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CASE REPORT

Cochlear implantation in cases with incomplete partition type III (X-linked anomaly)

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Abstract Incomplete partition-type III anomaly (X-linked deformity) is no common finding in a prospective candidate for cochlear implantation. In this paper, the problems about the cochlear implantation in cases with incomplete partition-type III anomaly (X-linked deformity) and profound sensorineural hearing loss is discussed. High-resolution multidedector computed tomography (MDCT) and magnetic resonance imaging were performed preoperatively in all patients. MDCT revealed that there was bulbous dilatation at the lateral ends of internal auditory canals (IAC) in all patients. There were also enlargements of labyrinthine segments of facial and superior vestibular nerve canals. Patients with the basal turns of cochlea incompletely separated from IAC were also presented. Patients with IP-type III (X-linked deformity) and profound SNHL were implanted. Standard transmastoid-facial recess approach was used and cerebrospinal gusher was encountered after the cochleostomy in all cases. Postoperative performance was very good in all patients. Two patients had complications, which are facial nerve stimulation and device failure. Both patients were reimplanted. Cochlear implantation is a good choice in the patients with IP-type III. However, this anomaly may have special potential risk than the other inner ear abnormalities. Therefore, the surgeon should be aware of them and must be ready to inform the patient and parents.

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Introduction

The interest of the otologists, scientists and radiologists in the inner ear has increased since the initiation of the studies on cochlear implantation. Several reports on various topics related to cochlear implantation have been published. One is on cochleovestibular malformations. The type of the malformations is highly important for the cochlear implantation. Total absence of inner ear is a contraindication whereas cochlear implantation is very successful in the case with a mild inner ear deformity.

Several classification systems based either on politomography or on high-resolution computerized tomography have been proposed [6, 10, 12, 14, 18]. X-linked deformity (the most widely known name) or incomplete partition type III (IP-type III),which has pathagnomonic computerized tomography (CT) findings, has not been included in them, except for Phelps' classification. This might be due to the rarity of the anomaly. Phippard addressed this anomaly as pseudo-Mondini stage II and described as partial hypoplasia of the cochlea, stapes fixation and dilated internal auditory meatus (IAM) and abnormal communication with the base of the cochlear duct [13].

Different genotypes and phenotypes were described in the literature under the name of X-linked deafness though patients demonstrated almost identical findings on high-resolution computerized tomography (HRCT). Mixed hearing loss, stapes fixation and CSF gusher during the stapedectomy is the most common forms of X-linked deafness

(DNF3). Nance et al. were the first to describe that this was transmitted as an X-linked trait in 1971 [8].

There have been numerous reports about the cochlear implantation in the cases with inner ear anomalies. However, there is no report of cochlear implantation in the patient with IP-type III to the best of our knowledge. In this paper, four cases with IP-type III, (X-linked anomaly), who received multichannel cochlear implant are reported.

Case reports

Case 1

A three and half year old girl with a congenitally profound sensorineural hearing loss, having parents with neither consanguinity nor hearing loss, and having a brother with mild mental retardation, paraparesis, agenesis of the corpus callosum and normal hearing, attended the hospital. Her preoperative radiological evaluation including temporal bone computerized tomography and magnetic resonance images (MRI) was reported as normal. In May 2000 she underwent cochlear implantation on the right ear using a Clarion 1.2 implant with HiFocus (Model AB-5100H)© multichannel cochlear implant. During the operation, cerebrospinal gusher was encountered. It lasted 15 min with the elevation of the head. Then a cochlear implant was placed and cochleostomy was sealed with pieces of muscles. Cerebrospinal gusher led us to reevaluate preoperative CT scan. However, specific details cannot be identified due to inappropriate technical parameters; only the vestibular aqueduct was seen larger on this CT. The patient's postoperative course was uneventful. She was doing very well with cochlear implant and was attending mainstream. In 2005, facial nerve stimulation started on the implanted side. All attempts on the programming system to eliminate facial nerve stimulation were unsuccessful and she could not use cochlear implants because of facial nerve stimulation. High-resolution multidedector computed tomography (MDCT) was performed to see the location of the electrodes. MDCT scan confirmed the correct electrode placement in the cochlea, but electrode array protruded to labyrinthine canal from the second turn of cochlea, which might be due to local dehiscence or weakness of the otic capsule, and this could lead to facial nerve stimulation (Fig. 1). Moreover IP-type III anomaly (X-linked anomaly) with the bulbous dilatation at the lateral ends of internal auditory canals (IAC) on both sides was diagnosed. Although she has used her conventional hearing aids on the left side, her speech development started to get worse. After the parental counseling, cochlear implantation was planned for the left ear. Due to delay of parental decision and official procedures, the operation took place a year after the

