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OTOLOGY

MRI of endolymphatic hydrops in patients with vestibular schwannomas: a case-controlled study using non-enhanced T2-weighted images at 3 Teslas

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Abstract

Objective Vestibular schwannomas (VS) may present with similar symptoms endolymphatic hydrops. Association between hydrops and internal auditory canal VS has been described by Naganawa et al. (Neuroradiology 53:1009–1015, 2011), but has never been confirmed since. The aim of this work was to study the prevalence of a saccular dilation on a T2-weighted sequence at 3 T MRI in VS compared to a control group.

Materials and methods All patients presenting with typical VS between May 2009 and July 2018 were included ($n=183$) and compared to a control group ($n=53$). All underwent a high-resolution T2-weighted 3D sequence (FIESTA-C). The height and width of the saccule were measured on a coronal plane by two radiologists.

Results The saccule was dilated on the side of the schwannoma in 28% of the cases ($p=2.81\times10^{-5}$), with 15.7% of bilateral dilation. Saccular dilation was correlated to sensorineural hearing loss (OR 3.26, $p=0.02$). There was also a significant correlation between saccular hydrops on the normal contralateral side of patients with VS and vertigo ($p=0.049$), and between saccular hydrops on the side of the tumour and tinnitus ($p=0.006$).

Conclusion A third (29%) of VS are associated with a saccular dilation on the side of the tumour, which is an MR sign of endolymphatic hydrops (bilateral in 15.7% of the cases) and it appears related to sensorineural hearing loss and tinnitus, as well as vertigo if a contralateral dilation is present. This opens new therapeutic potentialities with the use of anti-vertiginous drugs, which could have a beneficial effect on the clinical symptoms.

Keywords Magnetic resonance imaging (MRI) · Endolymphatic hydrops · Sensorineural hearing loss · Vertigo · Neuroma · Acoustic

Objective

Vestibular schwannomas (VS) are the most frequent tumours of the cerebellopontine angle (80%), arising from branches of the vestibulocochlear nerve [1–3]. They present with varied clinical symptoms, the classic triad being deafness, disequilibrium and tinnitus. These symptoms overlap those of other pathologies, such as Meniere's disease. In 2011, Naganawa et al. observed an endolymphatic hydrops in 30% of their patients with an internal auditory canal vestibular schwannoma on a non–injected 3D FLAIR sequence, but their results have never been confirmed since [4]. In Meniere's disease, two imaging techniques have been used to image the membranous labyrinth and the saccule, either a delayed-FLAIR sequence acquired 4h30 after a gadolinium injection or a non-contrast coronal heavily

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T2-weighted sequence at 3 T [5–14]. Such sequences are efficient to analyse inner ear fluids intensities and variations according to their biochemical composition, while offering enough spatial resolution to measure small endolymphatic structures, such as the saccule [4, 13–15].

The aim of our work is to study the prevalence of saccular dilation, as a sign of endolymphatic hydrops, in patients with a VS, by means of saccular measurements (height and width), using a 3D non-contrast heavily T2-weighted sequence at 3 T, compared to a control group. We also wanted to study if some of their clinical symptoms could be related to the presence of a hydrops.

Materials and methods

Study population

264 patients, who underwent an MRI imaging of the inner ear and internal auditory canal, were screened for our study from May 2009 to July 2018. The study has been approved by the Ethics committee of our institution (authorization number RNI 2018—HUS n°FC/dossier 2018-29).

