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Original article

Delayed endolymphatic hydrops. Special emphasis on nystagmus associated with episodes and contribution of chemical labyrinthectomy

P. Reynard , A. Karkas , M. Gavid , Y. Lelonge , P. Bertholon *

Département d'ORL, centre hospitalier universitaire de Saint-Étienne, 42055 Saint-Étienne cedex 2, France

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ABSTRACT

Objectives: The main objective was to describe spontaneous nystagmus characteristics during an episode of delayed endolymphatic hydrops (DEH), including an initial vertical upbeating nystagmus in one patient. The secondary objective was to highlight the contribution of chemical labyrinthectomy.
Methods: Episodic vertigo after a prolonged period of time of sensorineural hearing loss (profound or total) in one ear characterized ipsilateral DEH and was associated with the development of hearing loss in the opposite ear in contralateral DEH.
Results: Ten patients met the criteria for DEH: 7 ipsilateral and 3 contralateral. Three (all ipsilateral DEH) were examined during a vertigo episode. Two patients had a typical horizontal-torsional nystagmus beating contralaterally to the hearing loss. One patient showed atypic initial vertical upbeating nystagmus with a slight torsional component, which secondarily became horizontal-torsional beating contralaterally to the hearing loss. Four patients had disabling vertigo with unilateral total deafness (ipsilateral DEH), successfully treated by 1-3 transtympanic gentamycin (Gentalline®) injections.
Conclusion: Nystagmus direction during vertigo episodes varies, and may initially present as vertical upbeating nystagmus, which, to our knowledge, has not been previously reported in DEH or Menière's disease. This nystagmus might reflect an inhibition of the superior semicircular canal (on the hearing-impaired side), suggesting incipient hydrops in this canal. Chemical labyrinthectomy is a simple and effective procedure in unilateral DEH, especially as the patient often suffers from total deafness.

1. Introduction

Ipsilateral delayed endolymphatic hydrops (DEH) was first described in 1971 by Kamei [1], then by Nadol [2] and Wolfson [3], both in 1975. In 1978 Schuknecht described a contralateral form [4]. In the ipsilateral form, the delayed symptomatology consists in onset of vertigo episodes in patients with profound unilateral hearing loss after an interval of 1 to 68 years [2], for a mean 26.8 years [4]. In the contralateral form, it consists in fluctuating hearing loss in the healthy ear, with or without recurrent vertigo, with onset at a mean 17 years [4].

The initial underlying hearing loss is usually total, or at least profound [5]. In both ipsi- and contra-lateral DEH, the etiology of the primary hearing loss is usually unknown (in about 60% of cases), very often with onset in early childhood [5]. Mumps has, however, been implicated, notably in children (9–16% of cases). In adults, onset is sudden, with an equivalent frequency [5]. Many other etiologies have been reported: measles, varicella-zoster virus, flu, otitis/mastoiditis, meningitis, cranial or acoustic trauma, or otologic surgery [5–7].

Vertigo usually occurs in violent episodes, reminiscent of the symptoms of Menière's disease [3]. According to Kamei [7,8], in ipsilateral DEH nystagmus during a vertigo episode is horizontal-rotary beating in the direction of either the healthy or the impaired ear [9,10]. Vestibular exploration, caloric test and cervical vestibular-evoked myogenic potentials (cVEMP) usually show relatively conserved vestibular function, in contrast to the severity of hearing loss, enabling onset of vertigo episodes [3,11]. cVEMPs can be improved by glycerol test in both ipsilateral and contralateral forms of DEH, providing electrophysiological evidence for endolymphatic hydrops [12,13], as suggested in the first observations of surgical labyrinthectomy, which is highly effective in unilateral DEH [2,14]. The saccule appears dilated in contact with the stapes footplate [2], and vestibular fluid sampled

* Corresponding author.
E-mail address: pierre.bertholon@univ-st-etienne.fr (P. Bertholon).

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below the footplate (perilymph) is identical to the endolymph, as found in Menière's disease [2,14]. It was hypothesized that the initial episode led to secondary atrophy or fibrous obliteration of the endolymphatic pathways, causing defective endolymph resorption, inducing delayed hydrops [3,4]. In contralateral DEH, an anatomic study by Schuknecht found lesions typical of viral labyrinthitis on the impaired side and lesions typical of hydrops, as found in Menière's disease, on the other [15]. Schuknecht's hypothesis was that bilateral viral infection led to profound hearing loss on one side and infraclinical injury on the other with secondary fibrosis and atrophy of the endolymphatic pathways, causing hydrops [15]. Labyrinth MRI can now demonstrate endolymphatic hydrops in vivo [16,17].

We report the cases of 10 patients with rotary vertigo associated with DEH, focusing on the characteristics of the nystagmus during the episodes in 3 patients, and on the contribution of chemical labyrinthectomy by transtympanic gentamycin (Gentalline®) injection in 4 patients.

2. Material and methods

A retrospective study included patients with vertigo episodes corresponding to DEH on the diagnostic criteria adapted from the 1987 Committee of the Japan Society for Equilibrium Research criteria [5].