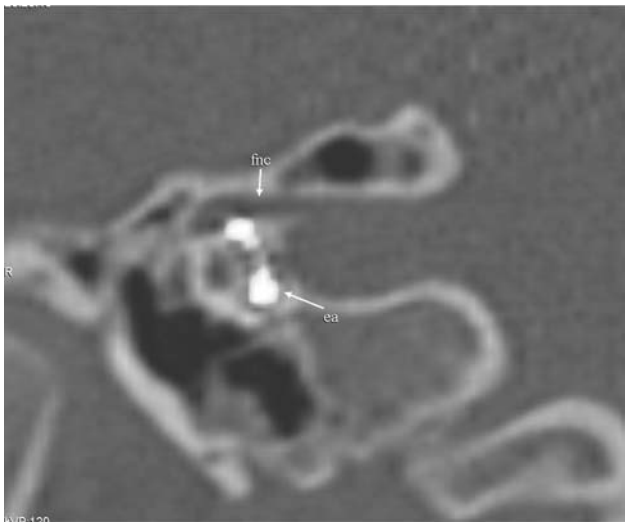

Fig. 1 Electrode array protrusion to labyrinthine canal from the second turn of cochlea in case 1. *ea* electrode array, *fnc* facial nerve canal

starting of facial nerve stimulation. During the operation, stapes was found fixed. Round window niche could not be identified clearly; therefore, cochleostomy was performed 2 mm inferior to the stapes tendon over the promontory. Cerebrospinal gusher ending in 15 min was encountered. Advanced Bionics HiRes™ 90 K© multichannel cochlear implant was placed and cochleostomy was sealed with pieces of muscles and bone dust. Immediately after the operation, she had dizziness, which was resolved on the second day. On the same day, she developed rhinorea and subfebril fever. In two days, rhinorrhea resolved with only conservative management such as strict bed rest, head elevation and reduced activity.

Case 2

A 19-year-old boy having a brother (case 3) with the same anomaly, presented with progressive sensorineural hearing impairment. He was born after a full-term uneventful pregnancy. The parents were not related. Hearing loss was recognized at 12 months and he started using bilateral conventional hearing aids and attended the parents' guidance sessions in a rehabilitation center for hearing impaired children. He graduated from primary and secondary schools in mainstream. Since his hearing loss has started to get worse he was evaluated to be a possible candidate for cochlear implantation at the age of 19. His audiological tests showed profound hearing loss in both ears. Incomplete type III (X-linked anomaly) deformity was diagnosed with preoperative radiological evaluation. He underwent cochlear implantation in the same year with the standart transmastoid-facial recess approach. In the middle ear, stapes had only anterior crura. Profuse cerebrospinal gusher was encountered at the opening of endostium. It ended with

the elevation of the head. Then Clarion 1.2 implant with HiFocus (Model AB-5100H)© cochlear implant with the positioner was placed and cochleostomy was sealed with pieces of muscles and bone dust. The postoperative course was quite stable. Postoperative Stanvers view showed that all electrodes were in the cochlea. His postoperative performance was very good. He could talk to the known people on telephone. In 2006, after 4 years, because of the device failure, reimplantation surgery was planned on the same side. After removing the previous device, the cochlea was found totally obliterated with fibrous tissue. The new implant could not be introduced into the cochlea on this side. Therefore, implantation was performed on the left ear in the same session, since the permission of the parents and patient has already been taken before the operation. Cerebrospinal gusher was also encountered. Elevation of the head was adequate to encounter a clear field, then Advanced Bionics HiRes™ 90 K© multichannel cochlear implant was introduced without any difficulty. Postoperative conventional radiography showed normal location of the implant.

