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ORIGINAL ARTICLE

Acoustic neuroma as first sign of inner ear functional disorders

Authors' Contribution:
A – Study Design
B – Data Collection
C – Statistical Analysis
D – Data Interpretation
E – Manuscript Preparation
F – Literature Search
G – Funds Collection

Marzena Bielińska[AC], Kalina Owczarek[B], Joanna Nowosielska-Grygiel[F], Jurek Olszewski[E], Piotr Pietkiewicz[D]

Department of Otolaryngology, Oncological Laryngology, Audioloogy, and Phoniatrics, Medical Univesity of Lodz, Poland, Head: Prof. Jurek Olszewski, MD PhD

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ABSTRACT:

Introduction. The aim of this work was to describe acoustic neuromas as the first signs of inner ear functional disorders.

Material and methods. The study covered 3,456 audiological and otoneurological patients, who were treated in the Department of Otolaryngology, Laryngological Oncology, Audiology and Phoniatrics Military Medical Academy University Teaching Hospital in Lodz within the period of 2011–2016. Among the studied subjects, an acoustic neuroma on the vestibulocochlear nerve was diagnosed in 13 cases (5,16%), including 9 women and 4 men. Each patient underwent a medical interview and an objective examination with static and dynamic tests, pure tone audiometry and speech audiometry tests, impedance audiometry tests, characteristics of tinnitus, videonystagmography and auditory brainstem evoked potentials (ABR) in crack. In each case of an incorrect ABR recording, contrast-enhanced MRI was performed.

Study results. The conducted tests showed an incorrect ABR recording in 252 patients (7,29%, including 54,37% women and 45,63% men).

In the performed contrast-enhanced MRI, 13 patients (5,16%), including 3,57% women and 1,59% men, had an image typical to neuroma of the vestibulocochlear nerve. Each one of the studied patients showed signs and symptoms typical to an acoustic neuroma such as hypoacusia and balance disorders, tinnitus in 12 subjects, headaches in the temporal and occipital area in 4 subjects, trigeminal neuralgia symptoms in 2 subjects and vision problems like scotoma and blurred vision reported by 1 patient.

Conclusion. Each case of sensorineural hearing loss, particularly unilateral sensorineural hearing loss, requires expanded hearing tests and auditory brainstem evoked potentials tests. An incorrect brainstem evoked potentials recording such as: an elongated latency of the 5th wave, an elongated interlatency of 1–2-3 and 1–2–3–4 waves, requires a contract-enhanced magnetic resonance imaging to diagnose cerebellopontine angle tumours.

KEYWORDS: neuroma, vestibulocochlear nerve, inner ear functional disorders

INTRODUCTION

The benign tumors of the vestibulo-cochlear nerve arising from the Schwann's sheath, and located in the cerebellopontine angle, are among the most common intracranial tumors. Eduard Snadifort was the first to describe them in 1777 [1]. In 1830, Charles Bell gave a description of the symptoms associated with them. In 1894, Sir Charles Ballance performed the first successful surgery [1].

Tumors of the cerebellopontine angle comprise approximately 6-10% of all central nervous system tumors. The most frequent type of tumor seen in this location is the neuroma of the vestibulo-cochlear nerve (vestibular schwannoma) accounting for 80-90% of cases. Meningiomas (3%), cholesteatomas (2-6%), and facial neuromas (1%) are less frequent [2,3].

Annually, in the USA there are 7-19 cases of primary brain tumors diagnosed per 100,000 people as well as 100,000 cases of metastatic brain tumors [4]. Among risk factors for neuromas there are tobacco smoking, epilepsy diagnosed at least 10 years earlier and giving birth. There is no significant relation between neuromas and allergy, antihistamine drug use, head trauma, acoustic trauma or other neoplastic disorders [5].

Tumors of the vestibulo-cochlear nerve involve primarily the vestibular part of it (usually the inferior vestibular nerve). The-

se tumors have their origin in the Obersterner-Redlich zone, a transitional zone between the glial tissue and the Schwann's sheath.

The following are the symptoms of cerebellopontine angle tumors: one-sided or asymmetric sensory hearing impairment, sudden hearing loss (ca. 25% of cases), tinnitus, gait instability, weakness of the trigeminal nerve, facial nerve dysfunction, and headaches.