In this case–control, non-interventional, monocentric study, we retrospectively included all patients presenting with a typical VS of the internal auditory canal, without intralabyrinthine extension, together with a heavily T2-weighted (3D FIESTA-C) sequence on their MR examination. The exclusion criteria were: atypical diagnosis, labyrinthine extension of the tumour, major motion artefacts on the T2-weighted sequence, concomitant middle or inner ear inflammatory pathologies and previously treated tumours without available preoperative imaging. To limit selection biases, we only included the first available MRI of each patient, with matching functional tests (performed by an otorhinolaryngologist). 183 patients met the inclusion and exclusion criteria (89 men and 94 women). They were then divided into 3 groups according to the degree of obstruction of the internal auditory canal by the tumour:

- Group 1 ($n=28$): "non-obstructive VS" did not have any direct contact with the internal auditory canal walls and a margin of cerebrospinal fluid > 1 mm remained between the walls and the tumour on all planes
- Group 2 ($n=28$): "mildly obstructive VS" presented with margin of cerebrospinal fluid < 1 mm between the tumour and the walls of the internal auditory canal
- Group 3 ($n=127$): "obstructive VS" were in close contact with the internal auditory canal wall, with no CSF margins between the tumour and the walls of the internal auditory canal

All patients were compared to a control group of healthy volunteers without any otological diseases, Group 0 ($n=53$).

The primary endpoint was to compare the size (in millimetres) of the saccule (height and width) on a coronal high-resolution heavily T2-weighted sequence (3D FIESTA-C) on the side of the VS and the tumour-free side (healthy contralateral side). It was compared to the size of the saccule in a control group of healthy volunteers. The aim was to highlight a possible statistical correlation between saccular hydrops and VS. The saccule was considered enlarged (dilated) if its height was > 1.6 mm and/or its width was > 1.4 mm [13].

The secondary endpoints were:

1. Bilaterality of the signs of hydrops (presence of a dilation on the tumour-free side).
2. Correlation between the presence of saccular hydrops and the tumour volume and/or degree of obstruction (between groups 1, 2 and 3).
3. Statistical link between saccular hydrops and clinical symptoms and/or auditory–vestibular abnormalities.

Imaging protocol

All patients and control group underwent a gradient-echo heavily T2-weighted 3D FIESTA-C sequence on a 3T MRI (Signa HDxt, General Electrics, Strasbourg, France) without gadolinium injection. Its parameters were as follows: field of view = 19.8 × 22 cm, flip angle = 60°, acquisition time 7 min 49 s, repetition time = 7 ms, echo time = 2.8–1.2 ms, bandwidth = 83.3 kHz, phase × frequency = 484 × 484, number of excitations = 1, thickness of the slices = 0.3 × 0.3 × 0.3 mm. The patients were asked not to swallow in order to avoid motions artefacts.

The diagnosis of typical vestibular schwannomas was made using a contrast-enhanced 2D spin-echo T1-weighted sequence 0.2 ml/kg − 0.1 mmol/kg of gadoterate meglumine (Dotarem®, Guerbet, Roissy, France). Its parameters are as follows: echo time = 14 ms, repetition time = 580 ms acquisition time 6 min 15 s, field of view = 20 × 20 cm, phase × frequency = 320 × 320, flip angle = 90°, bandwidth = 1.23 kHz, number of excitations = 2, slice thickness = 1 mm.

Clinical and imaging data

The gathered clinical data were: age, sex, presence/absence of hearing loss, type of hearing loss (sensorineural, mixed, conductive/acute or progressive), tonal audiograms, presence/absence of vertigo or tinnitus, caloric tests and V-HIT (video-head impulse tests) results.

Clinical data were available for 171 patients, representing an overall response rate of 93%. When available, the results of audiometric and vestibular tests were also

collected. Tonal audiometry data were available in 93% of cases ($n = 171$). Caloric test data were available for 96 patients (52%). V-HIT data were available for nine patients only.

The specific relevant characteristics of VS (in addition to the classification into three groups: G1, G2 and G3, according the degree of obstruction of the internal auditory canal) were gathered. This included the lateralization of the tumour, the volume and Koos' classification of VS.

Image analysis

All images were acquired directly in the plane of the lateral semicircular canal (axial) by placing the study box parallel to the roof of the orbit. Coronal and sagittal reconstructions were gathered from the 3D data set. All images were analysed using the Osirix® software (http://www.osirix-viewer.com). Two radiologists (a junior—4 years of experience—and a senior radiologist, with 35 years of experience in head and neck radiology) read the T2-weighted images.

