Case Report

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Mumps Labyrinthitis, Endolymphatic Hydrops and Sudden Deafness in Succession in the Same Ear

Key Words
Mumps
Endolymphatic hydrops
Sudden deafness

Abstract
Acute sensorineural hearing loss, mostly unilateral and reversible, is a well-known complication to mumps. Secondary endolymphatic hydrops, Ménière's syndrome, has rarely been associated with a previous mumps infection. This paper presents the case report of a woman who experienced unilateral hearing loss, vestibular symptoms and a caloric depression on the same ear during mumps. The symptoms and findings were reversible. Twelve years later she developed Ménière's symptoms in the same ear. This continued for 2 years after which she suddenly had a sensorineural hearing loss. This was localized in the mid- and high-tone area and was almost identical with the initial hearing loss 14 years earlier. Viral damage to the resorptive structures of the inner ear seems to have caused the hydrops. It also seemed to have weakened the neuronal structures of the ear, letting the initial damage become overt after repeated attacks of pathological pressure changes.

Introduction

Acute sensorineural hearing loss is a well-known complication to mumps [1]. Secondary endolymphatic hydrops, Ménière's syndrome, has also rarely been associated with virus, i.e. mumps. This paper presents the case report of a woman who experienced unilateral, reversible sensorineural hearing loss during mumps. Twelve years later she developed Ménière's symptoms in the same ear. This continued for 2 years after which she suddenly got a sensorineural mid- and high-tone loss in the same ear, which was almost identical with the initial hearing loss 14 years earlier.

Case Report

A previously healthy woman, born in 1949, fell ill with mumps in February 1976, at the age of 27. The symptoms started 18 days after her son got the disease. Six days after her initial symptoms of parotitis she experienced sudden vertigo, nausea and deteriorated hearing in her right ear. Four days afterwards she saught help at the Clinic for Infectious Diseases in Linköping. A lumbar puncture was performed, which showed a pathological mononuclear cell increase in accordance with a serous meningitis. Due to her audiovestibular symptoms the patient was referred to the ENT Clinic in Linköping, where she was hospitalized and investigated by the author.

At investigation the patient was somewhat pale, unsteady but without clinical signs of meningitis. The swelling of the salivary glands was on return. A left-beating nystagmus was observed in Frenzel's glasses. The initial pure-tone audiogram showed a mid- and high-frequency sensorineural hearing loss on the right side (fig. 1a). Speech audiometry was normal while the stapedius reflex test

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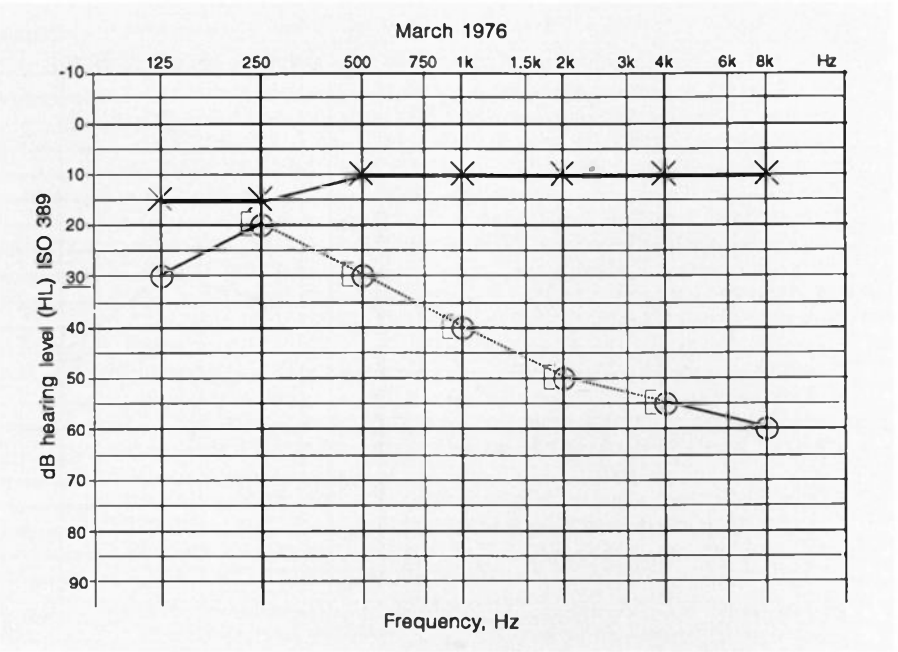

