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Unusual cases of delayed endolymphatic hydrops

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Abstract
The objective of this report is to emphasize the clinical features of delayed endolymphatic hydrops in comparison with Ménière's disease. Four cases were selected from a series of 16 examined between the years 1993 and 2005, due to their unusual characteristics. A short clinical history and significant tests are presented for each of the four cases. The cases of delayed endolymphatic hydrops discussed in this article suggest that hydrops is the most important underlying pathology that causes the hearing loss and the vestibular symptoms both in the better ear and in the ear with severe hearing loss. They also suggest that this condition probably occurs in patients with congenitally sensitive ears that make them prone to the development of the late hydrops.

Keywords: *Labyrinth diseases, endolymphatic hydrops, Ménière's disease*

Introduction

Delayed endolymphatic hydrops was first described in 1975 by Nadol et al. [1] and Wolfson and Leiberman [2].

The expression 'delayed endolymphatic hydrops' was proposed by Schuknecht in 1978 [3]. He classified the disorder as ipsilateral, contralateral, or bilateral. The ipsilateral type is characterized by the early onset of a profound hearing loss in one ear. Several years later the patient begins to present vertiginous attacks, often with nausea and vomiting. Tinnitus and intra-aural pressure may also be associated. In the contralateral type the patient presents fluctuant and progressive hearing loss, most often accompanied by vertiginous episodes, in the ear opposite to the one that experienced the early hearing loss. Patients with bilateral hydrops experience hearing loss in both ears, several years before the onset of vertigo.

Schuknecht et al. [4] made a histopathological analysis of the temporal bones of two patients with contralateral delayed endolymphatic hydrops. The findings in the deaf ears were consistent with viral labyrinthitis, and those of the hearing ears were similar to Ménière's disease.

From 1993 to 2005, 16 cases of delayed endolymphatic hydrops were seen and treated in my private clinic. Twelve of them were ipsilateral and four were contralateral; the bilateral type was not encountered. In the same period of time 411 patients with Ménière's disease were seen. This report describes four cases that presented unusual clinical findings.

Case report

Case 1. Female, born 2 December 1934. This patient became profoundly deaf in the right ear when she was 2 years old and had measles. At age 25 she began to present episodes of vertigo, fluctuant hearing loss and tinnitus in the left ear. Her hearing in this ear usually became better after the vertiginous attacks. She was medically treated for Ménière's disease and was asymptomatic for a period of 20 years. When seen in 1991 she had become obese and diabetic and in the last 4 years she again presented fluctuant hearing loss, tinnitus and vertiginous attacks. These symptoms subsided when she was treated for metabolic inner ear disorder, losing weight and controlling her diabetes more rigidly. Figure 1 shows the audiological findings for this patient.

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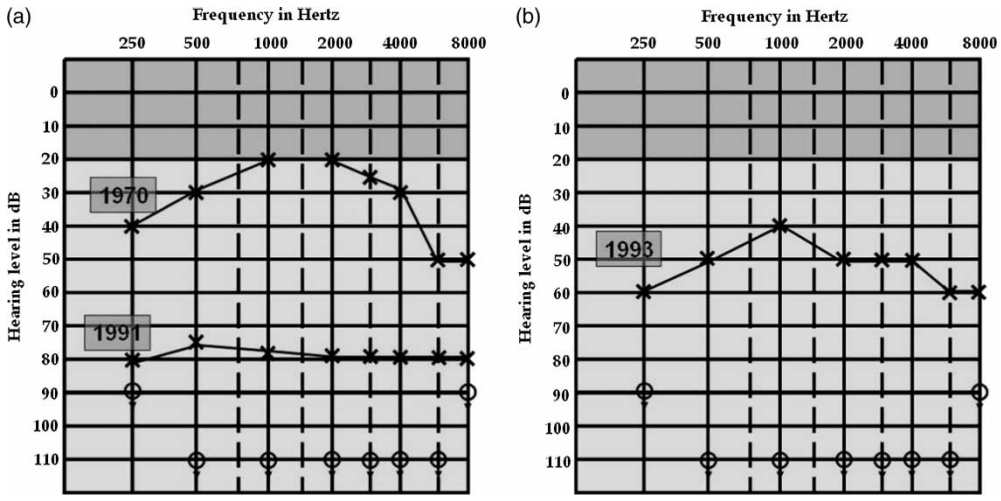

