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VESTIBOLOGY

Sensitivity and specificity of vestibular bed-side examination in detecting VIII cranial nerve schwannoma with sensorineural sudden unilateral hearing loss as presenting symptom

Sensibilità e specificità della vestibular bed-side examination nell'individuare lo schwannoma dell'VIII nervo cranico con ipoacusia improvvisa come sintomo di esordio

L. CALIFANO, F. SALAFIA, M.G. MELILLO, S. MAZZONE
SSD di Audiologia e Foniatria, Azienda Ospedaliera "Gaetano Rummo", Benevento, Italy

SUMMARY

The objectives of this study were to identify signs of vestibular nerve suffering through a bedside vestibular examination protocol in case of sudden sensorineural unilateral hearing loss without spontaneous signs of vestibular impairment and to propose a bed-side vestibular examination based protocol for the focused execution of gadolinium-enhanced magnetic resonance imaging (MRI) only if a vestibular schwannoma is suspected. 96 patients, 52 men, 44 women, mean age 57.73 +/- 12.85 years, suffering from sudden sensorineural unilateral hearing loss, which presented neither vertigo nor spontaneous nystagmus, were enrolled. Pure tone audiometry, tympanometry, measurement of acoustic reflexes and Anderson test to detect adaptation, bedside vestibular examination through head shaking test, vibration test, head impulse test, hyperventilation test and detection of nystagmus in supine and lateral decubitus to search for signs of vestibular impairment were performed. Patients with signs of vestibular impairment and pure tone audiometry threshold at high frequencies better than 70 dB nHL were subjected to auditory brainstem responses. Gadolinium enhanced MRI centred on internal acoustic canals was carried out in all patients with sudden sensorineural unilateral hearing loss. Main outcome measures were signs of vestibular impairment at vestibular bedside examination and presence of vestibular schwannoma on MRI. Signs of vestibular impairment were detected in 22/96 cases (22.9%); a vestibular schwannoma was detected by MRI in 5/96 cases (5.2%), always when vestibular impairment was present. In case of sudden sensorineural unilateral hearing loss, vestibular bedside examination seems to be useful to restrict the suspicion of a vestibular schwannoma to cases with signs of vestibular impairment, reducing the number of MRI exams, with considerable economic savings.

KEY WORDS: Vestibular Schwannoma • Sudden Sensorineural Hearing Loss • Vestibular bed-side examination

RIASSUNTO

Gli obiettivi dello studio sono stati: identificare segni di sofferenza vestibolare attraverso un protocollo di "bed-side examination" in caso di ipoacusia improvvisa monolaterale senza segni clinici di sofferenza vestibolare; proporre i risultati della bed side examination vestibolare come criterio per l'esecuzione mirata della RMN per i canali acustici interni in caso di sospetto di neurinoma dell' VIII nervo cranico. Sono stati valutati 96 pazienti, 52 uomini e 44 donne, con ipoacusia improvvisa neurosensoriale monolaterale che non presentavano né vertigine né nistagmo spontaneo. Sono stati eseguiti: esame audiometrico tonale, esame impedenzometrico con test di Anderson per la ricerca di adattamento, Head Shaking Test, Test Vibratorio, Head Impulse Test, Test di iperventilazione, ricerca del nistagmo posizionale in posizione supina e nei decubiti laterali; l'ABR è stato eseguito nei pazienti con segni di sofferenza vestibolare se con soglia tonale ai toni acuti migliore di 70 dB nHL; tutti i pazienti con ipoacusia improvvisa hanno eseguito RMN con gadolinio per i canali acustici interni. Segni di sofferenza vestibolare sono stati identificati in 22/96 pazienti (22.9%) e la RMN ha evidenziato la presenza di schwannoma dell' VIII nervo cranico in 5/96 casi (5.2%), tutti con segni di sofferenza vestibolare evidenziati alla "vestibular bed-side examination". I nostri dati hanno evidenziato che gli schwannomi dell' VIII nervo cranico sono stati individuati solo nei casi di ipoacusia improvvisa monolaterale con segni di deficit vestibolare omolaterale. L'indicazione alla RMN con gadolinio può quindi essere limitata solo a questi casi, con evidente beneficio organizzativo ed economico.

