Comparative Analysis of CT and MRI Diagnosis of Large Vestibular Aqueduct Syndrome (LVAS) in Children

Ling Yang[1] and Jie Liu[2]

ABSTRACT

Objective: To analyse application value of CT and MRI in the diagnosis of large vestibular aqueduct syndrome (LVAS) in children.

Study Design: A descriptive study.

Place and Duration of Study: Department of ENT, Xiangyang Central Hospital, Affiliated Hospital of Hubei University of Arts and Science, China, from January 2014 to January 2018.

Methodology: Children with LVAS confirmed by CT, with different degrees of hearing loss and vertigo, less than 18 years, with complete clinical and imaging data, were included. Children under 2 years, with other serious diseases in organs, poor compliance, with other genetic diseases, were excluded. CT and MRI diagnosis data of 25 cases (50 diseased ears) with LVAS were retrospectively analysed. CT and MRI imaging results were observed. Midpoint and external aperture diameter of vestibular aqueduct in CT, MRI diagnosis were compared.

Results: CT images showed visible vestibular aqueducts in different degrees of enlargement and bone defect shadow in different shapes. Vestibular aqueducts were enlarged in the 3D image after VR reconstruction. MRI images showed higher signal of endolymphatic sac and endolymphatic ducts in varying degrees of enlargement, and endolymphatic sac was enlarged in the 3D image after VR reconstruction. Midpoint diameter of vestibular aqueduct was larger in CT than in MRI diagnosis ($p<0.001$), external aperture diameter of the vestibular aqueduct was smaller in CT than in MRI diagnosis ($p<0.001$).

Conclusion: LVAS children diagnosed as vestibular aqueduct enlargement by CT should receive MRI scan to further clarify the enlargement degree of endolymphatic sac and endolymphatic duct to increase the diagnosis rate.

Key Words: *Children, Large vestibular aqueduct syndrome (LVAS), CT, MRI, Diagnosis.*

INTRODUCTION

Enlargement of the vestibular aqueduct (EVA) is a vestibular aqueduct diameter greater than or equal to 1.5 mm in the median point or greater than or equal to 2 mm for the operculum.[1] Large vestibular aqueduct syndrome (LVAS) is a type of deafness characterised by fluctuating, and progressive hearing loss, with vertigo and/or tinnitus.[2] LVAS is one major cause of deafness in children and a congenital inner ear malformation with a high clinical incidence. The typical clinical manifestations of LVAS in children is progressive or fluctuating hearing loss after birth or at a young age. Mainly attacking bilaterally, the disease can be sudden or invisible, and can attack at any stage from birth to puberty, mostly during 3-4 years of age.[3,4]

LVAS in children involves complex pathogenesis, and is a refractory disease due to the lack of specific and effective methods for early diagnosis and treatment.[5,6] For the imaging of LVAS in children, some scholars merely analyse the axial image.[7,8] Although, high resolution CT scan document the presence of bilateral LVAS,[9] Okamoto *et al*. discovered that the CT and MRI findings of LVAS, *i.e*. the large vestibular aqueduct varied in size, and MRI scans could clarify the lymphatics and lymph sac.[10]

The aim of study was to compare and analyse the diagnosis of CT and MRI for diagnosis of LVAS in children.

METHODOLOGY

This study was a descriptive study conducted in the Department of ENT, Xiangyang Central Hospital, Affiliated Hospital of Hubei University of Arts and Science, China, from January 2014 to January 2018. The study was approved by the Hospital Ethical and Research Committee. Clinical data of 25 children (50 diseased ears) with LVAS were retrospectively analysed. Inclusion criteria were children with LVAS confirmed by CT (Axial CT of temporal bone measured vestibular aqueduct diameter >1.5 mm), with different degrees of hearing loss and vertigo, less than 18 years of age, with complete clinical and imaging data. Exclusion criteria were children under two years of age, with other serious diseases in organs, poor compliance, and other genetic diseases.