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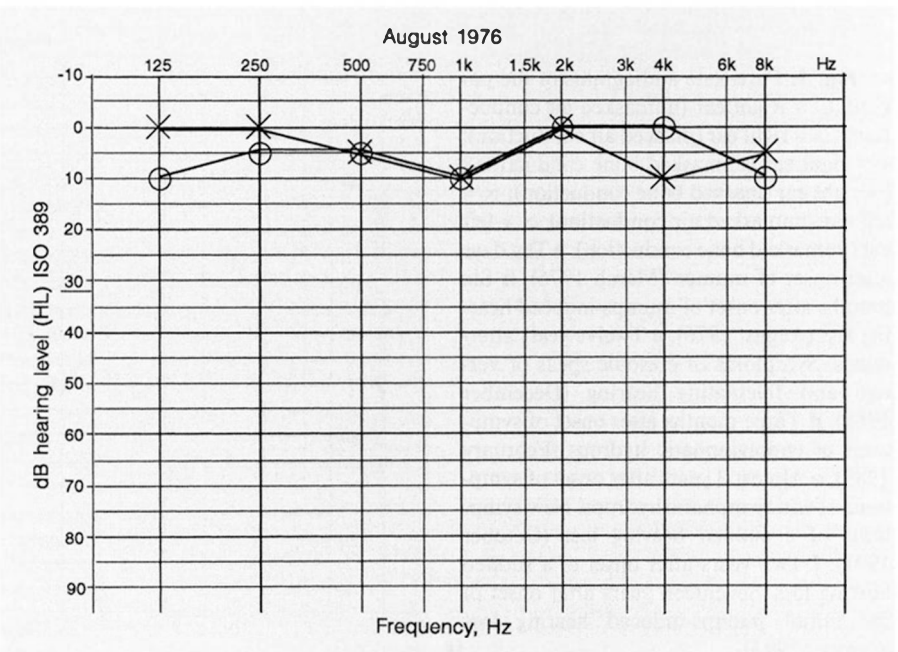

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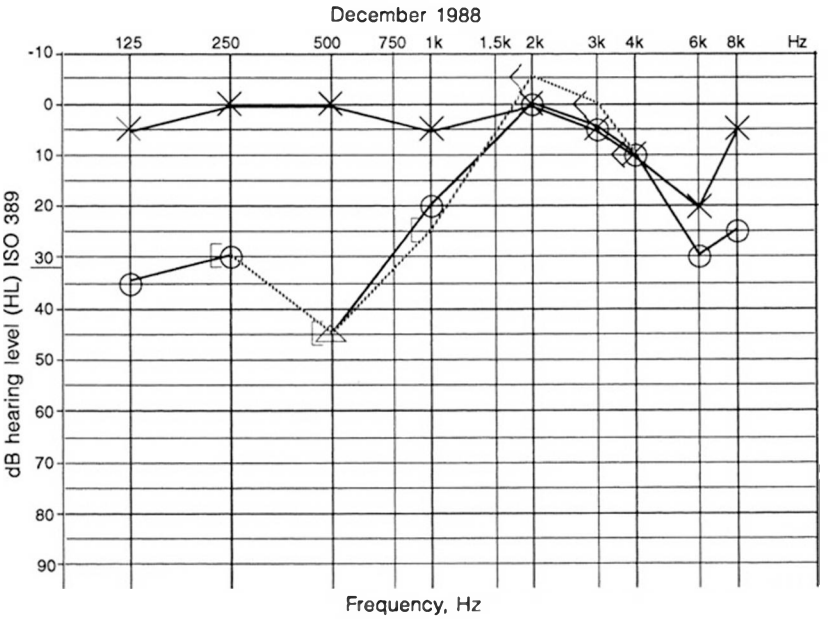
Fig. 1a–f (for legend see page 340).