PAROLE CHIAVE: *Schwannoma vestibolare • Ipoacusia improvvisa • Valutazione vestibolare "bed-side"*

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Introduction

Vestibular schwannoma (VS) represents about 80% of cerebellopontine angle tumours, 90% of intra-cranial schwannomas and 5-10% of all intra-cranial tumours. Its most common origin is from the superior branch of the vestibular nerve, but some studies on the temporal bone have identified its origin from the cochlear branch of the 8th cranial nerve in 24% of the cases [1].

The incidence of VS seems to be increasing, both because of improvements in diagnostic techniques and the presence of supposed favouring factors, such as electromagnetic pollution [2]; there is no sex predilection and the peak of incidence is between 50 and 60 years old; it usually occurs in isolated forms, as opposed to the multiple lesions often observed in neurofibromatosis type 2, which is caused by mutations in the NF2 tumour suppressor gene on chromosome 22.

The most common symptoms of VS are progressive unilateral sensorineural hearing loss, tinnitus and postural instability. Sudden sensorineural unilateral hearing loss (SSUHL) can be the presenting symptom in 2%-10% of the cases, whereas it occurs in 20-25% of cases during the natural history of the tumour [3-10]. Sudden hearing loss is defined as "a rapid onset, occurring over a 72-hour period, of a subjective sensation of hearing impairment in one or both ears. The most frequent used audiometric criterion is a decrease in hearing of ≥ 30 decibels (dB) affecting at least 3 consecutive frequencies" [11].

SSUHL seems to be more common in intracanalicular tumours [3 7] and in young patients [4]; its complete resolution is possible [12 13] so that even in the case of a recovered SSUHL, magnetic resonance imaging (MRI), which is the diagnostic gold standard for VS, is recommended.

It is reasonable to think that suffering of the superior vestibular nerve could cause clinical signs of vestibular impairment, recognisable through a vestibular bedside examination protocol.

The objectives were: 1) to identify signs of vestibular nerve suffering through a bedside vestibular examination protocol-head shaking test (HST), vibration test (VT), head impulse test (HIT), hyperventilation test (HVT) and detection of nystagmus in supine and lateral decubitus;

2) to propose a clinical protocol for the focused execution of gadolinium-enhanced MRI only if a VS is clinically suspected.

Materials and methods

From January 2013 to December 2015 we observed 96 cases (52 men, 44 women, mean age 57.73 ± 12.85 years) of apparently idiopathic SSUHL, that presented neither vertigo nor spontaneous nystagmus in the sitting position. In particular, we investigated in the absence of barotrauma, acute acoustic trauma or other trauma as causes of SSUHL. Seven individuals referred a previous diagnosis of benign paroxysmal positional vertigo. Pure tone audiometry, tympanometry, measurement of acoustic reflexes and Anderson test were performed in all patients. Diagnosis of SSUHL was based on the criterion exposed in the previously reported guidelines [11].

In SSHUL patients and in a homogeneous control group of 20 individuals (11 men, 9 women, mean age 56.75±14.53 years ($p = 0.76$)) with no medical history of audiological, vestibular or neurological diseases, we searched for signs of vestibular impairment under infra-red binocular videonystagmoscopy: head shaking induced nystagmus (HSIN), vibration induced nystagmus (VIN), hyperventilation induced nystagmus (HVIN), and positional nystagmus in both the supine position and lateral decubitus, and compensatory saccades evoked through HIT. Patients with one or more signs of vestibular impairment performed ABR, if the tonal threshold in the range 2000-4000 Hz was better than 70 dB nHL. Criteria of suspect for VS were considered: absence of waves in relation to the pure tone audiometry data; lengthening of I-III and I-V interpeaks and lengthening of wave V absolute latency. All patients affected by SSUHL underwent gadolinium-enhanced MRI centred on internal auditory canals.

Main outcome measures

We considered: the presence of signs of vestibular impairment and the presence of VS on MRI.