Measurements were made on both sides in all patients and control group. The radiologists, blinded to the clinical presentation, were presented with a coronal section at the level of the superior and lateral ampullas (through the anterior portion of the vestibule), which was used to analyse and measure the saccule.

- The visibility of the saccule was qualitatively assessed using a scale of 0–2: well defined (0), visible but with some artefacts (1) or too many artefacts for correct measurements (2)
- The quantitative analysis of the saccule was based on measurements in two different planes [13, 14, 16], as illustrated in Fig. 1.

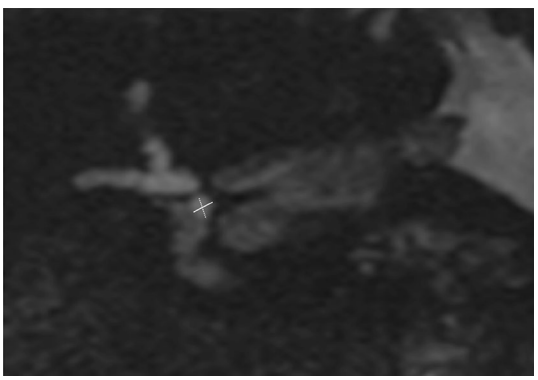

Fig. 1 Measurements of the height (dotted line) and the width (continuous line) of the saccule on the coronal plane of reference, at the level of the anterior and lateral ampullas

  - The length of the saccule was measured along its axis, from the inferior portion of the utricular macula down to the lowest portion of the saccule.
  - The width of the saccule was measured from the middle of the lateral wall of the saccule towards the medial wall of the adjacent vestibule.

Statistical analysis

Dispersion parameters, such as mean measures, standard deviation and minimum/maximum values, given as [mean ± standard deviation (range)], were calculated for all quantitative variables. Qualitative variables were expressed in terms of numbers and percentages. To search for a difference in height or width measurement, a mixed linear regression model was used taking into account the group variables (case/control), the side of the tumour and the sex and with a random effect for the subject variable. A McNemar test was used to compare the proportion of dilated saccules between the normal and pathological sides in patients with VS (G1 + G2 + G3). A Mann–Whitney test was used to investigate a possible correlation between the tumour volume and the presence of a saccular dilation. Finally, odds ratios were calculated for all clinical parameters as a function of the presence of a saccular dilation on the pathological or on the tumour-free side in all patients with a VS. The interobserver correlation was analysed by means of a Cohen's Kappa (K) for the visual analysis and intra-class correlation by means of a intraclass correlation coefficient (ICC) for the quantitative analysis. A Fisher test was used to calculate p values at $p < 0.05$ significance. All analyses were carried out with the R software (version 3.4.2) by a senior statistician.

Results

Patients characteristics

183 patients, who met the inclusion and exclusion criteria, were included and compared to a control group of 53 healthy volunteers (see flowchart). The clinical data of all patients and control group are reported in Table 1. The specific relevant characteristics of VS are reported in Table 2.

Qualitative and quantitative analysis

The saccule was visible (100%) on the coronal reference image in each of the 53 healthy volunteers. It had an ovoid shape, occupying the anterior and median part of the vestibule, with a high signal on the T2-weighted sequence, as featured in Fig. 2a. The saccular macula was not visible on MR images because of the hypointense signal of the bony wall of the adjacent vestibule on T2-weighted images.

