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Atypical Meniere Disease: Case Report of a Patient Treated as Sudden Hearing Loss

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Abstract

Meniere's disease is an idiopathic disease involving the inner ear, which is characterized by fluctuating low frequency sensorineural hearing loss, tinnitus, aural fullness, and recurrent vertigo. We could see the Meniere's disease without vestibular symptom in some cases. These cases have been reported as atypical (Cochlear) Meniere's disease. We have evaluated a patient of atypical Meniere's disease followed as sudden hearing loss with fluctuating low tone type sensorineural hearing loss without vertigo. Sudden deafness, cochlear sensorineural hearing loss, chronic progressive sensorineural hearing loss, presenile presbycusis, noise induced and autoimmune sensorineural hearing loss may be confused in diagnosis of Meniere disease.

Key Words: Atypical Meniere Disease; Hearing Loss; Diagnosis.

Atipik Meniere Hastalığı: Ani İşitme Kaybı Olarak Tedavi Edilen Olgu Sunumu

Özet

Meniere hastalığı, fluktuan, başlangıçta düşük sonradan tüm frekansları tutan sensörinöral tip işitme kaybı, tinnitus, aural dolgunluk ve tekrarlayan vertigo atakları ile karakterize bir iç kulak hastalığıdır. Bazı vakalarda vestibüler semptomlar olmaksızın Meniere hastalığı görülebilir. Bu vakalar atipik (koklear) Meniere hastalığı olarak adlandırılır. Bu makalede, vertigo semptomları olmaksızın ani başlangıçlı, fluktuan ve düşük frekansları tutan sensörinöral işitme kaybı olan atipik Meniere hastalığı olan bir vaka sunulmuştur. Meniere hastalığı tanısında, ani işitme kaybı, koklear tip sensörinöral işitme kaybı, kronik progresif sensörinöral işitme kaybı, presenil presbiakuzi, gürültüye bağlı ve otoimmün sensörinöral işitme kaybı ayırt edilmelidir.

Anahtar Kelimeler: Atipik Meniere Hastalığı; İşitme Kaybı; Tanı.

Olgu Sunumu/Case Report

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Introduction

Meniere's disease (MD) is defined by the American Academy of Otorhinolaryngology Head and Neck Surgery as an idiopathic disease involving the inner ear that is characterized by fluctuating low frequency sensorineural hearing loss (SNHL), tinnitus and aural fullness, and recurrent vertigo (1). We could see the MD with or without vestibular symptoms. Full complement of symptoms of MD does not develop simultaneously in some cases. These cases have been reported as atypical MD (Cochlear MD). Vestibular MD is characterized by recurrent episodic vertigo but without hearing loss. In contrast, cochlear MD is defined as having fluctuating low frequency hearing loss without vertigo. Idiopathic sudden sensorineural hearing loss of low tone type with preservation of high-tone hearing and without episodes of vertigo may be confused with atypical MD (2,3). In the present report, we have evaluated a patient of atypical MD followed as sudden hearing loss with fluctuating low tone type sensorineural hearing loss without vertigo.

Case Report

A 32-year-old male with fluctuating low frequency SNHL and tinnitus in his right ear, but without vertigo (i.e. not fulfilling the AAO-HNS diagnostic criteria of MD) was evaluated audiologically (1). The patient had experienced fluctuation SNHL without associated episodic vertigo four times for his left ear for 14 years and had been treated with different sudden hearing loss treatment protocols including steroids. He had a permanent SNHL during an attack in five years ago. He had no problems for his right ear since two years ago. He also experienced fluctuation SNHL without associated episodic vertigo three times for his right ear for two years. He had a permanent SNHL in his right ear during an attack in two months ago. He also had been treated with different sudden hearing loss treatment protocols including steroids for his right ear. His mother was MD in his history

The audiogram and hearing fluctuation of this patient in our laboratory are summarized on Figure 1. He had a pure-tone average of 55/53 dB in his left ear and 38/33 dB for air/bone way in his right ear. Normal speech discrimination and the recruitment investigation tests including Bekesy audiome try and short increment sensitivity index (SISI) test confirmed that there was cochlear involvement. The Electronystagmography (ENG) test battery and temporal Magnetic Resonance (MR) imaging were normal. The autoimmune SNHL test markers including sedimentation (SH), romatoid factors (RF), immunoglobulin's (IgG and IgM) were in normal range. Head shock protein 70 (HSP 70) could not examined for health insurance problems. A positive glycerol test (10% glycerol (Glyserine Pure, ORO co., 1mg/kg), intravenous administration for 2 h with hearing test performed before/after injection) suggested possible endolymphatic hydrops (Figure 2).