Figure 1. Audiograms of case 1: (a) in 1970 and in 1991, when she became diabetic and obese; and (b) in 1993, after treatment and weight loss.

Case 2. Female, born 28 November 1923. This patient had experienced profound sensorineural hearing loss in the right ear since childhood. When seen in 1968 she had been suffering from incapacitating vertiginous attacks for the previous 4 years. Her ear, nose and throat examination was essentially normal. Her audiograms are shown in Figure 2.

On 17 December 1968 she was submitted to a right ear labyrinthectomy, performed through a mastoid approach, with the destruction of the three semicircular canals.

She recovered well from the operation and, after the compensation period, remained asymptomatic until March 1973. At this time she began to complain of fluctuant hearing loss in the left ear and occasional episodes of unsteadiness. The audiological examination confirmed the presence of hearing loss. She received medical treatment for Ménière's disease. Her hearing improved and the unsteadiness was markedly reduced.

Case 3. Female, born 27 August 1956. This patient had experienced hearing loss since childhood and was seen in 1990 with a complaint of episodes of vertigo in the previous 3 years. Some of these episodes were accompanied by nausea and vomiting. The ear, nose and throat examination was essentially normal. The audiogram showed intense sensorineural hearing loss in the right ear (Figure 3). The vestibular tests showed a slightly hypoactive right labyrinth. She was advised to have surgery on the right ear but she preferred to try a medical treatment. In the following years she had other episodes of vertigo, including a very intense one that required hospitalization. In 1993 she underwent an endolymphatic subarachnoid shunt, performed with a

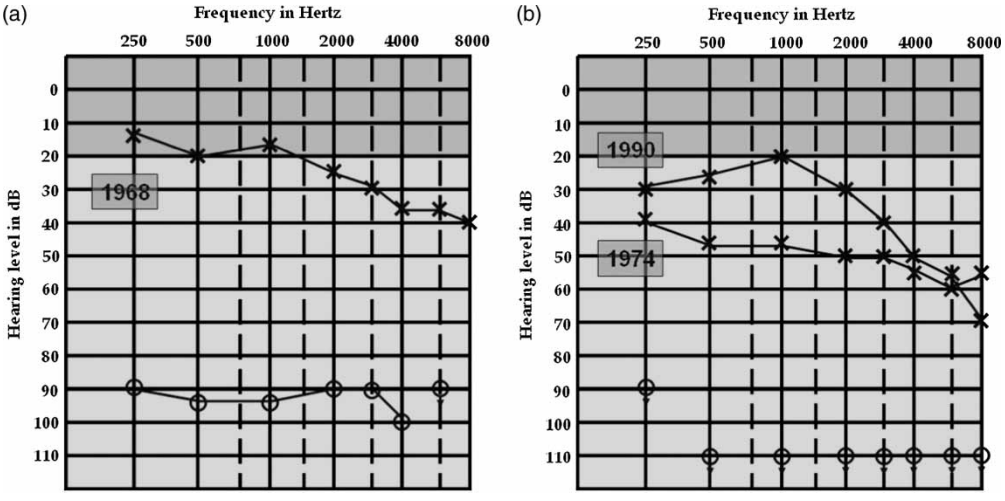

Figure 2. Audiograms of case 2: (a) the thresholds of her first consultation in 1968; (b) the thresholds in 1974 and 1990.