Table 1 Relevant characteristics of patients with VS compared to the control group

	Schwannomas (G1, G2, G3) $n = 183$	Control group (GO) $n = 53$
Mean age (years)	59.7 (24-92, median 60)	33.4 (23–57, median 30)
Sex		
Men	89	23
Women	94	30
Cochlear symptoms ($n = 168$)		
Normal	6 (3.6%)	53
Sensorineural hearing loss	146 (86.9%)	0
Conductive hearing loss	0	0
Mixed hearing loss	16 (9.5%)	0
Progressive hearing loss	134	0
Sudden hearing loss	28	0
Complete loss of hearing	14 (8.3%)	0
Tinnitus	59 (53%)	0
Vestibular symptoms ($n = 168$)		
Vertigo	61 (36.3%)	0
Balance disorders	54 (32.1%)	0
Availability of functional explorations		
Tonal audiometry	$n = 164$	0
Caloric test	$n = 96$	0
V-HIT	$n = 9$	0

Interobserver agreement for the visualization of the saccule within the control group was excellent ($\kappa = 1$), regardless of the side studied.

The mean height of the normal saccule was 1.41 mm ± 0.14 (1.1–1.6) and the mean normal width was 1.2 mm ± 0.13 (0.9–1.4). No saccule was > 1.6 mm long and/or > 1.4 mm wide (thresholds used to consider a saccule as "pathological" and therefore dilated). Interobserver correlation showed a strong correlation (Pearson coefficient > 0.5 for all measurements).

In patients with VS ($n = 183$), the saccule was visible in 97% of cases on the coronal reference images (354 out of 366 internal ears), as featured in Fig. 2b. For the visualization of the saccule, the interobserver agreement was mild on the pathological side ($\kappa = 0.41$), however, both observers attributed the same score (= 2) to subjects with major motions artefacts (all in the G3 group, four on the side of the tumour + 8 contralateral). On the tumour-free contralateral ear, interobserver agreement was satisfactory ($\kappa = 0.67$). On the side of the VS, the mean height of saccule was 1.43 mm ± 0.26 (0.9–2.1) and the mean width was 1.1 mm ± 0.21 (0.7–1.7). On the normal contralateral side of the tumour, the mean height of saccule was 1.4 mm ± 0.21 (0.8–2.3) and the mean width was 1.06 mm ± 0.19 (0.8–1.8). For saccular measurements, interobserver correlation showed a strong correlation (Pearson coefficient > 0.5 for all measurements).

28% of patients with internal auditory canal VS presented with a saccular dilation ($n = 59$ dilated saccules in 51 patients) with 38 dilated saccules on the side of the VS and 21 dilated saccules on the healthy side (contralateral side to the tumour), as featured in Figs. 3 and 4. There was a statistically significant difference for the presence of saccular dilation on the pathological side compared to the control group ($p = 2.81 \times 10^{-5}$), as featured in Table 3. There was also a statistically significant difference in the presence of saccular dilation on the healthy side compared to the control group ($p < 0.05$).

Subgroup analysis of G1, G2 and G3 also showed a statistically significant difference for a saccular dilation, compared to the control group with p values at 0.0002 (tumour side) and 0.004 (contralateral side), as presented in Table 4. On the contrary, saccular dilation did not appear to be correlated with the degree of obstruction of the internal auditory canal by the VS, as the proportions of dilated saccules within

Table 2 Specific relevant characteristics of VS between the three groups

	Group 1 ($n = 28$) Non-obstructive schwannomas	Group 2 ($n = 28$ mildly) Obstructive schwannomas	Group 3($n = 127$) Obstructive schwannomas
Tumour side			
Right	15	15	55
Left	13	13	72
Mean tumour volume (cm^3)	0.031 (0.002–0.247, median = 0.0195)	0.313 (0.018–1.023, median = 0.232)	1.498 (0.026–20, median = 0.524)
Koos' classification			
Tl	28	28	32 (25%)
T2	0	0	56 (44%)
T3	0	0	39 (31%)

