Abstract

**Objective**: To present the association of Ménière's disease (MD) in one ear and vestibular schwannoma (VS) in the contralateral ear as a rare clinical entity, and discuss management options.

**Methods**: A retrospective chart review of all patients diagnosed with MD over a ten year period was conducted for patients that met a definite diagnosis of MD in the ear contralateral to the ear with VS diagnosis, based on the 1995 MD criteria set by American Academy of Otolaryngology—Head and Neck Surgery.

**Results**: Of 974 patients that met the inclusion criteria for a diagnosis of VS, five patients had a diagnosis of contralateral MD. The age range was 32-55 years. The size of VS ranged from 3-25 mm. All patients had the MD component managed with an aggressive medical protocol, and short courses of prednisone. For VS, four patients underwent surgery (three translabyrinthine, one retrosigmoid), and one patient opted for stereotactic radiation.

**Conclusion**: The association of MD in one ear and VS in the contralateral ear represents a rare clinical scenario. Bilateral deafness and/or bilateral labyrinthine hypofunction are interventional risks that can lead to severe patient incapacitation. Tumor size, brainstem compression, tumor growth, residual functional hearing, and the degree of disability of MD vertiginous symptoms are important considerations in this management algorithm.

**Keywords**: Ménière's disease, vestibular schwannoma, sensorineural hearing loss, endolymphatic hydrops, intracanalicular enhancing lesion

Introduction

Ménière's disease (MD) was first described by Prosper Ménière in 1861 as a peripheral inner ear disorder, challenging the general terminology at the time that named this disease apoplectic cerebral congestion, implying a disorder of the brain. Epidemiological data reports a prevalence of 17-47 cases per 100,000. The disease is characterized by fluctuating sensorineural hearing loss (SNHL) accompanied by aural pressure, tinnitus, and episodic vertigo. The hallmark of the disease is the fluctuating, waxing, and waning nature of its symptoms. In its early stages, MD might present with only cochlear symptoms such as hearing loss and pressure in the ear without true vertigo. The natural history of the disease is a progression to permanent moderate to severe SNHL. Most patients develop unilateral symptoms, but a significant proportion of patients may develop bilateral disease many years after the onset of the unilateral symptoms; several studies have reported the rate of bilateral MD to be as high as 50%-5. No cure is available for MD; however, medical and surgical options are available in an escalating fashion of aggressiveness, guided by the severity of patients' symptoms and failure to respond to appropriate treatment.
Vestibular schwannoma (VS) is a benign tumor arising from the Schwann cells in the vestibular component of the statoacoustic nerve. It is the most common tumor of the cerebellopontine angle. The clinical incidence rate is 10–15 per million/year\(^6\)-\(^8\). The diagnosis of VS is usually made in adults in their fifth and sixth decade of life. In younger patients, the diagnosis of neurofibromatosis type 2 should be considered. The most common presenting symptoms include unilateral hearing loss, tinnitus, imbalance, and facial hypoesthesia. The growth pattern of VS is highly variable, ranging from spontaneous involution to rapid growth\(^9\)-\(^13\). However, no reliable clinical or radiographic predictors have been found\(^3\), \(^34\)-\(^36\). Surgical management of VS has made significant progress over the past century, with the introduction of the surgical microscope and advancements in microsurgical techniques. In the early 1970s Gamma knife radiosurgery was introduced, and has since been shown to be a viable option for VS management with the goal of tumor growth arrest. With more series published showing that VS may remain unchanged in size for years following diagnosis, serial follow-up by magnetic resonance imaging (MRI), so-called “watchful waiting,” has emerged as a third avenue for managing these tumors. While all three management strategies have focused on tumor control and preservation of cranial nerve function, health related quality of life has emerged as a new and important issue in VS management.

The association of MD in one ear and VS in the contralateral ear is an uncommon clinical entity and presents the treating neurotologist with a challenging management dilemma. The English literature contains eight cases of patients with concomitant MD and contralateral VS\(^17\)-\(^19\); presented in this paper are five cases that outline management strategies in these difficult diagnostic and treatment scenarios.