showed recruitment in the right ear. The ENG revealed a vivid left-beating nystagmus and the caloric response was severely reduced on the right ear. Her symptoms improved and she was sent home after 5 days in hospital. At a checkup 1 month after onset her hearing had improved. The spontaneous nystagmus was gone, while a severe caloric depression was present. Three months after onset there were no audiovestibular symptoms left. Pure-tone audiogram had further improved and the caloric response was better than before. Six months after onset the pure-tone audiogram was normal (fig. 1b) and the caloric response on the right ear was almost normalized. No further control was decided.

Twelve years after this episode the patient returned to the ENT Clinic due to renewed audiovestibular symptoms. In the period in between she had been completely free of such symptoms. In December 1988, however, she started to get recurrent spells of vertigo with a duration of some hours. At the same time hearing deteriorated in her right ear and she had a feeling of fullness in the same ear. A new pure-tone audiogram showed a basal-tone loss on the right side



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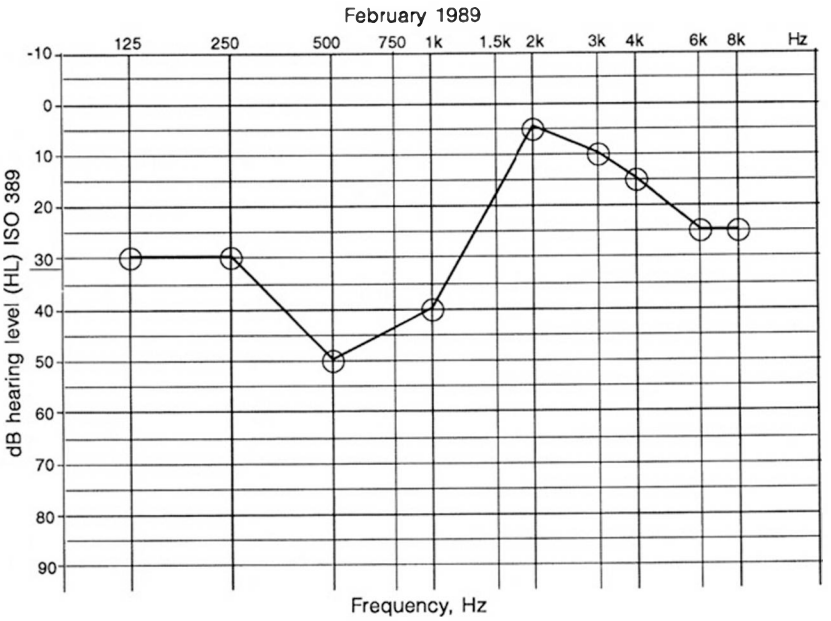