For the diagnosis of cerebellopontine tumors, methods such as pure-tone audiometry, speech audiometry, brain auditory evoked potentials and electronystagmography/videonystagmography are used. With respect to neuroimaging studies, the gadolinium-enhanced magnetic resonance imaging of the posterior cranial fossa is the gold standard for the diagnosis of the cerebellopontine angle tumors [6-9].

The diagnostic schedule proposed by Welling et al. is routinely used in the Department of Otolaryngology, Laryngological Oncology, Audiology and Phoniatry, Medical University of Lodz [2].

The classification of Koss and Perneczky, that of Samii and Mathies as well as the classification put forward at the International Symposium of the Keio University are used for staging the tumors of the cerebellopontine angle [8].

The aim of the study was to demonstrate neuromas of the vestibulo-cochlear nerve as the first symptom of inner ear dysfunction in our patients.

MATERIALS AND METHODS

We analyzed data from 3,456 patients diagnosed in the Department of Otolaryngology, Laryngological Oncology, Audiology and Phoniatrics, Medical University of Lodz in years 2011-2016. Of them, 13 (5.16%) patients were diagnosed with vestibulo-cochlear neuromas (9 women – 3.57%, aged 67-31 years, mean age 48.5 years; 4 men – 1.59%, aged 60-24, mean age 43.5 years).

In every patient we took medical history, performed physical examination and carried out static-dynamic tests, pure-tone and speech audiometry, impedance audiometry, analysis of tinnitus, videonystagmography, and auditory brainstem responses (ABR) with a crack of 11 or 37 Hz frequency and 70 or 80 dB nHL intensity.

In all patients with abnormal ABR results (prolongation of the following intervals: I-III above 2.55 ms, III-V above 2.35 ms, I-V above 4.6 ms), a gadolinium-enhanced MRI scan was performed.

Tab. I. Staging of vestibule – cochlear neuromas according to Koos and Perneczky

STAGING OF VESTIBULE – COCHLEAR NEUROMAS ACCORDING TO KOOS AND PERNECZKY	N	%
T1 – tumor limited to the internal acoustic meatus	9	69,23
T2 – tumor bulges into the cerebellopontine angle, less than 2 cm in diameter	3	23,08
T3 – tumor is in contact with the brainstem but not causing displacement, less than 2-3 cm in diameter	1	7,69
T4 – tumor compresses and displaces the brainstem, larger than 3 cm in diameter	-	-
Total	13	100,00

Tab. II. Staging of vestibule – cochlear neuromas according to Samii and Mathies

STAGING OF VESTIBULE – COCHLEAR NEUROMAS ACCORDING TO SAMII AND MATHIES	N	%
Class T1 - intrameatal tumor	9	69,23
Class T2 - intrameatal and extrameatal tumor	-	-
Class T3a – tumor bulging into the cerebellopontine cistern	4	30,77
Class T3b – tumor in contact with the brainstem	-	-
Class T4a – tumor displacing the brainstem	-	-
Class T4b – tumor displacing the brainstem and compressing the fourth ventricle	-	-
Total	13	100,00

RESULTS

An abnormal ABR test result was present in 252 (7.29%) patients, of whom 137 (54.337%) were women and 115 (45.63%) were men.

Based on gadolinium-enhanced MRI scans, vestibulo-cochlear neuromas were found in 13 patients (5.16%), of whom 9 (3.57%) were women and 4 (1.59%) were men.

According to the classification of Koos and Perneczky (Tab. I), 9 (69.23%) patients had a T1 tumor – limited to the internal auditory meatus, 3 (23.08%) patients had a T2 tumor – bulging into the cerebellopontine cistern but smaller than 2 cm in diameter, one patient (7.69%) had a T3 tumor – the tumor reached the brainstem but did not cause its dislocation, it had 2-3 centimeters in diameter.

The classification according to Samii and Matheis was as follows (Tab. II): T1 – 9 (69.23%) patients – neuroma limited to the internal auditory meatus; T3a – 4 (30.77%) patients – neuroma reaching the cerebellopontine angle.