Case 3

A 17-year-old boy was presented with progressive sensorineural hearing impairment. He was the brother of the second case. He was born after a full-term uneventful pregnancy with a cleft lip and a cleft palate. The moderate-to-severe hearing loss was diagnosed at 9 months and he had been using bilateral hearing aids since then. The operation for cleft lip and cleft palate was performed at 9 and 10 months, respectively. He attended a special school for hearing impaired with a strong aural/oral education. Then he graduated in primary and secondary education in mainstream. Since profound hearing loss was diagnosed at the age of 17, he underwent cochlear implantation. MDCT before the operation showed us the same inner ear abnormalities like that of his brother. Standart transmastoid-facial recess approach was used. Profuse cerebrospinal gusher, which lasted from 15 to 20 min, was encountered during the cochleostomy. Then Clarion 1.2 implant with HiFocus (Model AB-5100H) cochlear implant with the positioner was placed and cochleostomy was sealed with pieces of muscles and bone dust. Postoperative radiography was taken instead of intraoperative radiography because of the lack of the equipment in the operation theatre. The evaluation indicated that the electrodes in the cochlea extended into the IAC approximately upto 1 mm. Electrodes were pulled back slightly in the second operation after two days from the first operation. Little CSF oozing which lasted 2–3 min with head elevation was encountered. Postoperative Stanvers view confirmed that electrodes were within the cochlea without entering into the IAC.

Case 4

The patient was a two and half year old boy, born after a full term uneventful pregnancy. The profound sensorineural hearing loss was diagnosed in 14 months and he started using conventional hearing aids in both ears. No consanguinity was noted and no other family members had hearing loss. Since there was no benefit from the hearing aids, he was considered to be a candidate for cochlear implantation. During the evaluation procedure, MDCT showed us typical findings of IP-type III anomaly. He underwent operation on August 2006. Profuse CSF gusher was encountered. After the elevation of the patient's head, gusher resolved. Nucleus© CI24 (RE) implant was introduced into the cochlea and full insertion was achieved. Neural response telemetry was obtained. Postoperative course was uneventful.

Radiological findings

All patients underwent a high-resolution multi-dedector computed tomography (MDCT) (Toshiba Aquilion 64, Toshiba Medical Systems Corp. Tochigi-ken, Japan) in axial plane, with 120 kV, 300 mA and 240 mm FOV (field of view) settings. The slice thickness was chosen 0.5 mm. for each patient and slices were obtained with 0.641 pitch factor, while the helical pitch was set to 41. All images were reconstructed with bone algorithms in order to make proper otic capsule evaluations. High-resolution MDCT slices demonstrated that there was bulbous dilatation at the lateral ends of internal auditory canals (IAC) in all patients (Fig. 2). There were also enlargements of labyrinthine segments of facial and superior vestibular nerve canals. Singular nerve canals were also shown to be prominent and the canals were strikingly long due to proximal orifice formation. MDCT of all patients indicated that the basal turns of cochlea were incompletely separated from IAC and seemed to be a continuation of the IAC. Modioli were completely absent in all patients. Vestibular aqueducts (VA) showed great variation, but all were large and symmetrical in fashion. In none of the patients were VA's orifices large but they become cystic or enlarged from the middle parts to the distal ends near the vestibule. Other than otic capsule anomalies all of the patients had oval and round window and stapes anomalies. In two of the patients (Case 1 and 2) stapes has single thickened cruses with annular fixation and the oval windows were atretic bilaterally. In case 4, oval windows were thick posteriorly and posterior cruses of stapedes were thickened. In case 2, both oval windows were small, atretic and stapedial footplate was thickened on the left side while it was impossible to assess on the right side because of postoperative changes (dislocated?). In all

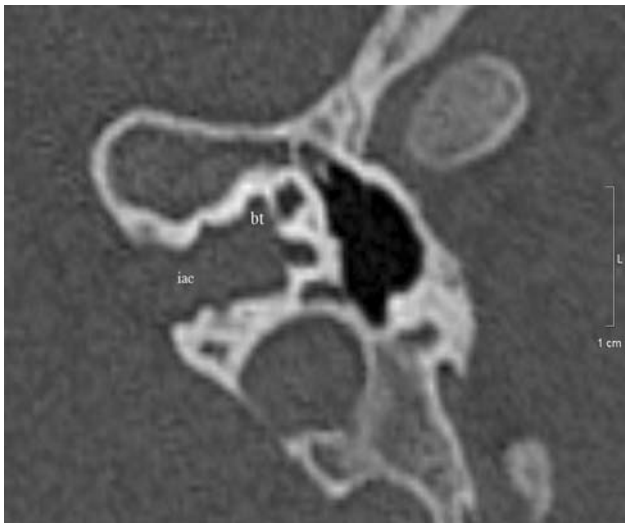

Fig. 2 Reconstructed oblique section showed bulbous dilatation of the IAC (*bt* basal turn, *iac* internal auditory canal)

patients round windows were small and atretic. In one patient with facial nerve stimulation after cochlear implantation on the right side, the implant was seen to be protruding to labyrinthine canal from the second turn of cochlea, which might be due to local dehiscence or weakness of the otic capsule. MRI confirmed that cochleovestibular nerves were present on both sides in all patients.

