EAR

MENIERE'S DISEASE

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MENIERE'S DISEASE

DEFINITION:

An inner ear disorder of unknown aetiology characterized by episodic vertigo with associated fluctuating sensorineural hearing loss, tinnitus and aural fullness.

First described by Dr. Prosper Meniere in 1861.

AETIOLOGY:

1) Idiopathic

2) Various theories:

a) Defective absorption by endolymphatic sac

- increased endolymphatic pressure (normally endolymph, secreted by stria vascularis fills the membranous labyrinth & is carried by the endolymphatic duct to the sac where it is absorbed).
- poor vascularity & so poor absportion

b) Distension of membranous labyrinth

- rupture of Reissner's membrane, mixing of endolymph with perilymph causing attacks of vertigo

3) Vasomotor disturbance

- increased sympathetic overactivity – spasm of internal auditory artery interfering the function of cochlear & sensory neuroepithelium.

4) Allergy – inhalant or food allergy , 50% cases

5) Sodium & water retention – increases extracellular fluid (perilymph)

6) Hyperthyroidism – increases hearing impairment

7) Autoimmunity & viral infections (e.g., measles, mumps)

8) Trauma – release of debris into the endolymph

Pathology:

Overproduction or malabsorption of endolymph – distension of endolymphatic system especially affecting cochlear duct (scala media) and saccule – dilatation of cochlear duct – fills the scala vestibule – bulging of Reissner's membrane may herniate through the helicotrema into the apical part of scala tympani – distended saccule comes to lie against stapes footplate. The increased endolymphatic pressure may lead to rupture of Reissner's membrane, mixing of endolymph & perilymph & chemical insult to the basilar membrane. This causes an alteration of in basilar membrane mobility resulting in deafness & tinnitus. This increased pressure also leads to distortion of the ampullae of the semicircular canals causing vertigo.

Pathogenesis:

Periodic rupture of membranous labyrinth is the main pathogenesis.

Has 3 phases:

1) Irritative phase -

Leakage of endolymph into the perilymph – excitation of hair cells due to increased potassium concentration (which is initially neuorotoxic) around the basal surfaces of hair cells producing attacks.

2) Paretic phase –

Due to blockage of neurotransmitter release

3) Recovery (Remission) phase –

Healing of rupture by restoration of normal chemical composition of endolymph & perilymph – termination of attack & improvement of vestibular & auditory functions.

CLINICAL FEATURES:

Age : 35 -60 years

Sex : Equal incidence

Race: Rare in Caucasians & Blacks

Family history : 14-20%, autosomal dominant

Usually unilateral

1) Episodic vertigo: Lasts for few minutes to 24 hours, sudden onset, feeling of rotation himself or his environment, increased intensity over a period of few minutes & then lasts for several hours, accompanied by nausea, vomiting abdominal cramps, diarrhea, cold sweats, pallor & bradycardia , then back to normal level of consciousness.

2) Hearing loss :

may persist for days

sensorineural hearing loss, ipsilateral, initially low frequency, fluctuant

later all frequencies affected leading to moderate degree flat curve in pure tone audiometry

3) Tinnitus :

non- pulsatile, continuous or intermittent

4) Aural fullness :

pressure sensation in the ear or head & neck

5) Somatopsychic effects :

fear of vertigo

An important sign of the disease is nystagmus (an involuntary, oscillatory movement of eyeball)

A typical attack has 3 phases regarding nystagmus:

1) Irritative phase –

Nystagmus usually horizontal towards the affected ear, lasting for <1 hour

2) Paretic phase –

Nystagmus beats away from the affected ear, lasting for several hours, even 1 or 2 days due to peripheral hypofunction

3) Recovery phase:

Nystagmus towards affected ear, lsts for several hours, 1 or 2 days due to brain-stem compensation

Following remission of attack:

Peripheral vestibular function recovers & nystagmus again beats towards the affected side.

In early stage :

Patient is well between attacks

Later stage :

Drop attacks known as Tumarkin or otolithic crisis due to otolithic dysfunction.

Patient just drops to the ground without warning with no loss of consciousness or vertigo.

Related syndromes to Meniere's disease :

Lermoyez syndrome : A syndrome with sudden sensorineural hearing loss which improves during or after an attack of vertigo.

Meniere's syndrome :

Many diseases of the inner ear or temporal bone e.g., CSOM, Syphilis, Cogan's syndrome, otosclerosis, Mumps, Trauma can produce the clinical picture of Meniere's disease after many years.

DIAGNOSIS:

Guidelines by the American Academy of Otolaryngology – Head and Neck Surger y

1) **Definite Meniere's disease :**

If patient has 2 or more spontaneous attacks of vertigo, each lasting 20 mins. or longer + Hearing loss documented by PTA on at least one occasion with other possible causes excluded.

2) **Probable Meniere's disease :**

Single spontaneous attacks of vertigo, each lasting 20 mins. or longer + documented hearing loss with unilateral hearing loss, tinnitus & aural fullness all at the same time.

3) **Possible Meniere's disease :**

2 or more episodic attacks of vertigo with no documented auditory symptoms during the attacks.

DIFFERENTIAL DIAGNOSIS:

1) Vestibular neuronitis
2) Migraine
3) BPPV
4) Sudden sensorineural hearing loss
5) Vestibular schwannoma
6) Cogan's syndrome

INVESTIGATIONS :

1) Pure tone audiometry – low frequencies affected with rising curve, then flat affecting both middle & low frequencies
2) Electrocochleography – best objective test, normally summating potential (SP) is 30% of action potential (AP). Here SP is >30% of AP
3) Recruitment test is positive
4) Speech discrimination is < 90%
5) Caloric test – reduced response on the affected side in 75% of cases
6) Glycerol test : A dehydrating agent reducing endolymphatic hydrops producing an improvement in the audiogram & the Electrocochleography ECochG.

PTA & SD scores are recorded before. Then 1.5 ml / kg of glycerol + equal amount of water + a flavouring agent e.g., lemon juice given orally.

Then PTA & SD scores recorded again 1-2 hours after ingestion – an improvement of 10 dB & a gain of 10% in SD scores seen.

TREATMENT:

A) Medical B) Surgical

A) Medical:

1) Reassurance & explanation that it is not life-threatening. Although there is no cure, modern treatment can improve the quality of life & lessen the severity & frequency of vertigo episodes. But there aren't any treatments for the hearing loss.

2) Bed rest with head supported on pillows.

3) Vestibular sedatives – Prochlorperazine (Stemetil) might control nausea & vomiting during an episode of vertigo.

4) Diuretic & limit salt intake to reduce fluid retention.

5) Hearing aid might improve hearing.

6) Positive pressure therapy: a device called Meniett pulse generator applies pulses of pressure to the ear canal through a ventilation tube at home 3 times daily for 5 mins. But effectiveness is not yet determined.

7) Limit caffeine, alcohol & tobacco which might affect the fluid balance

8) Benzodiazepines: Diazepam – reduces sympathetic activity, vestibular nuclear activity & relieves anxiety.

9) Cerebral vasodilator: (Beta) β- histine hydrochloride increases labyrinthine blood flow

10) Middle ear injections: Medications e.g., gentamicin, steroids injected into the middle ear and absorbed into the inner ear may improve vertigo.

B) Surgical Treatment:

When other treatments fail**, surgery** might be an option.

1) Endolymphatic sac decompensation
2) Labyrinthectomy
3) Vestibular nerve section.

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THANK YOU