CASE REPORT

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Cochlear implantation in a child with cystic cochleovestibular malformation: a case report

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ABSTRACT

Background: The inner ear and internal auditory canal (IAC) malformations account for 20% of congenital sensorineural hearing loss in children. The cystic cochleovestibular malformation is one of the congenital malformations in the cochlea, in which the cochlea is empty, unpartitioned, and cystic. The dimension of the cochlea is normal. This presents a significant challenge even to the most experienced clinicians because of difficulty in the surgery with facial nerve anomaly and gusher, choice and placement of electrode, and increased risk of meningitis after the procedure. This study aims to report the procedure of cochlear implantation in one case of cystic cochleovestibular (IP type I) malformation, which still became a challenge in the otology field.

Case Presentation: We reported one case of 5 years old boy with bilateral cystic vestibular malformation who underwent cochlear implantation on the left ear. The patient was diagnosed with bilateral profound congenital sensorineural hearing loss. He already used hearing aids on both ears but gained limited improvements. Peri-operative perilymph gusher was happened and was stopped by plugging the cochleostomy hole using fascia. The patient showed good condition after the procedure. Although facial nerve lesion occurred after the operation, it was improved by giving steroids. There is no sign of cerebrospinal fluid leaks such as rhinorrhea, otorrhea, or meningitis.

Conclusion: This report showed good outcomes following cochlear implantation in cystic cochleovestibular malformation.

Keywords: cystic cochleovestibular malformation, cochlear implantation, challenge, perilymph gusher.

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INTRODUCTION

Cochlear implant surgery is one of the best amongst the various management options available in children and adults with severe to profound sensorineural hearing loss.[1] About 20% of children with sensorineural hearing loss (SNHL) have associated temporal bone malformation. Increased experience in cochlear implantation has led to more children with abnormal cochlea-vestibular anatomy submitted to this procedure.[2]

The incidence of congenital sensorineural hearing loss in children varies from 1:1,000 to 1:2,000 depends on the observed population.[3] The study conducted by Aldhafeeri AM and Alsanosi AA in 2013 found that from a total of 316 patients who underwent cochlear implantation, they found inner ear malformation in 24 patients, with a prevalence of 7.5%.[4] In 2010, reported by Sennaroglu L the subtype and frequency of cochlear anomaly: 6% of Michel deformity, 5% of cochlear aplasia, 8% of common cavity malformation, 12% of cochlear hypoplasia, 41% of IP type I, 20% of IP type II and 2% of IP type III.[5]

Inner ear development started in the third week of gestation through the thickening of the hindbrain forming the otic placode.[6] The invagination of the otic placode becoming an otic pit that closes on its surface to form an otic cyst.[6] In the fifth week of gestation, the otic cyst divides itself into a vestibular and cochlear pouch. The cochlear pouch form the cochlear duct and saccule, the vestibular pouch is forming the endolymphatic duct, utricle, and semicircular canals.[6] The cochlear membrane will reach the 1-1.5 turn at the end of the sixth week of gestation and reach the 2.5 turns at the end of the seventh week of gestation. The semicircular canals will begin to form from the utricle segment of the otic cyst in the seventh-eight week of gestation. The inner ear structure will have an adult structure at the end of eight weeks of gestation.[6]

In 1987, Jaekler et al. proposed a classification of cochleovestibular malformations based on polytomography and related to embryological genesis.[6] More recently, in 2002, Sennaroglu L and Saatci I suggested an extension based on computed tomography (CT) findings and provided a detailed classification of cochlear malformations, which is particularly important in the field of cochlear implantation.[7] The malformation known as Mondini deformity was defined as incomplete partition (IP) and two types of IP were described by the authors: IP type I and type II.[7] Recently, X-linked deafness has been recognized as a third type of IP: IP type III.[7]

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IP type I is described as "cochlea-vestibular malformation." In this anomaly, there is a clear differentiation between cochlea and vestibule.[8] The cochlea is located in its usual location in the anterolateral part of the fundus of the IAC. It lacks the entire modiolus and interscalar septa, giving the appearance of an empty cystic structure.[8] External dimensions (height and length) of an IP-I cochlea are similar to the normal cochlea. An enlarged, dilated vestibule accompanies cochlea. Vestibular aqueduct enlargement is very rare.[8] There may be a defect between the IAC and the cochlea due to the CA's developmental abnormality and the absence of the modiolus and CSF may fill the cochlea.[8]