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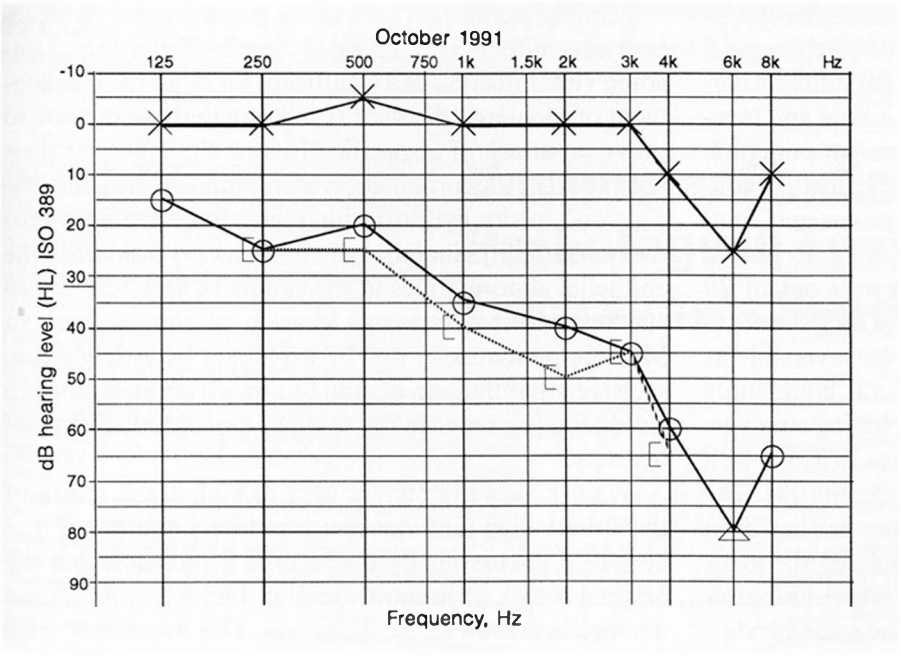
Fig. 1. Pure-tone audiograms of the patient. ○ = Right ear (unmasked air conduction); △ = right ear (masked air conduction); < = right ear (unmasked bone conduction); [= right ear (masked bone conduction); × = left ear (unmasked air conduction); > = left ear (unmasked bone conduction). **a** Ten days after onset of mumps (March 1976). **b** Six months after onset of mumps-induced hearing loss (August 1976). **c** Twelve years afterwards. Symptoms of episodic spells of vertigo and fluctuating hearing (December 1988). **d** Three months after onset of symptoms of endolymphatic hydrops (February 1989). **e** Almost 3 years after onset of symptoms of endolymphatic hydrops. New symptoms of a sudden hearing loss (October 1991). **f** Two years after onset of a sudden hearing loss. Seventeen years after onset of the initial mumps-induced hearing loss (October 1993).

(fig. 1c), which fluctuated between investigations (fig. 1d). The ENG showed no spontaneous nystagmus but a slight caloric depression (22% asymmetry) of the same magnitude as had been noted on the right ear at the last checkup in 1976. A BRA was normal. The patient was, due to her symptoms and findings of an endolymphatic hydrops in her right ear, put on diuretics (bendroflumethiazide with potassium chloride). The situation was, however, not stabilized. Slow-release furosemide was instituted in February 1989. From then on up to 1991 the patient's Ménière symptoms were fairly well under control. The pure-tone audiogram showed only a slight base-tone loss at a checkup in January 1991. She had some problem with tension headache which was relieved by physiotherapeutic treatment.

In October 1991, however, there was a sudden deterioration of hearing in her right ear over a day. At the same time she had a feeling of unsteadiness, but there were no vertiginous spells. A pure-tone audiogram (fig. 1e) was very similar to the initial one made in 1976 with a mid- and high-frequency loss in the right ear. Hearing was normal in the left ear. No nystagmus was observed, neither with



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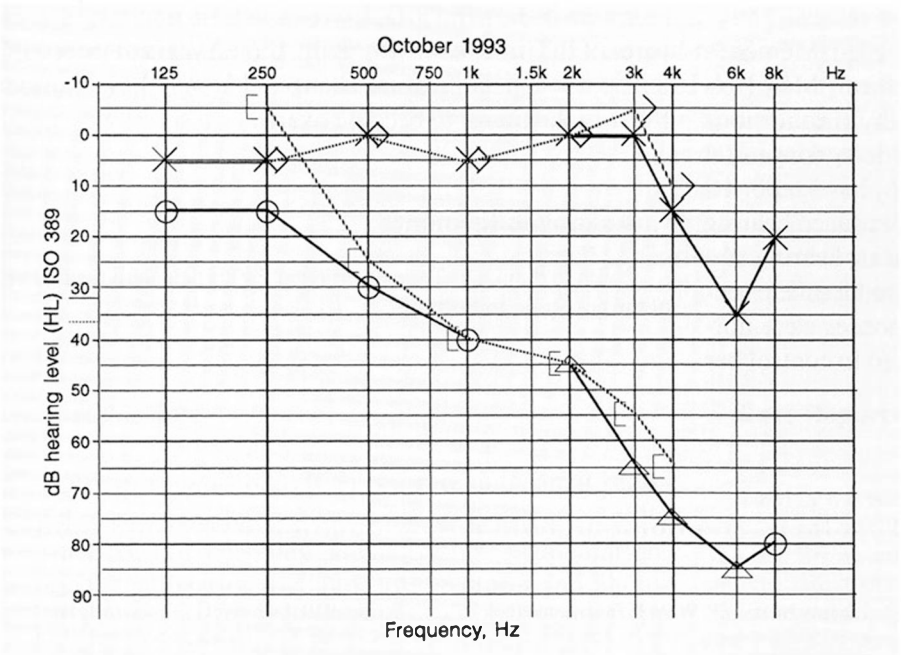