Statistical analysis

Fisher's exact test was performed to compare: a) diagnosis of VS by MRI with vestibular bedside examination results; b) data on vestibular impairment in the VS group: one sign vs. at least two signs of vestibular suffering and both groups with vestibular impairment vs. the group without vestibular impairment; c) vestibular bedside examination results of the SSUHL group vs. the control group. An unpaired t test was used to compare the mean

Table I. Results of vestibular bed-side examination in cases with at least two positive tests.

	HSIN	VIN	HVIN	HIT	Supine position nystagmus	Schwannoma (MRI)
Case 1	Paretic	Paretic	Absent	No saccades	Absent	No
Case 2	Paretic	Paretic	Paretic	No saccades	Paretic	12 mm
Case 3	Paretic	Excitatory	Paretic	No saccades	Absent	13 mm
Case 4	Absent	Paretic	Excitatory	No saccades	Absent	8 mm

HSIN: Head Shaking Induced Nystagmus; VIN: Vibration Induced Nystagmus; HVIN: Hyperventilation Induced Nystagmus; HIT: Head Impulse Test; paretic: nystagmus with fast phases directed toward the healthy side; excitatory: nystagmus with fast phases directed toward the affected side (hearing loss); MRI: Magnetic Nuclear Imaging

age of the SSUHL group with the control group. In both tests, a P value < 0.05 was considered significant.

Results

Audiometric tests. A flat configuration hearing loss was observed in 50/96 patients (pure tone average from 250 to 8000 Hz: 54.3 ± 10.24 dB; range: 45-94.5 dB); a sloping configuration in 28/96 patients (pure tone average from 250 to 8000 Hz: 38.5 ± 15.64 dB; range: 28.5-56.3 dB); a rising configuration in 10/96 patients (pure tone average from 250 to 8000 Hz: 33.5 ± 9.40 dB; range: 25.5-50.3 dB); a "U" shaped configuration- greatest hearing loss in the mid-frequency range- in 8/96 patients (pure tone average from 250 to 8000 Hz: 36.5 ± 12.65 dB; range: 30.5-55.3 dB). In all cases, the criterion for a decrease in hearing of ≥ 30 decibels (dB) affecting at least 3 consecutive frequencies was respected [11]. At least two signs of vestibular impairment were found in 4/96 patients (4.2%) (Table I). Only one sign of vestibular impairment was found in 18/96 patients (18.8%) (Table II). On the whole, vestibular impairment was observed in 22/96 patients (22.9%). In the control group, no signs of vestibular impairment were observed, with a significant difference vs. the SSUHL group (p = 0.02).

ABR was performed on 16/22 patients with vestibular impairment. In 2/5 of the cases in which MRI demonstrated a VS, we observed the lengthening of I-III and I-V interpeak latencies, which are the most specific signs of suspicion for VS; in three cases we observed the lengthening of Wave V absolute latency, which is a less specific sign of neural suffering [13]; in fact, it was the most frequent finding in the 11 cases that were MRI-negative for VS. In these cases, when evaluable, I-III and I-V inter-latencies were normal.

MRI revealed the presence of VS in 3/4 cases in the group with at least two signs of vestibular impairment (Table I) and in 2/18 cases in the group with only one sign of vestibular impairment. VS was never found in the 74 cases without signs of vestibular impairment. Incidence of VS in the population affected by SSUHL was 5.2% (5/96 patients). The differences were statistically significant: $p < 0.001$ for the group with at least two signs of vestibular impairment vs. the group without vestibular impairment; $p = 0.03$ for the group with one sign of vestibular impairment vs. the group with no sign of vestibular impairment; $p = 0.02$ for the group with at least two signs vs. the group with one sign of vestibular impairment. A flat configuration hearing loss (pure tone average 250-8000 Hz: 54 dB and 50 dB) was observed in two cases MRI positive for VS, in three cases a sloping configuration (pure tone average: 35.5 dB, 37.5 dB, 40.5 dB). No significant difference was noted for the pure tone average vs. the cases MRI negative for VS. The Anderson test was positive in 4/5 cases MRI positive for VS, but it was also positive in six cases that were MRI negative for VS (sensitivity = 80%; specificity = 64.7%). In relation to the detection of VS, vestibular bedside examination presented a sensitivity of 60%, a specificity of 98.8% and a positive predictive value of 75% in the group with at least 2 signs of vestibular impairment (4/96 cases);

Table II. Results of vestibular bed-side examination in cases with one positive test.