Cochlear implantation (CI) is an effective rehabilitation method for profoundly hearing-impaired patients who do not benefit from hearing aids.[1] It is a multi-component electronic device that provides auditory information by direct stimulation of auditory fibers in the cochlea. IP type I patients suffer from profound SNHL and gain little benefit from traditional hearing aids. Thus, cochlear implantation is an option for these individuals.[8]

A cochlear implant is an electronic device that works to replace the transducer system of non-functioning inner hair cells by converting the mechanical energy of sound into electrical signals delivered to the cochlear nerve.[9] The cochlear implant mechanism starts from capturing sound impulse by microphone then is forwarded to the speech processor through the connecting wire.[9] The speech processor will select the appropriate voice information and convert it into sound, delivered to the transmitter. The sound code will be converted into an electrical signal and sent to the corresponding electrodes inside the cochlea, causing nerve fibers' stimulation. Cochlear implants' success is determined by assessing hearing ability, increasing vocabulary, and understanding of language.[10]

During the operation, the most common complication was perilymph gusher, whether temporary or permanent, facial nerve injury, or wrong placement of the electrode into the inner ear canal in patients with fundus defect.[11] After cochleostomy, the perilymph or cerebrospinal fluid leakage happened because of bone defects on the inner ear canal's lateral end. These barriers could undergo deficiency or incomplete malformation in congenital ear dysplasia patients, which cause a meeting of perilymph and cerebrospinal fluids.[11,12]

Cerebrospinal fluid leakage, which occurred peri-operative on the cochlear malformation patients, were reported from a previous study. Buchman et al. found perilymph fluid leakage from cochleostomy in 6 patients from 28 patients with inner ear malformation. All the leakage was stopped using fascia or temporal muscle.[13] Based on those mentioned above, this case study aims to evaluate the cochlear implantation in a child with cystic cochleovestibular malformation.

CASE DESCRIPTION

A two years old male child reported with the complaint of delayed speech and language. The patient never complained of dizziness, brain infection history, and the history of teratogenic substance exposure during pregnancy. There is no family history of hearing impairment in the family. Baseline assessment revealed bilateral profound hearing loss on condition play audiometry, bilateral type A tympanogram on impedance measurement, and absent otoacoustic emissions. Findings were correlated with electrophysiological assessment using Auditory Brainstem Response (ABR), which revealed bilateral profound hearing loss. The patients than using hearing aids on both ears for 3 years and went to speech therapy.

After 3 years, there is an improvement in the hearing and speech ability, which is better on the right ear than the left ear. This shows in the pure tone audiometry report on the right ear, the hearing threshold is 38.75 dB, while the left ear is 48.75 dB. This result convinces the parents to consider cochlear implantation. Radiographic investigations included Magnetic Resonance Imaging (MRI) and Computerized Tomography (CT) was done. Their findings revealed cystic cochleovestibular

