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Hearing and balance in vestibular schwannoma – a 12-year clinical perspective

Слух и равнотежа код вестибуларног шванома – 12-годишња клиничка перспектива

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SUMMARY

Introduction/Objective Vestibular schwannoma is a benign tumor originating from Schwann cells, predominantly affecting the vestibular portion of the eighth cranial nerve. It is the most common tumor of the cerebellopontine angle, presenting with varying degrees of hearing loss, along with tinnitus and vestibular symptoms. This paper gives a comprehensive analysis of the degree of hearing and balance impairment in individuals presenting with vestibular schwannoma, along with a detailed clinical perspective.

Methods The research involved a retrospective analysis of the clinical database utilizing available medical records from the Audiology Department of a major tertiary care hospital in Serbia. It encompassed 83 patients diagnosed with vestibular schwannoma between 2011 and 2023, with presenting symptoms of hearing and/or balance impairment. The analysis included basic demographic data, presenting symptoms, and results of tonal liminal audiometry, alongside specific vestibular diagnostic tests and imaging.

Results A significant hearing loss was observed in the majority of patients with vestibular schwannoma ($p<0.01$), with 48.7% having a severe degree of sensorineural hearing loss. Increasing age was positively associated with higher degrees of hearing impairment, as expected ($p<0.05$). Dizziness was present in 15.6% of patients, while postural instability was reported by 29% of patients. The impairment of the vestibulo-ocular reflex of the lateral semicircular canal was observed in 62.7% of cases.

Conclusion The study provides insight into the hearing and balance of patients with vestibular schwannoma. The results highlight the need for continuous monitoring, both pre- and postoperatively, to understand the relationships between patients' characteristics, symptoms, degree of hearing impairment, and tumor localization. These data are crucial for improving diagnostic and multidisciplinary therapeutic approaches to enhance the quality of care and function in these patients.

Keywords: vestibular schwannoma; hearing loss; dizziness; cerebellopontine angle

САЖЕТАК

Увод /Циљ Вестибуларни шваном је бенигни тумор који потиче из Шванових ћелија и најчешће захвата вестибуларни део осмог можданог нерва. То је најчешћи тумор понтоцеребеларног угла, који се презентује са различитим степеном оштећења слуха, зујањем и вестибуларним симптомима. Ово истраживање анализира степен оштећења слуха и равнотеже код особа којима је постављена дијагноза вестибуларног шванома, заједно са детаљном клиничком перспективом.

Методе Истраживање је укључивало ретроспективну анализу базе података користећи доступну медицинску документацију из Одсека за аудиологију највеће болнице терцијарног нивоа неге у Србији. Обухватило је 83 пацијента са дијагнозом вестибуларног шванома постављеном између 2011. и 2023. године, који су имали симптоме оштећења слуха и/или равнотеже. Анализа је обухватила основне демографске податке, клиничку слику и резултате тоналне лиминалне аудиометрије, заједно са резултатима специфичних вестибулолошких дијагностичких тестова и радиолошком дијагностиком.

Резултати Значајно оштећење слуха примећено је код већине пацијената са вестибуларним шваномом ($p < 0.01$), при чему је 48.7% имало тежак степен сензоринеуралног оштећења слуха. Године старости су позитивно корелирале са тежим степеном оштећења слуха, као што се и очекивало ($p < 0.05$). Вртоглавица је била присутна код 15,6% пацијената, док је постуралну нестабилност пријавило 29% пацијената. Оштећење вестибуло-окуларног рефлекса латералног полукружног канала констатовано је у 62,7% случајева.

Закључак Наше истраживање пружа увид у клинички профил и презентацију пацијената са вестибуларним шваномом. Добијени резултати наглашавају потребу за континуираним праћењем и пре и постоперативно, како би се боље разумели односи између одређених карактеристика пацијената, презентујућих симптома, оштећења слуха и саме локализације тумора. Ови подаци су кључни за побољшање дијагностичких и мултидисциплинарних терапијских приступа, да бисмо унапредили квалитет неге и лечење ових пацијената.

