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CASE REPORT

Recurrent Meningitis due to Non-implanted Ear in Cochlear Implant Patient with Bilateral Inner Ear Abnormality: A case report

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Abstract: Recurrent meningitis occurs usually due to immunological or anatomical abnormality as well as chronic parameningeal infections. Anatomical abnormalities can be acquired or congenital and most of them are located in the head and neck region. Congenital defects include inner ear abnormalities, patent cochlear aqueduct, patent Hyrtl's fissure, abnormal developed facial canal and dehiscence at the lateral end of the internal auditory canal. In this case report, recurrent meningitis due to nonimplanted ear in a cochlear implant patient with bilateral congenital inner ear abnormality was discussed with surgical and radiological findings.

Key words: recurrent meningitis; congenital; inner ear abnormality; cochlear implant

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Introduction

Recurrent meningitis is described as two or more episodes of meningitis due to different bacteria or, second or further episodes due to same organism after a period of full recovery from the previous episode [1]. Recurrent meningitis should be warned to the physician about the possible immunological or anatomical abnormality as well as chronic parameningeal infections. Anatomical abnormalities can be acquired or congenital and most of them are located in the head and neck region. Acquired anatomical defects in this area can be caused by head trauma, iatrogenic surgical trauma or neoplastic invasion of the skull base. Congenital defects include inner ear abnormalities, patent cochlear aqueduct, patent Hyrtl's fissure, abnormal developed facial canal and dehiscence at the lateral end of the internal auditory canal [2-4]. All these conditions result in perilymphatic fistula which is an abnormal communication between perilymphatic space and middle ear. It is one of the potential causes of recurrent meningitis.

If any patient with recurrent meningitis has also severe sensorineural hearing loss or cochlear implant, the suspicion about the inner ear abnormalities or complication due to cochlear implant would be high. In this case report, recurrent meningitis due to nonimplanted ear in a cochlear implant patient with bilateral congenital inner ear abnormality was discussed with surgical and radiological findings.

Case Report

D.C. is an 8 and half years old girl. She was diagnosed with profound sensorineural hearing loss at age of one year. She was rehabilitated with bilateral conventional hearing aids. She attended to a rehabilitation center for hearing-impaired children. No consanguinity was noted and no other family members had hearing loss. Her mental and motor development was normal. Since she couldn't get enough benefit from conventional hearing aids, she was evaluated for cochlear implantation. Preoperative high resolution computed tomography (CT) showed bilateral severe inner ear abnormalities including defective modiolus, interscalar septa and cribriform plates, large cyst formation of cochlea and vestibule as well as dysplastic semicircular canals. Both internal auditory canals were larger and vestibular aquaducts were reported as normal. Magnetic resonance images confirmed the