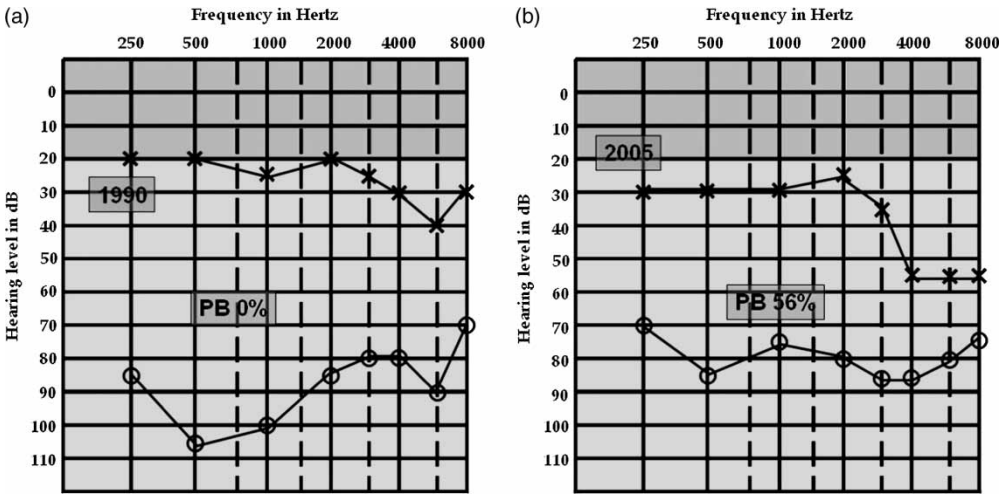

Figure 3. Audiograms of case 3: (a) before and (b) after surgical treatment.

House-type shunt tube. Her hearing thresholds improved on the operated ear and the discrimination changed from 0 to 56%. She has had no more episodes of vertigo up to her last visit in November 2005.

Case 4. Female, born 26 October 1972. This patient had experienced profound hearing loss in the right ear since childhood. She was married at age 22 and during her second pregnancy began to notice fluctuant hearing loss in the left ear. Gradually the hearing in the left ear deteriorated and she began to wear a hearing aid in the left ear. Hearing tests performed 2 months after childbirth indicated a profound hearing loss in the right ear and a moderate sensorineural loss in the left ear (Figure 4). Two months prior to her first consultation she began to have intense vertiginous episodes of short duration. Twenty days before the consultation she had a sudden deafness in the left ear. Her ear, nose and throat examination was normal and the hearing tests showed profound hearing loss in each ear. CT scans of the temporal bones showed no structural deformities. Vestibular tests showed marked bilateral vestibular hyperreflexia. She was given prednisone (60 mg/day) for 2 weeks but her hearing did not change. She was told that she might need a cochlear implant and that she could try an endolymphatic shunt operation in the left ear.

The therapy with corticosteroids did not improve her condition. In July 2001 she was submitted to an endolymphatic shunt, with insertion of a silicone sheet. The middle ear was also investigated to rule out the possibility of a perilymphatic fistula, which