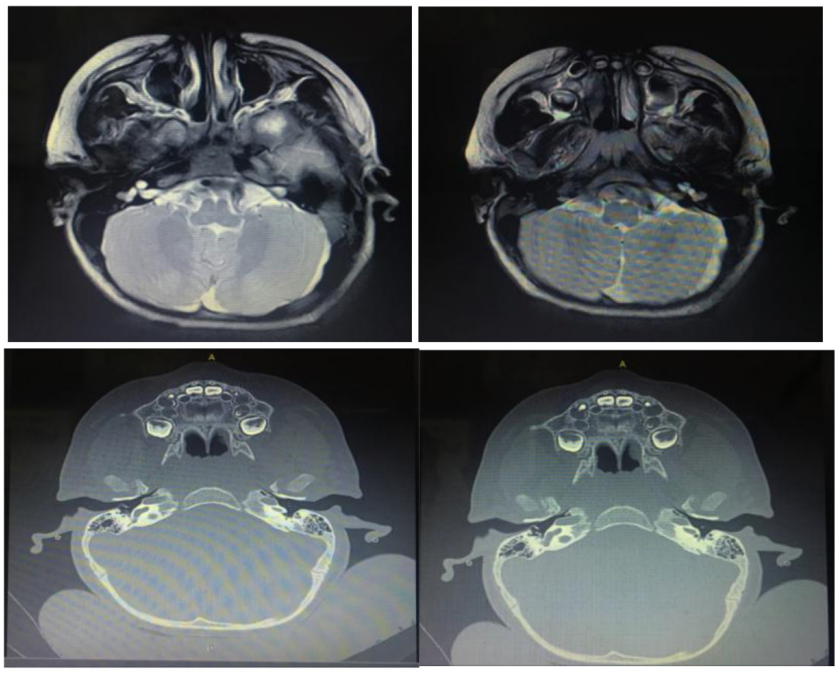

Figure 1. The MRI and CT showing the cystic cochleovestibular malformation on both ears

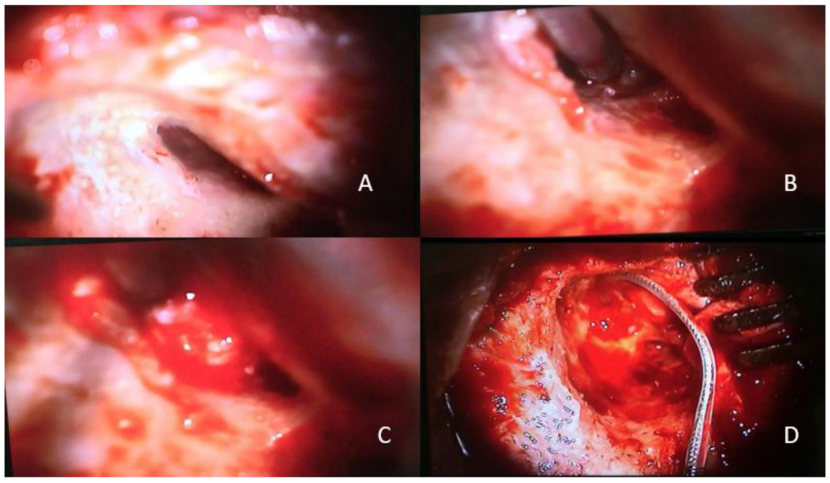

Figure 2. (A) Gusher from cochleostomy; (b) insertion of the electrode; (c) and (d) plugging using fascia and temporal muscle

malformation, an Incomplete Partition type I (IP-I) in both right and left ear. The result is dysplastic cochlea with a cystic appearance to the basal turn and apex. The vestibule demonstrates a globose appearance with the assimilation of the lateral semicircular canals, the vestibular aqueduct is not dilated, and mildly bulbous appearance to the IAC.

The left ear was implanted at the age of 5 years 1 month. The implant with 19 electrodes was inserted successfully during operation. A perilymph gusher happened during operation, which showed clear liquid from cochleostomy, which filled the tympanic space. After 10 minutes, the amount of liquor was decreased but not stopped entirely. Electrode insertion was successfully done in conjunction with the suction of the liquor. The gusher was stopped by plugging the cochleostomy and tympanic space obliteration using fascia and temporal muscle. Neural Response Impedance (NRI) post-implantation shows the good result and all of the electrodes are inserted successfully into the cochlea.

The post-operation patient was given antibiotics and analgesics. One day after the operation, there was facial paralysis. This complaint was improved by using steroids. This child underwent regular speech therapy for around 6 months post-implantation with regular mapping. Auditory verbal therapy and structured auditory training were the clinician's primary treatment strategies during the therapy sessions, and the mother was instructed to train the child with the same target goals during home training.

After 6 months of therapy, this patient was evaluated using the conditioned play audiometry, which shows improvements of the hearing threshold on the left ear of 31,25 dB. The speech ability is evaluated with CAP II and MSLDS. The child achieved a score of 7 (use of the phone with familial speaker) from a maximum score of 9 on CAP II by 6 months post-implantation, while achieved a score of 8 (able to use complex grammar and sentence structure) from a maximum score of 10 on MSLDS.

DISCUSSION

The IP-I patient in this report is a child with hearing impairment since birth. The patient never complained of dizziness or brain infection history. This is consistent with the literature where the sensorineural hearing loss is the main complaint in IP-I malformation but not always accompanied by vertigo. Clinical evaluation of the patient confirms the congenital sensorineural hearing loss with typical laboratory results and cystic cochleovestibular malformation (IP-I) on both ears, as seen in the CT and MRI.

