Meniere’s disease and Sudden Sensorineural Hearing Loss

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Meniere’s disease (MD) and idiopathic sudden sensorineural hearing loss (SSNHL) are two different diseases; however, they do overlap in some cases. Many patients with SSNHL, especially low-tone SSNHL, later develop MD. In this keynote lecture, I would like to describe the definition, epidemiology, image findings, and etiology and future prospects for MD and SSNHL.

Definition

The diagnostic criteria for MD defined by the American Academy of Otolaryngology–Head and Neck Surgery (AAOHNS) in 1995, which have been used worldwide for the past 20 years, classify MD into four different subtypes: certain, definite, probable, and possible MD. The diagnosis of certain MD requires the confirmation of endolymphatic hydrops (EH) by histopathological assessment of the temporal bone after death. Given the relationship between EH and MD, the functional and morphological diagnosis of EH has improved considerably with the development of MRI of the inner ear, although this technique is not widely used in clinical practice at present.

The 2015 criteria only include definite MD and probable MD and do not require the confirmation of EH. However, possible MD or borderline MD should be investigated to understand the overall features of MD with evaluation of the presence or absence of EH (Nakashima et al, 2016).

The criteria for diagnosing SSNHL, created more than 40 years ago in Japan, did not include a specific decibel range for establishing a diagnosis of SSNHL. In 2012, the Research Committee of the Ministry of Health and Welfare for Acute Profound Deafness established new criteria that defined hearing levels for SSNHL as 30 dB or more over three consecutive frequencies, in accordance with the definition widely used in other parts of the world (Nakashima et al, 2014).

Epidemiology

The prevalence of MD in the United States ranged from 9 per 100,000 in
patients <18 years of age to 440 per 100,000 in patients ≥65 years of age (Harris & Alexander). The prevalence increases with age, with a peak prevalence in those aged 61–70 years of age. Finnish data collected in a population-based study indicated a significantly higher prevalence (513/100,000) of MD than in previous studies collected from national data banks or tertiary referral hospitals (Havia et al).

The prevalence of bilateral MD increases with age and duration of MD. The overall frequency of patients with bilateral MD was 24%, of which 11% showed bilateral disease at the initial presentation and 14% progressed from unilateral disease to bilateral disease (House et al, 2006). Unilateral disease was reported to progress to bilateral disease in up to 35% of patients within 10 years and in up to 47% within 20 years during follow-up (Huppert et al).

The prevalence may depend on the diagnostic criteria. Recent MRI findings have revealed that the prevalence of asymptomatic EH is much higher than expected. The prevalence of possible MD or asymptomatic EH should be also studied.

The incidence of SSNHL in the United States increases with age, ranging from 11 per 100,000 for patients younger than 18 years to 77 per 100,000 for patients aged 65 years and older (Alexander & Harris).

In Japan, after the criteria for SSNHL changed, the epidemiological study performed from April 1, 2012 through March 31, 2013 revealed that the incidence of SSNHL was 60.9 per 100,000 population (Nakashima et al, 2014). The incidence was much higher than the rates reported previously in Japan. This difference may due to the inclusion of private ENT clinics in the survey. The average age was 54.2 years (SD = 17.1). The incidence of SSNHL increased with age, from 9 per 100,000 for patients younger than 20 years to 94 per 100,000 for patients in the age range 60–69 years. In the younger age groups, more females than males showed SSNHL. The survey revealed that 65% of SSNHL patients visited only private ENT clinics. Twenty-three percent of patients had been diagnosed with acute low-tone SSNHL.

**Imaging**

Evaluation of EH has been performed using intratympanic or intravenous administration of gadolinium contrast agents with 3D-FLAIR MRI. The intratympanic method provides good-quality images unless the permeability of the inner ear windows is blocked. The intravenous method can evaluate the status of the blood-labyrinthine barrier. The blood-labyrinthine barrier is often disrupted in ears with MD, especially in cases with significant EH. A characteristic sign of MD is EH. In our study, all patients with unilateral and bilateral MD showed significant or mild EH in the cochlea or the
vestibule of the affected side. In most patients, the clinical symptoms of MD present after considerable accumulation of endolymph has occurred. However, some patients develop symptoms in the early stages of EH (Nakashima et al, 2016).

