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Vestibular System Changes in Sudden Deafness With and Without Vertigo: A Human Temporal Bone Study

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Abstract

Objective—To investigate the vestibular system changes in sudden deafness with vertigo (SDwV) and sudden deafness without vertigo (SDwoV) and the cause of persistent canal paresis (CP) in SDwV patients.

Study Design—Retrospective study.

Materials and Methods—Four temporal bones from the affected ear in 4 patients with unilateral sudden deafness (SD), 2 SDwV and 2 SDwoV, were selected. Four contralateral temporal bones with normal-hearing ears were defined as the control. Morphologic findings of the labyrinth, the number of Scarpa's ganglion cells, and the density of vestibular hair cells were investigated in all temporal bones. Clinical data and the results of vestibular tests of 11 patients with unilateral SD, as a separate group, also were investigated.

Results—Atrophic change of the organ of Corti, tectorial membrane, and stria vascularis in cochlea, and deposits and atrophic otoconial membrane in vestibular sense organs were seen on affected ears more than control ears. The density of Type I hair cells seemed to decrease on the saccular macula and the posterior semicircular canal crista on affected ears, and there was no remarkable difference between SDwV and SDwoV. In 1 patient with SDwoV who died 10 months after the onset of SD, there were large amount of deposits on the cupula, the atrophied otoconial membrane was peeling off from the saccular macula, and the saccular membrane collapsed to the saccular macula in the affected ear. In the clinical data, all SDwV who were examined within 2 years from the onset had CP, and all SDwV had profound hearing loss.

Conclusion—There is no remarkable difference between SDwV and SDwoV in the number of Scarpa's ganglion cells and the density of vestibular hair cells. The damage of the extracellular superstructure is seen in SD with or without vertigo. The damage of extracellular superstructure is potentially one of the causes of persistent CP in patients with SD.

Keywords

Extracellular superstructure; Sudden deafness; Scarpa's ganglion cell; Temporal bone; Vertigo; Vestibular hair cell

Patients with sudden deafness (SD) often have accompanying vertigo. Kitahara et al. (1) suggested that patients with SD with vertigo (SDwV) had damage mainly in the labyrinth, which was concluded from the pattern of acceleration of central vestibular compensation. They also reported that 66.7% of SDwV with canal paresis (CP) at the onset of SD still had CP around 2 years later. Iwasaki et al. (2) reported that the lesion site of SDwV was within the labyrinth, which was concluded from the results of click- and galvanic-vestibular evoked myogenic potential, and the saccule could be involved more frequently than the semicircular canals, which was concluded from the findings of click-vestibular evoked myogenic potential and caloric test. However, it was reported that there were no remarkable histopathologic changes in the vestibular sense organs in patients with SD (3–8). Khetarpal (9) also reported that there was no significant difference in the number of Scarpa's ganglion cells or the density of vestibular hair cells among SDwV, SD without vertigo (SDwoV), and control. He suggested that SDwV was caused by ultrastructural changes in the vestibular nerves and sensory cells or alterations in their biochemical environment.

In this study, we correlated the vestibular system changes in SDwV and SDwoV and with the clinical data of patients with SD.

MATERIALS AND METHODS

The materials for this study were selected from the temporal bone collection of the University of Minnesota, Minneapolis. There were 4 temporal bones (TBs) from the affected ear in 4 unilateral patients with SD, 2 SDwV and 2 SDwoV, which had no history of otologic disease except SD. Four contralateral TBs with normal-hearing ears were defined as the control.

TBs were harvested at the time of autopsy, fixed in 10% formalin, decalcified with ethylenediamine tetra-acetic acid, dehydrated in graded concentrations of alcohol, and embedded in celloidin. Specimen was cut at a section thickness of 20 μ m with a sliding microtome in the horizontal plane from superior to inferior. Every 10th section was stained with hematoxylin-eosin and mounted on a glass slide.

The organ of Corti, tectorial membrane, and stria vascularis in cochlea, any deposits, and integrity of membranes and cupula in the vestibular sense organs were investigated under light microscopy.