Five (38.46%) of patients experienced significant noise in the workplace. Head trauma was reported by only 3 (23.08%) pa-

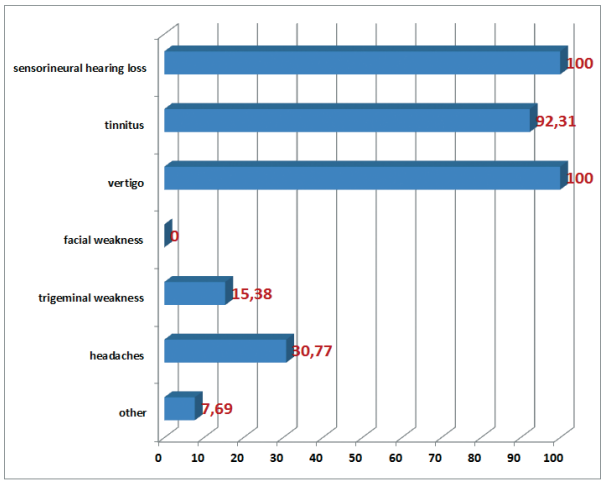

Fig. 1. Major symptoms reported by patients

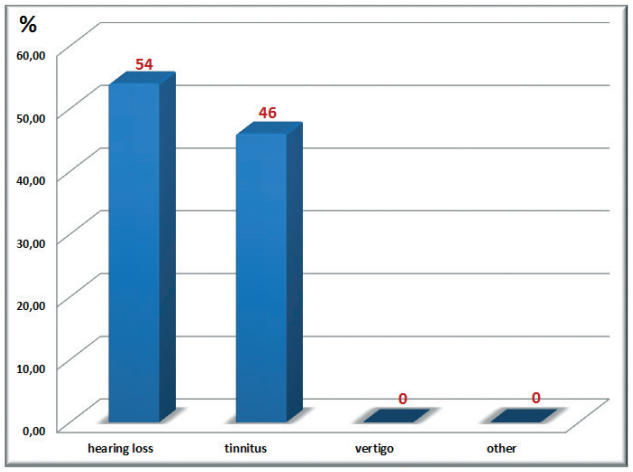

Fig. 2. Patients according to the initial symptom

tients. Among the patients there were 10 (76.92%) non-smokers.

As regards characteristic symptoms of vestibulo-cochlear nerve tumors (Fig. 1), all patients reported hearing impairment and gait instability. Tinnitus was present in 12 (92.31%) patients, which was described as unilateral (ipsilateral to the tumor) squeaky or ringing noise accompanied by a feeling of ear fullness.

Headaches in the temporal or occipital areas were present in 4 (30.77%) patients. Symptoms of trigeminal nerve weakness, i.e. paresthesia of the middle facial area, were reported by 2 (15.35) patients. One patient (7.69%) had visual symptoms – blurred vision with scotomas.

Most frequent initial symptoms were unilateral hearing impairment (7 patients – 54%) and tinnitus (6 patients – 46%), (Fig. 2).

Hearing loss is not defined unanimously. According to the guidelines of the National Health Fund, the WHO'97 norm is used most commonly in Poland. Hearing impairment is computed as the average of hearing thresholds at three of four of the following frequencies – 500 Hz, 1,000 Hz, 2,000 Hz, 4,000 Hz. This classification of hearing was used in our study in patients diagnosed with a neuroma (Fig. 3). Mild hearing loss was most common (5 patients, 38.46%), followed by moderate hearing loss (4 patients, 30.77%). Three patients (23.08%) had hearing loss in the range of 61 – 80 dB, and one patient (7.69%) had a complete hearing loss.

One of the most frequently reported symptoms was gait imbalance accompanied by paroxysmal and non-directional vertigo with no nausea or vomiting. Mixed vertigo with vestibular dysfunction unilateral to the tumor was found in the vast ma-

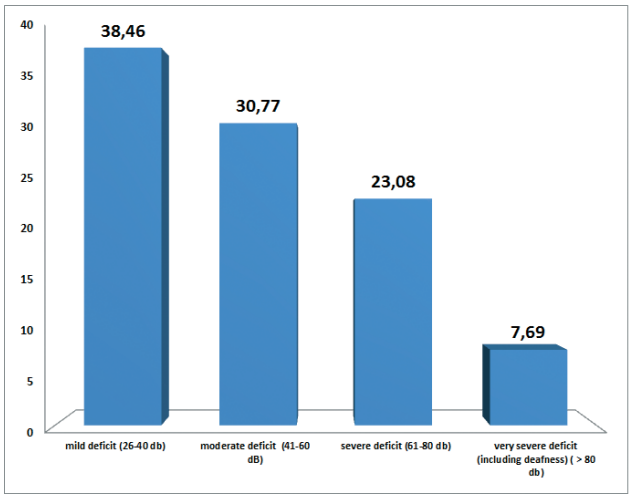

Fig. 3. Patients according to the WHO hearing impairment classification

jority of patients (11, 84.62%). Central vertigo was present in only 2 patients (15.38%).