Materials and Methods

The archival database at the Department of Otolaryngology-Head and Neck Surgery, University of Cincinnati, was accessed for retrieval of all patients diagnosed with VS during the 10-year period extending from January 1, 1999 to December 31, 2009. Inclusion criteria comprised of patients of 18 years of age and over, and a radiographic diagnosis of VS based on MRI. Patients that met these criteria were then filtered for a secondary diagnosis of “dizziness”, and the medical records of these patients were reviewed for the nature of their symptoms, their clinical examination findings, and their diagnostic workup results (Figure 1). Only patients that met a definite diagnosis of MD in the ear contralateral to the ear with VS diagnosis, based on the 1995 MD criteria set by the American Academy of Otolaryngology-Head and Neck Surgery\(^29\), were included in the study cohort.

Results

During the 10-year study period, 974 patients met the inclusion criteria for a diagnosis of VS, and of the patients with VS, 5 patients were diagnosed with concurrent MD on the contralateral ear.

Case 1

A 49-year-old female presented with a 4-year history of left sided fluctuating hearing loss, tinnitus and aural fullness, as well as episodic vertigo. Audiometric assessment showed normal hearing in the right ear and low frequency moderately severe up sloping to mild SNHL in the left ear. She had excellent word recognition scores (WRS) bilaterally. The clinical diagnosis was that of left ear MD. An MRI scan with gadolinium was undertaken, showing a right 3 mm intracanalicular-enhancing lesion. She was started on an aggressive MD medical regimen. The MRI scan was repeated at 6 months and showed a 1 mm increase in the size of the right VS, which stabilized thereafter on serial scans. She continued to be followed for the past two years with stable audiometric thresholds in both ears, and was doing well from a vestibular functional capacity.

Case 2

A 55-year-old female presented with a 2 year history of right sided progressive hearing loss. She was diagnosed 12 years prior with left sided MD. Her vestibular symptoms and audiometric thresholds in the left ear had been stabilized with strict adherence to a MD medical regimen. On her most

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**Figure 1.** A flow chart summarizing how patients met inclusion criteria.
recent audiometric evaluation, she demonstrated stable left-sided thresholds, mild low frequency SNHL up sloping to normal thresholds, and excellent WRS. On the right, there was elevation of the pure tone thresholds from her baseline, now demonstrating a moderate/severe SNHL with poor WRS. An MRI scan with gadolinium was performed which revealed a 2.5 cm right intracanalicular-enhancing lesion, with fundal extension and mild brainstem compression. She underwent translabyrinthine extirpation of this lesion, and pathological examination was consistent with VS. An aggressive medical MD protocol was resumed postoperatively, and she continued to have only a mild sensation of imbalance, with audiometrically stable hearing in her left ear.

Case 3

A 44 year-old male presented with subjective progressive fluctuating right-sided hearing loss and intermittent aural fullness for a number of years, and a more recent 4-month history of right-sided tinnitus. Audiometric assessment showed left mild to moderately severe SNHL, and right low frequency moderately severe rising to moderate then dropping to profound SNHL. WRS were 92% on the left and 60% on the right. Electrocochleography (ECOG) showed a significantly elevated summation potential (SP) to action potential (AP) ratio in his right ear. MRI scan with gadolinium showed a left 8 mm intracanalicular-enhancing lesion. The clinical impression was that of right MD, and he was started on an aggressive medical MD protocol. Serial audiometric assessments every three months thereafter continued to show fluctuating right-sided SNHL and WRS, the latter dropping to as low as 20%. Episodic fluctuations were managed with short courses of high dose oral steroids. In light of the uncertainty of the hearing in his MD right ear, and the risk of progressive or sudden hearing loss in his better hearing left ear, management of the left IAC lesion with fractionated radiation therapy or middle cranial fossa resection was recommended, and the patient opted for the former. Post radiation, his left ear pure tone thresholds elevated to a mild down sloping to a profound SNHL, with WRS diminishing to 72%. Over the ensuing three years, he continued to have fluctuating SNHL in his right ear, and stable hearing in his left ear. Serial MRI scans showed stability of the left IAC lesion. Hearing aids were successfully used bilaterally.