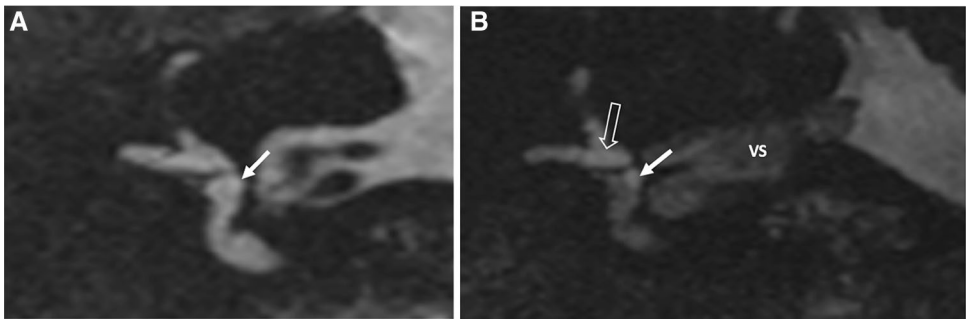

Fig. 2 **a** Normal saccule (arrow) and normal hyperintense signal of the perilymphatic and endolymphatic fluids on T2-weighted images. **b** Normal saccule in a patient with a VS, height ≤ 1.6 mm, width ≥ 1.4 mm (arrow). The endolymph, comprising the saccule (arrow) and the utricle (empty arrow), has a normal hyperintense signal on T2-weighted images, while the perilymph presents with a decreased signal intensity

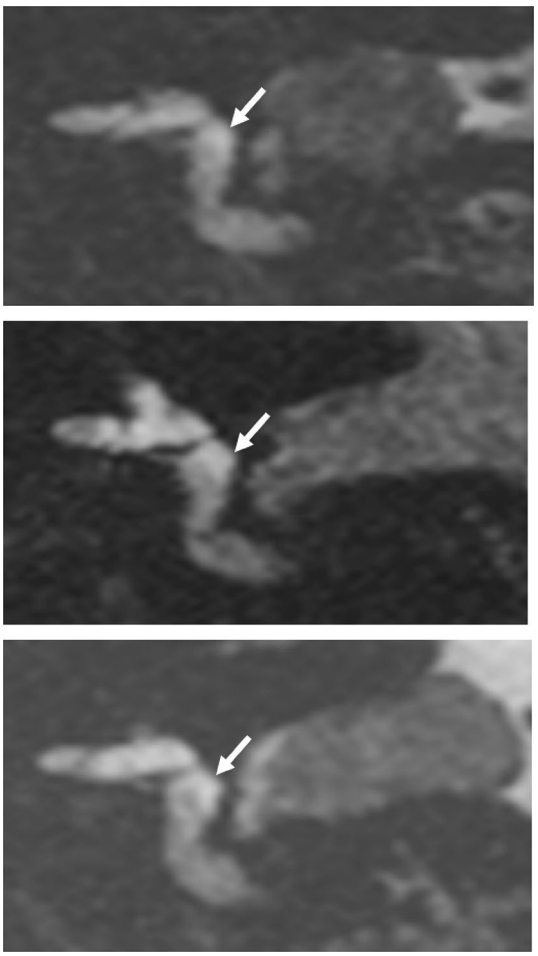

Fig. 3 Enlarged saccule with an increased height ≥ 1.6 mm, in three patients with a VS

groups G1, G2 and G3 were, respectively, 21.4%, 21.4% and 21.1%. The saccular dilation was also not correlated to the volume of the tumour, with p values at 0.8 on the pathological side and 0.6 on the healthy side.

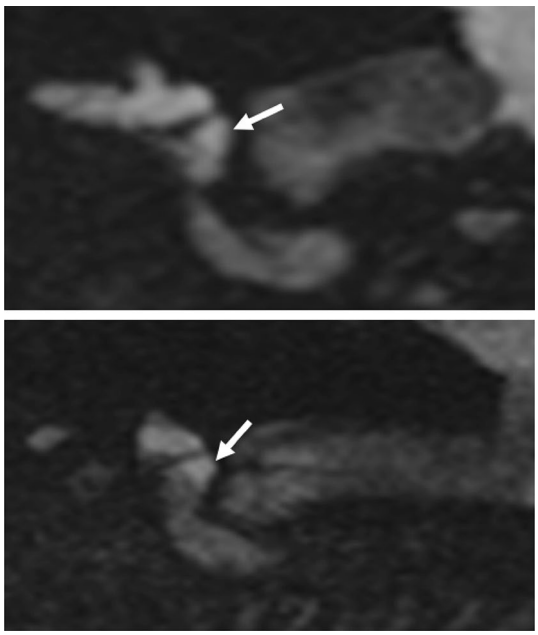

Fig. 4 Enlarged saccule with an increased width ≥ 1.4 mm, in two patients with a VS

