CASE REPORT

UNVEILING A RARE CAUSE OF CONGENITAL BILATERAL SENSORINEURAL HEARING LOSS

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ARTICLE INFO	ABSTRACT
	Congenital anomalies of the cochlea are important cause of congenital sensorineural hearing loss in children. Cochlear anomalies are also known to be associated with vestibulocochlear nerve aplasia or hypoplasia. Such children are ideal candidates for cochlear implantation surgery. We report here a rare case of a 5year old female child who presented with bilateral hearing loss and was planned for cochlear implantation. She was referred for pre-operative imaging evaluation of temporal bone. HRCT temporal bone and MRI inner ear were performed, which revealed in complete partition type I with cochlear nerve aplasia on the right side and common cavity cochlear malformation with normal vestibulocochlear nerve on the left side. It is vital that cochlear and associated nerve anomalies must be recognized prior to surgery as this would predict the feasibility and outcome of the cochlear implantation.

INTRODUCTION

Congenital anomalies of the cochlea are important cause of congenital sensorineural hearing loss in children. The condition may occur as an isolated anomaly or as part of inner ear malformations associated with a number of syndromes and present with congenital sensorineural hearing loss (SNHL), which can be bilateral (Huang, 2011). This has to be treated early in life to enable adequate speech development. An important method of treatment is cochlear implantation, which is being performed with increasing success in recent years (Daneshi, 2003). The cochlear malformations are also known to be associated with vestibulocochlear nerve aplasia or hypoplasia. It is vital that this be recognized prior to surgery as this would predict a poor prognosis for cochlear implant and auditory brainstem implant (Shelton, 1989). We report here a rare case of incomplete partition type I with cochlear nerve aplasia on the right side and common cavity cochlear malformation with normal vestibulocochlear nerve on the left side.

Case Report

A 5year old female child presented with bilateral hearing loss, was referred to us by ENT surgeon for pre-operative imaging evaluation of temporal bone that has been planned for cochlear implantation. The child had initially been evaluated at 18 months of age for non-development of speech. There was no

history of congenital hearing loss in the family. Prenatal and perinatal periods were uneventful. The mother had been on regular evaluation and follows up during pregnancy by a qualified obstetrician and had received adequate supplements during pregnancy. No history of any significant illness during pregnancy was present. Birth was by normal institutional vaginal delivery and child was stable after birth. On examination, the child's development was normal for age, except for lack of development of speech. The child was not responsive to auditory stimuli. Otological examination revealed normal external and middle ear structures. Brainstem evoked response audiometry (BERA) showed no response to click audiometry at maximal stimulation of 100 dB in bilateral ears suggestive of profound sensorineural deafness. Otoacoustic emissions were not detected in bilateral ears. HRCT of the temporal bones was performed for evaluation of the inner ear structures. On right side, the inner ear revealed a rudimentary cochlea consisting of a cystic cavity without a bony modiolus separated from the dilated vestibule by a constriction suggestive of type I incomplete partition (Figure 1a). The left inner ear showed no definite cochlea, with a cystic space replacing it, which was continuous with the vestibule suggestive of a common cavity malformation (Figure 1b). The semicircular canals on the both sides were normal. The internal auditory canals (IAC) on both sides were normal in width (Figure 2). MRI of the bilateral inner ears was performed which revealed similar findings to the HRCT (Figure 3a & 3b). In addition, it revealed non-visualization of cochlear nerve with presence of vestibular nerves and the facial nerve in right-sided IAC (Figure 3c). The left side showed normal visualization of the cochlear nerve in the left

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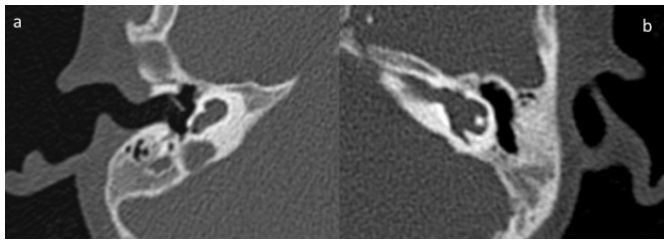IAC along with the left facial and vestibular nerves (Figure 3d).