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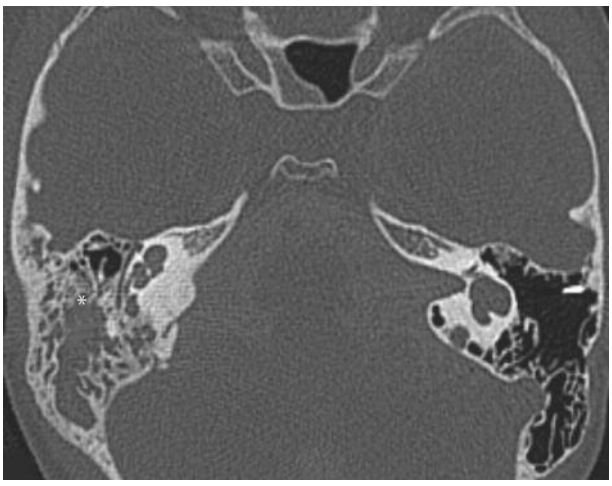
presence of the cochlear nerve in the both sides. At the age of 3 and half years, she underwent cochlear implantation on the left ear using standart transmastoid-facial recess approach. No anomaly was seen on the facial nerve. Promontory was flat than normal. Round window niche couldn't be identified clearly; therefore cochleostomy was performed 1.5 mm inferior to the stapes tendon over the promontory. Cerebrospinal gusher was encountered and lasted in 15 minutes with the elevation of the head. Med-El© short electrode was placed and cochleostomy was sealed with pieces of muscles. An intraoperative and postoperative antibiotic for five days was applied. Detailed information about the inner ear deformity, possible meningitic bouts, risks of head trauma were given to the parents. She had immunization against only Haemophilus influenza type B during the first year of age, because of the vaccination agenda in Turkey at that time. Her postoperative follow-up was uneventful in the first two years. Then, she had three episodes of meningitis in nine months in her hometown and cerebro-spinal fluid culture was positive for Streptococcus pneumonia in each of these episodes. Unfortunately, any information couldn't be obtained about the tympanic membrane appearance before or during the attacks. After successful treatment with ceftriaxone and vancomycin, pneumococcal vaccination was applied. Then she referred to our clinic to find the possible anatomical defect and treatment. CT-cisternography showed (Figure 1 and 2) high density fluid in the middle ear and mastoid cavity on the right ear. She underwent transcanal middle ear exploration on this ear. Two fistulae located above the stapes were seen and stapes was abnormal. Fistulae were obliterated with pieces of temporalis muscle, fascia and fibrin glue. Her postoperative period for ten months was uneventful until she had another episode of meningitis following head trauma. CSF culture was positive for Streptococcus pneumonia. Exploration revealed a profuse leak of CSF through the defects which had been sealed with the soft tissues and tissue adhesives. Continuous lumbar drainage was performed and strict bed rest put in force. Unfortunately, clear rhinorrhea was observed ten days later. Right ear was explored again. Incus was removed to recognize exact location of the defects. The bigger defect was located a few millimeters anterior the oval window. The other one was seen very close to oval window border. Since the parents did not accept the total obliteration of the inner and middle ear, simple obliteration of the defects was performed again using pieces of muscles, fascia, bone dust which was obtained from external auditory canal with very short drilling and fibrin glue. Continuous lumbar drainage lasted in nine days. Her 26 months follow-up was uneventful.



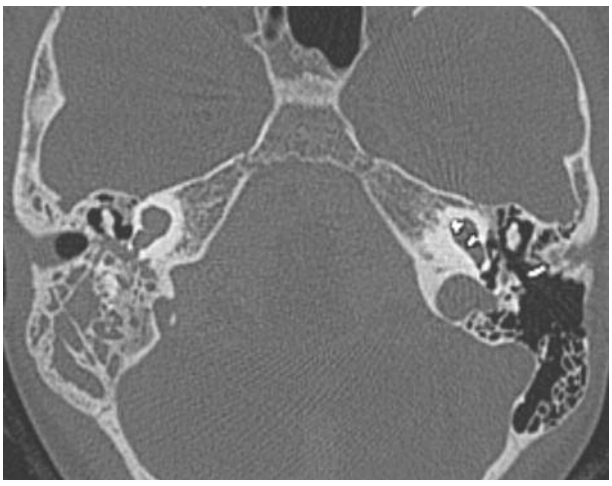


Figure 1 and 2. CT - cisternography showed high density fluid (Hounsfield value: 193.3±137.3, 11 mm²) in the middle ear and mastoid cavity on the right ear that supports abnormal communication between subarachnoid space and middle ear. (*) Hounsfield value: 193.3±137.3, 11 mm²

Discussion

Meningitis following cochlear implantation is an important problem since the number of the reported cases has been increased after 2002. The incidence of pneumococcal meningitis in children with cochlear implant was reported 138.2 cases in 2003 [5]. Several reasons such as using two component electrode system, incomplete cochlesotomy sealing, having an inner ear abnormality, meningitis in the past, previous surgery in inner ear or head, being a young child are considered to be risk factors for post-implant meningitis. Until now, only one case with bilateral inner ear abnormality and recurrent meningitis due to contralateral ear was reported to the best of our knowledge [6].

The consequence of recurrent meningitis might be fatal or highly morbid. Therefore, evaluation and intervention must be performed meticulously. The previous systematic literature review between 1978 to1988 revealed 47 patients with recurrent meningitis. Fifty five percent of them had congenital CSF fistula and the most commonly identified area for the defect was stapes footplate [7]. More recent English literature review between1998 to 2007 presented 363 cases. Inner ear anomalies were found in 55 out of 363 cases [8]. In the past, few cases with recurrent meningitis and congenital anomaly were also published (Table 1) [6, 9-22].

