CASE REPORT

Dissociated responses to caloric and head impulse stimulation in a case of isolated vestibule-lateral semicircular canal dysplasia

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ABSTRACT
Isolated vestibule-lateral semicircular canal dysplasia (LSCCD) is one of the most common inner ear malformations. We present a case of a 59-year-old patient with right vestibule-lateral canal dysplasia and a history of spontaneous vertigo spells without hearing loss. Vestibular assessment showed a dissociation between caloric responses and vestibulo-ocular reflex gain as measured with the video head-impulse test.

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Introduction

Inner ear malformations can display a great variability of clinical and radiological findings. Depending on induction time during embryological development, they may involve several structures such as the cochlea, vestibule, vestibular aqueduct or semicircular canals. Various classification systems for these malformations have been proposed, based on both temporal bone studies and radiographic findings [1,2].

Anomalies of the vestibular system are the most common inner ear malformations encountered in temporal bone studies [3]. In particular, isolated vestibule-lateral semicircular canal dysplasia (LSCCD) is the most common inner ear malformation [1,3] and it may present with no cochlear abnormalities nor dilation of the vestibular aqueduct [4]. Computed tomography and high-resolution magnetic resonance imaging can identify peripheral vestibular anomalies such as deformities of the bony labyrinth and pathological changes of the membranous labyrinth, respectively.

The reports on vestibular function in patients with inner ear malformations are scarce, limited to short series or case reports. Whether a malformed vestibular system is capable to generate a vestibulo-ocular reflex (VOR) suitable for a normal life or the deficit is compensated for by the visual and somatosensory systems in an unanswered question. Another unsolved issue is whether the malformed bony labyrinth may contain a normally structured membranous labyrinth with normal or near normal function. We report on the case of a patient with vestibule-lateral semicircular canal dysplasia admitted to our clinic with a history of spontaneous vertigo attacks in the last three years. The results of the audiometric and vestibular tests and radiological findings are presented and discussed.

Case report

A 59-year-old patient referred to our outpatient clinic with a three-year history of spontaneous vertigo spells, two to three times per year. The attacks were not triggered by changes of head position nor accompanied by vegetative symptoms, yet associated with some degree of instability. She denied hearing loss, but she mentioned a sporadic tinnitus on the left ear. Before being referred to us, a temporal bone computed tomography (CT) scan was performed which showed an absence of the central bony island of the lateral semicircular canal which was fused together with an enlarged vestibule on the right side (Figure 1). The middle ear, cochlea, vestibular aqueduct and internal...
auditory canal showed no anomalies and the left side was normal. The otoscopic examination showed normal eardrums and the only positive finding in the otoneurological examination was a left beating nystagmus with a vertical upward and a torsional component with the upper pole of the eye beating toward the left when visual fixation was suppressed and which was more intense with extreme left gaze and on the left lateral and left Dix–Hallpike positions.

Pure tone audiometry, speech audiometry, tympanometry and stapedial reflex were normal. Both cervical and ocular air and bone-conducted vestibular evoked myogenic potentials were within normal limits. Interaural amplitude ratios above 40% were considered abnormal (Figure 2).

Caloric test revealed an 82% right canal paresis (Figure 3). For this test, each ear was irrigated alternately for 40 s with a constant water flow at temperatures of 30°C and 44°C. The maximum slow-phase velocity of nystagmus was calculated, and the canal paresis and directional preponderance were determined according to Jongkees formula. An asymmetry between the left and right ear greater than 21% was considered as indicative canal paresis. The video-head impulse test showed normal gains on both lateral canals (Figure 4). A magnetic resonance (MR) with 3-dimensional reconstructions of the inner ear and specific sequences for evaluation of endolymphatic hydrops was performed which showed an enlarged vestibule forming a single fluid-filled cavity with the lateral semicircular canal on the right side and significant hydrops on the area of the vestibule (Figures 5 and 6). As such, this patient was diagnosed as having an episodic vestibular syndrome secondary to recurrent right vestibular dysfunction, probably caused by the hydrops within the dilated vestibule-lateral canal cavity.

Discussion

Vestibule and lateral semicircular canal anomalies are highly prevalent among inner ear malformations either with or without hearing loss [1,5]. The three semicircular canals are formed during the fourth to seventh weeks of the embryonic period. Vulnerability of the lateral semicircular canal may be due to its late formation during inner ear embryogenesis since it is the canal that develops last [1] or to dysfunction of genes that are expressed in the inner ear [6]. This also explains why it is the canal which may be absent on its own. Sennaroglu et al. [2] proposed that asymmetrical pathological conditions, like the case we present are due to an external factor that causes the developmental arrest and bilateral symmetric cases are most likely due to genetic defect as it would be expected to cause identical deformity on both sides. The mechanism of LSCCD involves the lack of formation of the central bony island (CBI) and the measurement of its size on axial temporal bone CT scan or axial MR cisternographic image has been used for diagnosis [7].

Like our case, Schuknecht [4] reported a patient with isolated LSCC malformation in both ears with normal hearing; on the contrary, Jackler et al. [1] reported an average hearing level of 35 dB in isolated vestibule-LSCC dysplasia. In a review of clinical records and imaging studies of 15 pediatric patients (28 ears) with LSCC malformations, Johnson et al. [5] found sensorineural hearing loss in 20 ears (71%), conductive hearing loss without external or middle ear findings in 4 ears (14%) and normal hearing in three ears (11%), they concluded as well that there was no correlation between extent of radiological LSCC malformation and level of hearing loss or vestibular symptoms.