History of teratogenic substance exposure during pregnancy and family history of hearing impairment are denied. Various syndromes associated with cystic cochleovestibular malformations were also not found in this patient. However, teratogenic or genetic factors are still suspected, although in literature, 30% cause of this malformation is idiopathic.[13] A study conducted by Philippon et al recommended early CI for patients with profound bilateral deafness secondary to meningitis owing to the risk of labyrinthitis ossificans.[14] Blamey et al reported below-average post-CI speech perception in post lingually deaf adults with meningitis, perhaps due to reduced ganglion cells' survival.[15]

The existence of a critical period for language development in children has been well documented. The best time to learn a language is during the first 5 years of life.[16] In terms of language development, the crucial period for cochlear implantation in children with profound SNHL is <3.5-5 years of age.[16] They considered that even adults with prelingual deafness significantly improved speech perception scores after CI compared with preimplantation scores in the study by Teoh et al. as much more did CI during the critical period.[15] These patients underwent cochlear implantation at the age of 5, so they were still critical when they underwent implantation. At the time of CI, age affects speech perception in children after CI, and younger children displayed more rapid post-CI improvement than older children in one study. A study by Kang DH et al. also shows no effect of delayed CI on post-CI speech perception.[17]

A perilymph gusher is the pulsatile egress of clear fluid for up to 1 minute during cochleostomy.[18] It differs from perilymph leakage, commonly occurs after cochleostomy associated with CI,

and is generally non-pulsatile. The likelihood of a perilymph gusher increases when cochleovestibular anomalies are present.[18] However, a study conducted by Adunka OF et al suggest that the type of labyrinthine anomaly has a more significant effect on postoperative performance than the presence or absence of an intraoperative perilymph gusher.[19] Therefore, rather than concluding that a perilymph gusher has a significant impact on speech perception after CI, the correlation between it and auditory perception in patients with inner ear anomalies should be explored. Incomplete electrode insertion may closely correlate with poor post-CI speech perception.

Although perilymph gusher incidence is infrequent, if not handled properly will cause complications of meningitis. This patient did not show cerebrospinal fluid leakage as rhinorrhea or otorrhea and not showing signs of meningitis post-operation. This indicates that the cochleostomy plugging was a success, although the risk of leakage is still possible.[20]

Numerous inner ear anomalies cause unwanted postsurgical outcomes. They are associated with the cochleostomy site, inadequate electrode placement and stability, facial nerve injury, perilymph leakage, and post-CI meningitis. It is universally accepted that children with more severe inner ear anomalies have poorer hearing outcomes after CI than those with less severe abnormalities. In this case, the patient could develop good speech perception, although he suffers from IP-I. After 6 months of evaluation, this patient was already showing good progress, where he could speak with better pronunciation and vocabulary than before the operation. The patient's speech ability based on MSLDS is in level 8, which can use complex grammar and sentence structure. While the patient's hearing ability is on level 7, which patient could use a phone with familiar speakers. A study by Papsin in 2005 reported on 42 patients with IP malformation, obtained that speech perception score in the IP malformation patients are higher than patients with other inner ear malformation like common cavity and cochlear hypoplasia because there is more nerve fiber, but there are no differences in IP-I and IP-II.[21]

Buchman et al. also report a better result in patients with IP malformation than other malformation without differentiating between two types of IP.[13] But, Kontorinis G et al. reported that there is better result in IP-I patients than IP-II.[22]

CONCLUSION

We reported one case of congenital sensorineural hearing loss with a cystic cochleovestibular malformation in 5 years old child who underwent cochlear implantation on the left ear. Intra-operative founded perilymph gusher and was plugged using fascia and temporal muscle, and electrodes were successfully inserted. Six months of evaluation after implantation, the patient able to develop better speech perception than preimplantation.

CONFLICT OF INTEREST

The authors declare that there is no competing interest regarding the manuscript.

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AUTHOR CONTRIBUTION

All of the authors equally contribute to the study from case selection and administer cochlear implantation in a child with cystic cochleovestibular malformation until reporting the case study outcome.

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