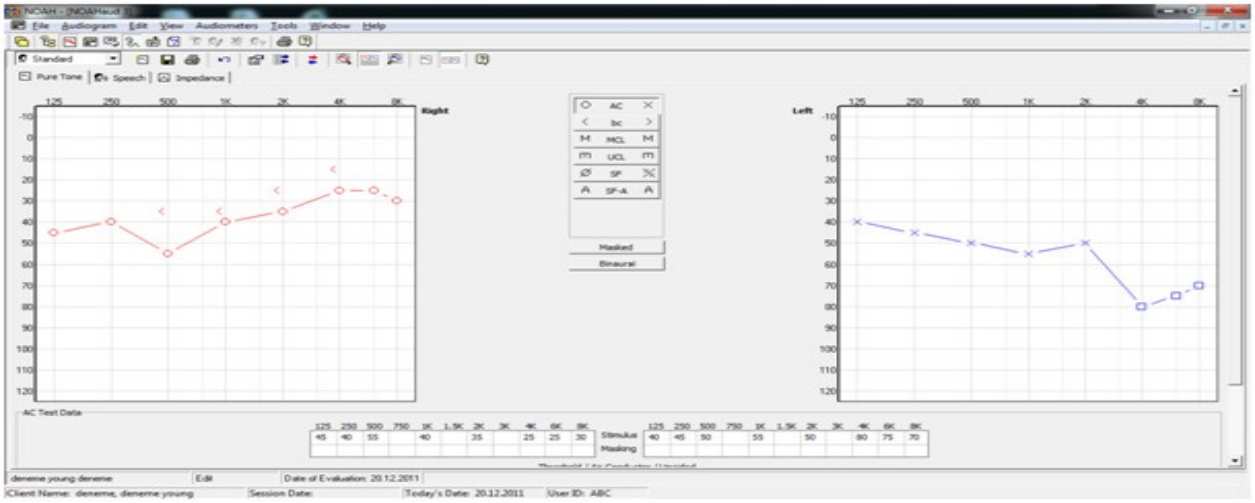

Figure 1. The patient had a fluctuating low frequency sensorineural hearing loss (SNHL) in his right ear for his first evaluation.

We have eliminated the autoimmune hearing loss and sudden hearing loss from the history and laboratory results of this patient. The initial diagnosis was atypical (cochlear) MD. The patient was treated with a loop diuretic and low salt diet consistent with the hydrocortisone sodium succinate (300 mg/day for 2 days, 200 mg/day for 2 days, 100 mg/day for 2 days). A repeat audiogram showed significant improvement in the low and middle frequency sensorineural thresholds. SNHL had got better from 38/33 dB to 20/14 dB for his right ear (Figure 3).