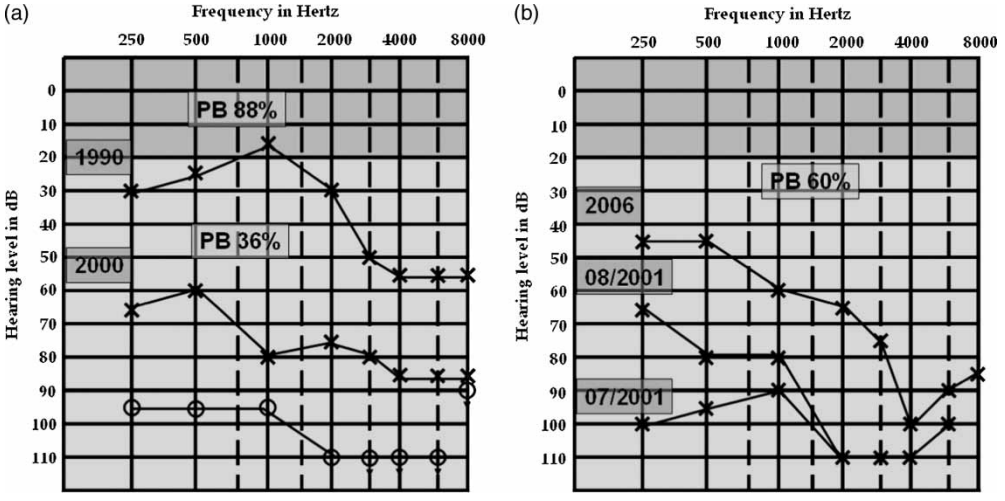

Figure 4. Audiograms of case 4. (a) The thresholds obtained at the first audiometry in 1990 and soon after she had her second child, in 2000. (b) The thresholds immediately after the sudden hearing loss (July 2001), after the endolymphatic shunt (August 2001), and the most recent audiogram (2006). The operation to close the perilymphatic fistula was performed in 2002.

was not found. In the following week the patient reported increase in the tinnitus but could hear with the hearing aid.

In January 2002 she had another sudden hearing loss that did not respond to medical treatment. Her operation was then revised and a fistula was found at the round window membrane, which was obliterated with a fragment of adipose tissue obtained from the ear lobe and biological glue. Her hearing improved and she regained good social hearing with her hearing aid.

Discussion

Endolymphatic hydrops is a histopathological entity most frequently associated with Ménière's disease, in spite of the fact that other inner ear disorders, such as Mondini dysplasia and cochlear vasculitis, both auto-immune and luetic, can also induce pressure changes.

Delayed endolymphatic hydrops has more similarities with Ménière's disease than the other hydrops-inducing disorders. The difference between them is the presence of sensorineural hearing loss, usually unilateral, since childhood. It would seem that the virus of epidemic parotiditis is the most common cause, although other viruses may be involved. In one of the cases described in this study the measles virus was the causative factor. It should be noted, however, that most of the patients who acquire unilateral hearing loss during childhood never develop delayed hydrops. The virus destroys the hair cells but usually does not destroy the vestibular receptors and vestibular disorders are rarely seen in these patients, except in those who develop delayed hydrops, several years later. However, the time lapse may be shorter and some cases of delayed hydrops have been described in children [5].

Delayed hydrops, therefore, is of unquestionable viral origin, although autoimmunity has been suggested as an additional contributing etiology in the contralateral cases [6].

Recently there has been a trend towards separating idiopathic Ménière's disease from the cases with identical symptoms in which an etiologic factor can be ascertained. From a strictly scientific point of view the reasons for this distinction are questionable. The cases of delayed hydrops suggest that a viral infection may eventually cause typical Ménière's disease in patients without hearing loss since childhood, and it would be expected that several years would elapse between the virus infection and the first cochlear and/or vestibular symptoms. It would be impossible to distinguish such cases from idiopathic Ménière's disease except through post-mortem histology. Patients who suffer from acoustic or physical trauma often present cochlear and/or vestibular symptoms several years after the trauma, implying that endolymphatic hydrops is a condition that develops slowly.

It is also known that the symptoms of endolymphatic hydrops are quite variable. There are benign cases that respond well to different kinds of treatment and there are severely incapacitating ones. Delayed endolymphatic hydrops is usually severe and comparable to the worst cases of Ménière's disease [7], in spite of the fact that the audiological and vestibular findings in each of the disorders show no statistically significant differences [8].

The cases selected for this study present some special characteristics that demonstrate some meaningful aspects of delayed hydrops.

Case 1 shows a patient with typical contralateral delayed endolymphatic hydrops. She responded well to medical treatment and remained asymptomatic for 20 years. There have been many clinical and investigational studies demonstrating the influence of disorders in carbohydrate metabolism in inner ear disorders, including Ménière's disease [9–11]. This case suggests that the influence may also occur in cases of delayed endolymphatic hydrops. This patient had a relapse of her fluctuating hearing loss and attacks of vertigo when she became obese and diabetic. The adequate control of her diabetes, in conjunction with weight loss, resulted in relief of her inner ear symptoms.