Table 3 Number of dilated saccules (height > 1.6 mm and/or width > 1.4 mm) in patients with VS

	Dilated saccules
Tumoral side	
Total	38 (21.2%, $p < 0.0001$)
Unilateral on tumoral side	30
Healthy side	
Total	21 (12% $p = 0.005$)
Unilateral on healthy side	13
Bilateral	In 8 patients
Total	59 (51 patients, 29%)

Table 4 Subgroup analysis of the measurements of the saccule and number of dilated saccules > 1.6 mm (height) and/or > 1.4 mm (width) between patients with the different groups of VS: G1, G2 and G3 (according to the degree of obstruction of the internal auditory canal) and the control group (G0)

	Gl		G2		G3		Control group GO
	Tumour side	Contralateral side	Tumour side	Contralateral side	Tumour side	Contralateral side	
Height (mm)							
means (± SD)	1.43 ± 0.24	1.44 ± 0.22	1.40 ± 0.25	1.45 ± 0.2	1.44 ± 0.26	1.38 ± 0 21	1.41 ± 0.14
Range	1–2	1.1–2.3	1.1–2	1.1–2	0.9–2.1	0.8–2.1	1–1.6
Width (mm)							
Means (± SD)	1.12 ± 0.18	1.05 ± 0.2	1.11 ± 0.21	1.11 ± 0.2	1.1 ± 0.21	1.06 ± 0.18	1.2 ± 0.13
Range	0.9−1.6	0.8−1.7	0.7−1.7	0.9−1.8	0.7−1.7	0.7−1.6	0.9−1.4
Total of dilated saccules	59						0
Dilated saccules							
Number	6	2	6	6	26	13	/
%	21.4%	7.1%	21.4%	21.4%	21.1%	10.9%	
	$p = 0.0002$	$p = 0.004$	p=0.0002	$p = 0.004$	$p = 0.0002$	$p = 0.004$	

Correlation with clinical symptoms (Fig. 5)

On the side of the VS, a statistically significant correlation was found between the presence of a saccular hydrops and:

- Sensorineural hearing loss [OR 3.26, $p = 0.02$, (1.2–8.85)]
- Progressive deafness [OR 2.7, $p = 0.03$, (1.1–6.54)]
- Tinnitus (OR 2.96, $p = 0.006$, (1.36–6.43)].

On the tumour-free side a statistically significant correlation was found between the presence of a saccular hydrops and:

- Sensorineural hearing loss (OR 5.09, $p = 0.03$, (1.14–22.65)]
- Progressive deafness (OR 3.95, $p = 0.03$, (1.12–14.01)]
- Vertigo [OR 2.61, $p = 0.049$, (1–6.79)].

On the other hand, no statistically significant correlation could be demonstrated between balance disorders and saccular dilation. Regarding vestibular functional tests data, no significant correlation was observed.

