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OTOLOGY

Malformations of the lateral semicircular canal correlated with data from the audiogram

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Abstract

Objectives Lateral semicircular canal (LSCC) malformations are one of the most common inner ear malformations. The purpose of this study is to analyze the prevalence and type of hearing losses associated with LSCC malformations, compared to a control group.

Materials and methods We retrospectively included 109 patients (166 ears) presenting with a CT-confirmed LSCC malformation, compared to a control group (24 patients). The bony island surface and the width of the inner portion of the LSCC were measured to confirm the malformation. There results were correlated to audiogram data: sensorineural (SHNL), mixed (MHL) or conductive hearing loss (CHL) by an otologist.

Results In the LSCC group, 60.9% of patients presented with an audiogram-confirmed hearing loss, especially SNHL (39.2%, $n=65$) and MHL (12.7%, $n=21$). Hearing was normal in 39.2% ($n=65$) of the cases. Bilateral LSCC malformations ($n=57$) were frequently associated with hearing loss (80.7%), SNHL in most of the cases (33.3%). Unilateral LSCC malformations were associated with hearing alterations (51.9%, $n=27$), but we also observed a high rate (81%, $n=42$) of contralateral abnormalities of the audiogram.

Conclusion LSCC malformations are commonly associated with hearing loss (61%), especially SHNL (39%). The high rate (81%) of contralateral hearing disturbances in unilateral LSCC malformations should be taken into account in the patient's daily life to avoid triggering or exacerbating any hearing loss. Otologists and radiologists must cooperate to ensure that all malformations are correctly described on CT, especially to improve the patient's education regarding hearing preservation.

Keywords Lateral semicircular canal · Congenital · Malformation · Hearing loss · Audiogram

Introduction

Labyrinthine malformations and especially posterior labyrinthine malformations (deformations and minor or major dilatations) are routinely observed on CT and MRI [1, 2].

Lateral semicircular canal (LSCC) is one of the most common inner ear malformation and represents 63% of posterior labyrinth malformations [1]. LSCC dysplasia has an embryologic origin [3]. During the sixth week of development, the budding semicircular canal (SCC) forms an evagination from the vestibular anlage. Thereafter, the central portion of the pocket-shaped protrusion adheres, leaving a peripheral semicircular tube. Failure in this central adhesion results in SCC dysplasia. In addition, lateral SCC malformations are more common than superior or posterior ones, since they appear earlier during embryogenesis [4, 5]. When exploring a congenital deafness, the knowledge of a family or genetic background (for instance, inactivation of the homeobox gene Nkx5-1) can immediately guide the diagnosis [6]. In other cases, clinical and audiological examinations by an ENT (ear, nose and throat) surgeon are usually followed by morphological explorations, to look for inner ear malformations.

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CT is usually the first imaging modality performed, as LSCC malformations are easily observed on axial CT images [1, 2]. The association of inner ear malformations with hearing loss remains underappreciated, with few studies published. Labyrinthine malformations were commonly thought to be associated with sensorineural hearing loss (SNHL). However, some authors such as Johnson et al. have studied 13 LSCC malformations and suggested that they may be associated with both SNHL and conductive hearing loss (CHL) [1, 2]. Other authors even reported cases of LSCC malformation with normal hearing [7, 8]. These findings have never been confirmed on larger cohorts.

The purpose of this study is to analyze the prevalence and the type of hearing losses (sensorineural, conductive or mixed hearing losses) associated with LSCC malformations on a larger cohort and compared to a normal reference population.

Materials and methods

Study population

We retrospectively included patients presenting with an isolated posterior labyrinthine malformation of the LSCC, described on the radiological report, on a temporal bone CT performed at our institution (Tertiary center, University hospital, reference center for ENT pathologies) between January 1st 2014 and January 1st 2016.

A posterior labyrinthine malformation was defined as a malformation of either the vestibule, the cistern and/or the semicircular canals (anterior, posterior or lateral), compared to an anterior labyrinthine malformation which occurs in the cochlea. All patients had to be refered by an otolaryngologist from our hospital, so that the results of the clinical and audiological examinations would be available and reproducible. In the majority of cases, the CT was requested by the otorhinolaryngologist for further assessment of the middle ear cavity because of ascertained hearing loss. Exclusion criteria were: the absence of audiometric data, the presence of defects of the semicircular canals, anterior labyrinth dysplasia, ossicular lysis or malformations, otosclerosis, inflammatory filling of the middle ear, chronic otitis media scars, history of previous surgeries on the head and neck, external auditory canal abnormalities and benign or malignant tumors. The absence of valid audiogram data was also an exclusion criterion. 109 patients (218 ears) met the inclusion criteria. Out of 109 patients, 166 ears (79 right and 87 left) were included, as featured in Table 1, and 52 ears were excluded.

