

Mumps-associated Sensorineural Hearing Loss: A Review

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Abstract

Mumps is one of the common causes of acquired sensorineural hearing loss (SNHL). Hearing loss following mumps infection is usually mild or moderate and severe. Bilateral hearing loss due to mumps is very uncommon. The diagnosis is primarily based on clinical presentations, supported by audiometric findings to confirm the degree and nature of deafness. The majority of mumps-related hearing loss manifests as unilateral significant SNHL, which is frequently unresponsive to therapy. Management of mumps-associated SNHL includes early detection and intervention to improve the outcome. Corticosteroids can be prescribed to decrease the inflammation and edema in the inner ear, although their efficacy in treating viral-induced SNHL is uncertain. The role of antiviral medications in mumps-induced SNHL has a limited role with doubtful effectiveness. For those with severe hearing loss, rehabilitation methods including hearing aids and cochlear implants may be beneficial. Cochlear implants improve speech perception in those with bilateral significant hearing loss. It is generally advised to get vaccinated against mumps to avoid hearing loss linked to the disease.

Keywords: Cochlear implantation, intratympanic steroids, mumps, sensorineural hearing loss

INTRODUCTION

Mumps is a viral illness with an acute onset caused by the mumps virus.^[1] It primarily affects the parotid glands, resulting in swelling and discomfort of the parotid gland.^[1] Mumps is usually associated with fever, headache, muscle aches, and parotid gland swelling but can also result in mild-to-severe sensorineural hearing loss (SNHL) due to damage to the cochlea and/or auditory nerve.^[1] The mumps usually spreads through saliva and reaches the inner ear/labyrinth through the bloodstream.^[2] In the inner ear, the virus causes a labyrinthitis, damaging the cochlear hair cells.^[2] It can also damage the vestibular component of the labyrinth.^[3] Hearing loss in mumps may be unilateral, sudden in onset, profound, and permanent.^[2] The pathophysiology of SNHL due to mumps is thought to be direct invasion of the virus into the cochlea damaging the organ of Corti, the cochlear nerve myelin sheath, and degeneration of stria vascularis, tectorial, and Reissner's membrane.^[4] There is a definitive treatment for mumps infection, so interventions are usually supportive. High dose of steroids can be prescribed to reduce inflammation and improve the chances of hearing recovery.^[5] Corticosteroids can be prescribed for preventing the mumps-induced hearing loss.^[5] For the treatment of SNHL in mumps, hearing aids can be used in case of mild-to-severe hearing loss.^[5] In case of bilateral severe to profound SNHL,

cochlear implantation has been shown to be effective.^[6] Mumps is a vaccine-preventable disease, and immunization is the important countermeasure to combat this infection. Mumps is known for its poor prognostic features in respect to restoration of hearing, especially those with profound hearing loss.^[4]

METHODS OF LITERATURE SEARCH

A search was conducted for research articles on the mumps-associated SNHL using various methods. This began with searching online databases such as Scopus, PubMed, Medline, and Google Scholar. A search strategy was created based on the Preferred Reporting Items for Systematic Reviews and Meta-Analysis guidelines. The search approach found published article abstracts, and citations were used to manually find more research publications. The suitability of observational studies, comparative studies, case series, case reports, and randomized controlled trials for inclusion in this review was

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evaluated. A total of 61 articles (23 case reports, 17 case series, and 21 original articles) were found across various databases, with 44 being included in this review [Figure 1]. This article discusses the prevalence, etiopathology, clinical presentations, diagnosis, current treatment, vaccination, and prognosis of mumps-associated SNHL.