Кључне речи: вестибуларни шваном; оштећење слуха; вртоглавица; понтоцеребеларни угао

INTRODUCTION

Vestibular schwannoma (VS), also known in the literature as acoustic neurinoma, is the third most common benign intracranial tumor after meningioma and pituitary adenoma, with the most common localization at the level of the cerebellopontine angle (CPA) [1, 2]. It originates from nerve sheath cells (Schwann cells) and usually involves the vestibular part of the vestibulocochlear nerve, although its most common clinical presentation is cochlear, i.e., ringing in the ear and varying degrees of hearing impairment [3, 4]. Although benign, a vestibular schwannoma, due to its localization, carries the danger of the compressive mass effect on other intracranial structures [3]. Clinically, most patients present with unilateral high-frequency sensorineural hearing loss (94%) and tinnitus (83%) [1, 3, 4]. Hearing loss in patients with vestibular schwannoma has traditionally been associated with direct pressure and stretching of the vestibulocochlear nerve during tumor growth, causing disruption of the vascular supply [5]. However, the onset and course of hearing loss often cannot be predicted; it occurs in very small tumors, and its progression can be observed even in patients with non-growing lesions [6]. The frequency of vestibular symptoms such as vertigo and unsteadiness of gait varies widely (from 17% to 75% of patients), and it is considered that symptoms are often underreported or unrecognized [7]. Furthermore, large tumors can cause trigeminal and facial neuropathies, such as paresthesias, and can exert pressure on the brain stem, leading to hydrocephalus and increased intracranial pressure [1, 3]. Most tumors are unilateral and sporadic, while bilateral localization is rare, present in less than 5% of cases, and characteristic of the hereditary disease, neurofibromatosis type 2 [1–4]. Symptoms usually appear between the fifth and sixth decades of life, with a tendency for symptoms to appear earlier in people with a hereditary form of the disease [1, 2, 3]. The reported incidence of vestibular schwannomas shows an increasing trend, but there is a general consensus that this increase may be attributed to improved detection of new cases and more frequent imaging [3].

The goal of this research is to perform a comprehensive analysis of the degree of impairment of hearing and balance in individuals presenting with vestibular schwannoma and to point out the importance of precise preoperative audiovestibular assessment both to improve diagnostic protocols and to better guide therapeutic approach (either "wait and scan", radiotherapy or microsurgery).

METHODS

The retrospective analysis of the available medical records of the Department of Audiology in the tertiary health care hospital was conducted. The study included 83 patients diagnosed with vestibular schwannoma between 2011 and 2023 who had symptoms and signs of hearing and/or balance impairment.

Basic demographic data, presenting symptoms, as well as the results of specific diagnostic tests such as pure-tone audiometry (measured by AC 40 clinical audiometer, Interacoustics, Denmark) were included. The average values of pure tone audibility thresholds for air and bone conduction were determined: air or bone Pure Tone Average (aPTA and bPTA), and the difference between the air and bone thresholds, i.e. Air-Bone Gap, ABG. PTA and ABG values are calculated as a mean value for frequencies important for speech understanding, according to the recommendations of the American Academy of Otorhinolaryngology and Head and Neck Surgery (at 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz, where ABG values >10 dB are defined as clinically significant due to normal test variability i.e. limitations of equipment and natural fluctuation of audiometric testing) [8]. Additionally, data on all tests performed during the neuro-otological examination, including the presence of spontaneous or gaze-evoked nystagmus, as well as provoked and induced nystagmus, were included (tested with Frenzel or videonistagmography goggles, Visual Eyes 515, Interacoustics, Denmark). The presence of

asymmetry in vestibular tonus was assessed using the head-shaking test, and the testing of the vestibulo-ocular reflex (VOR) of the lateral semicircular canal was performed both with low-frequency stimulus (bithermal Aqua Stim Caloric Irigator, Interacoustics, Denmark) and with high-frequency stimulus with clinical Head Impulse Test (HIT). Furthermore, the data obtained by imaging methods available in medical documentation were evaluated. Since the data in this study do not include patient-identifiable information, and no additional data collection was performed, no significant ethical concerns were identified (the study was formally approved by the Ethics Committee of the Faculty of Medicine, University of Belgrade). Descriptive and analytical statistics methods were used for analyses, while statistical hypotheses were tested at the $p < 0.05$ level of statistical significance.

