removed. The authors of the above-mentioned publication [41] concluded that bleeding can be stopped only with the help of intratympanic manipulations, and they decided to perform tumor removal (transmeatal resection) without preoperative embolization of the arteries feeding pathological formations. After creating a tympano-meatal flap and performing an atticotomy, several pieces of Spongel® sponge were inserted between the neoplasm and the wall of the tympanic membrane. The latter absorbed blood and created a space between the tumor and the wall of the tympanic membrane, which allowed the tip of the Vesalius® tip to be inserted to coagulate the tumor. Thus, the operating team of doctors managed to successfully achieve hemostasis. The patient was discharged from the hospital on the sixth day after the operation. One year after the operation, pure tone audiometry did not change the level of bone conduction.

Suffice it to note, that special attention in the literature was brought to the glomus tympanicum with pulsating tinnitus. As such, the author of a recent study reviewed the clinical characteristics and treatment outcomes of patients with glomus tumors of the tympanic membrane with pulsatile tinnitus and concluded that this symptom can be successfully treated surgically (by removing the neoplasm), and preoperative embolization reduces intraoperative bleeding. The topic of embolization has also been raised in other publications since paraganglioma is an extremely vascularized tumor due to the active production of vascular growth factors [42]. Embolization after angiographic examination helps to identify the arteries feeding the tumor with their subsequent blockade, which helps to reduce intraoperative bleeding. The literature describes the case of a 40-year-old woman with unilateral deafness and tinnitus without concomitant diseases. During the otoscopy and otomicroscopy, a red neoplasm was found behind a healthy eardrum. MRT photos showed a neoplasm, located on a cochlea. A glomus tumor was diagnosed on a basis of clinical, audiological, and visual data. Preoperational embolization was conducted two days before the surgery. However, unlike in the above-mentioned cases, the full removal of a tumor was achieved through a microsurgical method. The authors of the study [30] note that the treating physician should consider the differential diagnosis of a glomus tumor to prevent any catastrophic complications such as intraoperative bleeding. Surgeons should utilize preoperational embolization more often. This bleeding prevention method has shown to be a promising one, as the abovementioned case describes. It is also noted that preoperative planning and tactics should also be carried out with due regard to all associated risks.

Surgical methods of treating this pathology, although effective, are not the only ones. Other ways of paraganglioma treatment are currently being researched. Thus, the aim of a recent systematic review and meta-analysis [43] was to determine the effectiveness of stereotactic radiosurgery (SRS) for the treatment of paragangliomas. Information from 37 articles was reviewed, which described more than 11000 cases, in which a glomus tympanicum was diagnosed amongst 8.2%. The average age of patients

was 56 years. The results of this study confirm that stereotaxic radiosurgery for paragangliomas is associated with good clinical and radiographic outcomes. Conservative treatment is especially important for elderly patients. The long natural course and complications associated with surgery have led to controversy in the treatment of paragangliomas in this group of patients. At the same time, it is important to note that more and more evidence is accumulating about the benefits of conservative treatment and long-term follow-up of treatment outcomes in this population [44]. As noted earlier, most of the analyzed tumors are treated with surgical resection, endovascular embolization, radiation, or a combination of these methods. Two main types of stereotactic radiosurgery or stereotactic radiation treatment are gamma-knife and cyber-knife. In comparison with the SRS procedure, conducted with a gamma-knife, the results of hypofractionation stereotactic radiation treatment, conducted through a cyber-knife are insufficiently studied [45, 46, 47]. Gamma-knife appeared to be a good alternative or an adjuvant microsurgical tumor resection for those patients, that are unsuitable for total surgical removal of a tumor. A great neoplasm growth control level and small secondary disease risk after prolonged examination were achieved [48].