Department of ENT[1] / Radiology[2], Xiangyang Central Hospital, Affiliated Hospital of Hubei University of Arts and Science, Hubei Province, Xiangyang 441021, China

Correspondence: Jie Liu, Department of Radiology, Xiangyang Central Hospital, Affiliated Hospital of Hubei University of Arts and Science, Hubei Province, Xiangyang 441021, China
E-mail: rongjiang79319@163.com

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3.0-T MRI and HRCT examinations were performed in all patients. The children were given oral 10% chloral hydrate at a dosage of 40 mg/Kg per weight if they did not cooperate. Imaging examination was performed after the children fell sleep. CT examination was performed using low-dose spiral CT. The layer thickness and interlayer spacing were set at 0.45 mm, with screw pitch 0.562, 300 mAs, and 140 KV. After imaging acquisition, according to bone algorithm, bilateral reconstruction was performed by centering the promontorium tympani with a reconstruction interval of 0.3 mm. All data were transmitted to a supporting workstation for volume remodelling (VR) reconstruction to obtain high-resolution CT images. The optimal transversal image plane was selected for measurement of midpoint, external aperture diameter of the vestibular aqueduct. MRI examination was performed using 3.0-T magnetic resonance imaging instrument through 8-channel head phased array coil with scanning range above the orbitomeatal line. Scanning was in the sequence of transversal FSE T1WI (TR:525 ms, TE:15 ms), transversal and coronal FSE T2WI (TR: 4000 ms, TE: 200 ms), 3D-FIESTA (TR: 1200 ms, TE: 263 ms). The scanning layer thickness was set to 0.6 mm, and FOV was set to 180 mm x 180 mm. VR reconstruction was performed with the help of supporting workstation. The optimal transversal image plane was selected for measurement of midpoint, external aperture diameter of the vestibular aqueduct.

SPSS version 21.0 was used to analyse the imaging data. Measurement data such as midpoint, external aperture diameter of vestibular aqueduct obtained in CT and MRI diagnosis were expressed in $\pm$ standard deviation, and paired t-test was conducted. $P<0.05$ indicated statistically significant difference.

RESULTS

The 25 subjects (50 diseased ears) included 11 males (44.00%) with 22 (44.00%) diseased ears, and 14 females (56.00%) with 28 (56.00%) diseased ears. The age of onset varied from 2-9 years, average being 3.85 ±0.73 years. Seventeen (68.00%) cases showed progressive hearing loss in varying degrees at young age; five (20.00%) cases had history of significant trauma before the onset; three (12.00%) cases had a history of cold before the onset, with fluctuations in hearing changes.

CT scan showed deep triangular bone defect or trumpet-shaped, fish hook shaped or triangular bone defect shadow in varying degrees in the posterior edge of the bilateral petrous bone of the 25 children. With clear and sharp bone defect edge, outer terminus is directly connected to the vestibule or crus commune or immediately adjacent to its edge. Vestibular aqueduct enlargement was visible from 3D image after VR reconstruction by the workstation.

MRI scan showed different degrees of enlargement in endolymphatic sac and endolymphatic duct in the rear of the bilateral internal auditory meatus of the 25 children, which were capsular, elliptical, or arc-shaped, and mostly symmetric, while no obvious deformity was found in the cochlea. Most children had uniform signals, showing long T2 signals, high signal on T2 flair, and a few slightly shorter T1 signals after enlargement of endolymphatic sac and endolymphatic duct. Thickening was observed in endolymphatic sac of the front end. Endolymphatic sac enlargement was visible from 3D image after VR reconstruction by the workstation.

Midpoint diameter of vestibular aqueduct was larger in CT than in MRI diagnosis ($p<0.001$), external aperture diameter of vestibular aqueduct was smaller in CT than in MRI diagnosis ($p<0.001$, Table I).

DISCUSSION

LAVS is the most common inner ear malformation causing sensorineural deafness in children. About 81%-94% of children feature bilateral onset; whereas, unilateral onset is rare. The 25 studied children had bilateral disease. Most LVAS children manifest cold at the beginning of the disease, or have history of oral administration of aminoglycosides, and then show symptoms of dizziness, and tinnitus. Due to its hidden and sudden incidence, misdiagnosis or missed diagnosis is easy, if clinical diagnosis is not clear.[11,12] With the continuous development of modern medical imaging technology, high-quality imaging technology has enabled increased diagnostic rate of LVAS.[7,13]

The diagnostic advantage of CT lies in its capability in detailed observation of the ultrastructure of the bony vestibular aqueduct and the middle inner ear. By determining whether the vestibular aqueduct is enlarged from the transversal image in combination with unilateral multi-planar combination of different positions to accurately determine vestibular aqueduct structure and lumen, determine the inner diameter of the vestibular aqueduct of each section, and measure the inner diameter length, diagnosis can be improved and severity can be judged.[14,15] However, vestibular aqueducts can exist alone, or accompany other diseases such as inner

Table I: Comparison of midpoint and external aperture diameter of vestibular aqueduct in CT, MRI diagnosis .

