Case report

Sudden total bilateral deafness due to asymptomatic mumps infection

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Received 27 March 1998; received in revised form 17 June 1998; accepted 20 June 1998

Abstract

Mumps is the most common cause of unilateral acquired sensorineural hearing loss in children. Although it usually affects the salivary glands, the inner ear may be involved. Deafness is usually unilateral, sudden in onset, profound and permanent. Bilateral total sensorineural hearing loss had rarely been reported in English literature. We present a case of total deafness due to asymptomatic mumps infection. © 1998 Elsevier Science Ireland Ltd. All rights reserved.

Keywords: Mumps deafness; Sudden hearing loss; Autoimmune inner ear disease

1. Introduction

Mumps is an endemic disease in most urban populations all over the world, particularly occurring in children aged 5–10 years [1]. Although it usually affects the salivary glands, the inner ear may be involved [2]. Hearing loss associated with mumps is reported five out of 10000 cases [2]. Deafness is usually unilateral, sudden in onset and profound [2]. Bilateral total sensorineural hearing loss had rarely been reported in English literature. We present a case of total deafness due to asymptomatic mumps infection and our original ‘immunesuppressive’ therapeutic regimen.

2. Case report

A four year old girl was referred to our ENT department by her parents. They complained that
their daughter suddenly began not to answer to their sound stimuli 10 days previous. They had not noticed parotid swelling or tenderness, vertigo, nausea or meningeal signs but marked a mild fever and fatigue before the event. The remainder of the history (prenatal, delivery, postnatal, hereditary or systemic diseases, ototoxic medication, trauma) and ENT examination were negative, except the responsiveness to sound stimuli at the first meeting. Previously, the communicative abilities of the patient were well adjusted with her age, her parents did not notice any difference between the right and left ear’s hearing. This anamnestic data helped us to exclude a possible congenital (for example; congenital unilateral anacousis) or familial ethiology. She did not respond well to pure tone audiometric evaluation. Tympanogram was normal (Type A). Brainstem evoked response and audiology elicited no response at 100 dB HL bilaterally. Her responses in the sound field were not improved after the hearing aid. Complete blood count and urinalysis were within the normal limits. A viral antibody panel (Melotec Biotechnology, Melotest, Spain) was carried out for ethiological evaluation and the results are presented in Table 1. These results indicated a sudden total deafness due to asymptomatic mumps infection. Immediate therapy was instituted with our standard ‘sudden sensorineural hearing loss’ regimen (Table 2). Hearing loss did not resolve on this regimen after a week, and the drugs were discontinued. We did not observe any side effects.

Table 1
Serologic results

<table>
<thead>
<tr>
<th>Antibody</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anti-mumps IgM</td>
<td>Positive</td>
</tr>
<tr>
<td>Anti-mumps IgG</td>
<td>Positive (low titre)</td>
</tr>
<tr>
<td>Anti-measles IgM</td>
<td>Negative</td>
</tr>
<tr>
<td>Anti-measles IgG</td>
<td>Negative</td>
</tr>
<tr>
<td>Anti-rubella IgM</td>
<td>Negative</td>
</tr>
<tr>
<td>Anti-rubella IgG</td>
<td>Positive</td>
</tr>
<tr>
<td>Anti-CMV IgM</td>
<td>Negative</td>
</tr>
<tr>
<td>Anti-CMV IgG</td>
<td>Negative</td>
</tr>
</tbody>
</table>

Table 2
Istanbul faculty of medicine therapeutic regimen for sudden sensorineural hearing loss

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>Azathioprine</td>
<td>First day 4 mg/kg oral one dose</td>
</tr>
<tr>
<td></td>
<td>Other 6 day 1 mg/kg oral one dose</td>
</tr>
<tr>
<td>Methyprednisolone</td>
<td>Seven day 1 mg/kg t.i.d oral</td>
</tr>
<tr>
<td>Famotidine (H2 receptor antagonist for gastric protection)</td>
<td>40 mg oral daily</td>
</tr>
<tr>
<td>Duretic (volume protection against steroid usage)*</td>
<td>100 mg/d triamterene+50 mg/d hydrochlorothiazid oral</td>
</tr>
</tbody>
</table>

*These drugs are used for adults; in this case we added only an antacid to our azathioprine steroid combination.

3. Discussion

Mumps is the most common cause of unilateral acquired sensorineural hearing loss in children [2]. The reported incidence varies between 1/30000 and 1/2000 [3]. Hearing loss may occur before, during or after the parotitis, but also in subclinical cases [2]. In ~30–40% of the cases, the infection is in asymptomatic form [2]. The deafness is usually sudden in onset, severe, more pronounced in high frequencies and permanent [2,3]. Hearing loss may be seen in three different variety [3]. The most common form is sudden unilateral complete loss, the next is a unilateral partial deafness and rarest variety is bilateral complete deafness [3]. Bilateral deafness had been reported in only 22 patients until 1957 and since then very few cases have been added [3–5].

The hearing loss is sensorineural and may be due to direct viral invasion of the cochlea, affecting the organ of Corti and tectorial membrane, myeline sheath of the eight nerve [2]. Microscopic studies on humans showed degeneration and atrophy of the stria vascularis, tectorial membrane and organ of Corti and collapse of the Reissner’s membrane [3]. Tanaka et al. showed the degeneration of the organ of Corti which was usually found in the basal turn in experimentally mumps virus induced labyrinthitis [6].

Mizushima et al. presented their series of mumps deafness and suggests that the primary
route of invasion of the virus is hematogenous and proposes the term 'viral endolymphatic labyrinthitis' as the possible pathogenesis of the deafness. According to this study, both tympanicogenic and meningogenic routes of viral invasion to the labyrinth are excluded on the basis of the clinical and cerebrospinal fluid studies [7].

Many therapeutic regimens such as vasodilators, steroids, carbogen, hyperbaric oxygen, plasma expanders, etc. have been suggested for the sudden hearing loss. We have routinely practised azathioprine combined methyprednisolone regimen for idiopathic sudden sensorineural hearing loss since 1992 and have observed a satisfactory recovery (partial and complete). We believe that sudden hearing loss develops with an immune mediated reaction after a viral induction (except some clear vascular, traumatic, etc. etiologies). This 'immune mediated inner ear damage' theory is supported by our clinical, experimental and some international studies [8–10]. In this case, azathioprine combined methyprednisolone regimen failed. Failure may be concerned with:

1. Therapy has been started late,
2. cochlear damage due to mumps is severe and irreversible,
3. inner ear damage due to mumps is not associated with immune system.

The hearing aid was ineffective in our patient, and a rehabilitation therapy was introduced. There was no improvement 3 months later and she was accepted as a candidate for our cochlear implantation programme.

Although there have been some sensorineural deafness cases reported after measles–mumps–rubella (MMR) immunisation, the main therapy for mumps deafness is prevention with vaccination today [11,12]. In Turkey, a vaccination (MMR) programme is carried out at approximately 15 months.

References