1f

Frenzel glasses nor during ENG. The caloric reactions were within normal range (0–20% asymmetry). Posturography was also normal. An MRT was performed which excluded pontine angle pathology as well as other pathologies in the posterior fossa or brainstem. A complement fixation test for epidemic parotitis virus was negative. At follow-ups – the last one in October 1993 – she continued to have a nonfluctuating hearing loss with a slight tinnitus in her right ear. There were no balance problems. The pure-tone audiogram was unchanged at this last checkup compared to that 2 years earlier (fig. 1f).

Discussion

Total recovery from the initial sensorineural hearing loss was found in the present patient. In the Finnish study by Vuori et al. [1] a high incidence of recovery characterized their patients with mumps-induced hearing losses. They found total or almost total recovery in 12 out of 13 persons. Most of the cases showed, as in this patient, hear-

ing losses in the high-tone range. In the present case serous meningitis was diagnosed according to the cerebrospinal fluid investigation. However, in the Finnish study no correlation was found between hearing impairment and meningitis [1]. All their patients underwent lumbar puncture. Bilateral hearing losses have also been described in connection with mumps [2], vestibular symptoms and findings as well. We found partially impaired or totally absent caloric reactions on the ipsilateral side in 9 out of 20 patients (45%) with permanent hearing losses due to mumps [3]. Vestibular lesions can also be reversible as illustrated by the present case. A temporal bone study showed severe lesions of the inner ear including stria vascularis, tectorial membrane and the organ of Corti in a child who went deaf in conjuction with mumps [4]. The penetration of mumps virus into the inner ear has been verified by Westmore et al. [5]. They isolated the virus from the perilymphatic fluid in a patient who 1 year after a successful stapedectomy went deaf on the same ear during mumps. Exploration was done in order to exclude a perilymphatic fistula.

Secondary endolymphatic hydrops, called Ménière's syndrome in contrast to the idiopathic form, Ménière's disease, is mentioned together with a variety of conditions such as otosclerosis, autoimmune disorders, congenital lues and virus. Schuknecht and Gulya [6] have reported on a young patient, who 3 years a mumps-induced hearing loss experienced episodic vertigo, fluctuating hearing and tinnitus in the same ear. Audiogram showed a severe sensorineural hearing loss while caloric responses were normal. Labyrinthectomy had to be performed to control her symptoms.

Modern techniques such as the polymerase chain reaction have hitherto not revealed clear evidence for an ongoing viral infection as a significant factor in the development of Ménière's disease [7]. While a definite method to prove an attack of a specific virus on the inner ear does not yet exist, the combination of immunoelectron microscopy and in situ hybridization seems to be promising for this purpose [8]. Shambaugh and Wiet [9] think that the epithelial abnormalities in the endolymphatic sac as well as the abnormal size and location of the sac seen in Ménière patients can well be explained by a slowly progressive inflammation of viral (mumps?) origin starting in childhood [9]. Later in life this can cause clinical signs of hydrops.

What makes the present case special, is the return of the initial high-tone hearing loss after an intercurrent period of fluctuating base-tone loss. It indicates that the original attack of mumps virus on the inner ear caused damage in the resorptive structures. This manifested clinically as symptoms of endolymphatic hydrops. Also, it seems as if the viral attack weakened the neuronal structures of the inner ear. After more than 2 years of intermittent pathological pressure changes this may have caused the initial damage to become overt.

Acknowledgements

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