



Review Article

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Overview on Meniere Disease in a Primary Health Care setting

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ABSTRACT

Background: Meniere disease is a disorder, which characteristically affects the inner ear, and may present with tinnitus, hearing loss, and vertigo. The usual course of the disease is slow and progressive and thus a lot of patients present late to primary health care centers. The family physicians need to keep in mind the association between these symptoms in order to identify this disorder and manage it properly. Objectives: We aimed to review the literature reviewing the pathophysiology of Meniere disease, clinical features, risk factors, diagnosis, and management of this disease. Methodology: PubMed database was used for articles selection, papers on were obtained and reviewed. Conclusion: The clinician in a primary health care center needs to be able to take a thorough history and perform a thorough physical examination when the patient presents with vertigo. This is pivotal since the diagnosis is clinical, and early identification of the disease helps in the long-term outcome. Fortunately, there are many effective medical and surgical therapies that can control vertigo and preserve hearing in these patients.

Key words: *Meniere Disease, Diagnosis, Clinical Features, Management.*

INTRODUCTION

Meniere disease (MD) is a disorder that characteristically affects the inner ear, and may present with tinnitus, hearing loss, and vertigo. The disease name is after Prosper Ménière, who in 1861, noted that off-balance, vertigo, and hearing loss were all associated with a lesion in the inner ear. The usual course of the disease is slow and progressive and thus a lot of patients present late to primary health care centers. Moreover, due to the disorder course, it has a major effect on the social functioning of the affected individuals [1]. According to studies, around 50 to 200 per 100,000 adults will be affected by Meniere, with most patients between the ages of 40 and 60 years [2]. The family physicians need to keep in mind the association between these symptoms in order to identify this disorder and manage it properly. In this paper, we will review the pathophysiology, clinical features, evaluation, and management of this disease based on recent studies.

METHODOLOGY

PubMed database was used for articles selection, and the following keys were used in the mesh (“Meniere Disease”[Mesh]) AND (“Diagnosis”[Mesh] OR “Management”[Mesh]). In regards to the inclusion criteria, the articles were selected based on the inclusion of one of the following topics; Meniere disease evaluation, management, and diagnosis. Exclusion criteria were all other articles that did not have one of these topics as their primary endpoint.

Review

Meniere’s disease is a multifactorial, complex, and common inner ear disease. The prevalence of Meniere disease varies between countries from as low as 3.5 per 100.000 and up to 513 per 100.000. The incidence also varies and the cases reported in a year can vary between 4 cases and up to 20 according to studies. Some factors showed a higher incidence and prevalence of this disease, such as race (white), gender (women), and age (older people). However, some studies suggested that the old age factor is related to the late age of diagnosis and presentation to the clinic rather than the actual age of disease onset [3-5].

Pathophysiology

Multiple hypotheses have been suggested about the pathophysiology and etiology behind such disorder. However, until the present day, there is no globally accepted theory on the precise underlying pathophysiology. Some suggested endolymphatic accumulation in the cochlea and the vestibular organ revealed in some temporal bone studies in Meniere patients. Endolymphatic hydrops is generally attributed to the overproduction of endolymph, and/or decrease in absorption mechanisms in the inner ear. This will result in the distention of endolymphatic space because of the increased endolymphatic volume [6]. One of the consequences of this is Reissner’s rupture which would lead to leaking the (toxic) potassium-rich endolymph into the cochlear hair cells and vestibulocochlear nerve, which will damage them [7]. As a result, the association between endolymphatic hydrops with Meniere diseases and hearing loss (of more than 40dB) have been established [8]. However, association with vertigo have been controversial, thus endolymphatic hydrops is not entirely specific for all symptoms of Meniere disease. Moreover, endolymphatic hydrops have been found in patients with no Meniere disease, but suffering from other pathologies, such as idiopathic sensorineural hearing loss. However, all patients with Meniere disease have endolymphatic hydrops. Therefore, the exact etiology of this disorder is not certain and multiple factors can play a part in its pathophysiology such as genetics and environmental factors. Moreover, many autoimmune diseases were associated with this disease, including; systemic lupus erythematosus, ankylosing spondylitis, and rheumatoid arthritis [9, 10].