	HSIN	VIN	HVIN	HTT	Supine position nystagmus	Schwannoma (MRI)
Case 1	Absent	Absent	Excitatory	No saccades	Absent	6 mm
Case 2	Paretic	Absent	Absent	No saccades	Absent	No
Case 3	Absent	Paretic	Absent	No saccades	Absent	12 mm
Case 4	Absent	Paretic	Absent	No saccades	Absent	No
Case 5	Paretic	Absent	Absent	No saccades	Absent	No
Case 6	Absent	Paretic	Absent	No saccades	Absent	No
Case 7	Absent	Paretic	Absent	No saccades	Absent	No
Case 8	Absent	Absent	Absent	No saccades	Paretic	No
Case 9	Paretic	Absent	Absent	No saccades	Absent	No
Case 10	Paretic	Absent	Absent	No saccades	Absent	No
Case 11	Absent	Absent	Absent	No saccades	Paretic	No
Case 12	Absent	Paretic	Absent	No saccades	Absent	No
Case 13	Paretic	Absent	Absent	No saccades	Absent	No
Case 14	Absent	Paretic	Absent	No saccades	Absent	No
Case 15	Absent	Paretic	Absent	No saccades	Absent	No
Case 16	Absent	Paretic	Absent	No saccades	Absent	No
Case 17	Absent	Paretic	Absent	No saccades	Absent	No
Case 18	Paretic	Absent	Absent	No saccades	Absent	No

HSIN: Head Shaking Induced Nystagmus; VIN: Vibration Induced Nystagmus; HVIN: Hyperventilation Induced Nystagmus; HIT: Head Thrust Test; paretic: nystagmus with fast phases directed toward the healthy side; excitatory: nystagmus with fast phases directed toward the affected side (hearing loss); MRI: Magnetic Resonance Imaging

a sensitivity of 100%, a specificity of 81.3% and a positive predictive value of 22.7% % in the group with at least one sign of vestibular impairment (22/96 cases).

Discussion

Diagnosis of VS is sometimes "elusive" because its symptoms can be slowly progressive, especially unilateral hearing loss and postural instability. VS can be suspected through standard audiological and vestibular functionality examinations, but the diagnostic gold standard is gadolinium-enhanced MRI which must be performed in all cases with both clinical and functional suspicion.

In a low percentage of cases, SSUHL was identified as a possible presenting symptom of a VS [8 10]; however, since SSUHL can occur in a higher percentage of VS cases-up to 20% during the natural history of the tumour, an MRI of the internal auditory canals is recommended in cases of apparently idiopathic SSUHL [11]. Notwithstanding, since SSUHL is a revealing symptom of VS in only a few cases, most MRIs do not reveal the suspected disease, with an obvious waste of economic resources.

For this reason, it would be useful to develop a diagnostic protocol able to identify a suspect VS, with high sensitivity and specificity, especially when SSUHL is the presenting symptom.

Since the preferential origin of the VS is from the superior branch of the vestibular nerve, it makes sense to seek dynamic, static and metabolic signs of vestibular imbalance through a vestibular bedside examination that can be performed in less than 10 minutes. Teggi et al.[15] showed that intracanalicular length and intracanalicular diameter seem to be the main parameters that correlate with vestibular function and that also in case of small intracanalar VS a vestibular impairment is possible; Niu [16] observed caloric hyporeflexia in 50% patients with SSUHL without vertigo. Our data suggested vestibular impairment in 22/96 patients (22.9%). The vestibular bedside examination tests showed results in line with our previous studies [17]; VT presented the most frequent number of positive cases associated with VS (4/5 cases), even if the significance of the data is limited by the small number of VS cases it was related to. HIT did not show any positivity. This is not unexpected, because the occurrence of compensatory saccades to HIT is related to the presence of a serious vestibular hyporeflexia [4], which is not necessarily present in the small tumours that are the object of the present study (6-13 mm), as highlighted by MRI. Furthermore, HIT was performed in its clinical form; video-HIT could possibly improve its diagnostic sensitivity. A possible bias is that the positivity of vestibular tests could be related to a previous suffering of the system. The bias can be minimised by careful medical history. However, such a situation would lower the specificity and not the sensitivity of the vestibular bedside examination. In other words, we would have detected all the VS, carrying out most MRI.

