

# Catastrophic Bilateral Sudden Sensorineural Hearing Loss Due to Mumps: Report of Two Cases

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## ABSTRACT

Mumps is an acute self-limiting systemic disease that can potentially cause serious complications. One of its well-known complications is sensorineural hearing loss. Cases of SNHL associated with mumps are only reported in 1/1000 cases. This condition is also known for its poor prognostic feature regarding the restoration of hearing. Case report: patient 1: A 10-year-old immunized boy complaining of hearing impairment following an insidious onset parotid swelling with low-grade fever. Viral antibody panel serological results of anti-mumps IgM antibody were positive. Following the diagnosis of bilateral complete hearing loss, the patient was hospitalized and received treatment, but unfortunately, there was no improvement in hearing. He was sent home, and a rehabilitation plan was initiated. Patient 2: A 12-year-old non-immunized girl presented with hearing loss after experiencing bilateral tender parotid swelling and low-grade fever. She had complicated hearing loss of gradual onset. A week later a diagnosis of complete bilateral sensorineural hearing loss was confirmed hearing loss was confirmed on audiometry. Salvage therapy trial was initiated. However, there was no improvement. Mumps is highly contagious. In order to eliminate this disease, vaccination programs have been established around the world. The WHO advocates a coverage of 90% to prevent outbreaks. However, in Yemen, immunization programs have been suspended due to the political conflict. This has led to an increase in the number of endemic diseases, including mumps. Bilateral complete SNHL is the rarest form of SNHL, with only 22 cases until 1957. Very few cases have been added since then. Hearing loss may occur at any time before, during, or after the course of a mumps infection. In our cases, hearing loss occurred within the 3rd and 4th day, respectively. Many treatment strategies have failed to restore the hearing, and there is no treatment proven to be effective for sudden SNHL even if steroidal therapy is initiated immediately after the recognition of hearing loss. After hearing loss has occurred due to viral etiology, the retrieval of hearing using medical therapy is considered remote. The failure of treatment in our patient may also be related to the severity of cochlear damage. Hearing loss due to mumps infection is likely to be permanent regardless of treatment. Vaccination alone cannot prevent such deafness and outbreaks.

**Keywords:** Hearing loss, deafness, mumps

## INTRODUCTION

Mumps is an acute self-limiting systemic disease that can potentially cause serious complications, including inflammation of the salivary glands, pancreas, testes, meninges, and inner ear. One of its well-known complications is sensorineural hearing loss (SNHL) (1). Sudden hearing loss SHL is a significant matter of discussion. Bilateral SHL is considered rare in comparison to unilateral hearing loss and it is commonly related to an underlying disease rather than idiopathic etiology. The literature reporting infectious viral etiology causing bilateral SNHL represents only 10% of cases of SNHL, including mumps, HIV, HSV, and Viral Upper respiratory tract infections (2). Cases

of SNHL associated with mumps are only reported in 1/1000 cases. This disease is also known for its poor prognostic feature regarding the restoration of hearing, especially for those with profound loss (3-5).

## CASE PRESENTATIONS

### Patient 1

A 10-year-old immunized boy presented during an endemic period of mumps in a rural area in Yemen. He was brought to Al-Balasi Otolaryngology Hospital by his parents having complained of a hearing impairment for three days following

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an insidious onset of right-sided parotid swelling with low-grade fever which had been followed by a left-sided swelling on the following day. His condition was associated with nausea and vomiting on the first two days, and on the third day, his parotitis started to cease during which he began complaining of tinnitus and his parents started to notice that he had difficulty hearing. On the fourth day he had significant hearing loss, but at this point there was no headache, nor any signs of meningeal irritation. All the patient's family members had mumps during this endemic but none of them developed complications. Otherwise, the family history was negative for hereditary or systemic diseases, and also negative for ototoxic medication, or trauma. On examination, his tympanic membranes were normal, and mobile bilaterally with no evidence of ear inflammation. CBC was unremarkable. The viral antibody panel serological result of anti-mumps IgM antibodies was positive. Pure tone audiometry showed bilateral sensorineural hearing loss at 100 dB. On the auditory brain stem response, no waves were seen, confirming the diagnosis of complete hearing loss. After being diagnosed with bilateral dead ears the patient was hospitalized and received the following treatment plan: Prednisolone oral 60 mg for two weeks and then tapered over two weeks, Aspirin 75 mg three times a day orally for 10 days, Methylcobalamin 500 mcg twice a day intramuscularly, Acyclovir infusion 300 mg intravenously every 8 hours for 3 days, followed by 300 mg orally every 6 hours for another 7 days, and intratympanic methylprednisolone 0.5 ml buffered. During this time, hearing tests were repeated, and there was no improvement in his hearing. The patient was sent home, and a rehabilitation plan was initiated as we kept the patient's prognosis for 6 months with no alteration in hearing.

