Żebrowska Małgorzata, Nawrocki Adam, Kucharski Andrzej, Konopelko Michał, Trojanowski Piotr, Klatka Janusz. Diagnosis of vestibular schwannoma in modern clinical practice. Journal of Education, Health and Sport. 2019;9(9):797-803. eISNN 2391-8306. DOI http://dx.doi.org/10.5281/zenodo.3460354
http://ojs.ukw.edu.pl/index.php/johs/article/view/7518

The journal has had 5 points in Ministry of Science and Higher Education parametric evaluation. § 8. 2) and § 12. 1. 2) 22.02.2019.

The authors declare that there is no conflict of interests regarding the publication of this paper.

Received: 25.08.2019. Revised: 31.08.2019. Accepted: 22.09.2019.

Diagnosis of vestibular schwannoma in modern clinical practice

Małgorzata Żebrowska[1], Adam Nawrocki[1], Andrzej Kucharski[1], Michał Konopelko[1], Piotr Trojanowski[1], Janusz Klatka[1]

[1]Chair and Department of Otolaryngology and Laryngological Oncology, Medical University of Lublin

Abstract

The auditory nerve tumor is the most common tumor of the cerebellopontine angle region (CPA). The use of modern diagnostic methods allows to detect the tumor at an early stage of development, and consequently, the possibility of effective treatment. The clinical features and the most effective diagnostic procedures are presented.

Key words: vestibular schwannoma, acoustic neuroma, unilateral deafness, head and neck tumor diagnosis

Introduction

Vestibular schwannoma is a benign tumor originating from the Schwann cells of the Vestibular part of the VIII cranial nerve and is the most common cerebellopontine angle tumor (1 case per 100,000 population). Its early symptoms are caused by a progressive growth in the inner ear canal, most often causing sensorineural hearing loss (95% of patients) and tinnitus (70% of all cases). Sometimes it occurs as a form of sudden deafness (pressure of the tumor on the

vessels supplying blood to the inner ear) [1]. The less common symptoms are dizziness (18-58% of cases) and imbalance (16-70% of cases). Other sporadic symptoms (e.g. nerve V dysfunction, nerve VII paralysis) concern advanced and large tumors [2]. In the diagnostics of schwannoma, we mainly use auditory brainstem response (ABR) and MRI (the so-called gold standard in the diagnosis of small brain tumors). Once it has been diagnosed a small tumor may only be observed (MRI once in a year). An increased tumor growth of more than 2 mm per year or the appearance of symptoms other than hearing loss is an indication for the initiation of treatment - either surgical (transcochlear, middle cranial fossa or suboccipital approach) or radiotherapy (stereotactic radiotherapy).

Diagnostic scheme of vestibular schwannoma

The diagnostic scheme of vestibular schwannoma is based on the frequency of tumor occurrence for specific symptoms. Welling et al. (1990) divided the symptoms into three groups depending on the probability of neuroma detection. In the first group, where the risk of disease occurrence is lower than 5%, such symptoms as isolated dizziness, unilateral sensorineural hearing loss, tinnitus, symmetrical sensorineural hearing loss were distinguished. In the second group, where the risk is intermediate and ranges from 5 to 30%, symptoms such as sudden sensorineural hearing loss or permanent unilateral tinnitus of unexplained etiology were classified, while in the third group, where the risk is highest, there was a characteristic syndrome of schwannoma symptoms: unilateral, asymmetrical sensorineural hearing loss with tinnitus and speech understanding deterioration.

Fig. 1 Probability of vestibular schwannoma (n. VII) on the basis of clinical symptoms according to Welling et al.

Symptoms	**Propability of acoustic neuroma**	**Patient's risk category**
Classic unilateral asymmetric sensorineural hearing loss + tinnitus + deterioration of speech understanding	p > 30%	high
Sudden deafness or unexplained permanent one-sided tinnitus	5% < p < 30%	medium
Isolated dizziness, unilateral sensorineural hearing loss, tinnitus, symmetrical hearing loss	p < 5%	small

The diagnostic procedure differs depending on the group of symptoms. In the first group, auditory brainstem response (ABR) are the screening test, but if the result is normal, it should be repeated in six months, and if the result is abnormal, a contrast magnetic resonance imaging (MRI) test is recommended. In the second group, MRI with contrast is performed as a standard. In the third group, the examination of the first choice is MRI with contrast, and if the result is correct, follow up is based on ABR. [3,4]

Audiological diagnosis of acoustic neuroma

The audiological diagnosis of vestibular schwannoma (VS) is justified because of the symptoms of the disease, which mainly include hearing impairment. The most common symptom of the disease is unilateral or asymmetric sensorineural hearing loss, which occurs in approximately 92% of patients with diagnosed VS. Typically, the hearing loss is gradually progressive and rarely manifests itself as a sudden sensorineural hearing loss. The second most common symptom is tinnitus, which occurs in about 51% of patients. Other symptoms include dizziness and imbalance, headaches, both generalised and localized. Further and less frequent symptoms

resulting from tumor compression are trigeminal and facial nerve dysfunctions that may manifest themselves respectively in facial numbness in the trigeminal nerve innervation region and abnormal sensation of the auricle boat and parts of the external auditory canal, known as Hitselberger's symptom, characteristic for dysfunction of the sensory fibers of the intermediate nerve - parts of the facial nerve [4,5,6].

