Int. Adv. Otol. 2010; 6:(1) 109-111

CASE REPORT

Successful Cochlear Implantation in a Child Deafened by Mumps

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Our aim was to present a case who underwent cochlear implantation due bilateral total deafness resulted from mumps. A thirty-four-month- old girl was admitted to our clinic by her parents with the complaint of sudden deafness. Comprehensive audiologic examination showed bilateral total hearing loss. The physical examination, family history and serological tests suggested the diagnosis of deafness due to asymptomatic mumps infection. The child was implanted with a Nucleus CI 24 device after 6.5 months of deafness with a successful outcome. Last visit was conducted on 21st month after surgery. Average tone thresholds was 30 dB, and the child was using oral communication and made 3-word sentences at that time. Patients suffering from bilateral mumps deafness and unresponsive to medical treatment should be candidates for cochlear implantation, and surgery should be performed as early as possible once diagnosis is established.

Submitted : 14 May 2009 *Revised : 14 June 2009* *Accepted : 23 June 2009*

Introduction

Mumps is a common childhood infection caused by the mumps virus. The hallmark of the infection is swelling of the parotid gland. Aseptic meningitis and encephalitis are common complications of mumps together with orchitis and oophoritis. Other complications include deafness and pancreatitis [1]. Although deafness is a rare complication of mumps, it is still one of the most common causes of acquired sensori-neural hearing loss [2]. Herein, we present a 3.5-year-old girl with a bilateral total hearing loss due to asymptomatic mumps infection who was implanted successfully.

Case Report

A thirty-four month-old girl was admitted to our department with complaints of sudden deafness and balance problem on walking for last 5 days. The parents informed that communicative abilities of the patient had been consistent with her age previously and no difference between the right and left ear's hearing was noticed. Vaccination history for mumps was unclear. Family history revealed that two sisters had parotid gland swelling due to mumps infection within the last 20 days. Otologic and systemic physical examinations were within normal limits except mild ataxia. There was no swelling in the parotid region. Body temperature, complete blood count and blood biochemistry were within normal limits. Mumps IgM and IgG antibody titers were positive in blood serum. Child audiometry showed a bilateral total sensorineural hearing loss. There was no response in distortion product oto-acoustic emission (DPOAE) bilaterally. Both auditory brain stem response (ABR) test and auditory steady-state response (ASSR) test failed to detect a hearing threshold bilaterally. Patient was diagnosed as sudden bilateral total hearing loss due to asymptomatic mumps infection.

Steroid therapy with prednisolone (Table 1) was started immediately. Unfortunately, there was no hearing gain after completion of the treatment schema. Patient was included to our cochlear implantation (CI) program. However, it took 6.5 months until the patient

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has gone to CI surgery due to health insurance problems. The child was fitted with hearing aids bilaterally and started special CI speech training program 3 months before implantation. The child has became introverted and lost most of the speech ability during this waiting period.

Table 1. Prednisolone treatment schema of the patient

1st day	75 mg IV
2nd day	50 mg IV
3rd day	25 mg IV
After than	start 20 mg PO and decrease 5 mg in every 3 days.

Nucleus CI 24 device was used for cochlear implantation. We performed cochleostomy via posterior tympanotomy approach and electrode array was fully inserted without difficulty to the right cochlea of the patient. There was no fibrosis or occlusion in the cochlear lumen. There was no complication during surgery. CI device was activated on the 4th week after surgery, and the child continued speech training program. Last visit was conducted on 21st month of the surgery. Pure tone average was 30 dB and the child was using oral communication and able to make 3-word sentences on that time.

Discussion

Mumps deafness is still one of the most common causes of acquired sensori-neural hearing loss. Its incidence was reported to be 5.1 per 1,000,000 in Japan [3]. Okamato et al. (1994) showed that 6.9% of sudden deafness cases were accompanied by asymptomatic mumps infection [4]. Recently Dunmade et al. (2007) investigated the etiology of bilateral profound sensorineural hearing loss in 115 children. They found mumps infection as an etiologic factor in 8 of 115 children (6.9%) [5]. Riga et al (2005) reported only one patient who had bilateral sensorineural hearing impairment due to mumps in 36 hearing impaired children due to non-hereditary acquired causes in Greek population [6].

Onset of the mumps deafness is usually rapid, and it is unilateral in 80% of the cases. Hearing loss is often profound and usually permanent, but a prospective study by Vouri et al. (1962) found it to be transient in some cases. Hearing loss is usually most pronounced in the high frequency region. Tinnitus and fullness in the involved ear are common, and some patients develop vertigo. Disequilibrium usually resolves over several weeks [7]. Vertigo is thought to be secondary to inner ear dysfunction. However, recently Tsubato et al. (2008) reported a case of mumps deafness with acute vestibular symptoms secondary to both retrolabyrinthine and inner ear dysfunction [8]. In the presented case, there was a sudden bilateral permanent total loss of hearing accompanied with mild transient ataxia.

Route of dissemination of the virus into the inner ear is undefined. It may result from viremia that often occurs in mumps virus infection. It is also possible that virus may be transported to the inner ear by the way of the eustachian tube and the middle ear. A third possibility is the meningeal route. Examination of the temporal bones from patients with mumps associated deafness showed widespread degenerative changes in the tectorial membrane and stria vascularis, as well as in the organ of Corti. Nerve fiber loss has also been reported [7,9].

Definite diagnosis of mumps deafness in patients without evident clinical signs of mumps is made by looking at the increased serum antibody titer within 2-3 weeks after the onset of acute hearing loss [10]. In this case family history raised the suspicion of mumps deafness. Detection of the increased titer of serum IgM and IgG antibodies confirmed the diagnosis. Recently, Otake et al. (2006) showed the use of 3D-FLAIR Magnetic Resonance Imaging in the evaluation of mumps deafness. They showed high signals in the cochlea and vestibule indicating hemorrhage or a high concentration of protein in the inner ear of a child deafened due to mumps infection [11].

Although it is believed that hearing loss is permanent for mumps deafness we preferred to try medical treatment. We applied steroid treatment protocol for sudden hearing loss, but we could not observe any hearing improvement, and then CI surgery was performed. Full insertion of electrode array without difficulty indicated the absence of cochlear

110

ossification. Successful outcome of cochlear implantation showed that mumps infection had not destroyed the auditory neurons. Although evidence of nerve fiber loss has also been reported in mumps deafness [9], acoustic nerve was at least partly preserved in this case.

Recently Wang et al. (2003) reported three cases of bilateral total mumps deafness treated with CI in Chinese literature. Cochlear implantation has been successful in all 3 patients. The one who had been wearing bilateral hearing aids and received one-year language training before CI had the best outcome of hearing and speech than the other two [12]. Our patient was also fitted with bilateral hearing aids and received speech training for 3 months before implantation.

As a conclusion, mumps deafness is still a common cause of sudden hearing loss especially in children. Medical treatment does not improve the outcome usually. Patients unresponsive to medical treatment should be a candidate for CI surgery as early as possible. Bilateral hearing aids use and speech therapy will help to increase the success of CI in cases with a long waiting period before surgery.

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