Figure 1. A 5year old female child presented with profound congenital sensorineural hearing loss. Axial HRCT images of the temporal bone show incomplete partition Type I cochlear malformation in the right ear (a) and common Cavity cochlear malformation in the left ear (b)
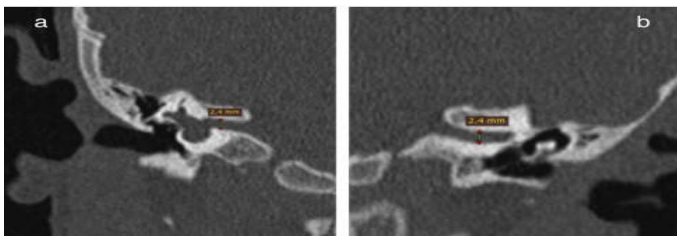

Figure 2. Coronal HRCT images of temporal bone of the same patient show that the right IAC (a) and the left IAC (b) are normal in width (> 2 mm)
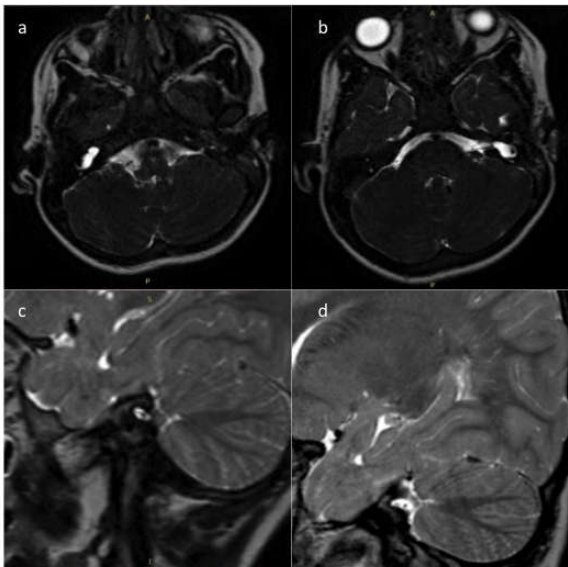

Figure 3. Axial (a&b) and sagittal (c&d) MRI DRIVE images show Incomplete partition Type I cochlear malformation in the right ear (a) and common Cavity cochlear malformation in the left ear (b). Right IAC in sagittal image (c) shows facial nerve seen anteriorly, vestibular nerves posteriorly with non-visualization of cochlear nerve anteroinferiorly. Left IAC in sagittal image (d) shows normal facial, cochlear nerves and Vestibular nerves

DISCUSSION

In children who are candidates for cochlear implantation surgery, imaging provides vital preoperative information about the inner ear, the vestibule cochlear nerve, and the brain. High resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) provide excellent delineation of the intricate anatomy of the inner ear. HRCT depicts the minute details of osseous structures, and MRI allows visualization of the fluid-filled spaces and the vestibule cochlear nerve. Together, these complementary modalities can aid decision

making about the best management strategy by facilitating the identification and characterization of inner ear malformations and any associated neurologic abnormalities (Joshi, 2012).
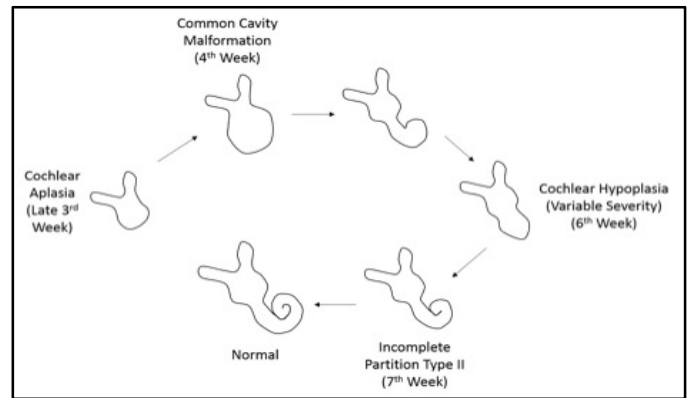

Figure 4. Schematic depiction of cochlear malformations in accordance to the week of insult in the intrauterine period

Table 1. Classification of cochlear malformations with week of insult, relative incidence and association with cochlear nerve aplasia

WEEK OF INSULT	INNER EAR MALFORMATION	RELATIVE INCIDENCE	A/W COCHLEAR NERVE APLASIA
Early 3rd week	Complete labyrinthine aplasia (Michael's aplasia)	1%	100%
Late 3rd week	Cochlear aplasia	3%	100%
4th week	Common cavity of the cochlea and vestibule	26%	100%
5th week	Incomplete partition Type I	24%	29%
6th week	Cochlear hypoplasia	15%	11%
7th week	Incomplete partition Type II	31%	0%