Table 1. Findings in some literatures about the case with recurrent meningitis and inner ear abnormality

Reference	No. of case	Age at first attack of menengitis	No. of attacks	Organism(s) isolated	Radiology	Operative findings
Maxwell et al, 1953, (9)	2	Case1: 5 years	4	None (2) S. pneumoniae (2)	A transverse fracture through the left petrous apex, (X-ray)	Fracture through the left petrous apex
		Case 2: 2 years	3	S. pneumoniae (3)	Destructive process in the region of the right petrous apex, (X-ray)	A meningocele stalk
B. Barr et al, 1965,(10)	1	3 1/2 years	2	N. menengitidis	N/A	N/A
William J. Rice et al, 1967, (11)	1	2 1/2 years	3	S. pneumoniae	Increase in density in the right mastoid air cells, (X-ray)	N/A
Paul Schultz et al, 1970, (12)	1	27 months	2	S. pneumoniae	N/A	A small fistula in the anterior portion of the stapes footplate.
Hipskind M, et al 1976, (13)	2	Case 1: 5 years	1	S. pneumoniae	NarrowOW,hypoplastic cochlea, large vestibule, short superior and horizontal canal (X-ray)	Unusual shaped of the stapes partially dehiscent fascial canal, a leak of clear fluid
		Case 2: 41 years	3	S. pneumoniae	N/A	N/A
Gacek R, et al. 1979, (14)	1	16 months	3	S. pneumoniae	Unremarkable except for wider internal auditory canal on the right side, (HRCT)	N/A
Luntz M. et al, 1986, (15)	1	3 years	4	H. influenzae (1) S. pneumoniae (2) None (1)	Enlarged right vestibule, superior and lateral SSC, partially developed cochlear turns, (HRCT)	(3 operations) 1.Soft cystic formation bulging through a hole on the FP 2. A fissure in front of the RW 3. CSF leakage from the OW, RW, Hyrtl's fissure, fissure between the fascial nevre and the lateral SSC

Table 1. Findings in some literatures about the case with recurrent meningitis and inner ear abnormality

Reference	No. of case	Age at first attack of menengitis	No. of attacks	Organism(s) isolated	Radiology	Operative findings
Claros P et al, 1993, (16)	1	2 years	4	N. menengitidis(2) S. pneumoniae (1) None (1)	Extensive dehisence of the left cribriform plate, (HRCT)	Dehisence with multiple perforations of the left cribriform plate
Stevenson DS et al, 1993, (17)	1	17 months	6	None (4) Actinobacter spp (1) P.aeruginosa (1)	Bilateral severe cochlear dysplasia, (HRCT)	Dehisence in the OW niche
Rupa V et al, 2000, (18)	2	Case1: ? Case 2: ?	4 5	None S. pneumoniae	Mondini's dysplasia, (HRCT) Normal, (HRCT)	A small central dehisence over the FP Defects at the RW and on the promontory
Rupa V et al, 2001, (19)	1	29 years	3	None	Possible fracture of the petrous tempoaral bone (HRCT) Dialated vestibule, Mondini type,CSF in widened IAC (MRI)	Flow of CSF into middle ear revealing a circular defect in the center of the stapes FP
Belmont MJ et al, 2004, (6)	1	12 years	3	S.pneumoniae(1) None (1) H. influenzae(1)	Cochleo- vestibular dysplasia and middle ear- mastoid opacification on the right side and a left cochlear implant, (HRCT)	CSF leak at the anterior aspect of the oval window
Kitazawa K et al, 2004, (20)	1	2 years	2	None (1) S. pneumoniae (1)	Mondini dysplasia in the right iner ear, small right cochlea and dysplastic apical turns, (HRCT)	A defect on the promontorium
Soshan MB et al, 2007, (21)	3	Case 1: 2 yrs 5 months Case 2 : 2 years Case 3: 1 ¾ years	3 2 1	S.Pneumoniae S.Pneumoniae (1) None (1) S.pneumoniae	Fusion of the right cochlea and vestibule with aberrant track of the right facial nevre, (HRCT) Enlarged cavity in cochlea, abnormal semisircular canals and abnormal location of the seventh nevre, (HRCT) Incomplate partition of the cochlea bilaterally and saccular dilatation of the vestibule, (HRCT)	Defect in the oval window of the right side N/A N/A
Torkos A. et al, 2009, (22)	1	2 1/2 years	4	H. influenzae (1) S.Pneumoniae (2) H. influenzae and S.Pneumoniae (1)	Labyrinthine dysplasia on both sides, determined as cochlear dysplasia, (HRCT)	(3 operatrions) 1. A cystic sac filling the aditus ad antrum, CI was performed on the right ear 2. Left mastoidectomy 3. Sacced filled with CSF in the tympanic cavity and aditus ad antrum , partialy developed FP, defect on promontorium