In case 2 there were both ipsilateral and contralateral symptoms, with an interval of several years between the onset of the respective symptoms. This is not the condition that Schuknecht [3] classified as bilateral; the patients thus classified had had sensorineural hearing loss in both ears during childhood. When this patient began to have episodes of vertigo, which were not accompanied by any hearing changes in the left ear, she was considered a good candidate for labyrinthectomy – the treatment recommended for ipsilateral cases with profound deafness. After the labyrinthectomy she remained well for 5 years, after which she began to have symptoms of Ménière's disease in her only hearing ear. Fortunately she responded well to medical treatment. The fact that she responded well to the labyrinthectomy demonstrates that the vestibular symptoms, at the time, originated from the deaf ear. The presence of fluctuant hearing loss in the hearing ear and vestibular symptoms 5 years after the operation demonstrate that the second ear also became involved by the disease.

Case 3 presented a hearing loss in the right ear that was not profound. This is not the usual finding in cases of delayed hydrops. The hearing loss was observed when she was 5 years old, but the etiology

and precise time of onset are unknown. The first vertiginous episodes occurred 25 years later. After undergoing an endolymphatic-subarachnoid shunt she no longer had dizzy spells and her hearing improved. It has been suggested [7] that in patients with this disorder, hydrops (when present), occurs in the better ear. In this patient with ipsilateral delayed hydrops it is quite clear that the pathology underlying her symptoms was of a pressure nature.

Case 4 is a contralateral case in which multiple situations occurred. This patient began to suffer from fluctuant hearing loss, without vestibular symptoms, during her second pregnancy. After the baby was born the vestibular symptoms subsided but she had to wear a hearing aid. A few years later she presented dizzy spells which were followed, a month later, by a severe sudden hearing loss that did not respond to medication. She then underwent an endolymphatic sac decompression, after which she regained some hearing and again derived benefit from the hearing aid. During the sac procedure a tympanotomy was performed to rule out the presence of a perilymphatic fistula, which was not found. The possibility of a perilymphatic fistula was considered due to the sudden character of the hearing loss and the circumstance that another patient with delayed endolymphatic hydrops whose first symptom was a sudden hearing loss with vertigo had a surgically confirmed fistula.

Six months later, however, she had a second episode of sudden hearing loss and when her operation was revised a perilymphatic fistula was found in the round window membrane and obliterated. In the last 4 years her hearing has stabilized and she derives good social benefit from her hearing aid.

Perilymphatic fistulas as a cause of sudden hearing loss are a controversial issue. There is no question that many patients have good surgical results from closure of these fistulas. However, they occur in circumstances that do not cause any problems in other persons, which suggests that some ears are more sensitive to pressure changes in the cerebrospinal, perilymphatic, and endolymphatic spaces. This sensitiveness may be congenital, since no previous pathology has been correlated with the incidence of fistulas. On the other hand, fistulas are frequent in patients with structural defects such as Mondini dysplasia [12] and perilymphatic hypertension syndrome [13].

The occasional presence of fistulas in patients with delayed endolymphatic hydrops suggests that these patients also have particularly sensitive ears. Even in the absence of fistulas this sensitiveness might explain the late development of cochlear and/or vestibular symptoms that do not occur in the majority of patients with viral hearing loss in childhood.

Conclusions

Delayed endolymphatic hydrops is a labyrinthine disorder of viral origin that occurs several years after the initial viral infection. The cases discussed in this article suggest that hydrops is the main cause of the hearing loss and the vestibular symptoms and can be present in the ear with severe hearing loss. They also suggest that this condition probably occurs in patients with congenitally sensitive ears that make them prone to the development of the late hydrops.

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