The normal cochlea develops during the 3rd to 8th weeks of pregnancy. Any insult during this period can lead to malformations of the cochlea and the labyrinth. The cochlear anomaly thus occurring depends upon the stage of development at which the insult occurs. The insult may be in the form of congenital infections such as rubella, cytomegalovirus infection, teratogenic drugs or due to genetic factors and syndromes (Bartel-Friedrich, 2007), Various classifications of congenital cochlear malformations exist, such as those proposed by Jackler (Jackler, 1987), Marangos (Marangos, 2002), Sennarogluand Saatci (Sennaroglu, 2002). A widely accepted classification in use is the one proposed by Jackler *et al*. in 1987, incorporating some modifications by Sennaroglu and Saatci, which classifies the congenital malformations of the inner ear based on the week of pregnancy at which the insult occurs or the timing of the developmental arrest. Starting from the third week, each week produces a characteristic malformation of decreasing severity as complete labyrinthine aplasia, cochlear aplasia, common cavity of the cochlea and vestibule, incomplete partition type I, cochlear hypoplasia or incomplete partition type II as depicted in Figure 4 and tabulated in Table 1 (Jackler, 1987; Sennaroglu, 2002 and Giesemann, 2012). Relative incidence of cochlear malformations reported by Jacker *et al*. is as incomplete partition defects 55%, common cavity 26%, cochlear hypoplasia 15%, cochlear aplasia 3%, complete labyrinthine aplasia 1% (least common), (Jackler, 1978), also tabulated in Table I. The relative incidence of incomplete partition as 24% Type I and 31% Type II out of the 55% incidence is based on extrapolation of the results of the study by Sennaroglu and Satacci (Sennaroglu, 2002). The vestibulocochlear nerves develop from the neuroblasts of the cochlear ganglion. The

neuroblasts of the cochlear ganglion disaggregate from the otic epithelium and the fibers of the cellular bodies of such ganglion grow from the outside into the otic epithelium and into the brainstem (Hemond, 1991). Hence the development of the vestibulocochlear nerve is closely related to the development of the cochlea itself. Giesemann *et al*. found that certain types of cochlear malformation were strongly associated with vestibulocochlear nerve aplasia or hypoplasia. In particular, complete labyrinthine aplasia, cochlear aplasia and common cavity malformation were found to be strongly associated. None of the subjects in their study with these malformations showed visualization of all four nerves in the IAC. Incomplete partition defect Type I and cochlear hypoplasia were also associated with vestibulocochlear nerve malformation, but to a lesser extent (29% and 11% respectively in their study). Incomplete partition defect Type II was not associated with cochlear nerve aplasia. These are also tabulated in Table 1 illustrating the Classification of cochlear malformations with week of insult, relative incidence and association with cochlear nerve aplasia (Jackler, 1987; Sennaroglu, 2002 and Giesemann, 2012) Internal auditorymeatal stenosis was also found to be strongly correlated with vestibulocochlear nerve aplasia.

This was found to be 100% in the study by Giesemann *et al*. (Giesemann, 2012). Internal auditory canal stenosis is considered when the vertical height of the canal is less than 2 mm (Baek, 2003). However, the nerve may be absent even when the IAC is completely normal and vestibulocochlear nerve deficiency can be definitively assessed only on high resolution MRI images (Adunka, 2006). In the present case, an incomplete partition defect Type I is found on the right side while a common cavity malformation of the cochlea and the utricle is found on the left side which indicates the timing of the insult as between 4th and 5th weeks of pregnancy according to Jackler's classification. Based on these findings a better result would have been expected from cochlear implantation on the right side than on the left. Also in accordance with Giesemann's findings (Giesemann, 2012), vestibulocochlear nerve aplasia would be expected much more on side (left) with common cavity malformation (100%) than on the side (right) with the incomplete partition type I (29%). However, MRI surprisingly revealed a cochlear nerve aplasia on the right side with a normal cochlear nerve on the left side. This means that a cochlear implant on the right side would have likely led to lack of response to auditory stimulus post implantation and failure of cochlear implantation on that side. Auditory brainstem implant would be an appropriate treatment in the setting of cochlear nerve aplasia (Shelton, 1989). In recent times, cochlear implantation is being increasingly performed in children with cochlear nerve deficiency. Although most children with cochlear nerve deficiency rarely achieve speech perception, many do benefit in terms of auditory awareness (Buchman, 2011). Complete absence of cochlear nerves is, however, a clear predictor of poor cochlear implantation candidacy as in the present case in the right ear, however in the setting of cochlear nerve hypoplasia, stimulation of the even a small number of nerve fibers may provide beneficial given the plasticity of the auditory cortex in young children (Kutz, 2011).

Conclusion

HRCT and MRI are important modalities for assessment of congenital malformations of the inner ear and the radiologist plays an important role in the assessment and prognostication of the post-surgical auditory outcome in these patients prior to cochlear implantation. While HRCT enables assessment and classification of the osseous malformation and inference of the possible timing of the intrauterine insult, associated vestibulocochlear nerve malformations can only be assessed on MRI. Further, in rare instances, vestibulocochlear nerve malformation can be present on the side of the less severe malformation among, as is the case here. This underlines the importance of performing MRI for assessment of the vestibulocochlear nerve in all cases of cochlear malformations detected on HRCT prior to cochlear implantation.

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