In the videonystagmography, the Fitzgerald-Hallpike caloric test with the use of 30 degree Celsius warm water showed only a 2.4 o/s difference between vestibuli (the average reaction on the side of the tumor was 4 o/s, and on the opposite side 6.4 o/s). A significantly more pronounced difference was seen when 44 degree Celsius water was used. The maximal speed on nystagmus on the side of the neuroma was 3.8 o/s and 15.3 o/s on the opposite side. This gives a difference of 11.5 o/s (Tab. III)

The results of the videonystagmography for the evaluation of vertigo type in patients with vestibulo-cochlear neuromas are presented in Tab. IV. The average vestibular deficit on the side

Tab. III. Results of the Fitzgerald-Hallpike caloric stimulation test

PATIENT	EAR UNILATERAL TO THE TUMOR (O/S)	EAR CONTRALATERAL TO THE TUMOR - COLD - (O/S)	EAR UNILATERAL TO THE TUMOR - WARM (O/S)	EAR CONTRALATERAL TO THE TUMOR - WARM (O/S)
1	10,6	9,2	8,1	19,2
2	0,4	3,7	0,5	4,6
3	2,8	-1,7	-2,5	3,9
4	4,6	8,1	1,8	7,7
5	6,0	6,7	4,9	27,7
6	1,8	6,5	0,5	2,9
7	6,1	6,6	7,5	6,0
8	4,0	8,1	11,4	8,8
9	4,2	4,2	1,1	6,3
10	6,7	1,8	12,1	39,2
11	1,9	8,3	3,9	18,1
12	1,8	20,8	1,2	51,4
13	1,0	0,8	-0,5	2,8
Mean	4,0	6,4	3,8	15,3
Max.	10,6	20,8	12,1	51,4
Min.	0,4	-1,7	-2,5	2,8

Tab. IV. Vestibular function in the Fitzgerald-Hallpike caloric stimulation test

PATIENT	RESPONSIVENESS (O/S) (6 TO 80 O/S)	RELATIVE DIRECTIONAL PREPONDERANCE (%) (MAX.:11%)	ABSOLUTE DIRECTIONAL PREPONDERANCE (O/S) (MAX.:2O/S)	DEFICIT (%) (15%)
1	8,4	9,0	0,00	80,0
2	28,4	27,0	2,80	21,0
3	2,2	100,0	2,60	76,0
4	15,8	11,0	1,00	43,0
5	34,5	49,0	2,90	52,0
6	9,4	19,0	0,20	60,0
7	13,6	7,0	0,50	4,0
8	16,9	20,0	1,70	5,0
9	10,4	33,0	1,40	32,0
10	58,0	20,0	0,40	51,0
11	26,4	24,0	0,00	64,0
12	72,2	41,0	0,90	92,0
13	3,6	70,0	0,80	77,0
Mean	23,1	33,1	1,17	50,5
Max.	72,2	100,0	2,90	92,0
Min	2,2	7,0	0,00	4,0

of the tumor was 50.5% (maximal 92%). In two patients, the reaction to the caloric stimulation was symmetrical. The average relative directional preponderance was 33.1% (normal value below 11%) and the average absolute directional preponderance 1.17 o/s (normal value below 2 o/s). Therefore, in the vast majority of patients the vertigo was fully or partially compensated for, which can reflect a slow development of the pathological reaction.

DISCUSSION

An abnormal ABR test result was present in 252 (7.29%) patients, of whom 137 (54.337%) were women and 115 (45.63%) were men. In the performed contrast-enhanced MRI, 13 patients (5.16%), including 3.57% women and 1.59% men, had an image typical for neuroma of the vestibulo-cochlear nerve.

According to retrospective studies, the sensitivity of the ABR test is 92-98% in tumors larger than 2 cm and around 83% in tumors smaller than 1 cm. Therefore, this test is the "gold standard" for the diagnosis of vestibulo-cochlear neuromas [10].

According to most authors, the most frequent location of these tumors is the internal auditory meatus, the cerebellopontine angle, the jugular foramen, and only rarely the inner ear [10]. Neuromas are tumors of the central nervous system that originate from Schwann cells.