The contralateral normal ear (free from lesions) of 24 patients, included during the same period, with a unilateral temporal bone fracture and no abnormalities on the audiogram, served as the control population.

CT protocol

All participants underwent a non-enhanced CT of the temporal bone. All patients were imaged on a 128-section CT scanner (Definition AS, Siemens, Strasbourg, France). The direct axial multidetector temporal bone CT was performed with 120 kVp, 250 mAs/section. Images were reconstructed with a 0.4–0.5 mm section thickness and a 0.1–0.2 mm section interval. The acquisition box was placed parallel to the orbital roof to reduce lens exposure. Axial reconstructions were parallel to the lateral semicircular canal. Sagittal and coronal reconstructions were also performed.

Image analysis

All images were analyzed by a junior and a senior radiologist specialized in head and neck imaging. Axial plane was chosen as reference for the image analysis and the images were oriented in the axis of the lateral semicircular canal. Additional coronal and sagittal reconstructions were obtained from the original axial dataset.

We performed two measurements:

- The width of the medial part of the LSCC. We measured the width of the canal between its walls, as illustrated in Fig. 1.
- The surface of the bony island at the center of the LSCC was obtained by manual segmentation using Osirix® software and measured in square millimeters (Fig. 2).

We also visually analyzed the shape of the external wall of the LSCC, which must be regular. If the shape of the

Table 1 Distribution of the types of hearing losses in the LSCC population and control group

Type of hearing loss on tonal audiogram	Right ear ($n=79$)	Left ear ($n=87$)	Total ($n=166$)	Control group ($n=24$)
Normal hearing	30	35	65 (39.2%)	21 (87.5%)
SNHL	31	34	65 (39.2%)	3 (12.5%)
MHL	10	11	21 (12.7%)	0
CHL	8	7	15 (10.1%)	0

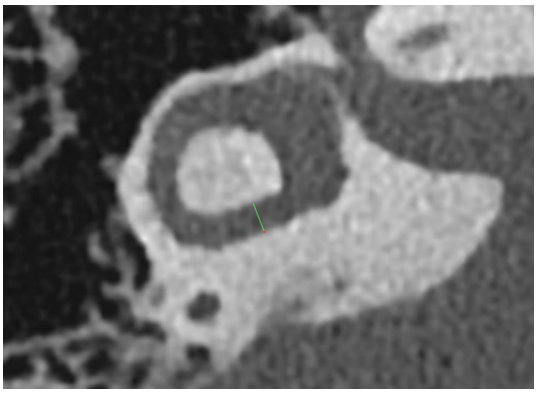

Fig. 1 Normal measurement of the inner portion of the LSCC

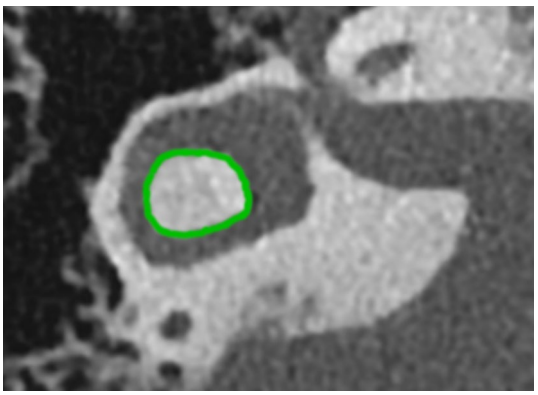

Fig. 2 Surface of the bony island at the center of the LSCC

LSCC appeared irregular, the LSCC was considered to be deformed.

Audiogram analysis

All audiometric data from the LSCC group and the control group were analyzed by an otorhinolaryngologist, blinded to the radiological presentation. The tonal audiogram was analyzed at 250–500–1000–2000–4000 KHz frequencies. Hearing thresholds were measured for each frequency. Thresholds were addressed at 5 dB HL steps (increasing and decreasing intensity) for both air (AC) or bone conduction (BC). Hearing losses were categorized as sensorineural (SNHL), conductive (CHL) and mixed hearing loss (MHL).

Statistical analysis

Continuous variables are given as mean ± standard deviation, median (range), in millimeters for the width of the medial portion of the LSCC and in square millimeters for the surface of the central bony island. Categorical variables were expressed in terms of numbers and percentages. A Student *t* test was performed to compare the means between patients with LSCC malformation and the control group. The significance was set at $p = 0.05$. All tests are performed using the SPSS software (Statistical Package for the Social Sciences, version 22.0 IBM, Inc., Armonk, New York, USA). Intraclass correlation coefficient (ICC) was calculated for inter-observer agreement using MedCalc Software, version 18.11.