The gold standard in VS diagnostics is to perform magnetic resonance imaging with contrast. [7] The audiological examinations which should be performed during the evaluation of hearing loss are tonal audiometry (AT), speech audiometry (AS), auditory brainstem response (ABR). [4,7]

Tonal audiometry is mainly associated with sensorineural hearing loss at high frequencies - 66%, less frequently with sensorineural hearing loss at low frequencies - 10%, or flat audiogram - 10%. There is also a statement called the 3000 Hz rule that in the case of asymmetric sensorineural hearing loss, where the difference between one ear and the other at 3000Hz is at least 15dB, the risk of VS is increased and in such cases an MRI should be performed. [4,7,8]

Speech audiometry reveals impaired speech intelligibility compared to tonal audiometry which is a characteristic feature of extra cochlear sensorineural hearing loss. [4,7]

Before magnetic resonance became the gold standard for VS diagnostics, the most useful study was to perform auditory brainstem response. [9]

Standard ABR (ABR STD) is obtained by the administration of a crack-type stimulus, typically between 80 and 100 dB nHL, which is characterized by a short rise in acoustic wave pressure and a high degree of synchronization of nerve pulse discharges. This results in a high amplitude of ABR STD waves after averaging. It should be noted that the crack-type stimulus stimulates the vestibulocochlear nerve fibers in the range above 2kHz, which may result in an increase in false negative results at small tumors located differently tonotopically. In extra cochlear pathology, this test may reveal various differences depending on the size of the tumor. In the case of smaller tumors, the interval of I-III waves is prolonged and the interval of I-V is prolonged secondary to this phenomenon. With increasing tumor size, only wave I may persist and the waves may desynchronize completely. The sensitivity of the ABR STD method is over 95% for tumors over 1cm in size. This sensitivity for tumors smaller than 1cm drastically decreases and amounts to about 50-60%, which may reflect the reduction in the number of nerve fibres affected by the pathology. [10]

The stacked ABR test (ABR STOS) was designed because of the above limitations and low sensitivity of the ABR STD as an alternative screening tool for small tumors by Don et al. This test is based on the summation of narrowband responses from different regions of the cochlea.

It assumes that if tumor compression occurs on any vestibulocochlear nerve fibers, the amplitude of the combined wave will decrease. In order to obtain narrow-band responses for individual cochlea tonotopic representations, high-frequency masking crackles are applied in different ranges, which are subtracted from each other. According to Don et al., the sensitivity of ABR STOS in small VS is comparable to that of magnetic resonance. This method, despite its satisfactory sensitivity, has numerous disadvantages resulting from the time consuming nature of more averaging devices, higher intensity of stimuli administered and acting on the patient, and a large variety of measurements of V-wave amplitudes. [10,11]

An alternative method is ABR TON developed by Kochanek et al. It is based on ABR registration not only for crash, but also after giving short tones with long rise times at frequencies 1000, 2000 and 3000Hz. The short tone method gives low ABR wave amplitudes and allows easier detection of desynchronizing factors such as small vestibulocochlear nerve tumors and fiber demyelination. The study analyzes V-wave latency and the interaural difference between V-wave latency and IT5. An abnormal test result is a V-wave deviation at one or more frequencies and is an indication for further diagnosis. [10]

ABR STD is characterized by good specificity, but sensitivity is low in tumors of less than 1cm in size. High sensitivity is characterized by ABR STOS, while the specificity of the test is low, which gives a large number of false-positive results. The highest sensitivity and specificity is obtained in the ABR TON test. Taking into account the advantages and disadvantages of all tests, the best solution is a two-stage screening test consisting of ABR STD and ABR TON. [12]

Radiological diagnosis of acoustic neuroma

The most common pathological lesion of the cerebellopontine angle region, which represents 70-80% of tumors, is vestibular schwannoma. In differential diagnosis, cerebellopontine angle meningioma is also taken into account as the second most common tumor in this area (10-15% of cases, respectively). [13]

The diagnostic procedure for suspected CPA tumor is currently based mainly on magnetic resonance imaging (MRI), which, as the method of choice, shows almost 100% sensitivity and specificity in the diagnosis of vestibular schwannoma. It is the "gold standard" of diagnosis according to the National Institute of Health [15,7]. The importance of computed tomography (CT) was limited to pre-operative planning and postoperative period control due to the masking of small lesions, even after the administration of a contrast by a strong signal from the surrounding bone[14]. However, it shows indirect features of the tumor such as: dilatation of the internal auditory canal at the site of the expected anomaly or local bone destruction.

