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Original Article

The Isolated Vestibular Anomaly with Normal Cochlea: An Analysis of Radiologic and Clinical Findings

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OBJECTIVE: The clinical reflection of isolated vestibular anomaly with normal cochlea is poorly understood due to the fact that isolated vestibules have been rarely reported to date. This study aimed to describe the radiological and clinical findings of isolated vestibular anomaly.

MATERIALS and METHODS: Eleven ears of eight patients with only a radiological diagnosis of complete fusion of the vestibule and lateral semicircular canal with normal cochlea and normal vestibular aqueduct on high-resolution computed tomography were identified. The clinical records were analysed.

RESULTS: Three patients were affected bilaterally and five patients were affected unilaterally. One ear was excluded due to chronic middle ear disease. The fused vestibule-lateral semicircular canal was associated with superior semicircular canal and/or posterior semicircular canal dysplasia in eight ears. The hearing capacity was as follows: normal in four ears; mixed hearing loss in four ears; conductive hearing loss in one ear; and sensorineural hearing loss in one ear. Vestibular tests were normal in seven patients. One patient had orthostatic hypotension.

CONCLUSION: Isolated vestibular anomaly may be associated with normal hearing, sensorineural hearing loss, conductive hearing loss, or mixed hearing loss, and may not cause imbalance.

KEY WORDS: Vestibule, semicircular canal, inner ear, anomaly, computed tomography

INTRODUCTION

The inner ear consists of the cochlea, the vestibular system, and the endolymphatic duct and sac. The vestibular system comprises the vestibule, with the saccule and utricle, and three semicircular canals (SCs; lateral, posterior, and superior), which are oriented perpendicularly to each other [1].

The association between sensorineural hearing loss (SNHL) and cochlear anomalies or large vestibular aqueduct is well known. However, isolated vestibular anomalies (IVAs) have been rarely reported to date, and for this reason the clinical reflections of these anomalies are poorly understood. A few reports on IVA state that this anomaly may be associated with normal hearing, SNHL, conductive hearing loss (CHL), mixed hearing loss (MHL), and imbalance [2-6]. This paper analyses the radiological and clinical findings of a group of eight patients with IVA, studied with high-resolution computed tomography (HRCT) of the temporal bone.

MATERIALS and METHODS

This study was approved by the ethic committee of Eskisehir Osmangazi University (12 June 2013, 80558721). Between 2007 and 2012, 11 ears in eight patients with only a radiological diagnosis of complete fusion of the vestibule and Lateral-semicircular canal (LSCC), without other associated inner ear anomalies on HRCT were selected to form a homogenous Group. The patients with other associated inner, middle, or external inner ear anomalies were not included, to better understand clinical outcomes of IVA.

All patients underwent temporal bone HRCT imaging performed with a 64-row multiple detector omputed tomography (MDCT) unit (Aquilion 64; Toshiba, Tokyo, Japan) using a 0.5 mm thickness, 0.5 mm collimation, 500 ms rotation time, 120 kV tube voltage, and a tube current of 300 mA. The images were retrospectively evaluated by a radiologist with 4 years of experience in head and neck radiology.

The clinical records of identified patients were analysed. All available audiometric and vestibular findings were recorded.

RESULTS

The age of the patients ranged between 2 and 25 years (M/F: 3/5, mean: 12.37±7.57 years). Six patients were of childhood age and two patients were adults. The main complaint of the patients was hearing loss, with the exception of one child patient (patient 2)

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who was referred for suspicion of middle ear disease. No patient complained of imbalance, vertigo, or tinnitus, with the exception of one patient (patient 7) who complained of imbalance. Seven patients were nonsyndromic. One child (patient 4) had several features of the CHARGE syndrome (truncus arteriosus, right renal agenesis).

Three patients were affected bilaterally and five patients were affected unilaterally. In 11 ears with IVA, the cochlea, vestibular aqueduct, and inner internal auditory canal were normal. One ear was excluded due to chronic middle ear disease associated with incus resorption. The superior semicircular canal (SSC) and posterior semicircular canal (PSC) were normal in two ears. The fused vestibule-LSC was associated with posterior limb dilatation of the SSC in three ears, with anterior and posterior limb dilatation of the SSC in three ears, with both the absence of PSC and posterior limb dilatation of the SSC in one ear, and with the absence of PSC and both anterior and posterior limb dilatation of the SSC in one ear (Figure 1).

