International Journal of Pediatric Otorhinolaryngology 114 (2018) 76–79

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Case Report

Outcomes of cochlear implantations for mumps deafness: A report of four pediatric cases

Masako Katsushika, Akinori Kashio*, Erika Ogata, Yusuke Akamatsu, Yujirou Hoshi, Shinichi Iwasaki, Tatsuya Yamasoba

Department of Otolaryngology and Head and Neck Surgery, Graduate School of Medicine, The University of Tokyo, 7-3-1 Hongo, Bunkyo-ku, Tokyo, 113-8655, Japan

ARTICLE INFO

Keywords:
Mumps
Deafness
Cochlear implantation
Vestibular

ABSTRACT

Mumps virus occasionally causes bilateral hearing loss. We report 4 cases of bilateral mumps deafness in whom cochlear implantations (CI) were performed. The age at the onset of hearing loss was 1–9 years. CI surgery was performed within 6 months from the onset of hearing loss in 3 cases and after 9 years in the other case, showing good speech perception in the early intervention cases and a poor outcome after later implantation. Early CI surgery is highly recommended in sudden onset deafness by mumps in childhood.

1. Introduction

Mumps is an infection caused by the mumps virus, typically presenting with flu-like symptoms, followed by bilateral swelling of the parotid glands [1]. However, subclinical infection is a possible presentation and some patients may not experience parotid swelling [2]. Mumps infection can lead to a number of complications including meningitis, encephalitis, pancreatitis, orchitis, oophoritis, infertility, epididymitis, and deafness [3,4].

The prevalence of mumps deafness is estimated to be as low as 0.5–5.0 per 100000 cases [5]. However, due to the low penetration of mumps vaccination in Japan, mumps is endemic with an incidence of deafness as high as 1 per 1000 cases [6].

Most cases of mumps deafness present with a profound unilateral hearing loss; however in some cases, it is bilateral [2]. There are few studies examining the pathogenesis of mumps deafness. The human temporal bone findings, as reported by Lindsay [7], were primarily of the cochlear duct. The stria vascularis, organ of Corti, and tectorial membrane were severely degenerated in the basal coil, diminishing progressively towards the apex. On the other hand, ganglion cells were near normal with minimal degeneration of the peripheral cochlear nerve in the basal coil. Experimental studies using guinea pigs and monkeys, revealed that the mumps virus has a high affinity for the stria vascularis and the outer hair cells of the ear, mainly disrupting the stria vascularis and the organ of Corti [8,9]. Although steroids have been clinically administered as a treatment for mumps deafness, the prognosis for patients is generally poor [2]. As a result, CI surgery is the only effective treatment for mumps deafness. However, its performance has rarely been reported [10–15].

Herein, we present 4 pediatric cases with bilateral mump deafness who underwent CIs, and report their clinical findings and outcomes.

2. Case reports

2.1. Current history

2.1.1. Case1

A 6-year old girl had bilateral hearing loss and vomiting, 3 days following the onset of right parotid swelling. Mumps-specific IgM and IgG antibody levels were high (13.8 mg/dL and 28.1 mg/dL, respectively; Positive assessment was made at ≧1.21 IgM and ≧4.0 IgG antibody levels). She had no history of mumps vaccination. The patient was diagnosed with mumps deafness and treated with intravenous prednisolone (21 mg/day). However, treatment was not effective and she was referred to our hospital for a CI consultation.

2.1.2. Case2

A 9-year old girl developed a right parotid swelling. Fourteen days later, she suffered from bilateral profound hearing loss, nystagmus, and vomiting. She had no history of receiving mumps vaccination. Her serum mumps-specific IgM antibody level was high. She was diagnosed with mumps deafness and treated with intravenous steroids. However, her hearing remained profoundly impaired. One month following the onset of hearing loss, she was fitted with hearing aids, which was not

* Corresponding author.
E-mail addresses: yuu-ykh@umin.ac.jp (M. Katsushika), kashioa-tky@umin.ac.jp (A. Kashio), ogatae-tky@umin.ac.jp (E. Ogata), aka-tky@umin.ac.jp (Y. Akamatsu), hoshi-tky@umin.ac.jp (Y. Hoshi), iwashin-tky@umin.ac.jp (S. Iwasaki), tyamasaba-tky@umin.ac.jp (T. Yamasoba).

https://doi.org/10.1016/j.ijporl.2018.08.027
Received 4 May 2018; Received in revised form 22 August 2018; Accepted 22 August 2018
Available online 27 August 2018
0165-5876/

effective. Two months later, the patient was referred to our hospital. Following hearing loss, her speech articulation deteriorated.

2.1.3. Case3

A 6-year old girl had bilateral parotid swelling, vomiting and dizziness. She developed hearing loss in the right ear 7 days later, followed by bilateral profound hearing loss on day 14. Mumps vaccination history was negative. Both serum mumps-specific IgM and IgG antibody levels were high (8.39 mg/dL and 128 mg/dL, respectively). She was diagnosed with mumps deafness and treated with steroids, which were not effective. She received hearing aids, which were deemed not effective, and was referred to our hospital one month after the onset of deafness.

2.1.4. Case4

A 15-months old boy complained of ear ringing and exhibited signs of a gait disorder. Ten days later, he underwent auditory brainstem response testing in a clinic, revealing a profound bilateral hearing loss. The patient had no history of mumps vaccination, and despite of the absence of a parotid swelling, the serum mumps-specific IgM antibody level was high. He was diagnosed with mumps deafness and treated with steroids, which were not effective. However, his gait improved a month later allowing him to walk again.

He was fitted with hearing aids, and began to participate in an early intervention program for deaf children after 6 months. Although he enrolled in a mainstream elementary school, he mainly used lip reading for communication. At 10 years of age, he was referred to our hospital for a CI consultation.