In our study, the following characteristic symptoms of vestibulo--cochlear nerve tumors were present: hearing loss and gait imbalance (all patients), tinnitus (12 patients), headaches in the temporal and occipital areas (4 patients), trigeminal nerve weakness (2 patients), and blurred vision with scotomas (1 patient).

The most frequent initial symptoms of the vestibulo-cochlear neuromas were one-sided hearing loss (7 patients, including one patient with complete hearing loss), and tinnitus (6 patients).

According to recent data, sudden hearing loss is present in as much as 25% of patients with a neuroma [10]. However, in our study we observed that finding in only 7.7% of patients.

One has to remember that in a small proportion of patients (3 – 12%) hearing remains normal. Some patients, even with large tumors (in the T3 stage) do not report problems with hearing [10].

About 90 percent present with a one-sided, slowly progressive hearing impairme

In our study, one of the most common symptoms in patients with vestibulo-cochelear neuromas was gait imbalance accompanied by mixed vertigo with unilateral vestibular deficit present in the majority of patients (11 patients). Central vertigo was present in only 2 patients, which is in line with earlier studies [10].

Physicians often try to estimate the risk of neuroma based on hearing loss. Approximately 2/3 of patients have hearing loss with respect to the high frequency range, whereas the remaining 1/3 have hearing loss within low frequencies. However, this should not be the basis for diagnosing vestibulo-cochlear neuromas. The only necessary study that should be performed in such cases is an objective hearing study, especially the brainstem auditory evoked potentials that indicate the location of the deficit and can necessitate a further work-up including magnetic resonance imaging in order to confirm the diagnosis [11-13].

According to the classification of Koos and Perneczky, the majority of tumors diagnosed in our patients were staged at T1 (9 patients) and T2 (3 patients) that were smaller than 2 cm in diameter. One patient had a T3 tumor that was 2-3 cm in diameter. This tumor reached the brainstem but did not cause its dislocation.

The results of retrospective studies comparing ABR with MRI are less encouraging. Ruckenstein et al. [14] estimated the sensitivity of ABR at 63% and specificity at 64%, which is in line with our study. Therefore, this method is used as a screening tool.

According to the NIH guidelines (Consensus Development Conference on Acoustic Neuroma, 1991), the recommended treatment for neuromas is microsurgery that should aim not only at resection of the lesions but also at preservation of neurological function [15]. In selected cases, radiation therapy or watchful waiting with follow-up imaging ("wait and scan policy") can be used.

The following factors should be taken into account when choosing an optimal method of treatment for a given patient – patient age, patient consent, tumor size, and previous treatment strategies.

Indications for the "wait and scan strategy" are old age, contraindications to general anesthesia, tumor recurrence after surgery, tumor in the only functioning ear, asymptomatic neuroma [16].

The aim of radiation therapy (gamma knife, linear accelerators) is the inhibition of tumor growth through the induction of cancer cell necrosis. Radiation therapy is indicated in older patients, patients in a bad general condition, tumor recurrence [17].

Surgical treatment should aim not only at a complete resection of the lesion but also at preservation of neurological function [18, 19].

As regards our patients, the largest neuroma was 24x28x22 mm in size (T3) and was compressing a cerebellar hemisphere. It was removed by microsurgery, whereas the remaining tumors (92.3%) were treated by radiation therapy.

CONCLUSION

- In our study, an abnormal ABR test result was present in 252, which constituted 7.29% of all patients who underwent audiological work-up because of sensorineural hearing loss,
- In the performed contrast-enhanced MRI, 13 patients (5.16%), including 9 (3.57%) women and 4 (1.59%) men, had an image typical for neuroma of the vestibulo-cochlear nerve.
- Every patient with sensorineural nearing loss should undergo a broad audiological work-up with the performance of brainstem auditory evoked potentials.

An abnormal result of brainstem auditory evoked potentials such as prolongation of the V wave in the left ear, prolongation of the I-III and I-V intervals necessitates contrast-enhanced magnetic resonance imaging, which is the gold standard for the diagnosis of vestibulo-cochlear neuromas.

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Corresponding author: Jurek Olszewski, Department of Otolaryngology, Oncological Laryngology, Audiology, and Phoniatrics, Medical University of Lodz, Poland, 90-549 Łódź, ul. Żeromskiego 113, Tel./fax 42/6393581; email: jurek.olszewski@umed.lodz.pl

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