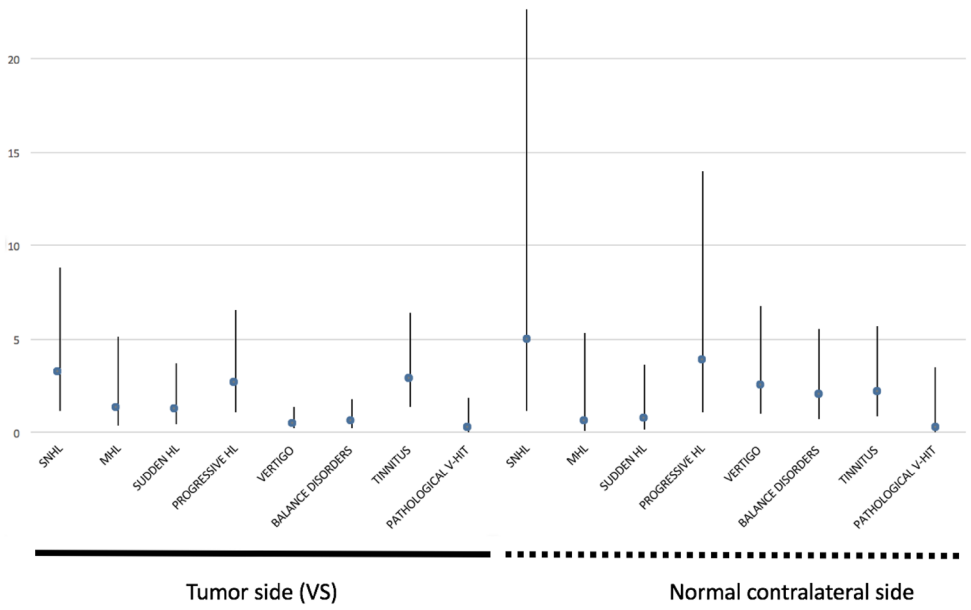

Fig. 5 Odds ratio (OR) with their confidence intervals (IC) for sensorineural hearing loss (SNHL), mixed hearing loss (MHL), sudden hearing loss (sudden HL), progressive hearing loss (progressive HL), vertigo, balance disorders, tinnitus and pathological results of the V-HIT for saccular dilation between the saccule on the side of the tumour and the saccule on the normal contralateral side

Discussion

In our study, around a third (28%) of vestibular schwannomas ($n = 51/183$) presented with a dilation of the saccule, regardless of the degree of obstruction of the internal auditory canal by the tumour and without influence of the tumour volume. This confirms—on a bigger cohort—the findings of Naganawa et al. [4] who found the presence of endolymphatic hydrops in 30% of patients with vestibular schwannoma on a 3D FLAIR sequence without contrast injection in 13 patients. Other authors, such as Ralli et al. [17] and Jerin et al. [18], also reported cases ipsilateral endolymphatic hydrops in vestibular schwannomas.

If delayed-FLAIR sequences have been used over the years to image hydrops [4, 8, 11], different research teams are now using heavily T2-weighted sequences for the visualization of the membranous labyrinth, and especially the saccule and the utricle [9, 12, 13). Our study found matching results [13] regarding the visualization of the saccule and its measurements in terms of height even though the measurements are submillimetric. One of the major interest of such a MRI protocol is the limited time required for the performance of the sequence (< 8 min) compared to 4 h 30 min with delayed FLAIR images [4, 8, 11] and the absence of gadolinium injection.

Several studies reported higher levels of protein in the perilymph of vestibular schwannomas [19–21], which resulted on MRI in an increased FLAIR signal or a decreased signal on T2-weighted sequences [15, 22, 23]. We showed in another article that decreased signal intensity on T2-weighted sequences correlated with the degree of obstruction of the internal auditory canal by the schwannoma [15]. Some authors such as Shinomori and al. [24] or Asmar et al. [25] suggested that accumulation of proteins in the perilymph could represent a cellular stress factor for different support cells of membranous system and lead to a deregulation loop of ion transporters and/or aquaporin system, favouring the occurrence of a hydrops. Increased levels of perilymphatic proteins could result in an increased oncotic pressure in the perilymph, responsible for water movements from the interstitium and the plasma to the perilymph, and as endo and perilymphatic sectors are closely linked there could also be movements of water towards the endolymph. However, an obstructive aetiology does not appear possible, since our study found a similar proportion of dilated saccules (respectively, 21.4%, 21.4% and 21.1% in groups G1, G2 and G3), regardless of the degree of obstruction of the internal auditory canal and no correlation with the volume of the tumour. In addition, we also found dilated saccules on the healthy side (12% of cases, $n = 21$, $p < 0.05$ compared to the control group). This contralateral dilation to the tumour suggests an association with a more general mechanism, which remains unclear.