Results

Study population

166 ears with LSCC malformations, in 109 patients [53 men, 56 women, median age 44 years (range 5–84)], were analyzed. The control group comprised 24 patients (16 men and 8 women), with the same median age of 44 years (8–84). There were no significant differences in terms of sex and age between LSCC patients and the controls.

The majority of patients (52%, $n = 57$ patients, 114 ears) presented with bilateral LSCC malformations and only 52 patients (52 ears) presented with unilateral LSCC malformations (48%).

Image analysis results

The medial portion of the LSCC was wider [1.84 mm ± 0.45; 1.8 mm (1.1–6 mm)] in the LSCC group compared to the control group [1.42 mm ± 0.16; 1.4 mm (1.1–1.7 mm)] ($p < 0.05$). The LSCC was never ≥ 1.8 mm in the control group, thus the medial portion of the LSCC was considered pathological when ≥ 1.8 mm (Fig. 3). The inter-observer agreement was good (0.89 for the control group, 0.98 for LSCC malformations).

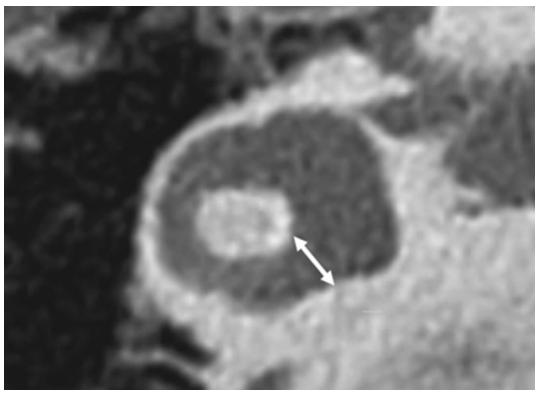

Fig. 3 In the LSCC group, the medial portion of the LSCC was wider (mean = 1.84 mm) than in the control group (1.42 mm, $p < 0.05$) (Fig. 4b). Thus, the medial portion of the LSCC was considered pathological when ≥ 1.8 mm. See text for average values and ranges

The average surface of the bony island was smaller [6.43 $mm^2 \pm 2.0$; 6.45 mm^2 (0.4–13.2 mm^2), Fig. 3] in the LSCC group compared to the control group [9.99 $mm^2 \pm 1.40$; 10.35 (7.1–12 mm^2), ($p < 0.05$)]. The central bony island was never < 7 mm^2 in the control group, thus its surface was considered pathological when < 7 mm^2 (Fig. 4). The inter-observer agreement was good (0.94 for the control group, 0.99 for LSCC malformations).

The majority 96% ($n = 161$) of the LSCC malformations presented either a decreased surface of the bony island < 7 mm^2 and/or a widening of the inner part of the LSCC ≥ 1.8 mm:

- A decreased bony island surface (< 7 mm^2) with a normal medial portion of the LSCC (< 1.8 mm) in 30% of the cases (49 ears: 22 right, 27 left)
- A normal bony island surface (> 7 mm^2) associated with a widened (≥ 1.8 mm) medial portion of LSCC in 35% of the cases (57 ears: 24 right, 32 left)
- A decreased bony island surface (< 7 mm^2) associated with a widened (≥ 1.8 mm) medial portion of LSCC in 35% of the cases (55 ears: 24 right, 28 left)

In 4% of the cases ($n = 5$), the shape of the LSCC was abnormal, with one bilateral deformation of the LSCC (1 patient) and three unilateral deformations of the LSCC (3 patients) without abnormalities of the surface of the central bony island or any abnormalities of the size of the inner part of the LSCC.

Audiometric findings

The audiometric data are featured in Table 2. In the LSCC group, 60.9% ($n = 101/166$) of patients presented with a hearing loss. SNHL was observed in 39.2% of the cases ($n = 65$), MHL in 12.7% of the cases ($n = 21$) and CHL in 9% of the cases ($n = 15$). The hearing was normal in 39.2% ($n = 65/166$) of the cases.