Regarding the vestibular function, the patient did not complain of vertigo until adulthood and this situation has been previously reported [2,4,8]. Dizziness appears to be an uncommon complaint and isolated uni or bilateral semicircular canal malformations may go clinically undetectable because of complete compensation. Jackler [1] reported that dizziness was present in only 19% of patients with inner ear malformations. Two findings in the vestibular examination of our patient were of interest and worthy to
Figure 2. (A) BC-oVEMP (asymmetry ratio 20.79%). (B) AC-cVEMP (asymmetry ratio 37.78%).

Figure 3. Caloric test. Right (A) and left (B) ear caloric responses. (C) Summary of maximal slow phase velocities (RE: right ear; LE: left ear).
Spontaneous and direction fixed positional nystagmus is a very common finding in patients with chronic dizziness and in those with recurrent nonpositional vertigo spells; in the later, intensity and direction are related to the time elapsed since the last vertigo spell. It is a non-localizing finding that even during an acute crisis of Ménière’s disease is difficult to correlate with a specific peripheral vestibular deficit [9]. In our patient seen in an intercritical period late after the last vertigo spell the VEMP tests and the horizontal canals VOR gains, as measured with the vHIT, demonstrated responses within normal limits. It could be argued, thus, that it could be a manifestation of habituation once the acute dysfunction has recovered. Likewise, Yukawa et al. [10] described a case of bilateral posterior semicircular canal and lateral semicircular canal dysplasia with dilation of the vestibule on both sides and with normal cVEMP and VOR elicited by horizontal rotational stimulus, along with decreased caloric response in the same ear.

The second finding in the vestibular work-up results raises a very current question in vestibular testing as it is the dissociation between caloric and vHIT results, being the former abnormal and the latter normal. This pattern has been found in 20% of patients with dizziness [8] but is more frequent in patients with Ménière’s disease in which it is hypothesized to be due to hydropic distention of the membranous
labyrinth in the LSC [11] and ampulla [12]. It has also been reported to be a frequent finding in patients with enlarged vestibular aqueduct [13]. We have shown in our case that this could be a possible explanation based on the MRI findings that were similar to those reported by others [7]. Technical limitations in voxel dimension in our study reduce the ability to image the membranous canal properly. Both techniques differ significantly, but regarding our findings we should also mention another possible source of a dissociated response. Angular VOR fails to compensate at lower frequencies of stimulation as in the caloric test; in order to prolong the peripheral vestibular signal (as seen in constant velocity rotations) a central processing, a velocity storage integrator, has been proposed. Different conditions can modify its function well characterized by a time constant: this is significantly reduced in MD as in other peripheral vestibulopathies [14]. The nystagmus response to the caloric test should be then also reduced as seen in our case. High-frequency angular VOR does not needs of the velocity storage to correctly compensate for head rotations [15].

Otopathological reports of clinical and experimental subjects also contribute to sustain this hypothesis. Normal differentiation studies have shown that canal development occurs independently of hair cell differentiation accounting for the presence of normal cristae and maculae despite vestibule-lateral SCC malformation, as it has been shown on morphological examinations of the inner ears of Ecl mouse, a mutant that displays a circling phenotype and hyperactivity associated with bilateral malformation of the horizontal semicircular canal and duct. [6]. Johnsson [16], in microdissections of temporal bones with LSCC malformations, reported a case which showed lack of development of lateral bony canal, but the bony canal anlage enclosed a membranous ampulla and canal of normal tubular morphology. Ohtani et al. [17] reported on a patient with trisomy 22 and Mondini dysplasia whose histological assessment showed the presence of a macula within a dilated utricule and a hypoplastic LSCC containing a crista ampullaris. Nevertheless, a normal sensory epithelium alone seems insufficient to generate a normal VOR since quantitative vHIT demonstrates that superior canal VOR gain is significantly reduced after plugging in treating a superior canal dehiscence syndrome [18]. Likewise, the VOR gain as measured with scleral search coils was also reduced in the posterior semicircular canal after occlusion for intractable benign paroxysmal positioning vertigo [19]. These findings suggest that, at least, a canal-shaped membranous labyrinth is needed to allow for pressure gradients to occur across the cupula during caloric testing. Johnsson [16] and McGarvie et al. [11], propose both in the malformed labyrinth and in the case of patients with Ménière’s disease, respectively, that the response’s dynamics to temperature change in the external auditory canal are disturbed; in the former due to the pouch-shaped horizontal canal and ampulla in which the cupula is not properly enclosed by a membranous wall and in the latter due to distention of the membranous canal as has been recently modeled [20].

In summary, vestibule-lateral semicircular canal is one of the most frequent inner ear malformations. Patients are mostly asymptomatic, but vestibular symptoms may appear with late onset in life. Vestibular testing results may range from normal to absent responses and a dissociation between caloric responses and VOR gain as measured with the vHIT has been reported. Image studies with CT and MRI are useful diagnostic tools and complementary to each other. In the future, new MRI sequences might bring detailed knowledge about the structure of the membranous labyrinth inside a malformed bony labyrinth and this could contribute to elucidate the physiopathology of our findings.

**Disclosure statement**

The authors declare no conflicts of interest.

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