PREVALENCE

The exact number of mumps cases is difficult to determine as many patients may not require medical attention, and clinical diagnosis may be inaccurate.^[4] The incidence of mumps-associated deafness is uncertain and usually unilateral, which can be overlooked. If hearing loss occurs later, identifying the etiology can be challenging.^[4] The implementation of widespread vaccination program has significantly reduced the incidence of mumps worldwide.^[7] However, mumps remains a significant health concern in the countries with lower immunization rates and among people with hesitancy of vaccination or lower access to healthcare.^[7] Epidemiologically, mumps-induced SNHL is more common among children and young adults, although it can affect people of any age. The viral etiology causing bilateral SNHL shows only 10% of cases of SNHL, including mumps, herpes simplex virus, human immunodeficiency virus, and viral upper respiratory tract infections.^[8] It has been estimated that the incidence of hearing loss due to mumps ranges from 1 per 1000 to 1 per 20,000 cases of mumps.^[9] Another study shows that SNHL occurs in 0.005%–0.3% of all mumps patients.^[10] There is a higher incidence of postmumps SNHL reported (4% of adult mumps).^[11] One report shows that hearing loss associated with mumps is approximately 5 out of 10,000 cases.^[12]

ETIOPATHOLOGY

Mumps is a common viral infection in pediatric and young adult age groups that is caused by a paramyxovirus, a single-stranded RNA virus of the Paramyxoviridae family.^[4] It is an acute, self-limiting systemic disease that may cause serious complications.^[4] Mumps can attack several organs

such as the salivary glands, pancreas, testes, meninges, and inner ear.^[13] This viral infection can directly damage the inner ear structures.^[13] Mumps is highly contagious and can have a long list of complications.^[4] SNHL is a well-known side effect of mumps.^[14] Hematogenous invasion is the main way that viruses enter the cochlea, and hearing loss can happen before, during, or after a mumps infection.^[15] A study revealed that the primary route of invasion of the virus is hematogenous and offers the word “viral endolymphatic labyrinthitis” as the likely pathophysiology of the hearing loss in mumps.^[15]

According to this study, the clinical and cerebrospinal fluid examinations rule out both tympanogenic and meningogenic pathways for viral invasion of the inner ear.^[15] It is believed that direct infiltration of the cochlea damages the organ of Corti, the auditory nerve myelin sheath, and degenerates the stria vascularis, tectorial, and Reissner’s membrane, which is the pathophysiology of SNHL in mumps.^[16] One study provided experimental evidence that the degeneration of the organ of Corti is the source of hearing loss associated with mumps.^[17] The tectorial membrane, organ of Corti, and stria vascularis are severely deteriorated at the cochlea’s basal turn and gradually deteriorate as it approaches the apex.^[18] However, there is a very little degeneration of the peripheral cochlear nerve at the basal coil, and the ganglion cells are almost normal. Some cases of asymptomatic or mild infection of mumps may present with hearing loss.^[19] According to one study, histological analysis of mumps deafness revealed degeneration of the stria vascularis and organ of Corti.^[20] Meningoencephalitis is another complication due to mumps. Meningoencephalitis can also manifest hearing loss due to damage to the vestibulocochlear nerve. Isolated mumps virus was detected from the perilymph of patients with sudden hearing loss following mumps infections.^[21] Thus, labyrinth might be the site of lesion following mumps in most of the patients.

CLINICAL PRESENTATIONS

Mumps is an endemic disease in most urban population all over the world. It is more common among children with age between 5 and 10 years.^[1] The flu-like illness and bilateral parotid gland swelling are the most typical clinical signs of mumps.^[22] Fever, headache, and muscle pain are typical features of mumps, which are followed by enlargement of the parotid and/or submandibular salivary glands. The swelling of the parotid gland is one of the most typical presentations of the mumps. Nonetheless, the subclinical infection is known to be widespread, and about 30% of patients may not have any parotid gland swelling at all.^[23]

Pancreatitis, orchitis, oophoritis, aseptic meningitis, encephalitis, and SNHL are among the complications that can infrequently result from mumps.^[23] The inner ear may be affected by mumps, even though the salivary glands are frequently affected. Hearing loss brought on by a mumps infection can happen prior to, concurrently with, or even