2.2. Preoperative examination

Each case's pure tone audiometry (PTA) and hearing aid thresholds are shown in Fig. 1. Hearing aids were not effective in all cases.

In all the cases, we did not detect any abnormal findings in the middle and inner ears on computed tomography (CT) or magnetic resonance imaging (MRI). (Fig. 2). The results of caloric testing with ice water and cervical vestibular evoked myogenic responses (cVEMPs) to air-conducted sound (4 ms tone burst, 500 Hz, 105 dBHL) are in Table 1. Three of the four cases showed peripheral vestibular dysfunction on caloric testing or cVEMPs to air-conducted sound. We did not perform genetic tests or tests for other viruses such as CMV.

2.3. Postoperative results

Case 1 received CI522 (Cochlear Co., Ltd, NSW, Australia) in the left ear after two and a half months following the onset of deafness, Case 2 received CI24M (Cochlear Co., Ltd) in the left ear after four months. Case 3 received CI422 (Cochlear Co., Ltd) in the right ear after two months. Case 4 received CI24M (Cochlear Co., Ltd) in the right ear after nine years of deafness.

All cases achieved a good hearing threshold of around 30 dB at all frequencies with CI. Postoperative Japanese Fukuda monosyllable test scores were 90% in case1, and 96% in case2 and case3, respectively. These three patients were able to attend a mainstream elementary school. However, case 4 scored only 30% on the 67-s Japanese monosyllable test. Two years after the CI surgery, he dropped out of our outpatient clinic.

3. Discussion

We performed unilateral CI in 4 cases of bilateral mumps deafness. No cases and their families requested bilateral CI. Of the 4 cases, 3 had CI surgery performed within 6 months from the onset of hearing loss, and 1 after 9 years. Good speech perception was achieved in the early intervention cases, while a poor outcome was found after late implantation. Our results suggest that early cochlear implantation in sudden onset deafness by mumps in childhood is highly recommended.

Table 2 lists the 14 reported cases of mumps deafness that underwent CI surgery, including our 4 cases [10–15]. Out of the 14 cases, 11 (79%) scored over 60% in the post-operative speech discrimination test, suggesting that mumps deafness is generally a good indication for CI. This is along with the pathology that mumps virus injures mainly the inner ear [7–9].

One of the cases reported (Case 4) had a poor outcome following CI. There are two possible reasons for this: one being the patient's age at the time of the CI surgery and the long period of deafness preceding it. It was shown that in pediatric CI surgeries, the patient's age and duration of deafness are inversely proportional to postoperative outcomes [16]. In case 4, the patient lost his hearing ability at one year of age, which was prior to the language acquisition period, leading to his dependency on lip reading up to nine years of age. Insufficient acoustic input during the maturation period might be related to the poor speech discrimination after CI. The other possible cause is that the deafness in this case could be caused by a retro-labyrinthine disorder. Lindsay suggested that viral invasion via the meningeal-subarachnoid system and through the internal meatus as a cause for deafness [17]. In this situation, the neural elements in the modiolus are directly exposed to the inflammatory processes causing severe degeneration of the cochlear nerves. Given that 1–10% of mumps-infected children develop aseptic meningitis [18], there is likely to be patients with a retro-labyrinthine disorder among children with mumps deafness. Noda et al. reported a case of mumps deafness with a poor CI outcome, proposing that the patient had a retro-labyrinthine disorder caused by meningitis [10]. Firrisi [19] reported a case with profound unilateral hearing loss due to mumps and demonstrated the preserved otoacoustic emissions. Comacchio [20] conducted enhanced MRI examination for a case with sudden deafness and vertigo due to mumps and demonstrated that both the labyrinth and nerve bundle were enhanced, suggesting retro-labyrinthine involvement. Unfortunately, we have not conducted further testing for retro-labyrinthine disorders. Higher-order processing testing such as electric-auditory brainstem response (E-ABR) testing that demonstrate waveforms originating from the retrocochlear area could be useful in exploring the exact mechanism for the poor outcomes in case 4.

However, we cannot definitively determine whether patients with mumps deafness have a retro-labyrinthine disorder preoperatively, it is necessary to discuss its possibility and obtain full informed consent before performing CI surgery in patients with mumps deafness.

It is difficult to determine the optimal timing of CI surgery for patients with mumps deafness. Since ossification of the cochlea is rarely seen in cases of mumps deafness [10-15], there is no need for immediate CI surgery as in patients with deafness caused by meningitis. However, even a short period of deafness at school age can result in language collapse and delay in its acquisition, as was seen in case 2. Additionally, unlike the congenitally deaf child, pediatric cases with mumps deafness are usually participants in normal hearing educational programs. Hence, their educational institution cannot promptly support a sudden loss of hearing. The aforementioned points suggest that early implantation is recommended. Nevertheless, it is necessary to carefully consider the optimal timing for surgery in each case; since it might take some time for the patient and their guardians to accept the diagnosis of profound hearing loss, evaluate hearing aids, and decide on CI surgery. From our experience and from other published reports [11,13], at least 3 months may be necessary before CI surgery is decided upon, during which steroid therapy and hearing aids are evaluated.

Of the 14 patients who had CI surgery for treatment of mumps deafness (Table 1), 8 (67%) had vestibular symptoms such as dizziness, gait disorders, or nystagmus. Previous studies reported that the incidence of vestibular symptoms in patients with mumps deafness ranged from 45 to 60% [21,22]. In the present study, three of the four cases showed peripheral vestibular dysfunction as shown by the caloric testing or cVEMPs to air-conducted sound, whereas the other case with