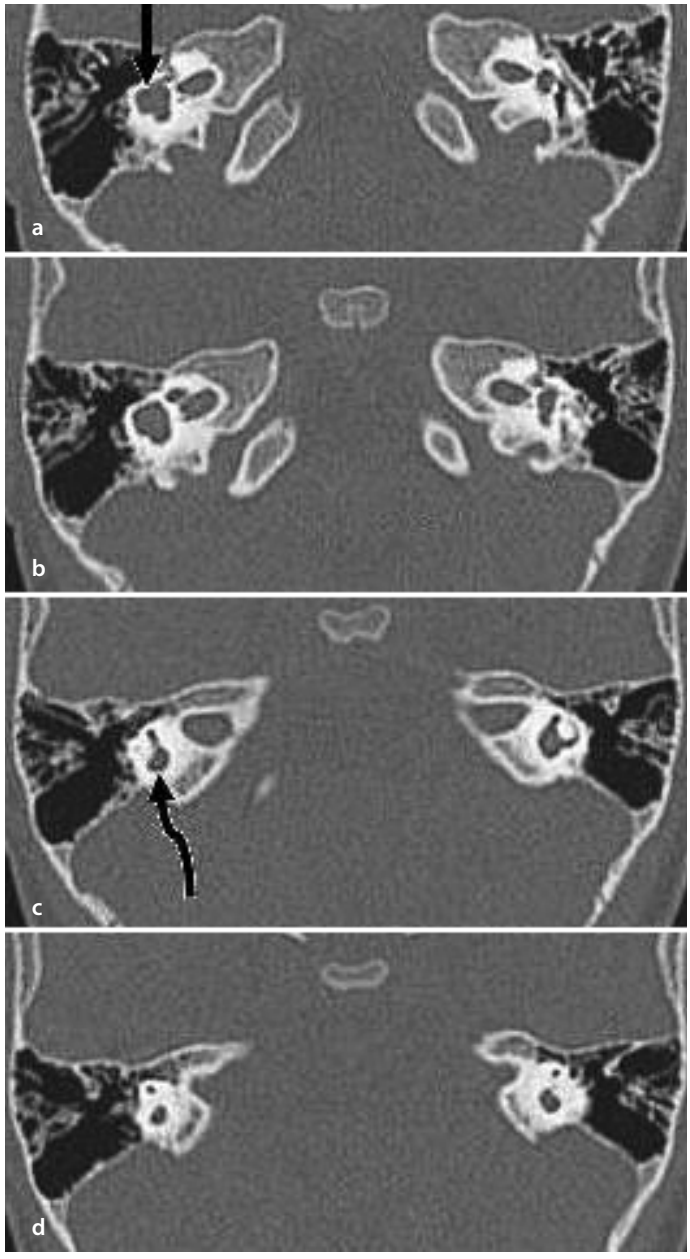

Figure 1. a-d. Consecutive temporal bone CT images from the caudal to cranial areas show complete fusion of the vestibule and LSC (straight arrow) on the right side. Both absence of the PSC and posterior limb dilatation of the SSC (*curved arrow*) are also shown

CT: computed tomography; LSC: lateral semicircular canal; SSC: superior semicircular canal

In the patients with unilateral IVA, contralateral inner ear findings were as follows: nearly completely fused cochlea-LSC and posterior limb dilatation in one ear; narrow internal auditory canal, narrow VII nerve entrance, with rudimentary LSC in one ear (Figure 2); and near completely fused cochlea-LSC and the absence of PSC and posterior limb dilatation of the SSC in two ears. According to HRCT imaging, only one patient with unilateral IVA had a completely normal inner ear on the contralateral side.

Although the bony labyrinth anomalies were similar to each other, different hearing loss types and levels were found among the patients (Figure 3). Hearing capacity of the patients was as follows: normal in four ears; profound MHL in two ears; moderate MHL in two ears; mild CHL in one ear; and profound SNHL in one ear.

Vestibular tests including video electronystagmography and rotational testing were normal in seven patients. Patient 7 was diagnosed with orthostatic hypotension after otological, neurological, and physical evaluation. A summary of each patient is provided in Table 1.