VS, identified through gadolinium-based MRI of internal auditory canals, presented an incidence of 5.2%-5/96 cases, all in the group of 22 patients with one or more signs of vestibular impairment, whereas VS was not found in patients with no sign of vestibular impairment ($p < 0.0001$). The above percentage is in agreement with previous studies that identified SSUHL as a rare VS revealing symptom [4 7 8 10 13]. Nevertheless, we must again consider that our results are limited by the low number of VS identified, even if their number is consistent with the predictable one.

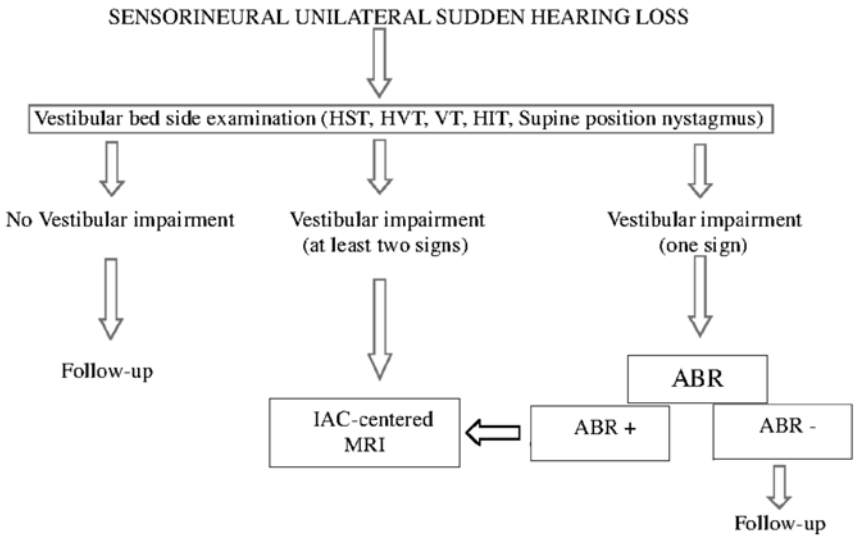

Fig. 1 Decision making flow-chart for sensorineural unilateral sudden hearing loss.

Conclusions

In case of unilateral sensorineural hearing loss it is mandatory to exclude VS as its cause. SSUHL is reported as the presenting symptom of VS in > 10% of the cases, but MRI is considered to be the gold standard test to exclude VS in case of SSUHL, too.

We propose to immediately perform MRI to detect a possible VS in case of SSUHL as presenting symptom only when a vestibular impairment is present; a cut-off value at two signs makes the protocol very specific (98.8%), but less sensitive (60%); nevertheless, a cut-off value at one sign decreases its specificity to 81.3%, whereas its sensitivity is increased to 100%. If a cut-off value at two signs is chosen, in cases with only one sign of vestibular impairment, to increase the sensitivity of our analysis, we recommend performing ABR before an MRI is carried out. In any case, even the decision to carry out MRI on all patients with at least one sign of vestibular asymmetry, would decrease significantly its number: according to our results, MRI would have been carried out only in 22/96 cases, with considerable economic savings. Audiometric and clinical follow-up will suggest MRI if new data eventually augment the suspect of the presence of VS.

In our opinion, for its simplicity of execution, sensitivity and specificity, vestibular bedside examination is indicated in all cases of apparently idiopathic SSUHL. A possible decisional flow-chart is represented in Figure 1.

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Address for correspondence: Luigi Califano, SSD di Audiologia e Foniatria, Azienda Ospedaliera "Gaetano Rummo" Benevento, via dell'Angelo 1, 82100 Benevento, Italy. Tel. +39 0824 57407. Fax +39 0824 57430. E-mail: luigi.califano@tin.it, vertigobn@hotmail.com