OW: *Oval window,* ***RW:*** *Round window,* ***FP:*** *Foot plate,* ***SSC:*** *Superior semicircular canal,* ***IAC:*** *Internal auditory canal,* ***CSF:*** *Cerebrospinal Fluid,* ***CI:*** *cochlear implant,* ***X ray:*** *X-radiation,* ***HRCT:*** *High Resolution Computerized Tomography,* ***MRI:*** *Magnetic Resonance Imaging*

An abnormal connection and perilymphatic or CSF gusher due to spontaneous or surgical fistula between subarachnoid space and middle ear may exist in the inner ear malformations. Although the defect in several cases with recurrent meningitis were reported as Mondini deformity in the literature, the risk of the recurrent meningitis and congenital perilymphatic fistula is expected very rare in type-II anomaly (classic Mondini deformity) because of normal basal turn and intact cribriform plate.

Preoperative CT and MRI scan shows the type of anomalies, however, they may not be enough to show the defect or spontaneous CSF leak. CT cisternography may give more information about the defect location, although it is an invasive method. 3D fast speen echo T2 weighted imaging MR scan and 3D FIESTA sequences were recommended for the assessment of CSF fistula [11]. In our case, high resolution CT scan showed bilateral incomplete partition type I anomaly (Fig 1). In which, there is no modiolus, cochlea is a cystic cavity accompanying cystic vestibule. Moreover, neuroradiologist was able to show the defect side in the nonimplanted ear in this scan. CT cisternography also confirmed the location of the defect.

Closure of the defect is possible in one operation although failure rate is not very low because of the high pressure of cerebrospinal fluid. Luntz et al.[15] checked the twenty-one cases with recurrent meningitis and large perilymphatic fistula in sixteen literatures. They found that fistulas were successfully repaired in the first operation in only 23.8% of cases. Obliteration of vestibule with multilayer technique including pieces of muscle, fascia, fibrin glue and reinforced by a pedicled temporalis muscle graft was recommended [23-25]. In addition to total obliteration of vestibule; middle ear and blind sac closure of the external auditory canal was also recommended. Since parents of our patient were hesitated about the aggressive surgical intervention including total obliteration of vestibule, middle ear and they wanted to preserve inner ear integrity for possible future developments in the cochlear implant technology, conservative surgical management was performed for our patients. Unfortunately, she had another bout after the head trauma and she underwent three surgical attempts for successful closure. Several materials such as soft tissue, fibrin glue and bone pate have been recommended sealing of the defect. The study of Gstoettner et al.[26] showed that boné pate provide new bone formation includes laminar and compact bone structures on the cochleostomy side around the cochlear implant electrode. Since transcanal middle ear exploration performed in our patient, we didn't attempt to obtain bone dust during the first two surgeries, but finally we decided to drill in small area in the external auditory canal to obtained bone dust and it used as well as pieces of muscles and fascia used to close the defect.

Implanted ear is the reason of meningitis in the most of the cases in the literature. However, contra lateral ear must be evaluated intensely in this group of patients; especially if there is any inner ear abnormality in the implanted cases. Follow-up, intense consultation with the parents about the early sign of middle ear infections and meningitis, avoidance from head trauma, vaccination should recommend. Several methods such as clinical examination, tympanometric evaluation to exclude inner ear fluid, periodical CT scan can be used for follow-up. Periodical CT scan is not a cost/effective approach; moreover the radiation from the scan may be harmful for the child. Periodical ear examination is easy method to see the middle ear fluid if the total closure of external auditory canal was not performed. The most important point for us is informing the parents about the early sign of infections.

Recurrent meningitis after the cochlear implantation is not very common. In any case, implanted and non-implanted ear should be evaluated carefully. Aggressive surgical procedure to close the defect may be necessary to avoid surgical failure.

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