The number of Scarpa's ganglion cells was counted in all TBs. Scarpa's ganglion cells were divided into 2 cell groups, superior and inferior. Cells with nucleoli were counted separately in every 10th section under a magnification of $\times 200$. The formula, $Ni = ni \times t/(t + d)$, was used to compensate for double counting of cells lying at the interface between sections (10). Ni is the estimated number of Scarpa's ganglion cells, ni was the counted number, t was the thickness of section (20 μ m), and d was the mean value of nucleolar diameters in 100 Scarpa's ganglion cells. Finally, the number of Scarpa's ganglion cells was given by multiplying by 10 to account for unstained sections.

The density of vestibular hair cells in the cristae of the lateral semicircular canal (LSC) and the posterior semicircular canal (PSC), and utricular and saccular maculae were examined in all TBs. According to the criteria of Merchant (11), Types I and II hair cells with nucleus

were counted separately under differential interference contrast (Nomarski) microscopy at a magnification of $\times 1,250$. The results were expressed as the number of hair cells per 0.01 mm^2 of surface area, which was determined by dividing the number of counted hair cells by the surface area of the sensory epithelium. Surface area was determined by multiplying the thickness of the section ($20 \text{ }\mu\text{m}$) by the length of the sensory epithelium where the count was made. The counts were performed in areas where the cut was perpendicular to the surface of the sensory epithelium. The formula, $N_i = ni \times t/(t + d)$, was again used. N_i is the estimated density of vestibular hair cells, ni was the raw density, t was the thickness of section ($20 \text{ }\mu\text{m}$), and d was the mean value of nuclear diameters in 100 vestibular hair cells. The density of total hair cells was calculated by summing the densities of Types I and II hair cells.

In addition, there were 11 patients with SD, 6 SDwV and 5 SDwoV, to whom pure tone audiometry and caloric test had been performed at the Paparella Ear Head and Neck Institute between July 2001 and October 2008. The criteria for SD included more than 30-dB sensorineural hearing loss occurring in at least 3 contiguous frequencies, and in addition, SDwV patients had a single attack of rotatory vertigo occurring almost simultaneously with the onset of hearing loss, and no other neurologic signs (2). Caloric weakness that was more than 25% was defined as CP. Pure tone average was calculated as the average threshold at 500, 1,000, and 2,000 Hz. Profound hearing loss was defined to be more than 60 dB in pure tone average. The recovery was defined as hearing improvement, which was more than 10 dB comparing the pure tone average with the previous one. This study was approved by the institutional review board of the University of Minnesota (0206M26181).

RESULTS

Histopathologic changes are shown in Table 1. Cochlea hair cells and supporting cells were more atrophic in affected ears than in control ears. In addition, the loss of inner hair cells was found in 3 of 4 affected ears. Stria vascularis was atrophic in all affected ears. The tectorial membrane was atrophic in all turns of the cochlea in 3 of 4 affected ears. Deposits were seen in the vestibular endolymphatic space and were more commonly attached to the cupula in affected ears than in control ears. The findings, which suggest previous rupture of the membranes, were not seen. There was no significant difference between SDwV and SDwoV groups regarding inner ear changes.

As for the number of Scarpa's ganglion cells and the density of vestibular hair cells in the utricular macula and the LSC crista, there was no remarkable difference between affected ears and control ears (Table 2). The density of Type I hair cells in the saccular macula and the PSC crista seemed to be lower in affected ears than control ears. There was no remarkable difference in the density of vestibular hair cells between SDwV and SDwoV.

Case 3 was a SDwoV who died 10 months after the onset of SD. In this case, there was a remarkably large amount of deposit (otolith) attached to the cupula (Fig. 1). The otoconial membrane was atrophied and peeling off from the macula in the saccule. The collapse of the saccular membrane also was seen (Fig. 2).

In the clinical data (Table 3), there were 4 patients with CP (CP+) and 2 patients without CP (CP-) in SDwV. The CP+ were examined within 2 years from the onset of SD, and the CP- were examined after 5 years and 17 years from the onset. All 6 patients had a profound hearing loss, and the recovery was seen in 2 CP+ and 1 CP-. In SDwoV, there were 3 CP+ and 2 CP-, all of which were examined within 2 years from the onset of SD. Profound hearing loss was seen in 2 CP+ and 1 CP-, and recovery was seen in 1 CP+ and 1 CP-. Mild hearing loss was seen in 1 CP+ and 1 CP-, and no recovery was noted.