The reference standard is currently MRI of the posterior cranial fossa in the high frequency T1-sequence dependent sequence after intravenous administration of gadolinium contrast (Gd-DTPA). This examination is particularly good for small lesions limited to the internal auditory canal and enables differentiation with other proliferative lesions (e.g. meningiomas) or ischaemic lesions of the respective cerebellum part. In T1 dependent images, the visible lesion exhibit iso- or hypodensity and is clearly visible when contrast is administered. In T2 sequences the changes are iso- or hypotensive, but they are more intense than meningioma, so we can perform a preliminary differential diagnosis at this stage.

Summary

In the era of increasingly accessible and more accurate imaging diagnostics, the diagnosis of vestibular schwannoma is becoming progressively easier and the changes are detected in more advanced stages. It should be emphasized that the importance of screening tests, especially confirmatory audiological diagnostics, which nevertheless still constitutes an important component of both the diagnostic process and the control of the therapeutic process is gradually diminishing.

Among audiological tests, ABR STD is characterized by significant specificity and low sensitivity in tumors below 1 cm in size. ABR STOS is characterized by high sensitivity, while the specificity of the test is low, which gives a large number of false positive results. The highest sensitivity and specificity is obtained in the ABR TON test. Taking into account the advantages and disadvantages of all tests, the best solution is a two-stage screening test consisting of ABR STD and ABR TON. [12]

The golden standard of diagnostics at the moment is to perform a resonance imaging of the posterior fossa of the cranium, because this examination shows almost 100% sensitivity and specificity in detecting the discussed changes. The reference standard is currently MRI of the posterior cranial fossa in the high frequency T1-sequence dependent sequence after intravenous administration of gadolinium contrast (Gd-DTPA). Computed tomography, on the other hand, is only helpful in pre- and postoperative management.

References

1.Niemczyk K., Vaneecloo T.-M., Skarżyński H., Podgrodzki P., Bruzgielewicz A. - Obraz kliniczny guzów nerwu VIII w erze nowoczesnych badań diagnostycznych. Audiofonologia, Tom XIII, 1998, 243-251.

2.Jamróz B., Niemczyk K. - Nerwiaki nerwu słuchowego – diagnostyka i leczenie. Otolaryngologia, 2013, 12 (1), 8-18.

3. Niemczyk K. Guzy nerwu przedsionkowo-ślimakowego. (w) Otolaryngologia praktyczna. Janczewski G (red.). Warszawa, Viamedica 2005; 142-50.

4. Jamróz B, Niemczyk K. Nerwiaki nerwu słuchowego – diagnostyka i leczenie Otorynolaryngologia 2013, 12(1): 8-18

5. Foley R W, Shirazi S, Maweni R M, et al. (November 15, 2017) Signs and Symptoms of Acoustic Neuroma at Initial Presentation: An Exploratory Analysis. Cureus 9(11): e1846.

6. Hitselberger WE, House WF. Acoustic neuroma diagnosis:External auditory canal hypesthesia as an early sign. ArchOtolaryngol 1966;83:218–221

7. Jamróz B, Niemczyk K. Nerwiaki nerwu słuchowego: zalecane postępowanie diagnostyczne i terapeutyczne. Pol. Przegląd Otorynolaryngol 2012;1 (1): 12-20

8. Saliba I, Martineau G, Chagnon M. Asymmetric hearing loss: rule 3,000 for screening vestibular schwannoma. Otology & Neurotology. 30(4):515-521, JUN 2009

9. von Kirschbaum C, Gürkov R (2016) Audiovestibular function deficits in vestibular schwannoma. Biomed Res Int. 10.1155/2016/4980562

10. Kochanek K i wsp. Koncepcja metody słuchowych potencjałów wywołanych pnia mózgu z wykorzystaniem krótkich tonów dla potrzeb wykrywania zaburzeń pozaślimakowych. Otorynolaryngologia 2015, 14(3): 127-135

11. Don M, Masuda A, Nelson R, Brackmann D. Successful detection of small acoustic tumors using the stacked derived-band auditory brain stem response amplitude. Am J Otolaryngol 1997; 18(5): 608-21.

12. Kochanek KM, Sliwa L, Golebiowski M, Pilka A, Skarzynski H. Comparison of 3 ABR methods for diagnosis of retrocochlear hearing impairment. Med Sci Monit. 2015;21:3814–3824.

13. Xu XQ, Li Y, Hong XN, Wu FY, Shi HB, Radiological indeterminate vestibular schwannoma and meningioma in cerebellopontine angle area: differentiating using whole-tumor histogram analysis of apparent diffusion coefficient. International Journal of Neuroscience 2017 Feb; 127(2): 183-190

14. Strasilla C., Sychra V., Imaging-based diagnosis of vestibular schwannoma. HNO 2017 May; 65(5):373-380

15. Roswell E, Parry D. Summary: Vestibular Schwannoma (acustic neuroma) Consensus Development Conference. Neurosurg, 1992;30(6): 962–964