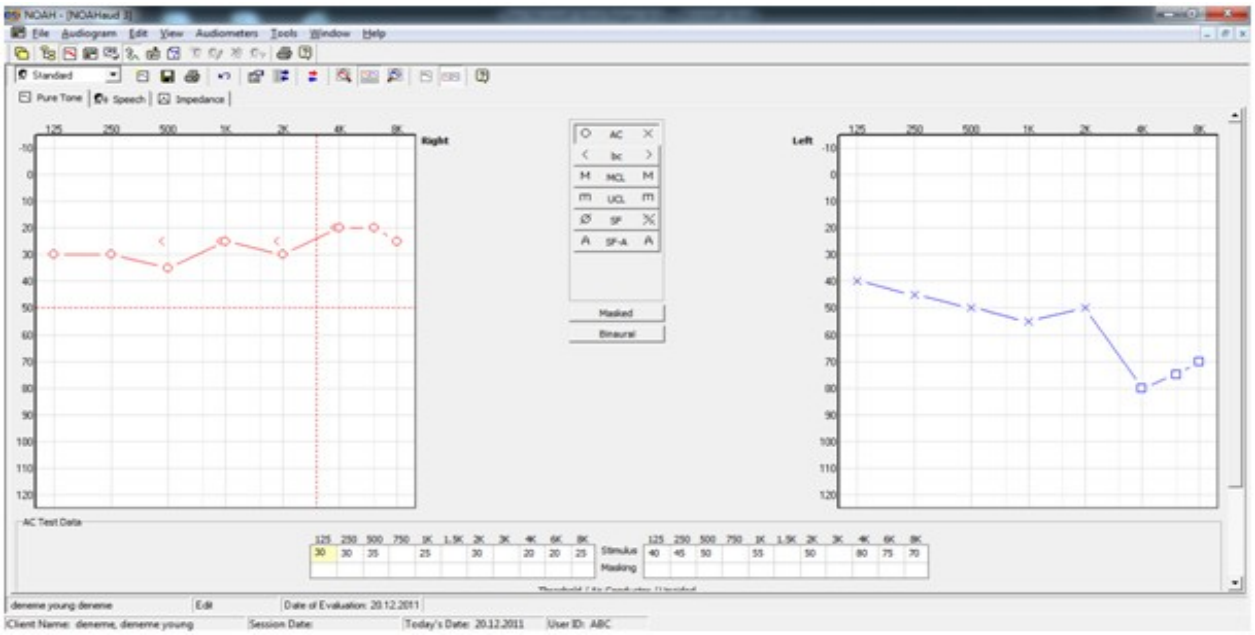

Figure 2. A positive glycerol test suggested possible endolymphatic hydrops

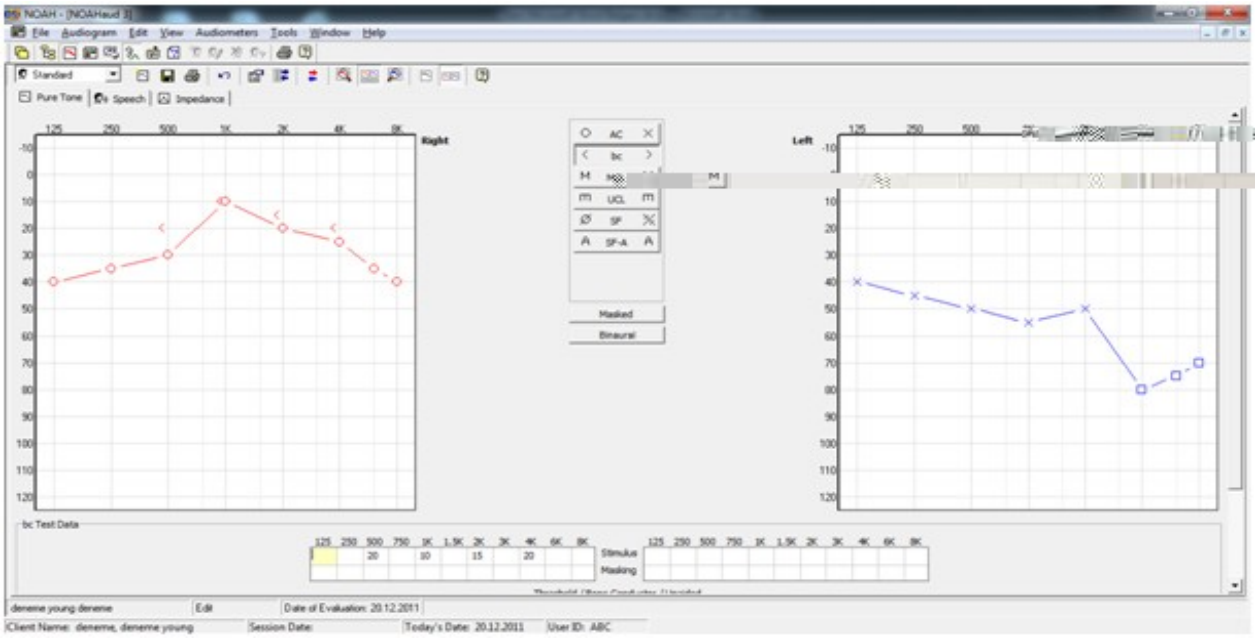

Figure 3. A repeat audiogram showed significant improvement in the low and middle frequency sensorineural thresholds.

Discussion

Idiopathic SNHL with and without endolymphatic hydrops and vestibular alterations has a high recurrence rate. The history of patient and characteristics of hearing loss are important for diagnosis. Sudden deafness, cochlear MD, cochlear SNHL, chronic progressive SNHL, presenile presbycusis, noise induced SNHL and autoimmune SNHL may be confused in diagnosis.