Diagnostic method	Number of ears diseased	Midpoint diameter of the vestibular aqueduct (mm)		External aperture diameter of the vestibular aqueduct (mm)	
		Mean ±SD	p-value	Mean ±SD	p-value
CT	50	1.96 ±0.40	<0.001	2.91 ±0.21	<0.001
MRI	50	1.85 ±0.32		3.14 ±0.23	

ear deformities. The limitation of CT is that it cannot accurately show the enlarged endolymphatic sac and endolymphatic duct, so the lesion degree cannot be accurately determined.[16,17]

MRI can directly observe the internal and external structures of the endolymphatic sac, measure the broadening of the endolymphatic duct, while endolymphatic sac enlargement can be used as the main basis and sign for MRI diagnosis of LVAS. As the abnormal signal shadow close to the cerebellar hemisphere surface is visible from the axial view, which is in sharp contrast with the adjacent structure signal, missed diagnosis will not easily occur. Moreover, 3D-FIESTA features fast imaging speed, showing significant comparison of soft tissue.[18] Modern MRI imaging believes that multi-directional scanning can intuitively determine the condition of enlarged endolymphatic duct in LVAS, so whether there is abnormality in cochlea and inner ear can be observed through three-dimensional reconstruction.[19]

This study evisaged that CT images show visible vestibular aqueducts in different degrees of enlargement, and bone defect shadow in different shapes. The vestibular aqueducts are enlarged in the 3D image after VR reconstruction. MRI images showed higher signal of endolymphatic sac and endolymphatic ducts in varying degrees of enlargement, and endolymphatic sac is enlarged in the 3D image after VR reconstruction. The results of this study were basically consistent with previous studies.[20] Relevant data show that in case of vestibular aqueduct enlargement, the hyperosmotic fluid in the endolymphatic sac backflows to the basal gyrus and vestibule of the cochlear duct through the enlarged vestibular aqueduct and ductus reunions, damaging the sensory nerve epithelium of basal gyrus and vestibule of the cochlear duct, inducing degenerative changes in hair cells and then causing sensorineural hearing loss and dizziness.[21]

The vestibular aqueduct is a bony duct starting from the vestibular medial wall and is located between the vestibule and endolymphatic sac. The endolymphatic duct joins the endolymphatic sac under the auditory canal mouth in the posterior of the talus cones. There are many folds called wrinkled part in epidermis of the endolymphatic sac, which contains a large number of small blood vessels and connective tissues. The wrinkled part in the bone tube is an important site for endolymph absorption. The vestibular aqueduct and endolymphatic duct, endolymphatic sac play an important role in maintaining tiny pressure balance in the inner ear as a whole. Studies have shown that the area, external aperture diameter and length of vestibular aqueducts are positively correlated with the area of wrinkled part in the endolymphatic sac.[22] This fully demonstrates developmental parallel relationship, morphological consistency and functional dependence of the two. The shape and length of the vestibular aqueduct can indirectly reflect the status of the endolymphatic duct, endolymphatic sac. This study shows that the midpoint diameter of the vestibular aqueduct is larger in CT than in MRI diagnosis, the external aperture diameter of the vestibular aqueduct is smaller in CT than in MRI diagnosis, indicating that CT and MRI have respective advantages in diagnosing midpoint and external aperture diameter of vestibular aqueduct. It suggests that CT and MRI should complement each other in diagnosis of LVAS in children to reduce the rate of missed diagnosis and misdiagnosis.

CONCLUSION

Children diagnosed as vestibular aqueduct enlargement by CT should receive MRI scan to further clarify the enlargement degree of endolymphatic sac and endolymphatic duct, thereby increasing the clinical diagnosis rate, which will provide imaging basis for clinical diagnosis and treatment of LVAS in children.

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