In ipsilateral DEH, inclusion criteria comprised:

- profound or more severe unilateral sensorineural hearing loss (mean > 90 dB at 500, 1000 and 2000 Hz);
- delayed onset of vertigo without hearing level fluctuation in the contralateral healthy ear;
- absence of central nervous system lesions, cochleovestibular nerve tumor or other cochleovestibular pathology on appropriate examination including at least brain MRI.

In contralateral DEH, inclusion criteria comprised:

- profound or more severe unilateral sensorineural hearing loss (mean > 90 dB at 500, 1000 and 2000 Hz);
- delayed onset of vertigo and sensorineural hearing loss in the contralateral initially healthy ear;
- absence of central nervous system lesions, cochleovestibular nerve tumor or other cochleovestibular pathology on appropriate examination including at least brain MRI.

Only patients with vertigo were included: contralateral forms of DEH thus associated vertigo with contralateral hearing loss: i.e., there were no cases of purely auditory contralateral DEH without vertigo; this was the main adaptation made to the 1987 Committee of the Japan Society for Equilibrium Research criteria [5].

All patients underwent interview and full otoneurologic work-up, systematically including videonystagmoscopy (VNS) at each consultation or in case of emergency. Head-shaking test and bone vibration test (100 Hz) were performed under VNS. Pure-tone and speech audiometry were performed at each consultation. Videonystagmography (VNG) (Ulmer, Synapsis) and cervical vestibular-evoked myogenic potentials (cVEMP) (Neuro-audio, Collin Medical; Beyerdynamic DT48 headphone) were performed, and in many cases repeated according to clinical findings.

All patients underwent brain MRI, particularly screening for pontocerebellar angle and/or labyrinth lesions; in case of doubt, temporal bone CT was performed, being more precise for detecting labyrinth deformity.

In disabling vertigo, chemical labyrinthectomy was proposed. Using a titration technique, this consisted in a transtympanic gentamycin (Gentalline®) injection under local anesthesia, repeated after about 1 month in case of persistence. Repeat injections were performed only after full audiovestibular work-up.

3. Results

Ten patients were managed for DEH: mean age, 56.4 years [range, 21–73 years]; 7 (mean age, 53.4 years [range, 21–73 years]) with ipsilateral DEH, 3 (mean age, 63.3 years [range, 57–71 years]) with contralateral DEH (Table 1). Initial hearing loss showed childhood onset in 7 of the 10 patients, although exact date of onset was unknown. Etiology was unknown in 7 patients, implicated mumps in 1 case (no. 8) and was probably viral or post-otitis in 2 cases (no. 5, 6). Initial hearing loss was total in all but 1 case (no. 10) of contralateral DEH with profound hearing loss. In contralateral DEH, contralateral hearing loss was either moderate (no. 8, 9) or severe (no. 10), and was fluctuating in 2 patients (no. 9, 10) and progressive in 1 (no. 8). In line with the inclusion criteria, all patients showed Menière-type vertigo. Mean age at onset of vertigo in ipsilateral forms was 47.3 years [range, 20–69 years], and 61 years [range, 52–71 years] in contralateral forms, with a mean age of onset of fluctuation in the healthy ear of 46 years [range, 33–57 years]. The precise interval between onset of initial hearing loss (often unknown) and onset of vertigo could not be determined, but was at least 7 years (case no. 3). Three patients were examined during a vertigo episode; 2 (cases no. 1 (see below) and no. 3) showed horizontal-torsional nystagmus during the episode (beating contralaterally to the hearing loss), without change in direction, and 1 (case no. 2; see below) showed essentially vertical nystagmus becoming horizontal (beating contralaterally to the hearing loss). Vestibular exploration found persistent vestibular function, in contrast to the severity of hearing loss. Caloric test found symmetric reflex in 5 of the 9 patients tested and deficit on the hearing-impaired side in 4 of the 9. cVEMPs were present bilaterally in 8 of the 10 patients tested, absent on the hearing-impaired side in 1 (no. 9) and on both sides in 1 patient (no. 8, contralateral form), although a technical problem could not be ruled out here. All patients underwent brain MRI, which found no abnormalities; 2 underwent temporal bone CT, which was also normal.

Therapeutically, 4 patients with disabling vertigo and unilateral DEH underwent chemical labyrinthectomy (1–3 injections; cases no. 1, 2, 3, 4), and showed no recurrence, although follow-up was short (1–4 years). The 3 patients with contralateral DEH, managed medically, showed no recurrence of vertigo.

Below, we present 2 cases of unilateral DEH examined during a vertigo episode.

3.1. Case 1

A 69-year-old man suffering from migraine without aura was admitted to hospital after 5 recent vertigo episodes of more than 20 minutes, accompanied by nausea and sometimes vomiting, obliging him to lie down onto his left ear. He showed no auditory signs during vertigo, but reported sudden onset of definitive right-side total deafness at the age of 30, accompanied at the time by sensations of vertigo. Intra-episode clinical examination found very marked left horizontal-torsional nystagmus under VNS, associated with 100° rightward axial deviation on Fukuda test with eyes closed.

Pure-tone audiometry showed right-side total deafness and normal hearing in the left ear.

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