Clinical Features

Usually, the patient will present to the primary health care center due to vertigo. Thus, the clinician shall be able to differentiate between peripheral, central, and cardiovascular causes of vertigo [11, 12]. Some signs and symptoms which are dangerous and indicate a probable central origin of vertigo are acute deafness, acute headache (or migraine), new onset of neurological symptoms (or signs), and nystagmus (vertical torsional or rotatory) [13]. The vertigo is characteristically episodic, and thus the family physician shall further investigate any old episodes, the character of the present (and old) vertigo. Other clinical features can be noted clinically or identified by history such as sensorineural hearing loss, tinnitus, and/or aural fullness are critical for diagnosis. Moreover, some neurovegetative symptoms can be seen in these patients including; sweating, nausea, vomiting, and diarrhea. Tinnitus is reported commonly by patients and usually is ipsilateral [14-16]. The family physician shall take a full otologic history and perform the related examination as well. The examination shall include facial nerve testing, assessment of nystagmus (Frenzel goggles, head impulse testing), and full otologic examination (Rinne and Weber tests). Frenzel goggles may not be available in all primary health care settings, and the head impulse test can be done at least but with less sensitivity. Both tests in an acute patient may show horizontal nystagmus. In the Rinne and Weber test, a Meniere patient in an acute or advanced state will usually show a sensorineural hearing loss [17]. The natural course of this disease is usually progressive with unpredictable fluctuations. However, the frequency of vertigo attacks is higher in the first few years, and in some cases, it may decline and even surcease. Nevertheless, some patients with MD still report severe attacks of vertigo even after 20 years from diagnosis, thus, the vertigo natural history is variable and not fully understood. Unfortunately, the hearing loss will almost always worsen in a progressive matter. Another difficult issue for clinicians is the episodic feature of this disease, making it difficult to differentiate between episode symptom-free period, and positive effects of treatment and even the possibility of alternative diseases that may mimic MD [18].

Differential Diagnosis

The primary health care physician must keep in mind multiple diseases and causes that may present with similar signs and symptoms in order to diagnose accurately. Some central neurologic conditions such as basilar migraine will present with vertigo similar to MD but with no aural symptoms. Similarly, vestibular neuronitis may present with vertigo and balance issues, but no aural symptoms. Some other conditions such as benign paroxysmal positional vertigo will be noted upon history and physical examination. The patient in this disease will present with vertigo lasting seconds to minutes, which is associated with head movements, but with no aural symptoms. Some other conditions such as otosclerosis and vestibular labyrinthitis may present with aural symptoms and must be ruled out accordingly. Some medications; such as aminoglycosides and loop diuretics have been reported to give similar clinical symptoms and drug history must be taken thoroughly [19].

Diagnosis

The diagnosis of Meniere disease is mainly clinical and two widely used and recognized criteria used are the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) criteria developed in 1995, and Barany society developed in 2015. The Barany society diagnostic criteria can differentiate between a probable and a definite Meniere's patient. According to this criteria, a definitive Meniere's disease case will have two or more spontaneous episodes of vertigo, with each lasting between 20 minutes to 12 hours, and fluctuating aural symptoms (such as tinnitus, hearing loss, and fullness) in the affected ear. Other diagnostic criteria include a documented low (or medium) frequency sensorineural hearing loss in the affected ear at least once with relation to episodes of vertigo and the clinician cannot explain the symptoms with any other vestibular diseases. Having two (or more) episodes of vertigo or dizziness lasting 20 minutes to 24 hours, with fluctuating aural symptoms in the affected ear, with other causes excluded will make the patient "probable" to have the disease from a clinical point of view [20]. These recent criteria are simpler and thus easily conducted to have a more unified way of diagnosing the disease globally. Nevertheless, some additional tests such as Magnetic Resonance Imaging (MRI), can be carried out which can identify and visualize the endolymphatic hydrops that provide useful information in patients with characteristic symptoms. Ruling out retro cochlear pathology can also be done with MRI in patients with unilateral hearing loss [21]. Moreover, audiometric evaluation is essential in all patients, and a fluctuating low-frequency unilateral sensorineural hearing loss is a hallmark of the disease. However, this hearing loss can affect all frequencies, especially in progressive and severe cases. The hearing loss can progress to all frequencies. Some other advanced studies such as brainstem evoked response audiometry (BERA), cochlea function assessment, and auditory nerve overlapped waveform (ANOW) can be done. In addition, utricle and saccule function can be assessed with vestibular-evoked myogenic potentials (VEMP), and semicircular canal function can be measured with a caloric test and video-head impulse test (vHIT). The results usually show an under-functioning in these organs in up to 74% of patients, with complete loss of function in up to 11%. The timing of such tests does not need to be in the emergency setting, and thus the family physician can refer these patients to higher centers to carry out such advanced tests [22, 23].