### Patient 2

A 12-year-old non-immunized girl from the highlands of Yemen presented after a 12-day history of hearing loss, having experienced bilateral tender parotid swelling and low-grade fever for three days prior to the onset of otologic symptoms. On the fourth day she started to have hearing loss of gradual onset, and one week later, profound hearing loss was documented on an audiometry. Before being referred to our hospital, she received oral prednisolone 80mg and Vinpocetine for a few days with no improvement. The patient was otherwise healthy and had no significant past medical history. Her family history was also negative for hereditary or systemic diseases apart from the patient's younger siblings who had mumps but none of them was hospitalized or had any complications. On examination, tympanic membranes were normal, and mobile bilaterally with no evidence of ear inflammation. Her CBC was unremarkable. Viral antibody panel serological results of anti-mumps antibodies IgM were positive confirming recent infection. The diagnosis of complete bilateral sensorineural hearing loss was confirmed on pure tone audiometry and on brainstem response as no waves were seen. Hence, we were aware of the association. Another salvage therapy trial for hearing was initiated with intratympanic corticosteroid along with oral vasodilator, anti-inflammatory, and anticoagulants,

and we followed up with her for six months. However, there was no improvement in her hearing.

### DISCUSSION

Mumps is a disease of children and young adults caused by of an enveloped, single-stranded RNA virus of the paramyxoviridae family. It is an acute self-limiting systemic disease that can potentially cause serious complications. Mumps can attack multiple organs including the salivary glands, pancreas, testes, meninges, and inner ear. One of its well-known complications is SNHL (1). Mumps is highly contagious and can have a long list of complications. In order to eliminate this disease, a program offering the mumps vaccination has been established around the world. The WHO advocates a coverage of 90% to prevent outbreaks and states that mumps hearing loss has almost vanished after vaccination coverage reached this level (3, 6). However, in Yemen, the immunization programs have been suspended due to the political conflict, which has led to an increase in the number of endemic diseases, including mumps. Mumps deafness per se is rare because immunization programs prevent outbreaks. However, unilateral hearing loss is a known complication in comparison with bilateral SNHL which is very rare (1, 2).

Hearing loss due to mumps can present in three different ways: complete unilateral, partial unilateral, and bilateral complete SNHL. The bilateral complete SNHL is the rarest form, with only 22 cases until 1957. Very few cases have been reported since then (7, 8). Hiromi et al. described the incidence of hearing loss in their prospective study as being from 0.5 to 5.0 per 100,000 cases of mumps in 2008. This incidence in Japan was reported because immunization had not been a routine practice at that time, while in Belgium an outbreak in 2004 occurred due to vaccine failure (5, 9, 10). The primary route of viral invasion to the cochlea is hematogenous, and hearing loss may occur at any time before, during, or after the course of a mumps infection (3, 11). In our cases, the hearing loss occurred after the course of the disease was subsiding, within the third and fourth day, respectively. The pathophysiology of hearing loss due to mumps is thought to be by direct invasion of the cochlea damaging the organ of Corti, the cochlear nerve myelin sheath, and degenerating the stria vascularis, tectorial and Reissner's membrane (4, 12, 13). Tanaka et al. experimentally confirmed that mumps-related deafness is caused by the degeneration of the organ of Corti (12, 14). Even asymptomatic or mild mumps infection can cause hearing impairment (5). Many treatment strategies have failed to restore hearing, and there is no treatment proven to be effective for sudden SNHL. However, a trial of steroidal therapy has been implemented recently. Intratympanic steroidal injection has become more popular in recent years. This method delivers more concentrations of steroids to the affected tissue, as well as treating patients with vasodilators, anticoagulants, and hyperbaric oxygen therapy (15). Shinya et al. reported that 36 patients with SNHL due to mumps received steroidal therapy and only one patient showed improvement (3). In our cases, steroidal therapy was initiated immediately after the recognition of hearing loss. However,

after hearing loss has occurred due to viral etiology, the retrieval of hearing using medical therapy is considered remote. The prognosis for patients with profound SNHL is very poor apart from cochlear implantation surgeries, which achieved good results regarding speech and sound perception in patients who do not have central nervous system damage involved (3, 16). Our patients would have to go overseas to receive a cochlear implant. Failure of treatment in our patients may also be related to the severity of cochlear damage that occurred in both cases. Informing the children and their guardians that the damage was going to be permanent with no further resources available for hearing retrieval, e.g. cochlear implantation, and counseling them about this sudden unexpected complete hearing loss at this age with all its upcoming difficulties and quality of life challenges, was a very traumatic period for both of them, their families, friends and for the working staff as well. It is difficult to know the actual number of cases of mumps-related deafness in Yemen since the outbreaks can trigger such conditions. The majority of hearing loss is unilateral profound or mild to moderate hearing loss. Therefore, many patients, especially children, may not have otologic symptoms severe enough to warrant medical attention for further assessment and diagnosis. These two cases underline the importance of immunization and the effect of wars on developing countries.

## CONCLUSION

When SNHL occurs due to mumps it is likely to be permanent regardless of treatment. Mumps induced SNHL is preventable, as vaccination alone could prevent outbreaks. The social and psychological cost of hearing loss is a significant matter and for that reason, stakeholders in Yemen should consider implementing both vaccine programs and cochlear implant centers.

**Informed Consent:** Parents agreed and gave written consent to the publication of this study.

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