Sudden SNHL is defined as an idiopathic disorder with sensorineural hearing loss of at least 30 dB over three contiguous frequencies on pure tone

audiometry occurring acutely under 72 hours from initial onset (3). It has mostly involved for one ear, but sometimes bilaterally. Associated symptoms include tinnitus, vertigo, and aural fullness. The suspected etiologies for patients suffering sudden sensorineural hearing loss included idiopathic (71.0%), infectious disease (12.8%), otologic disease (4.7%), trauma (4.2%), vascular or hematologic (2.8%), neoplastic (2.3%), and other causes (2.2%) (4).

Patients should begin treatment with oral steroids within three weeks. One study found that patients with minimal hearing loss, no vestibular symptoms, and early treatment have better outcomes (4).

Autoimmune SNHL is usually bilateral. Patients present with rapidly progressive bilateral SNHL and poor speech discrimination scores, and they also may have vertigo or disequilibrium. Hearing loss progresses over three to four months, and an associated autoimmune disorder may be present. Symptoms usually improve with the administration of oral prednisone, and response to this steroid is currently the best way to make the diagnosis. Low-dose methotrexate therapy is becoming an accepted alternative to long-term prednisone therapy (5).

Meniere's disease is another cause of sensorineural hearing loss. Patients report unilateral fluctuating hearing loss with aural fullness, tinnitus, and episodic vertigo. Initially, the hearing loss is in the low frequencies, but higher frequencies are affected as the disease progresses. Treatment includes a low-salt diet, diuretics, and vestibular suppressants. Hearing aids are often ineffective because patients suffer from poor speech discrimination, as well as diminished tolerance to amplified sound. Chemical labyrinthectomy with gentamicin is now a common nonsurgical option for control of vertigo if medical management fails (6). Full complement of symptoms of MD does not develop simultaneously in some cases. We could see the MD without vestibular symptoms. The SNHL configuration of atypical MD is not standard (7). The audiogram and hearing fluctuation of this patient is including 500,100 and 2000 Hz low frequencies.
The association of endolymphatic hydrops with fluctuating hearing loss has been shown using positive glycerol tests and histological confirmation at autopsy (2-4). Although MD has been attributed to endolymphatic hydrops, only post-mortem histopathological confirmation has been available (8). Electrocochleography, glycerol test, or other functional testing has been utilized to estimate endolymphatic hydrops, but these do not give direct prof (1). These limited options of clinical diagnosis and functional testing were all that were available, thus making precise diagnosis of MD difficult. A positive glycerol test suggested possible endolymphatic hydrops (Figure 2).

The initial diagnosis was atypical (cochlear) MD for this patient. The patient was treated with a loop diuretic and low salt diet consistent with the hydrocortisone sodium succinate (300 mg/day for 2 days, 200 mg/day for 2 days, 100 mg/day for 2 days). The patient has been treated and SNHL had got better after the medical therapy. Sudden SNHL, and autoimmune SNHL may be confused in diagnosis of the atypical MD. The history of patient and characteristics of hearing loss and differential diagnosis are important for the diagnosis of atypical MD. Right diagnosis reaches us to the right treatment options and better results as could be seen in this case.

References

1. Sajjahi H, Paparella MM. Meniere's disease. Lancet 2008;372:406-14.
2. Yamasoba T, Sugasawa T, Yagi M, Harada T, Futaki T. clinical observations of acute low tone sensorineural hearing loss considered as cochlear hydrops. Nippon Jibiinkoka Gakkai Kaiho 1990;93:219-28.
3. Hughes GB, Freedman MA, Haberkamp TJ, Guay ME. Sudden sensorineural hearing loss. Otolaryngol Clin North Am 1996;29:393-405.
4. Wilson WR, Byl FM, Laird N. The efficacy of steroids in the treatment of ıdiopathic sudden hearing loss. a double-blind clinical study. Arch Otolaryngol 1980;106:772-6.
5. Salley LH Jr, Grimm M, Sismanis A, Spencer RF, Wise CM. Methotrexate in the management of ımmune mediated cochleovestibular disorders: clinical experience with 53 patients. J Rheumatol 2001;28:1037-40.
6. Blakely BW. Update on intratympanic gentamicin for meniere's disease. Laryngoscope 2000;1102:236-40.
7. Miyagawa M, Fukuoka H, Tsukada K, Oguchi T, Takumi Y, Sugiura M, et al. Endolymphatic hydrops and therapeutic effects are visualized in 'atypical' meniere's disease. Acta Otolaryngol 2009;129:1326-29.
8. Pirodda A, Ferri GG, Raimondi MC, Borghi C. Diuretics in Meniere disease: a therapy or a potential cause of harm? Med Hypotheses 2011;77:869-71.