RESULTS

Main demographic characteristics of the sample

The average age of the patients was 52 years (SD±15.6), with the youngest patient being 11, and the oldest patient being 83 years old. The majority of included patients were female, comprising 51 patients (61.4%). The median time from the onset of symptoms to the first medical examination was 12 months. In four patients, vestibular schwannoma was incidentally detected during a routine examination or imaging for other reasons. The most common initial complaint was impaired hearing (49.4%), followed by tinnitus (26.5%), while tingling in half of the face and vertigo were the least reported initial complaints, found in only two and 1 patient (2.4% and 1,2% respectively). For detailed patient characteristics, please refer to Table 1.

Hearing impairment

Age is positively associated with the severity of hearing impairment (right ear, $r=0.461$, $p<0.05$; left ear, $r=0.220$, $p<0.05$). As expected, there was a statistically significant relationship and high correlation between the severity of hearing impairment and the value of the air conduction thresholds at frequencies from 1000 Hz to 4000 Hz, with coefficients between 0.836 and 0.926 for the left ear and between 0.904 and 0.916 for the right ear ($p<0.001$).

We examined 83 patients; however, one patient had incomplete data on the tumor side, and another had bilateral tumors. Therefore, the correct number of patients evaluated is 82, with 83 ears included in the final analyses. When the tumor was on the right, pure-tone audiometry showed that the largest number of patients (48.7%) had a severe degree of hearing impairment, and complete deafness was found in 5.2% of patients. Mild sensorineural hearing impairment was present in 17.9% of patients, while the same percentage of patients had normal hearing thresholds. Hearing in the opposite ear was preserved. In cases with a tumor on the left, slightly different patterns of hearing impairment were observed, with severe impairment in 30.2% and complete deafness in 11.7% (see Table 2 and Figure 1 and 2 for further details on hearing impairment).

Vestibular impairment

Vertigo was present in 13 patients (15.6%), while 24 patients (29%) reported instability while walking. Only one patient had spontaneous nystagmus at the initial examination. Impairment of the angular VOR of the lateral semicircular canal, confirmed by a positive HIT, was recorded in 62.7% of patients, while positive post-Head Shaking nystagmus was present in 68.1% of the patients (as a sign of vestibular tonus imbalance). Based on the analysis of the results of the bithermal caloric test performed in 25 patients, hyporeflexia was observed on the side of the tumor in 13 patients, while areflexia was present in an additional 9 patients. Symmetric

(normal) findings were noted only in 3 patients. By analyzing the correlations between the results of different vestibular tests, a statistically significant positive correlation was observed between the positive findings in the Head Shake test and the Head Impulse test ($r=0.451$, $p<0.01$). However, a significant difference in the frequency of vertigo, instability while walking, results of the Head Shake test, HIT, or caloric test in relation to the degree of hearing impairment in both ears was not found ($p > 0.05$).

Tumor size and location

The average tumor size measured by magnetic resonance imaging (MRI) was 17.6 mm (SD ± 9.6 mm). There was a significant difference in the size of the tumor in relation to the localization, namely that tumors in the cerebellopontine angle (CPA) and at the CPA-IAM level (IAM = internal auditory meatus) are larger compared to tumors localized at the level of IAM only ($p < 0.001$) (Figure 1). In our sample, no statistically significant difference was found between the size of the tumor and the degree of hearing impairment ($p < 0.05$).