Case 4

A 32 year-old female presented with a 6-month history of progressive left hearing loss and spatial disequilibrium. Audiometric assessment showed normal hearing on the right ear, and absence of WRS on the left. Neurotologic examination demonstrated significant vestibular compromise. MRI scan revealed a 1 cm left VS. Following discussion of the different management strategies, the patient opted for translabyrinthine resection. Her postoperative course was unremarkable, highlighted by significant improvement in her balance function. On routine post-operative follow-up one year later, she noted a 6-month history of right-sided tinnitus, aural fullness and progressive hearing loss, as well as intermittent vertigo. Audiometric assessment revealed diminution of her hearing on the right to a moderate SNHL. Clinical impression was that of right MD, and she was started on an aggressive medical MD protocol. A short course of high dose oral steroids was commenced, to which her right pure tone thresholds improved. She continues to be managed conservatively on a medical regimen.

Case 5

A 51 year-old female presented with a one-week history of new onset vertigo for which she was evaluated by an MRI scan with gadolinium. This revealed a left 7 mm intracanalicular VS. Audiometric assessment demonstrated normal puretone thresholds and WRS bilaterally. She opted for surgical extirpation via the retrosigmoid route. Postoperatively, there was diminution of her left thresholds to a mild to moderate SNHL, with WRS of 88%. Four years following her surgery, she presented with new onset intermittent vertiginous episodes, and right aural fullness and tinnitus. Audiometry revealed right borderline to mild SNHL in the low to mid-frequencies. ECOG showed elevated SP to AP ratio in her right ear, and ENOG showed right-sided caloric weakness. The clinical impression was that of right MD, and she was started on a medical MD protocol. She remained stable from an audiovestibular perspective on follow-up over the ensuing 7 years.

Discussion

To the otologist and neurotologic surgeon, MD and VS represent unique diagnostic entities with challenging management algorithms. The occurrence of these diagnoses simultaneously in a patient in separate ears constitutes a rare clinical dilemma. Both disorders have been challenged with controversies regarding diagnosis and management; various factors play a role in the solitary management of these disorders, which substantially confound their dual management in the same patient.

Although nonfatal, the subjective manifestations of MD can be so incapacitating that sufferers may experience significant deterioration in physical, mental and social well being. There are rarely any clinical findings in MD patients who are not actively suffering from an acute
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episode, and there are no definitive diagnostic tests for MD. Between 35 and 70% of patients with MD show an abnormal result on electronystagmography, and only 54% of patients with unilateral MD demonstrate unilateral vestibular hypofunction on electronystagmography. In an attempt to standardize the diagnosis and reporting of MD, the American Academy of Otolaryngology–Head and Neck Surgery Committee on Hearing and Equilibrium have published recommended guidelines for the diagnosis of MD in 1972, 1985 and 1995. Nevertheless, the wide disparity amongst clinicians in both the diagnosis of MD and the undertaking of treatment options has made the standardization of clinical reporting challenging. The diagnosis of MD is, in most instances, inappropriate to make based solely on a fixed protocol or diagnostic guidelines. This stems mainly from the fact that MD, otherwise considered to be an entity of idiopathic endolymphatic hydrops, exists across a spectrum of clinical presentations. Many patients do not initially present with the entire set of symptoms comprising definite MD; only 27% of patients do ultimately fulfill these criteria satisfy such criteria at first presentation to an otolaryngologist. As such, in many instances it is left to the vigilance and experience of the treating clinician to recognize signs and symptoms of other conditions in the differential diagnosis, and to pursue appropriate investigations and treatments when warranted. The episodic nature of MD makes critical evaluation of therapeutic responses difficult. Torok suggested that most forms of therapy only have a placebo effect and the response to therapy simply reflects the natural history of MD. Management of MD is aimed primarily at vertigo control, as there are not equivalent concepts. Management of MD is primarily at vertigo control, as it remains the most incapacitating symptom. While some patients have noted improvement in their hearing with better control of their vertigo, this is not predictable. Despite the lack of randomized controlled trials evaluating the effectiveness of dietary or lifestyle modifications in controlling MD, these conservative treatment strategies remain the primary pillars in MD management, rationalized by the notion that decreasing the endolymphatic hydrops can be achieved by these strategies. Current medical regimens can control MD (as defined by the vertiginous episodes) in approximately 80% of patients. These include avoidance of triggers (e.g. emotional stress, fatigue, allergies), dietary sodium restriction (most regimens restrict to less than 2000 mg/day), caffeine and alcohol reduction, stress amelioration, psychological support, and diuretics induced by a dietary agent (most commonly triamterene and hydrochlorothiazide).