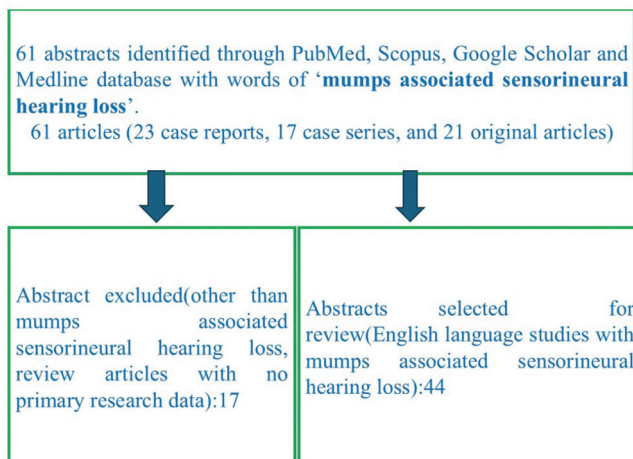


Figure 1: Methods of literature search

without parotid gland swelling.^[24] In majority cases of mumps, the symptoms are mild, but some present serious outcomes such as meningitis, encephalitis, and deafness. Viral-induced hearing loss can range from mild-to-severe vertigo and dizziness, which have also been reported in roughly half of post-mumps SNHL patients.^[25] Therefore, after a mumps infection, the vestibular system and the cochlea may be affected.

Hearing loss

SNHL is a well-known complication of mumps.^[4] The hearing loss is often sudden onset, unilateral, profound, and permanent.^[24] Hearing loss due to mumps can manifest in three different ways such as complete unilateral, partial unilateral, and bilateral complete SNHL.^[24] The bilateral complete SNHL is very uncommon type. The hearing loss in mumps is a serious and lifelong complication that occurs from 4 days before to 18 days after the swellings of the parotid and/or submandibular glands.^[26] One study suggests that SNHL might develop irrespective of the severity of the mumps infections.^[27] However, the incidence of vaccine-induced SNHL is considered to be lower than that from natural infection.^[28]

DIAGNOSIS

Management of mumps-induced SNHL includes early detection and intervention of mumps to improve the outcomes.^[5] The diagnosis is mainly clinical, supported by audiometric testing for confirming the degree and type of hearing loss. If a child is diagnosed with mumps, the hearing tests should be performed as soon as possible.^[5] If at any time mumps infections occur and hearing loss is suspected, it should be investigated earliest. In young children, single-sided deafness may go unnoticed for a long time. Hearing loss may be found in three different types. The most common type of hearing loss is sudden unilateral complete loss, next is unilateral partial deafness, and the rarest type is bilateral complete deafness.^[25] The measurement of anti-immunoglobulin M (IgM) antibody for mumps is helpful to detect silent mumps infection with SNHL. The positive anti-IgM antibody in SNHL suggests silent mumps infection in 7.2% of cases.^[29] Magnetic resonance imaging and other imaging can be performed to rule out other potential causes of SNHL. The diagnostic criteria for mumps associated deafness is provided in Table 1.

TREATMENT

Despite various treatment options, hearing often has not been regained in mumps.^[30] There is no proven treatment for sudden SNHL.^[30] Corticosteroids are helpful to reduce the inflammation and swelling within the inner ear, although their efficacy in the treatment of viral-induced SNHL remains uncertain. The use of intratympanic steroidal injection has increased recently. With this method, the afflicted tissue receives higher steroid concentrations. The vasodilators, anticoagulants, and hyperbaric oxygen therapy are also very helpful to restore hearing loss in mumps.^[30] Although corticosteroids

Table 1: Criteria for mumps-associated deafness

Criteria for deafness in mumps

Definite

Patients with evident clinical signs of mumps, such as swelling of the parotid and submandibular glands, and sudden severe hearing loss during the period from 4 days before to 18 days following such swellings

Patients without evident clinical signs of mumps, but have a significant increase in antimumps serum antibody titer within 2–3 weeks following onset of sudden hearing loss

Almost definite

Patients in whom IgM antibody to mumps virus is detected within 3 months following onset of sudden hearing loss