DISCUSSION

It is known that vestibular schwannoma is a slow-growing, benign tumor, usually unilaterally localized, with a tendency to involve the vestibulocochlear nerve and an overall incidence of 2.2 cases per 100,000 inhabitants per year (reaching 20.6 per 100,000 person-years for those over 70) [9, 10]. The main and most commonly reported symptom is asymmetric or unilateral hearing loss. It is also the most common initial manifestation of the disease and the main reason for diagnostic MRI in most cases [1, 3, 4, 9, 10, 11]. Similarly, impaired hearing was the most common initial complaint present in 49.4% of our patients. Wagner et al. [12] stressed that the age of patients significantly influences the degree of hearing loss before treatment. This is

confirmed in our sample, where older patients had a more severe degree of hearing impairment, while tumor size had no significant effect on hearing loss. The findings of asymmetric sensorineural hearing impairment in our study meant that the largest number of patients had moderately severe and severe hearing impairment on the affected side, with preserved hearing thresholds in the other ear. The hearing level in the contralateral ear is very important when considering serviceable hearing and in deciding on best treatment modality [1, 13]. This severity of hearing impairment can be explained by the relatively long period from the onset of symptoms to visiting a doctor, which is in accordance with the results of previously published research [11].

Although vestibular schwannoma more often involves the vestibular part of the vestibulocochlear nerve, vertigo is often not the primary symptom, and was present in only 15.7% patients included in this study, plus up to 30% of them reported gait instability. Since vestibular schwannomas grow very slowly over the years, progressive dysfunction of the vestibular nerve is inevitable and causes a gradual increase in the use of central adaptive mechanisms, known collectively as vestibular compensation [14]. Even if patients experience acute vertigo due to axonal stretching, vestibular nerve compression, or compromised blood supply to the labyrinth induced by tumor growth, compensation develops over time and symptoms diminish [7, 14, 15]. It is important to emphasize that vestibular symptom severity does not increase linearly with size but may be most pronounced in small–medium tumors (where partial nerve injury is occurring), and in very large tumors, often near-complete vestibular loss with central compensation is present, paradoxically reducing patient's symptoms. Pathological results of the caloric test were recorded in 22 out of 25 patients who underwent the test. Borgmann et al. pointed out that the caloric test should always be performed since it can be useful in predicting the origin of the vestibular schwannoma (superior vs. inferior vestibular nerve) and could serve as an indirect predictor of hearing preservation after surgery

(this was based on significantly smaller hearing loss in patients with tumors of the superior vestibular nerve) [16]. In addition to the caloric test, HIT is used to assess the high-frequency horizontal VOR and is currently the only bedside test that enables the identification of unilateral hypofunction of the peripheral vestibular system [15]. In our study, the results were positive in 62.7% of patients.

It remains unclear whether and how tumor size influences the degree of hearing loss in patients with vestibular schwannoma. Some studies support this idea, while others have shown that the tumor size does not correlate well with the level of hearing impairment [17, 18]. Our results indicate that there is no statistically significant relationship between hearing impairment and tumor size. Nonetheless, there are multiple published audiometric protocols for guidance on obtaining gadolinium-enhanced MRI in patients with asymmetrical hearing loss, and an interaural difference of at least 15 dB averaging in the 0,5 and 3 kHz frequency range was recommended as optimal by AAO-HNS (sensitivity 87.4%, specificity 65.4%) [19, 20].

Limitations of the study

In addition to being retrospective and susceptible to recall and information bias, this study has several other limitations that warrant mention. To accurately determine hearing functionality, it is essential to consider the results of the Speech Discrimination Test, which were not included in this assessment due to the unavailability of data [12, 13]. Consequently, we relied solely on pure tone audiometry data. Furthermore, as this study was conducted at a single tertiary center, the generalizability of the data may be limited. Future studies should focus on including the results of Speech Discrimination Tests and complete vestibular assessment to better elucidate any influence of tumor parameters on hearing and vestibular function.

CONCLUSION

This study provides insight into the initial clinical profile of patients with vestibular schwannoma. Given that hearing loss does not necessarily correlate with tumor size and vestibular symptoms may be mild or even paradoxically subclinical in advanced cases, it is crucial to screen for vestibular schwannoma every patient presenting with asymmetric sensorineural hearing loss (interaural difference of at least 15 dB averaging in the 0,5 and 3 kHz frequency range), unilateral tinnitus or vestibular findings. Our findings underscore the importance of detailed pre-treatment audiovestibular assessments and continued follow-up to improve diagnostics and develop personalized therapeutic approaches to preserve auditory and vestibular function, and enhance the quality of life of these patients.