DISCUSSION

The inner ear arises from the otic placode in a process that begins early in the 3rd week of gestation. By the 8th week, the development

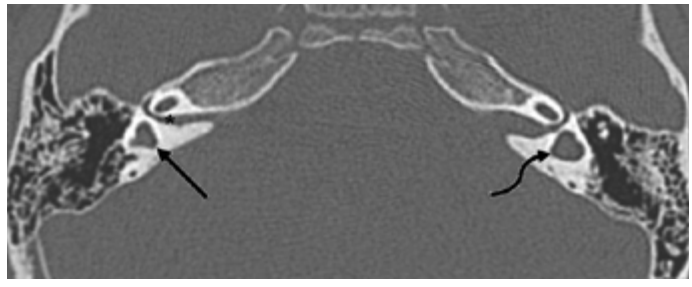

Figure 2. Axial CT image reveals fusion of the vestibule and LSC, forming a cystic lumen on the left side (curved arrow). On the contralateral side, there is both a narrow internal auditory canal and dysplastic vestibule-rudimentary LSC.

CT: computed tomography; LSC: lateral semicircular canal

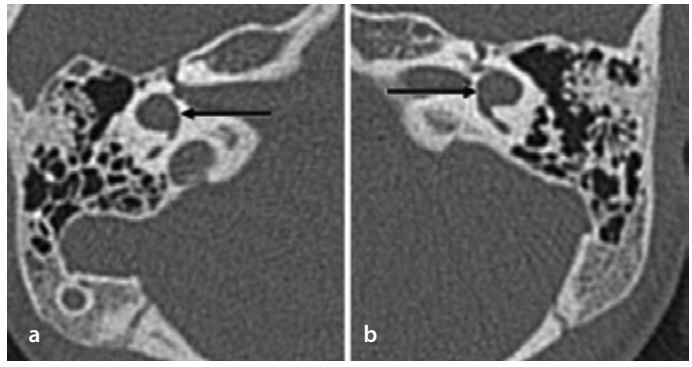

Figure 3. a, b. Similar malformations (*arrows*) are seen in the right ear (a) and the left ear (b) of patient 5. However, the right ear presented with CHL, whereas the left ear presented with normal hearing

CHL: conductive hearing loss

Table 1. Radiological and otological findings of each patient

PT No	Age	Sex	Radiological Findings: Fusion Side	Radiological Findings: Ipsilateral PSC And SSC	Radiological Findings: Contralateral inner Ear Findings	Audiological Findings at Isolated Vestibuler-SSC Anomaly Side
1	16	F	Right	SSC anterior and posterior limb dilatation		Profound mixed Type HL
			Left	SSC anterior and posterior limb dilatation		Profound mixed Type HL
2	10	F	Right	SSC posterior limb dilatation	Nearly completely fused vestibule-LSC and SSC posterior limb dilatation	Normal
3	7	M	Left	SSC posterior limb dilatation	Narrow internal auditory canal and narrow VII nerve entrance associated with rudimentary LSC	Normal
4*	7	M	Left	SSC posterior limb dilatation	Normal	Moderate mixed Type HL
5	12	F	Right	Normal		Mild conductive Type HL
			Left	Normal		Normal
6	2	F	Right	Absence of PSC, anterior and posterior limb dilatation of SSC	Nearly completely fused vestibule-LSC, absence of PSC, posterior limb dilatation of SSC	Profound sensorineural HL
7	20	F	Right	Absence of PSC, posterior limb dilatation of SSC	Nearly completely fused vestibule-LSC, absence of PSC, SSC posterior limb dilatation	Normal
8	25	M	Right	SSC anterior and posterior limb dilatation		Moderate mixed Type HL
			Left**	SSC anterior and posterior limb dilatation		Profound mixed Type HL

*The case with several features of the CHARGE syndrome (truncus arteriosus, right renal agenesis)
**This ear was excluded due to chronic middle ear disease associated with incus resorption

of the cochlea is complete. The vestibule is completely developed by the 11th week, and the semicircular canals, between the 19th and 22nd weeks; the lateral canal or duct is the last to form. Ossification of the labyrinth is complete by the 23rd week [1]. The LSC is the last complete ossification among the SCs and is the most susceptible to anomalous development. Disturbance of normal fusion and ossification may lead to a short LSC complex or to a persistent anlage of the LSC, with the lack of a central ossified bony island [4].

Vestibular malformations rarely occur in isolation [1]. Several terms to describe vestibular malformation, such as dysplastic, dilated, or enlarged vestibular anomalies are found in the literature. Commonly encountered anomalies include mild or globose dilatation of the vestibule, with nearly complete or complete assimilation of the LSC into the vestibule. The term 'fused vestibule' is used in this study to describe complete assimilation of the LSC into the vestibule. In order to form a homogenous Group, only fused vestibule anomalies were selected for this study.