With a diagnosis of VS in the contralateral ear, the remaining 20-25% of MD patients who have progressive disease, despite maximal medical therapy, present a significant clinical challenge. Of primary consideration is the functional disability of the patient, which in most instances is reflective of the inability to control the vertiginous component of MD. The American Academy of Otolaryngology–Head and Neck Surgery have published a functional disability scale that assists in the objective evaluation of MD patients. If the VS is small and stable in size, emphasis then is focused on control of the vestibular component of MD to improve quality of life and functional capacity. Next, the hearing status of the hydropic ear as well as that with the VS is important. The natural history of MD demonstrates progressive decline in hearing function to a variable severity; some patients progress to complete deafness. Even if the MD ear is the worse hearing ear, the unpredictability of the hearing status in the ear with VS mandates that management of the hydropic ear is akin to that of an only hearing ear. From an MD management standpoint, available options include the Meniett device (Medtronic), transtympanic corticosteroid perfusion, transtympanic gentamicin perfusion, and surgical interventions. In 1999, the U.S. Food and Drug Administration approved the Meniett as a Class II device, and it has been available from Medtronic Xomed since 2001. Randomized double-blind, placebo controlled clinical trials have demonstrated that the device is safe and effective in the short term. In 2004, the Hearing Committee of the American Neurotology Society reviewed the literature regarding transtympanic corticosteroid perfusion, and concluded that the evidence supporting use in MD is weak. A randomized, double-blind study of transtympanic corticosteroid use in MD failed to show any efficacy over placebo, although the study was primarily investigating its impact on hearing loss and tinnitus, and not vertigo. Chemical ablation with gentamicin inner ear perfusion is hindered by the absence of acceptable double-blind or blinded, prospective control trials, and subsequently lack of standardization of concentration, dose, frequency, and duration of treatment. In the management of patients with MD and contralateral VS, specific issues arise pertaining to gentamicin use. Firstly, the risk of sensorineural hearing loss with transtympanic gentamicin has been cited as high as 35%. Secondly, it is expected that the contralateral ear with VS will have reduced vestibular function, and following gentamicin treatment, bilateral labyrinthine hypofunction and subsequent disequilibrium and oscillopsia can be incapacitating. In the setting of contralateral VS, surgery on the hydropic ear should be considered with extreme caution given the risk of iatrogenic SNHL and bilateral deafness. Surgical interventions can be categorized as ablative or non-ablative. Ablative procedures should be avoided in this patient population to avoid loss of bilateral labyrinthine function, unless there is electrophysiologic
documented in the literature. A labyrinthectomy precludes the future option of cochlear implantation if hearing loss in the VS side occurs. Endolymphatic mastoid shunt (EMS) procedures carry a low risk of SNHL, cited as 1 to 3% in the literature. Many studies have documented improved long-term control of vertigo in patients with MD undergoing EMS surgery. A recent survey of 165 active members of the American Otological Society and the American Neurotology Society asked respondents for their choice of second line treatment in patients with MD in an only hearing ear who failed first line medical therapy. Overall, 33% chose the Meniette device as their second-line choice; 30% chose transtympanic steroids and 29% chose endolymphatic sac surgery (with or without shunt placement) as their first option for second line therapy.