Conflict of interest: None declared.

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Table 1. Detailed patient's demographic characteristics

Patients' characteristics (83 overall, unless otherwise specified)	**Results (%)**
Age	Mean ± SD: 52.34 ± 15.62
Sex	
Male	32 (38.6%)
Female	51 (61.4%)
Duration of symptoms	Median: 12 months
Presenting symptom	
Tinnitus	22 (26.5%)
Hearing loss	41 (49.4%)
Instability	4 (4.8%)
Vertigo	1 (1.2%)
Paresthesias	2 (2.4%)
Missing data	13 (15.6%)
Affected side	
Left	42 (50.6%)
Right	39 (47%)
Bilateral	1 (1.2%)
Missing data	1 (1.2%)
Tinnitus on the affected side	
Present	58 (70%)
None	25 (30%)
Vertigo	
Yes	13 (15.7%)
No	61 (73.5%)
Missing data	9 (10.8%)
Instability	
Yes	24 (28.9%)
No	47 (56.6%)
Missing data	12 (14.5%)
Average tumor size based on MRI findings	Mean ± SD: 17.6 ± 9.6 mm
Tumor localization	
IAM	22 (26.5%)
CPA	17 (20.5%)
IAM-CPA	21 (25.3%
Missing data	23 (27.7%)

IAM – internal auditory meatus; CPA – the cerebellopontine angle

Table 2. Pure Tone Average (PTA) in decibels for both ears and the degree of hearing impairment

Degree of hearing loss	**Side affected**			
	Right		Left	
	PTA in dB HL right ear	Percent of patients	PTA in dB HL left ear	Percent of patients
Normal hearing	16.04	17.9%	17.5	16.3%
Mild	33.75	17.9%	33.2	20.9%
Moderate	52.50	10.3%	49.4	20.9%
Severe	80.72	48.7%	76.06	30.2%

PTA on four frequencies important for speech comprehension (500, 1000, 2000, and 4000 Hz), dB; HL – decibel hearing level; total deafness was found in 5.2% of patients with right vestibular schwannoma, and 11.7% with vestibular schwannoma localized in the left ear

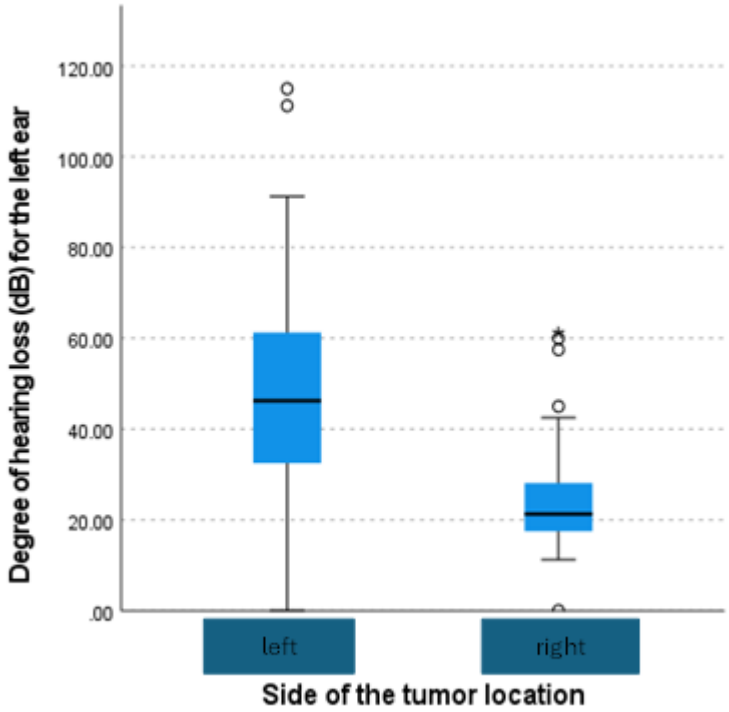

Figure 1. Average values of the degree of hearing loss in the left ear, expressed in decibel hearing level, in relation to the tumor localization