In another research, a retrospective patient review of, those who were treated with a cyber-knife between 2010 and 2019, was conducted. Medical documentation regarding 34 paragangliomas in 29 patients was reviewed. The average age of patients was 50±16 years. Half of the patients were treated with surgical tumor removal. In four cases, tumors were functional and produced hormones. According to Fish classification, cases of B, C, and D types at a rate of 1 (13%), 12 (42%), and 14 (48%) were found. Amongst those 2 (7%) were not identified. The average dose of radiation, which was supposed to cover 95% of the target dose, was 25 Gy. The local control level was 97%. The average period, when the tumor did not progress, was 5 years (inter-quarter diapason was 28-95 months). An important achievement is that a tumor did not grow for 8 years in 96% of patients. After radiation treatment, in 1 case, a permanent facial nerve paralysis (stage II by House-Brackmann), while in another case – an asymptomatic cerebellum radiation necrosis. After reviewing the data, the authors noted that a lack of prior surgery was not a positive predictor of symptomatic improvement. The researchers concluded that the use of cyber-knife was an effective treatment with minimal side effects in the long term and could play a role as first-line therapy, especially for symptomatic non-functional tumors, to better control symptoms [45].

The features of recurrence of glomus tumors of the head and neck were studied, as well as factors associated with the progression of the disease after treatment. Amongst those, 173 adult patients with 189 paragangliomas (41.3% of carotid body, 29.1% of jugular glomus, 19.0% of eardrum glomus, and 10.6% of vagus glomus), who were treated in a period from 1990 to 2010. The frequency and risk of secondary disease were also evaluated. The average treatment period of patients was 8.6 years. Around 2.92 secondary cases per 100 people per year were noted. As such, a secondary case percentage was 8.2% after 4 years and 17.1% after 10 years. Authors have concluded that the second case appears more often during jugular glomus tumors and less often during carotid body tumors. Radiation therapy can achieve a lower risk of tumor recurrence or progression than surgery for some types of paragangliomas. Tumor growth is a rare occurrence, although it is more frequent after each recurrence. Control neck examination is recommended at least once in several years in the upcoming decades after therapy [49]. A recent systematic review [50] has shown, that the main frequency of pheochromocytoma/paraganglioma recurrence was 0.98 cases per 100 human years. However, other research results [51] show, that recurrence of analyzed tumors after removal was in 13.3% of patients, who were monitored five years after the surgery. Authors note that this data show, that recurrence risk after a full recession earlier was underestimated. Other numbers were described in earlier research [52]: the main frequency of recurrence was less than 5% of patients, who had glomus tympanicum. Since the results of the literature are based on retrospective studies, comparisons of different treatment options for glomus tumors and prognostic factors should be considered with caution.

With careful dissection and hemostasis, successful surgical resection of the tumors in question can be performed with minimal complications [12]. It is also important to adapt the treatment of ear glomus tumors depending on the patient's comorbidities. Doctors should also be open to other treatments, including the use of the gamma-knife and radiation therapy [53].

**CONCLUSION**

1. Ear and skull base paragangliomas are rare, typically benign, and highly vascularized neuroendocrine tumors. Surgical intervention remains the primary treatment method, though alternative therapies such as cyber-knife or gamma-knife radiosurgery are increasingly discussed, especially for patients who cannot undergo surgery due to advanced age, comorbidities, or aggressive tumor characteristics.
2. Despite advancements in surgical techniques to manage bleeding, excessive blood flow remains a

significant challenge during the operation. The use of endoscopic and transcanal approaches is effective for small glomus tumors confined to the middle ear and can reduce surgical risks.

3. The choice of treatment should be tailored to each patient, considering factors such as age, tumor size, anatomical issues, and overall health. A personalized approach is crucial for optimizing outcomes and addressing the specific needs of each patient.

4. Stereotactic radiosurgery is a safe and effective treatment option for glomus tumors, offering high levels of tumor control with minimal complications. This method is particularly valuable for patients who are not suitable candidates for surgical intervention.

5. The frequency of recurrence and progression of neck and skull base paragangliomas post-treatment remains unclear, with significant variability in reported data. Continued research is necessary to better understand these aspects and improve treatment strategies for these tumors.

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