Conversely, approximately 40-50% of patients afflicted with VS report unsteadiness; vertigo is infrequently experienced in those patients. Treatment of VS is aimed mainly at tumor extirpation or growth stabilization to prevent potential neurologic complications from brainstem compression. In patients with VS and contralateral MD, management of large tumors, those with brainstem compression, or tumors that demonstrate growth on serial MRI scans takes precedence over MD management. Elucidating the natural history of VS has been the objective of many studies in the literature. VSs may remain dormant or grow slowly enough to never require treatment. The percentage of tumors that grow remains unknown and has varied widely in studies from 40% to 80%. In 2006, Stangerup et al. published a long-term follow-up of 14 years in more than 1800 patients with observed VS. In 83% of intrameatal tumors remained intrameatal and 70% of extrameatal tumors did not grow more than 2 mm. All tumors which exhibited growth did so in the first four years and maintained consistent growth rates throughout observation. It is thus appropriate in select patients with VS to undergo "watchful waiting" with serial MRI scans. Nevertheless, there are potential disadvantages to this approach that warrant consideration. Besides the cost of serial MRI scanning, tumors observed may go through accelerated growth despite a period of stable size. With growth, the risk of facial nerve injury with surgery may increase, and hearing preservation may no longer become a viable option. Furthermore, many patients observed are older with more co-morbidities, and if these patients need surgical intervention later on, their operative risks are potentially higher. Of particular relevance to a patient with a VS and contralateral MD is hearing preservation. Up to 25% of patients with VS will have sudden sensorineural hearing loss in the afflicted ear.

In the management of VS, alternatives to observation include radiation therapy and surgery. Reports on hearing preservation with radiation therapy are inconsistent amongst published reports; according to the Gardner-Robertson scale (grade A–C), hearing preservation is reported in 50% to 89%. Furthermore, surgery on previously irradiated VS carries virtually no chance of hearing preservation. Two approaches are generally considered for surgical removal of VS with the intent of hearing preservation: the middle cranial fossa and retrosigmoid approaches. Glasscock et al. stated that preservation of hearing is unlikely when the VS is larger than 20 mm. Yates et al. were unable to preserve hearing in tumors ≥25 mm in the CPA. Consequently, in patients with VS and contralateral MD, earlier tumor surgical intervention aimed at hearing preservation may be warranted, with concurrent aggressive medical management of MD in an attempt to stabilize the hearing in that ear. If surgical intervention is contemplated for management of MD, non-ablative, hearing-preserving procedures should be strongly pursued, regardless of the hearing status of the VS ear. This is because, even in the presence of good hearing in the contralateral VS ear, there exists a risk of complete hearing loss with surgical tumor extirpation or sudden SNHL.

Conclusion

The association of MD in one ear and VS in the contralateral ear represents an extremely rare clinical scenario, and the treating neurotologist is confronted with very challenging management options. Many factors play a critical role in guiding management algorithms. In the presence of a large VS, one that compresses the brainstem, or demonstration of tumor growth on serial MRI scans, treatment of the VS takes precedence, with an attempt at hearing preservation if clinically deemed feasible. With small, stable tumors, management paradigm shifts to that of MD control, with aggressive, conservative, medical management as the first line therapy. For those who fail and continue to have debilitating vertiginous symptoms, other surgical and non-surgical options can be contemplated, with the emphasis on non-ablative, hearing-preserving options.

Acknowledgements

An appreciation to the Department of Otolaryngology-Head and Neck Surgery, University of Cincinnati for providing access to their archival database and to the patients that participated in this case study. Also, a special thank you to the support and mentorship given by Dr. Nael Shoman and the Royal University Hospital Audiology Department.
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