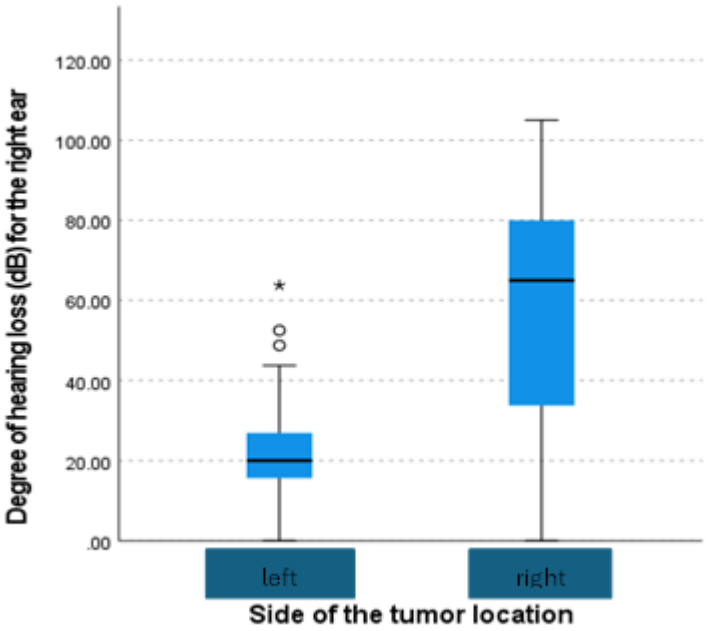

Figure 2. Average values of the degree of hearing loss in the right ear expressed in decibel hearing level in relation to the tumor localization

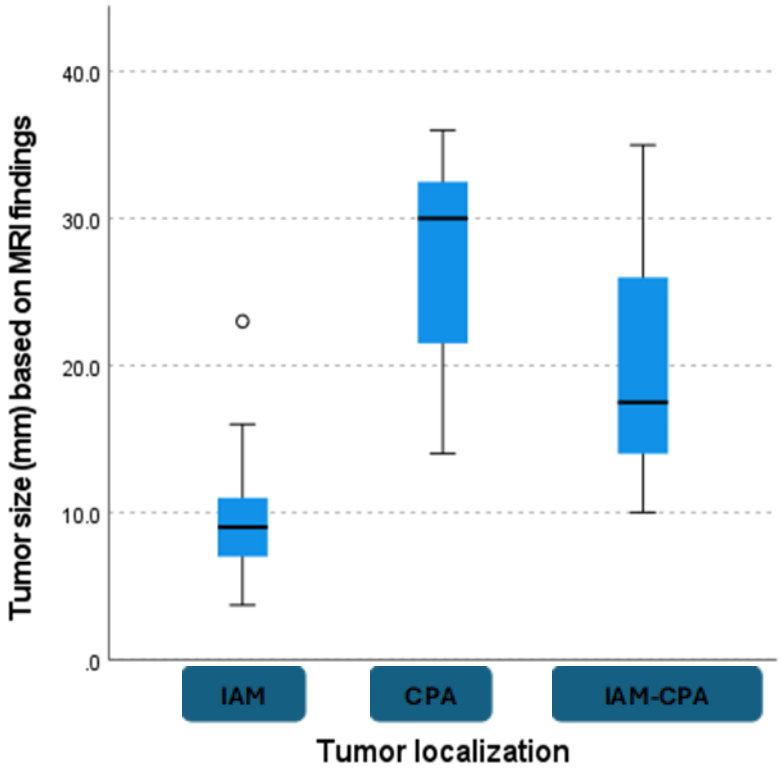

Figure 3. Average tumor dimensions (in mm) by tumor localization; IAM – internal acoustic meatus, CPA – cerebellopontine angle