Discussion

X-linked anomaly is not very common finding in a prospective candidate for cochlear implantation. However, results of our cases indicated that this anomaly has some features similar to the other inner ear abnormalities as well as some unique problems.

X-linked deafness shows X-recessive type of inheritance and the expectancy is severe hearing loss in male patients and mild to moderate or delayed onset hearing impairment in females. However, in one of our cases, a female who had congenital severe hearing loss is presented. Her CT scan demonstrated pathagnomonic findings of X-linked anomaly as stated in the literature. Moreover, stapes was found fixed in the operation. Papadaki et al. has also reported two females from a family who had mixed severe hearing loss associated with stapes gusher during the stapes operation and X-linked deformity in the inner ear on HRCT. Eleven male members of a family had normal hearing with normal HRCT [9]. X-linked deafness with typical CT findings cannot be seen in females in the X-linked heritage; therefore, these findings could be attributed to another category. This phenotype mimicking X-linked deafness is most probably inherited in an autosomal recessive manner although in the literature typical CT findings of this entity are still called as X-linked anomaly. This nomenclature may be considered as incomplete partition type III [15].

The presence of additional stapes malformation in the second case and additional cleft plate in his brother besides X-linked deafness can be evidence for a syndromic form of X-linked anomaly. Since the inner ear and stapes crura have different embryologic origins, complex genetic and exogen factors may be important for the development of this anomaly.

High-quality CT and MRI scans are very important to diagnose any inner ear abnormalities. Graham et al. suggested that four or five 2-mm axial sections were enough to exclude congenital dysplasia [4]. However, because of our experience from case 1, the statement "all high resolution CT scans should be obtained with at least 1-mm thick slices which were reconstructed with bone algorithms and an experienced neuroradiologist must evaluate the CT and MRI images" must be emphasized once again. Moreover, surgeons should collaborate with neuroradiologist to evaluate the problematic cases. After our experiences in children with inner ear abnormalities, we started to order MDCT scan for all children with sensorineural hearing loss; even when they were not evaluated to be a possible candidate for cochlear implantation. The reason of this policy was high percentage of the patient with inner ear abnormalities and the risk of meningitis due to this anomaly in this group of patients [11]. Detection of any malformation led us to advise the parents about risk of meningitis. MRI scan is important for the evaluation of membranous malformations and the cochlear nerve. Any inner ear abnormality is diagnosed with CT scan; preoperative MRI scan is becoming more important to see the presence of cochlear nerve. In the case with inner ear malformations, MR imaging with constructive interference in steady state (CISS) sequence in which multiplanar reconstruction must be obtained to see cochleovestibular nerve in the IAC. MRI confirmed the presence of cochlear nerve in all our patients.

Normally, internal auditory canal is ended in the bony plate of lamina cribrosa where the nerves pass through and this barrier separates perilymphatic space and subarachnoid space. On the other hand, an abnormal connection between these two spaces may exist in the inner ear malformations. Therefore, perilymphatic or CSF gusher is the result of an abnormal bony defect at the lateral end of the internal auditory meatus due to spontaneous or surgical fistula. Since internal auditory canal is ballooned and widely open in incomplete partition type III deformity, profuse CSF gusher was encountered in all our cases. Elevating the head of the operating table and waiting for a while were enough to obtain a clear surgical field. The surgeon must pay precise attention for the firm sealing of the cochleostomy in this group of patients. It will prevent the open communication between inner and middle ear. In this step, the size of the

cochleostomy is also important. Although, some authors suggested possible smallest cochleostomy in such cases [17], we recommend opening the relatively large cochleostomy and to place the pieces of muscles around the electrode array and put bone dust over them. In our first case, we didn't use bone dust but only pieces of muscles. Following the study of Gstoettner et al. [5], we started to use pieces of muscles and bone dust together. In spite of large cochleostomy and using of pieces of muscles and bone pate, after the second operation of the first case, rhinorrhea occurred. Since we thought that CSF drainage with lumbar puncture is an invasive procedure and it may increase the risk of meningitis, we preferred conservative management and kept the CSF drainage as a final choice. The using of bone dusts around the cochleostomy has a potential risk of ossification in the cochlea and obliteration of cochlea with fibrous tissue was encountered in our second case. Bone pate might be a good fixation material, but this risk must be considered.