Recently EH has been reported in some normal controls. The prevalence of asymptomatic EH may be much higher than that of MD in the population, just as the prevalence of asymptomatic glaucoma is much higher than that of symptomatic glaucoma. Hydrops imaging may be useful to estimate the prognosis of MD. Among patients with unilateral MD, some have bilateral EH and the others have EH only in the affected side. It is assumed that the probability of immediate progression to bilateral MD from unilateral MD is very low in patients without EH on the non-affected side (Morimoto et al).

Contrast enhancement of the labyrinth on MRI in patients with SSNHL was described in 1991 (Seltzer & Mark). Using three-dimensional fluid-attenuated inversion recovery (3D-FLAIR) MRI at 3 Tesla, pre- and post-contrast enhancement of affected ears in SSNHL was reported (Sugiura et al). Use of 3D-FLAIR MRI at 3 Tesla may contribute to the elucidation of pathologic conditions in the inner ears of patients with SSNHL and provide new radiologic indicators of mild hemorrhage, acute inflammation, and the presence or absence of blood-labyrinthine barrier breakdown (Berrettini et al). Two-thirds of SSNHL ears showed high signals on pre-contrast 3D-FLAIR. Pre-contrast signals may reflect minor hemorrhage or an increased concentration of protein in the inner ear, which has passed through blood vessels with increased permeability or has originated in disrupted cells in the inner ear. One-half of the cases with the pre-contrast signals showed gadolinium enhancement (Yoshida et al). Positive contrast enhancement may indicate breakdown of the blood-labyrinthine barrier. High signals in the affected inner ear on 3D-FLAIR MRI closely correlate with vestibular dysfunction and poor hearing recovery in patients with SSNHL, especially when the vestibular apparatus is involved (Ryu et al).

**Etiology**

Polymorphisms in genes involved in blood vessels or blood flow regulation have been reported to be associated with MD and/or SSNHL. However, obtaining direct evidence of disturbed blood flow in the inner ear is now difficult because it is not possible to observe blood flow in the inner ear or to measure endolymphatic pressure in clinical practice. This is a disadvantage for the treatment of inner ear diseases compared with eye diseases, in which observation of retinal blood flow and measurement of the intraocular pressure is clinically possible. Recent developments in MRI techniques have contributed to our understanding of the etiology of inner ear diseases including the
cochlea and the vestibular apparatus. Further developments in MRI techniques are expected to elucidate inner ear diseases.

**Future prospects**

Not all patients have all the typical symptoms of MD (tinnitus, hearing loss, vertigo and aural fullness) and may only have one or some of these symptoms for years. Although a characteristic sign of MD is EH, the relationship between clinical symptoms and EH needs to be studied further.

Because the etiology of SSNHL may be multifactorial, understanding the etiology is essential to improve the treatment results. SSNHL may be caused by inner-ear blood flow disturbance (hemorrhage and occlusion), breakdown of the blood-labyrinthine barrier, and sudden disruption of the organ of Corti in a manner similar to retinal detachment in the eye. Abnormal passage of the intravenous gadolinium reflects increased permeability of blood vessels or disruption of the blood-labyrinthine barrier in the inner ears. We hypothesized that glucocorticoids, which have anti-inflammatory actions and suppress the increased permeability of blood vessels, are effective in some cases showing post-contrast enhancement (Nakashima, 2009). Prospective studies including MRI before the treatment may be necessary to elucidate the relationship between the glucocorticoid effect and the disruption of the blood-labyrinthine barrier.

The total blood flow volume of the inner ear is <0.001% of the total cardiac output in humans, suggesting that systemic drug therapy might require extremely high doses. Suitable methods for the topical application of medication to the inner ear should be developed (Nakashima et al, 2016).

**References**