For reference

Patients in whom mumps-associated deafness is suspected clinically

IgM: Immunoglobulin M

have been administered in case of SNHL in mumps as in idiopathic sudden SNHL, the prognosis for patients with profound hearing loss is often poor.^[31] However, spontaneous recovery in patients of mumps with mild hearing loss has been documented.^[32] The unilateral profound SNHL is often refractory to several treatments such as steroids, vasodilators, Vitamin B12, and hyperbaric oxygen therapy.^[19] Meanwhile, patients with severe hearing loss present improvement. In some cases, antiviral agents may be considered, although there is limited evidence supporting their effectiveness specifically for mumps-induced SNHL. The rehabilitations of mumps-induced SNHL include hearing aids and cochlear implants and may be required for individuals with significant or permanent hearing loss. Patients with mumps-related deafness are considered good candidates for cochlear implantation, although central nervous system damage is typically linked to conditions such as meningitis and encephalitis. Cochlear implantation is useful for bilateral profound hearing loss and provides a good outcome for speech perception. Early cochlear implantation is strongly recommended for children with sudden hearing loss. However, the potential for a poor outcome should be communicated to the parents. Early implantation leads to good speech perception, while late implantation is associated with less favorable results.^[18] However, the central nervous damage associated with meningitis and encephalitis should be ruled out before cochlear implantation. In case of single-sided deafness, contralateral routing of signals hearing aids and bone-anchored hearing aids are good options. Mumps cannot be treated with a specific antiviral medication. It is commonly acknowledged that the mumps immunization is efficacious.

VACCINATION

There is no effective treatment for deafness due to mumps, so the importance of prophylaxis by vaccination should be emphasized. Preventing measures remain the most effective option for reducing the incidence of mumps and its complication. The vaccination coverage with measles-mumps-rubella (MMR) vaccine is crucial in preventing outbreaks and protecting individuals from severe

consequence of mumps-induced SNHL. A report from the World Health Organization documented that the countries that have received high vaccine coverage have shown a rapid decline of mumps morbidity, and deafness associated with mumps have almost vanished.^[33] A second dose of the mumps vaccine is typically recommended for children to prevent mumps, with the first dose given at 12–18 months of age and the second dose at 4–6 years old.^[29] However, there has been a reported case of sudden unilateral profound hearing loss affecting cochleovestibular function following the administration of the mumps vaccine.^[29] The incidence of deafness caused by the vaccine is believed to be lower than that resulting from natural mumps infection. One study documented that it is surprising for a developed nation to lack a prophylactic vaccination against mumps.^[34] Routine vaccination against mumps should be considered to decrease the incidence of mumps-related deafness. While the MMR vaccine is used in many countries, routine vaccination against mumps is also implemented to prevent mumps infections. In several regions, the MMR vaccine has been offered as an alternative to the monovalent mumps vaccine for routine immunization.

PROGNOSIS

Mumps is an acute, self-limiting systemic disease that can result in complication of SNHL. The failure of treatment is related to the severity of the cochlear damage.^[5] Once SNHL occurs due to a viral cause, restoring hearing through medical treatment is unlikely. SNHL caused by mumps is typically permanent, regardless of the treatment. The prognosis for patients with profound SNHL is generally poor, except for those who undergo cochlear implantation surgery, which offers a positive outcome in terms of speech and sound perception for patients without central nervous system damage.^[35] Mumps-induced SNHL can be prevented through vaccination, which helps avoid outbreaks. The psychological and social impact of SNHL is considerable for both patients and their families.^[36] The clinicians should inform the parents or guardians about the permanent damage of the cochlea by mumps infections and counseling them about sudden unexpected complete hearing loss that affect the qualities of life.^[37]

CONCLUSION

SNHL is a serious and permanent complication of mumps. Mumps virus can damage the labyrinth through direct viral invasion and inflammation. The majority of deafness in mumps presents as unilateral SNHL with sudden in onset. The hearing loss usually occurs without warning, ranging from mild to profound in severity. The unilateral profound SNHL due to mumps is common and refractory to treatment. Treatment includes high-dose steroids to reduce inflammation and improve the chances of hearing restoration. In cases of bilateral profound hearing loss, cochlear implants are beneficial. The vaccination against mumps is safe and effective and is recommended to prevent loss associated with mumps.

Mumps must be considered for the vaccination program because of its complications such as hearing impairment and infertility.

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Conflicts of interest

There are no conflicts of interest.

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