DISCUSSION

In the histopathologic findings of the cochlea, the atrophic change of organ of Corti, stria vascularis, and tectorial membrane and a significant decrease in the number of the spiral ganglion cells and cochlea nerve fibers have been reported in SD (3–8,12). These changes also were seen in the current study. It was reported that there were no remarkable histopathologic changes in the vestibular sense organs in patients with SD (3–8). Also, there was no remarkable histopathologic change in TBs of patients with SD in the current study, except for deposits on the cupula.

Khetarpal (9) has found no significant difference in the number of Scarpa's ganglion cells among SDwV, SDwoV, and controls. Our findings were consistent with Khetarpal's findings in this regard. We can speculate that the Scarpa's ganglion cells are not involved in the formation of vertigo in patients with SD.

Although several literatures reported the loss of vestibular hair cells in patients with SD (3–7), which was seen mostly in saccular macula, Khetarpal (9) has found no difference in terms of vestibular sensory epithelia. In the current study, the density of vestibular hair cells seemed to decrease on the saccular macula and the PSC crista in patients with SD. However, it is not the main reason for the presence of vertigo in patients with SD because these findings were irrespective of vestibular symptoms.

In the clinical data, all CP+ were examined audiologically within 2 years from the onset of SD, and there was no CP+ examined more than 2 years after the onset of SD in SDwV. On the other hand, there was CP+ even in SDwoV. It is suspected that the damage might be more or less accompanied to the vestibule in patients with SD and that the damage is large enough to cause the vertigo in SDwV. In fact, all SDwV had a profound hearing loss in the current study. There was not a clear relationship between the caloric weakness, the hearing level, and the recovery.

A recoverable structure that can cause the vestibular dysfunction is thought to be the lesion site of SD. Merchant et al. (8) investigated the TB of a SDwoV who died 9 days after the onset of SD, and swelling of the organ of Corti and small spherical tectorial membrane were reported. However, they did not report the vestibular findings. In Case 3, we can see a similar change in the cochlea, the damage of otoconial membrane, and cupula deposits. Otolith is usually covered with a supraotolithic cupular zone on maculae (13). Large amounts of cupula deposits seem to be the evidence of damage of supraotolithic cupular zones. All of the tectorial membrane, otoconial membrane, including the supraotolithic cupular zone, and cupula make up the extracellular superstructure covering each neuroepithelium (14). Therefore, we suspect that the extracellular superstructure, consisting of an acellular gelatinous membrane, is one of the lesion sites of hearing loss and vertigo in patients with SD. Although it is difficult to evaluate cupula histopathologically because of its structure, Suzuki (15) reported that the shape of the cupula was changed after injecting gentamicin into the perilymphatic space in the experiment using bull frogs. The atrophic changes of extracellular superstructures also change the reactivity of the semicircular canal. It is potentially one of the mechanisms of the persistent CP in SDwV.

CONCLUSION

The density of vestibular hair cells seemed to decrease on the saccular macula and the PSC crista; however, the number of Scarpa's ganglion cells did not change in patients with SD. There is no remarkable difference between SDwV and SDwoV.

Hearing loss and vertigo in patients with SD may be due to the damage of the extracellular superstructure, and it may potentially be one of the causes of persistent CP in patients with SD.

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FIG. 1. Ampulae of LSC and PSC in Case 3. This figure shows the crista and cupula of LSC on the affected (right) ear (A) and control (left) ear (B), and the crista and cupula of PSC on the affected ear (C) and control ear (D). Large amount of deposits (*arrow*) are attached to cupula in the affected ear. (Nomarski microscopy; original magnification, $\times 1,250$).

FIG. 2.

Vestibular sensory epithelia in Case 3. This figure shows the utricular macula on the affected (right) ear (A) and control (left) ear (B) and the saccular macula on the affected ear (C) and control ear (D). On the affected ear, otoconial membrane is atrophied (*arrow*) and peeling off from the saccular macula (*double arrow*) and the saccular membrane collapsed to saccular macula (*white arrow*). (Nomarski microscopy; original magnification, $\times 1,250$).