81% of patients ($n = 46$) with bilateral malformations (57 patients, 114 malformed ears, Table 2), presented

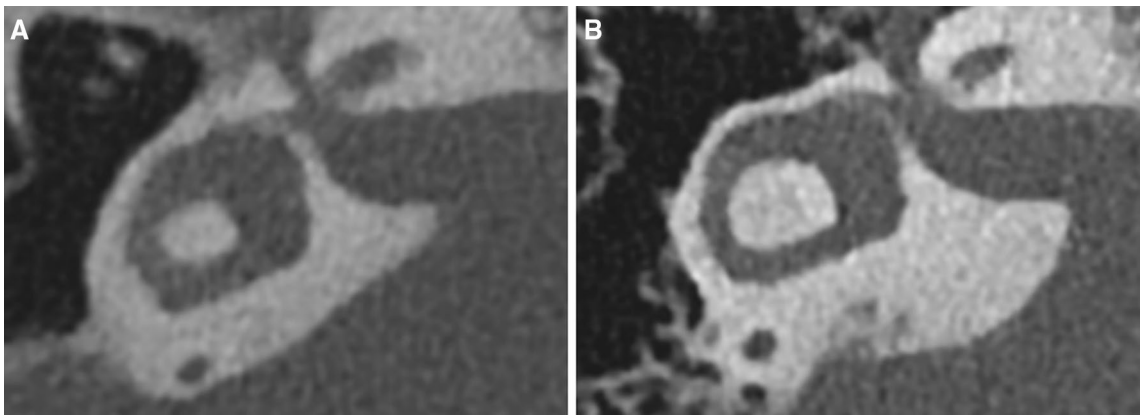

Fig. 4 The average surface of the bony island was smaller (6.43 mm^2) in the LSCC group (**a**) compared to the normal group ($p < 0.05$) where the average surface was 9.9 mm^2 (**b**). The surface of the bony island was considered pathological when < 7 mm^2

Table 2 Types of hearing losses associations in bilateral LSCC malformation

Type of hearing loss in bilateral LSCC malformation	Number of patient involved ($n = 57$)	%
Normal hearing	11	19.3
SNHL (bilateral)	19	33.3
MHL (bilateral)	4	7
CHL (bilateral)	1	1.8
SNHL (one side) + normal hearing (one side)	7	12.3
SNHL (one side) + MHL (one side)	6	10.5
MHL (one side) + normal hearing (one side)	2	3.5
CHL	7	12.3

SNHL sensorineural hearing loss, *CHL* conductive hearing loss, *MHL* mixed hearing loss, *NHL* no hearing loss

with hearing loss on the audiogram. Bilateral SNHL was observed in 33.3% of the cases ($n=19$), bilateral MHL in 7% of the cases ($n=4$) and bilateral CHL in 1.8% ($n=1$). The hearing was normal in 19.3% of the cases ($n=11$). Moreover, in 22 patients, associations of different hearing types were observed:

- SNHL + no hearing loss (NHL) in 12.3% of the cases ($n=7$)
- SNHL + MHL in 10.5% of the cases ($n=6$)
- CHL + NHL in 12.3% of patients ($n=7$)
- MHL + NHL in 3.5% of patients ($n=2$).

The hearing was normal in the majority of patients (51.9%, $n=27$) with unilateral LSCC malformation (52 ears: 22 right, 26 left, Table 3). SNHL was observed in most cases (26.9%, $n=14$), CHL in 11.5% of the cases ($n=6$) and MHL in 9.6% of the cases ($n=5$). A contralateral abnormality (on the non-malformed ear on CT images) of the audiogram was observed in 81% of cases ($n=42$).

Conversely, in the control group, only three patients presented hearing abnormalities. These three patients (> 60-year-old men) presented with SNHL on high frequencies (> 4 kHz) with normal hearing at low frequencies, the rest of the audiograms were normal.

Discussion

LSCC malformations are one of the most common inner ear malformations (representing around 63% of posterior labyrinth malformations and 39% of all malformations), occurring at early stages of embryogenesis (during the sixth week). The correlation between LSCC malformation and hearing loss remains controversial. In our study, LSCC malformation is characterized by either an increased width of its medial portion (≥ 1.8 mm) and/or a lowered central bony island surface (< 7 mm^2). In the literature, most authors describe the bony island in terms of its width, with a decreased width in case of LSCC malformation, but a surface measurement is more precise [9–11]. Johnson et al. classified their LSCC malformations between "anterior limb dilation", "posterior limb dilation" and "dysplasia", but did not give any measurements [1].