The relationship between inner ear anomalies and hearing status has been extensively studied. However, there are few studies focused on anomalies of the vestibular system in the literature [2, 5-8]. Since the first report by Parnes and Chernoff [7], it has seemed clear that selective vestibular anomaly can coexist with a normal or near-normal cochlea. Jackler [9] has stated that ears with malformations that are limited to the vestibular system often have normal and or near-normal hearing. Normal hearing in vestibular-lateral semicircular canal dysplasia patients may be explained by the peripheral involvement of the vestibular system. However, in the largest series reported by Yu et al. [8], all patients with LSC anomalies had hearing loss. In the largest series reported by Johnson and Lalwani [5], four patients with isolated LSC malformation of 15 patients with LSC malformation with or without other inner ear malformation such as cochlear hypoplasia, large vestibular aqueduct presented either with SNHL or CHL. SNHL may be attributed to anomalous membranous labyrinth development [5]. The pathophysiology of CHL associated with IVA remains speculative. Jackler [9] concluded that SCC dysplasia appeared to be associated with CHL due to congenital stapes fixation. It was observed in this study that IVA could be associated with normal hearing and SNHL, CHL, or MHL in different patients. For example, the anomalies on the right and left sides in patient 5 were very similar to each other. However, the right ear presented with CHL, whereas the left ear presented with normal hearing. These differences in hearing between IVA ears may be explained by abnormalities of the bony labyrinth, but may not always correlate with them.

Dallan et al. [2] reported a patient with isolated LSC malformation with vestibular complaints and normal hearing. However, the majority of the patients in the current study did not present with vestibular complaints. The observations of Johnson and Lalwani et al. [5], who did not report vestibular findings in patients with isolated LSC malformation in the series, were confirmed in this study.

In conlcusion, isolated vestibular anomaly may be associated with normal hearing, SNHL, CHL, or MHL, and may not cause imbalance.

Ethics Committe Approval: Ethics committee approval was received for this study from the ethics committee of Eskisehir Osmangazi University Ethics Committee (12 June 2013, 80558721).

Informed Consent: Written informed consent was not obtained due to the retrospective nature of the study.

Peer-review: Externally peer-reviewed.

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Conflict of Interest: No conflict of interest was declared by the authors.

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REFERENCES

1. Joshi VM, Navlekar SK, Kishore GR, Reddy KJ, Kumar EC. CT and MR imaging of the inner ear and brain in children with congenital sensorineural hearing loss. Radiographics 2012; 32: 683-98. [CrossRef]
2. Dallan I, Berrettini S, Neri E, Casani AP. Bilateral, isolated, lateral semicircular canal malformation without hearing loss. J Laryngol Otol 2008; 122: 858-60. [CrossRef]
3. Emmrich JV, Fatterpekar GM. Dilated dysplastic vestibule: a new computed tomographic finding in patients with large vestibular aqueduct syndrome. J Comput Assist Tomogr 2011; 35: 674-8. [CrossRef]
4. Yamashita K, Yoshiura T, Hiwatashi A, Tuvshinjargal D, Kamano H, Inoguchi T, Honda H. Sensorineural hearing loss: there is no correlation with isolated dysplasia of the lateral semi-circular canal on temporal bone CT. Acta Radiol 2011; 52: 229-33. [CrossRef]
5. Johnson J, Lalwani AK. Sensorineural and conductive hearing loss associated with lateral semicircular canal malformation. Laryngoscope 2000; 110: 1673-9. [CrossRef]
6. Satar B, Mukherji SK, Telian SA. Congenital aplasia of semicircular canals 2003; 24: 437-46.
7. Parnes LS, Chernoff WG. Bilateral semicircular canal aplasia with near-normal cochlear development. Two case reports. Ann Otol Rhinol Laryngol 1990; 99: 957-9.
8. Yu KK, Mukherji S, Carrasco V, Pillsbury HC, Shores CG. Molecular genetic advances in semicircular canal abnormalities and sensorineural hearing loss: a report of 16 cases. Otolaryngol Head Neck Surg 2003; 129: 637-46. [CrossRef]
9. Jackler RK. Congenital malformations of the inner ear. In: Cummings CS, et al, (eds): Otolaryngology-Head and Neck Surgery, 2nd ed. Chicago: Mosby-Yearbook; 1993; 152: 9.