TABLE 1

Histopathologic findings

Case	Cochlea						Vestibular system				
	Hair cell			Supporting cell	Tectorial membrane	Stria vascularis	Portion	Deposit	Integrity of membrane	Cupula	
	Turn	Inner	Outer								
Affected ear	1	SDwV	Apical		2+	Damage	Atrophic	Thin	Sacculle	+	—
			Middle	loss	3+	Damage	Atrophic	Thin	Utricule	+	—
			Basal	loss	3+	Damage	Atrophic	Thin	LSC	+	Removed
	2	SDwV	Apical		1+			Thin	Sacculle	+	Removed
			Middle	loss	3+	Damage		Thin	Utricule		—
			Basal	loss	4+	Damage		Thin	LSC	+	
	3	SDwoV	Apical	loss	4+	Damage	Atrophic	Thin	Sacculle	+	Collapse
			Middle	loss	4+	Damage	Atrophic	Thin	Utricule		—
			Basal	loss	4+	Damage	Atrophic	Thin	LSC	+	Deposit
Control ear	1	SDwoV	Apical		1+		Atrophic	Thin	Sacculle	+	Deposit
			Middle		3+		Atrophic	Thin	Utricule		—
			Basal		4+		Atrophic	Thin	LSC		Deposit
	2	SDwoV	Apical		1+			Thin	Sacculle		—
			Middle		2+			Thin	Utricule		—
			Basal		3+	Damage	Atrophic	Thin	LSC		Removed
	3	SDwoV	Apical		1+			Thin	PSC		Removed
			Middle		2+			Thin	Sacculle		—
			Basal		2+			Thin	Utricule		—
3	SDwoV	Apical		1+			Thin	LSC	+		
		Middle		1+			Thin	PSC		—	
		Basal		2+			Thin	Sacculle		—	

Case	Cochlea				Vestibular system					
	Turn	Hair cell		Supporting cell	Tectorial membrane	Stria vascularis	Portion	Deposit	Integrity of membrane	Cupula
		Inner	Outer							
4	Middle		1+			Thin	Utricle		D ^a	—
	Basal		1+			Thin	LSC	+		Deposit
							PSC			
	Apical		2+				Sacculle			—
	Middle		2+				Utricle			—
	Basal		2+				LSC			
							PSC	+		

Outer hair cell: 1+, less than 30% loss; 2+, 30% to 60% loss; 3+, 60% to 90% loss; 4+, greater than 90% loss.

LSC indicates lateral semicircular canal; PSC, posterior semicircular canal; SDwoV, sudden deafness without vertigo; SDwV, sudden deafness with vertigo.

^aD: the processing damage.

TABLE 2

The number of Scarpa's ganglion cells and the density of vestibular hair cells

Scarpa's ganglion cells					Vestibular hair cells (/0.01 mm ²)								
Case			Superior	Inferior	Total	Saccular macula		Utricular macula		LSC crista		PSC crista	
						Type I	Type II	Type I	Type II	Type I	Type II	Type I	Type II
Affected ear	1	SDwV	10814	7995	18810	21.6	19.1	25.2	20	30.5	20.7	D ^a	D ^a
	2	SDwV	8439	D ^a		8.4	11.6	18.9	17.4	17.5	17.4	14.7	18.5
	3	SDwV	9170	8404	17577	19.3	18	28.6	21.3	33.6	18.9	21.8	15.7
	4	SDwV	9013	5455	14472	10.1	17.7	26	20.4	37.1	21.1	29.4	23.5
Control ear	1	SDwV	10527	7386	17914	27.5	18	24.4	23.5	31.5	23.2	D ^a	D ^a
	2	SDwV	9413	D ^a		12.6	13.7	16.8	14.4	14.7	19.6	29.4	17.4
	3	SDwV	10344	7969	18316	26.6	18.9	24.4	23.1	37.1	20.3	35	24.6
	4	SDwV	9970	5090	15064	24.6	17.7	27.7	23.1	35	19.6	32.6	19.6

^aD: the processing damage.

TABLE 3

Clinical data

	Examined date		Hearing	
			Pure tone average	
	<2 Years	>2 Years		R/NR
Caloric test (n = 11)	CP+	4	0	4
	SDwV (n = 6)		Profound	2/2
			Mild	0
				—/—
	CP+	0	2	2
			Profound	1/0
			Mild	0
				—/—
	CP+	3	0	2
	SDwoV (n = 5)		Profound	1/1
			Mild	1
				0/1
	CP+	2	0	1
			Profound	1/0
			Mild	1
				—/—

CP indicates canal paresis; NR, no recovery; R, recovery.