Management

The main goals of treatment that the clinician shall focus on are reducing the severity and frequency of vertigo attacks, hearing loss, aural fullness, tinnitus, and improving the overall quality of life (QOL). The family physician shall refer patients, once stabilized, with suggestive signs of Meniere disease for otolaryngologic consultation. The treatment approach in Meniere disease can be either conservative or surgical. The choice of the appropriate options is based on previous treatment history and the severity of their symptoms. However, a general approach is to start with conservative as the first option for the newly diagnosed patients, in order to avoid possible side effects. Conservative treatment first-line option includes dietary modification (e.g. restriction of salt, caffeine, alcohol intake), and several drugs. Some other dietary changes such as abundant water intake (35 mL/Kg/day), which will result in ADH decrease has been suggested to improve and prevent hearing loss. Caffeine cessation along with a gluten-free diet has been suggested as well in some studies as ways to prevent triggering new episodes (caffeine) and induce remission [24]. Multiple drugs have been suggested in the management of MD when the dietary modifications fail. For acute attacks, dimenhydrinate and benzodiazepines have been used with good outcome in patients. Moreover, prophylactic therapy with betahistine, Intratympanic steroid injections, β -blockers, and diuretics have been proposed, but their efficacy remains inconclusive. Intratympanic gentamycin injections have been used to treat vertigo in these patients even though it has toxicity towards cochlear cells and strong ablation towards vestibular cells. As a result, one of the side effects of gentamycin is sensorineural hearing

loss and that is why the exact dosage of this drug is still controversial [25]. When all of the previous methods fail, the patient is offered the surgical option once all the advanced diagnostic tests mentioned earlier are done. Moreover, when ablation is considered a vestibular function of both ears shall be assessed. Endolymphatic sac surgery (ELSS) is the preferred surgery in early-stage refractory cases because it preserves the vestibular function and well-hearing function [26]. More destructive surgeries are to be considered in the more severe and late stages of refractory MD, these include the vestibular nerve section (neurectomy) and labyrinthectomy. However, these surgeries have a major effect on the patients, since labyrinthectomy will lead to a complete hearing loss in the operated ear. Thus, the choice of the operation depends on the hearing status of the patient, if the hearing is adequate (even if poor) vestibular neurectomy is performed, and if not, then labyrinthectomy is done. Some vestibular neurectomy side effects include cerebrospinal fluid leak, meningitis, and epidural hematoma. Generally, both of these destructive operations achieve total control of vertigo but they still to be done as a last resort [27].

Prognosis and complications

Patients usually have the highest episodes of vertigo in the early years, but in later years, many have no vertigo reach a steady-state phase with no vertigo. Some studies have suggested that this result has been achieved regardless they received treatment or not [28]. On the other hand, loss of hearing is notable with fast progression in the early years but stabilizes in later years, but unfortunately, the loss is permanent. The disease may affect both vestibular organs in almost half of the patients (47%) within twenty years, and in late stages, some patients experience Tumarkin's Otolithic Crisis (sudden drop with no loss of consciousness). Patients with MD have reported significant reduced quality of life when compared to healthy people of the same age [27, 29, 30].

CONCLUSION

One of the most common inner ear pathologies is Meniere disease affecting a wide range of people. The clinician in a primary health care center needs to be able to take a thorough history and perform a thorough physical examination when the patient presents with vertigo. This is pivotal since the diagnosis is clinical, and early identification of the disease helps in the long-term outcome. Fortunately, there are many effective medical and surgical therapies that can control vertigo and preserve hearing in these patients. However, multiple studies are still needed to establish enough evidence regarding multiple issues regarding the disease and the full effect of some suggested treatment options.

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