Vestibular schwannomas symptoms and endolymphatic hydrops symptoms overlap and our study was able to demonstrate a statistically significant association between saccular hydrops and each cardinal symptom of endolymphatic hydrops, such as sensorineural hearing loss, vertigo and tinnitus. There was a statistically significant relationship between sensorineural progressive hearing loss and saccular dilation, whether on the side of the VS or on the tumour-free ear. These results are consistent with at least two recent studies: (1) Sepahdari et al. [8] who found a correlation between increased endolymphatic volume and the degree of the hearing loss and (2) Attyé and al. [11] who found a correlation between saccular volume and sensorineural hearing loss. In another study, Eliezer et al. [9] also described the common occurrence of dilation of a membranous labyrinth structure (the utricle) and sensorineural hearing loss in vestibular schwannomas, but failed to obtain a significant correlation with saccular dilation. However, it was performed on a limited number of cases (32 patients included, 23 analysed) and was based on a volume analysis of the membranous structures (analysis still not validated, without concept of threshold values) [9]. Our study also showed a statistically significant correlation between the presence of a saccular hydrops on the normal tumour-free ear on MRI and the presence vertigo/disequilibrium. Such a correlation was not observed on the side of the VS. It could be secondary to decreased vestibular function. We also had missing vestibular functional tests (48% of the caloric tests and 95% of the V-HIT) which have not been performed in patients whose symptoms were already well compensated at time of diagnosis and imaging assessment. 85% of the schwannomas in our study belonged to group 2 or 3 and thus included all branches of the cochleovestibular nerve. Tinnitus appeared related to saccular dilation the side of the schwannoma. It should be noted, however, that the odds ratio of the healthy side, even in the absence of significant p value remains high (OR 5.85). Tinnitus has been described in VS on the side of the tumour, but its exact aetiology remains to be demonstrated, and several potential mechanisms of tinnitus generation in VS, which are not mutually exclusive, have been proposed: ephaptic coupling of cochlear nerve fibres secondary to compression, cochlear dysfunction, efferent system dysfunction following compression of nerve fibres or tinnitus occurring after cortical reorganization due to the hearing loss [26].

Our study is limited by its retrospective monocentric design, but our results confirmed the only previously published article on the subject by Naganawa et al. Myogenic vestibular evoked potentials, which appear to be more accurate regarding saccular explorations, are now performed at our institution, but at the time of this study, they were only

available for 11 patients (and none of the patients with a hydrops) thus, we did not include them.

Our secondary results regarding deafness, vertigo/disequilibrium and tinnitus would be promising—if confirmed—for therapeutic innovation. The current therapeutic options in case of VS are based on a tumour volume control strategy, which would not be sufficient in cases where the tumour itself does not account for all the symptoms presented by the patient, such as the presence of signs of associated endolymphatic hydrops. The radiological description of a saccular dilation, if associated to presence of para-clinical assessments (functional audio and vestibulometric tests), could suggest a loss of the homeostasis of endolymphatic system to the clinician, and thus lead to the prescription of drugs used in pressure-related disorders in order to alleviate the symptoms.

Conclusion

Around a third (28%) of vestibular schwannomas of the internal auditory canal presented with saccular dilation (bilateral in 15.7%) regardless of the degree of obstruction exerted by the tumour on the internal auditory canal or the tumour volume, with a statistically significant association between saccular dilation and progressive sensorineural hearing loss. There was also a statistically significant association with the presence of tinnitus on the ipsilateral side of the tumour and vertigo on the contralateral side. These findings are promising in terms of therapeutic management since the above-mentioned symptoms overlap the classical triad of primary pressure-related disorders.

Compliance with ethical standards

Conflict of interest The authors have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all participants included in the study.

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