Meningitis after cochlear implantation is very important issue since the number of the reported cases has increased since 2002. Moreover, the risk of meningitis is higher in children with inner ear abnormalities even they are not implanted [2, 16]. The firm sealing of cochleostomy and vaccination as well as intense consultation with the parents about the early sign of middle ear infections and meningitis are extremely important. Moreover, Food and Drug Administration (FDA) study showed that cochlear implants with electrode positioners were associated with a greater risk of developing meningitis than implants without positioners [3]. However, cochlear implant with positioner in our two cases before this information was used. The firm sealing or being an adult might be effective.

Selection of electrode is very important in the cases with inner ear abnormalities. In most cases with inner ear abnormalities, the number of the spiral ganglion cells is likely to be enough to obtain benefit. However, location of neural tissue may be different. Since modiolus is absent in the case with IP-type III, the use of circumferentially stimulating electrodes would be reasonable [10]. On the other hand, the full-banded electrodes would be very risky because of facial nerve stimulation. Therefore, we preferred to use half-banded electrodes, which have contacts on only one side in our last case and the left ear of the first case. In the pediatric population, incidence rate of 1.89% was reported for the facial nerve stimulation after the cochlear implantation [1]. The possible reason of postoperative facial nerve stimulation in children with inner ear malformations was the proximity of the facial nerve to the electrode array, aberrant course or dehiscence of facial nerve [7] or placement of the electrode into the internal auditory canal. Fibrous tissue growth around the electrode array resulted in high electrical threshold level, which may stimulate the facial nerve in time. Usually, switching off the responsible electrodes can overcome this problem. In our first case, electrode array protrusion into the labyrinthine canal resulted in facial nerve stimulation five and a half years after the first operation. It might be congenital dehiscence, which was not detected preoperatively or it might be due to local erosion due to high stimulation current to obtain sufficient loudness perception. Reimplantation of the same ear would be risky. Therefore, we preferred to implant the other ear.

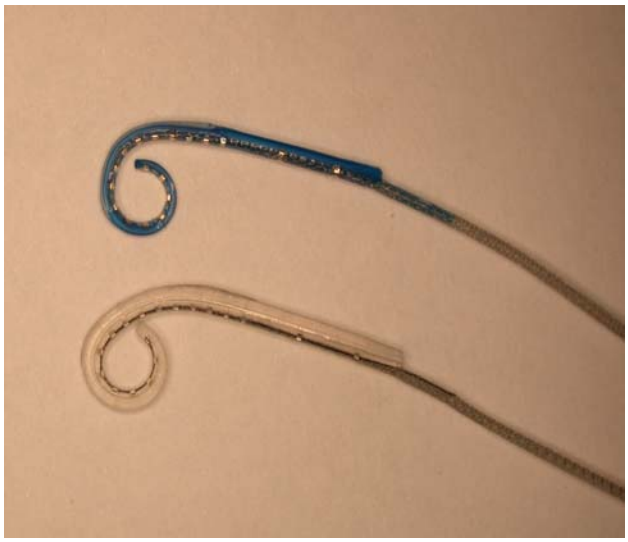

Fig. 3 Slim (*top*) electrodes and conventional (*bottom*) electrodes

Companies try to investigate different electrode designs for complicated cases. Slim electrodes, which are under investigation by Advanced Bionics Company, would be recommended to this group of patients (Fig. 3) to prevent unwanted facial nerve stimulation. Reimplantation can be performed due to several reasons but intracochlear fibrous tissue scars may impede insertion of the electrode array into the cochlea. We encountered this problem in the second case. After the device failure, reimplantation was not possible in the same ear due to severe fibrosis. This condition has two potential risks. The first one is the possibility of the future device failure. In this case, reimplantation in the second ear may not be achievable due to the same reason and patient may lose the chance of benefiting from cochlear implant. Auditory brainstem implant would be an alternative in this situation, but all we know is that the benefit from the cochlear implant is higher than the benefit from auditory brainstem implant in most of the cases. The second problem is auditory deprivation in the other ear due to none of the stimulation with conventional hearing aids. In our first case, it was not a concern, because she has used a conventional hearing aid. Fortunately, the second case is doing very well after implantation of the second ear, even though he has not used conventional hearing.

Conclusion

Cochlear implantation is a good choice in the patients with IP-type III (X-linked deafness). We used standard transmastoid-facial recess approach and complete electrode insertion was achieved in all cases. However, this anomaly may have special potential risks than the other inner ear abnormalities. Therefore, the surgeon should be aware of them and be ready to inform the patients and parents.

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