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Short Note

Meniere's Disease

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DEFINITION

Meniere's disease (MD) is a clinically defined disorder, characterized by recurrent episodic vertigo with aural fullness, tinnitus, and sensori neural hearing loss [1]. It affects approximately 0.2% of the world's population. The term Meniere's syndrome is used if an identifiable cause of this increased pressure is present. The term Meniere's disease is used if there is no identifiable cause [2].

EPIDEMIOLOGY

Meniere's disease (MD) can begin at any age but patients typically present with symptoms between ages of 20 and 40. Meniere's syndrome in children is rare and is often associated with congenital malformations of the inner ear. The estimated annual incidence of the disease is two per 1,000 [3]. There is no predilection between races; a predisposition for females and industrialized countries has been reported. MD has a high familial aggregation, and even though a genetic heterogeneity is observed, most families have an autosomal dominant inheritance pattern [4,5].

PATHOPHYSIOLOGY

Several theories regarding the pathogenesis of Meniere's disease have been proposed. The most accepted theory is the endolymphatic hydrops with the vertigo attacks being caused by contamination of perilymph with potassium rich endolymph due to rupture of Reissner's membrane [6]. High concentrations of extracellular potassium depolarize the cochlear and vestibular hair cells, resulting in acute loss of function [7]. The endolymphatic hydrops can be caused by malabsorption of endolymphatic fluid in the endolymphatic sac or duct [8]. The endolymphatic hydrops theory has been questioned, because hydrops could be a coincidental finding in asymptomatic patients. In a comparative human temporal bone analysis with MD, the volume of the vestibular aqueduct, endolymphatic duct, and intratemporal endolymphatic sac was lower, and the external aperture of the vestibular aqueduct was smaller as compared with nondisease bones from donors with endolymphatic hydrops without vestibular symptoms. These anatomic reported findings could explain why some patients with hydrops can be asymptomatic whether others have vestibular symptoms [9]. Another Proposed etiologies include hypoplasia of the vestibular aqueduct, immunological and viral mechanism (herpes simplex, varicella zoster, and cytomegalovirus) and ischemia of the endolymphatic sac or inner ear [10,11].

Migraine occurs more commonly in patients with MD than in the general population, supporting a vascular etiology [12]. In spite of the multiple theories that have been proposed, the development of MR imaging of endolymphatic hydrops supports the central role of endolymphatic hydrops in the pathology of MD, and confirms the same result from temporal bone studies [13].

SYMPTOMS

Recurring attacks of vertigo (96.2%) accompanied with low pitch tinnitus (91.1%) and ipsilateral sensori neural hearing loss (87.7%) [14]. Aural fullness and nausea may be present with these symptoms. Patients describe vertigo as a rotatory spinning sensation for about 20 minutes to 24 hours duration [15]. Hearing loss is usually fluctuating, and often initially affects the lower frequencies due to the important affection of the apical portion of the cochlea. Hearing loss progresses over time, and often results in permanent hearing loss at all frequencies in the affected ear over an 8 to 10 year period [16].

DIAGNOSIS

Horizontal rotatory nystagmus during attacks is a consistent finding; the direction varies over the course of the attack. In the initial phase, nystagmus beats toward the affected ear. Later a paretic nystagmus phase appears, thus beating towards the healthy ear, finally there is a recovery phase where nystagmus beats back to the affected ear [17]. The most common audiometric pattern in early MD is a low frequency or combined low and high frequency sensory loss with normal hearing in the mid frequencies. Over time the hearing loss flattens [18].

Vestibular testing may be normal in the initial phase of the disease. The caloric test can often localize the involved ear, and a significant caloric response reduction is found in 48% to 73.5% and head- impulse tests are usually normal in 65% to 100% [19,20]. According to the American Academy of Otolaryngology Head and Neck Surgery (AAOHNS) a definite diagnosis of MD is when two or more definitive spontaneous episodes of vertigo lasting at least 20 minutes along with audio metrically documented hearing loss on at least one occasion and the presence of tinnitus or aural fullness in the suspected ear. Recent developments of MR imaging provide a tool for visualizing endolymphatic hydrops with gadolinium chelate as the contrast agent, optimizing the differential diagnosis in patients with MD [13]. At the present day high resolution imaging of the inner ear is not available in all institutions.

Differential diagnosis

Vestibular schwannoma, multiple sclerosis, transient ischemic attack, labyrinthitis, sudden hearing loss, cogan's

syndrome and perilymphatic fistula, diabetes, thyroid disease, migraine [17].

TREATMENT

The goal is to reduce symptoms, stop tinnitus, and reverse hearing loss. Non interventional treatment for MD includes lifestyle adjustments, medical therapies, and rehabilitation. Infrequent vertigo attacks should be treated with vestibular suppressants. If attacks occur more than a few times a year additional intervention is indicated. Traditional treatment includes dietary sodium restriction, diuretics, betahistine and steroids (orally or intratympanic) due to the possible immunologic basis for MD [21]. With medical management about 60% to 87% of patients with MD disease are able to maintain their normal daily activities [22].

Interventional treatment is considered when vertigo attacks occur monthly or more frequently. Although controlling vertigo is the primary goal of treatment, hearing preservation is also important. If the patient's hearing is socially adequate (arbitrarily set at 50 dB and 80% speech discrimination) a non destructive surgical procedure is advised [23]. Destructive procedures include: elective vestibular nerve section, cochlea vestibular nerve section, labyrinthectomy, insertion of aminoglycosides or other medicine into the middle ear to perform a chemical labyrinthectomy [22]. Non destructive procedures include: endolymphatic sac decompression, insertion of ventilation, lateral semicircular canal plugging [22]. Recently new alternatives have emerged such as the positive pressure pulse generator (Meniett®). This device could be an option for patients who fail medical therapy or as an adjunct to medical therapy and has a significant favorable effect on hearing [24]. Loader et al. have presented promising results of the effectiveness of tenotomy of the stapedius and tensor tympani muscles with significant vertigo control rates, decreased postoperative symptoms and important hearing preservation rates [25,26]. More patients and longer follow-up are needed to support this procedure.

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