Table 3 Hearing losses in unilateral cases of LSCC malformations and contralateral normal ear audiogram data

	Unilateral malformations ($n=52$)	Contralateral ear
Normal hearing	27 (51.9%)	10 (19%)
Hearing loss		
Total	25 (48.1%)	42 (81%)
SNHL	14 (26.9%)	
MHL	5 (9.6%)	
CHL	6 (11.5%)	

LSCC malformations were associated with audiogram-confirmed hearing loss in 61% of the cases in our study. The prevalence of hearing loss raised up to 80.7% in case of bilateral LSCC malformations. The most frequent type of hearing loss was SNHL (39.2%), with a lower proportion than in Johnson's study. Compared to Johnson et al., our cohort is eight times larger and their LSCC malformations were not isolated, as some of their patients also presented with enlarged vestibular aqueduct, which could be a source of bias [1]. Similarly, Lan et al. described an association between a smaller width of the bony island and SNHL [10]. MFL (12.7%) and CHL (9%) were also observed, which is in agreement with the literature. Both Johnson et al. and Phelps et al. have described an association between LSCC and CHL, in four and two patients, respectively [1, 5]. Furthermore, in our study, 38.5% of the patients with bilateral LSCC presented with an association of different types of hearing losses. In our study, the hearing was normal in 39.2% ($n=65$ ears) of the cases of LSCC malformations, which is higher than reported in the literature, where Johnson et al. found a normal hearing in only three ears (out of 13 patients, 11%) [1, 5, 6].

Johnson et al. first suggested that CHL associated with LSCC malformation, as well as SNHL, could be of inner ear origin [1]. The occurrence of different types of hearing loss in LSCC malformations has a possible embryological origin. The anterior and posterior crus of the stapes derive from mesoderm, while the footplate, as well as the labyrinth, share a common ectodermal origin [11]. We could speculate that malformations occurring during the embryogenesis could affect multiple structures sharing the same origin (ectoderm) with various degrees of clinical or radiological impairment (apparent or not on CT/MRI). For instance, SNHL could be explained by associated anterior labyrinth abnormalities, some of which may be infra-radiological [1]. SHNL also occurs in enlarged vestibular aqueduct syndrome, which is another type of posterior labyrinthine malformation [12]. Enlarged vestibular aqueducts are often associated with hypoplasia of the cochlea, but SNHL may occur even in cases of isolated malformations [12]. CHL and MHL occurrence may also be explained by the ectodermal origin of the footplate, which shares the same origin as the labyrinth: one could speculate that if the labyrinth is affected by malformations, the footplate may also be, as they derive from same primary germ layer of the embryo. Infra-radiological malformations, occurring in the ectoderm during embryogenesis and affecting ectoderm-derived structures of the ear, could also explain the surprising high rate (81% of cases, $n=42$) of contralateral hearing disturbances (on the

non-malformed ear), without any signs of malformation on CT images, in patients with unilateral LSCC malformations.

Our study has several limitations. For data access purposes, we retrospectively enrolled patients based on the results of their radiological reports and limited our selections to outpatients from our referring clinicians. We could not have performed the study on all patients who underwent a CT in our hospital, as the clinical data would have not been available for all patients. Our control population was also small. Another limitation is the spatial resolution of the CT compared to flat-panel CT as reported by recent studies. The resolution of the flat-panel CT is indeed higher than the 128-slices CT, but for the assessment of the LSCC and the bony island (1–10 mm^2), the resolution of the CT remains sufficient [13, 14]. In the control group, only three patients presented hearing abnormalities. These three patients were > 60-year-old men and presented with SNHL on high frequencies (4000 Hz), which is compatible with presbycusis-related SNHL.

Labyrinthine malformations, despite their frequent pathological audiometric profile, often go unrecognized and therefore underdiagnosed, even though the CT-based diagnosis is easy and does not require any contrast-media injection. Particular attention should be paid in the presence of labyrinthine malformations whenever stapes surgery is planned (otosclerosis, chronic otitis media, perilymphatic fistulas…) [10]. The surprising high rate for contralateral hearing disturbances in LSCC malformations must also be taken into account in the patient's daily life to avoid triggering or exacerbating any hearing loss.

Conclusion

Lateral semicircular canal (LSCC) malformations are one of the most common radiological inner ear malformations, usually bilateral and associated with hearing loss in 61% of the cases. The most common type of hearing loss is SNHL (39.2%), followed by MHL. Hearing loss is even more frequent (80.7%) in case of bilateral LSCC malformations. The hearing was normal in 39.2% of LSCC malformations and in unilateral LSCC malformation, we found a surprising high rate (81%) of contralateral hearing disturbances, which should be taken into account in the patient's daily life, to avoid triggering or exacerbating any hearing loss. Otologists and radiologists must fully cooperate to ensure that even minor labyrinthine malformations are correctly described on CT, especially to improve the patient's education regarding hearing preservation.

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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