The ischemic mechanism of Meniere attacks

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Course objectives

At the conclusion of this course you should be able to:

- Compare various theories of Meniere attack causation
- List vascular risk factors for Meniere attacks
- Explain the relationship of hydrops to inner ear ischemia
Classic Meniere’s attack

- Starts with a stuffy, full feeling in one ear
  - Can be present for days
- Ear seems deadened to sound
  - Audiogram shows a low tone sensorineural hearing loss
- A low, roaring tinnitus develops
- Over minutes to hours, a vertigo spell begins
  - Lasts several hours
  - Environment rotates
  - Vomiting
- The affected ear eventually dies in a progressive fashion
Older models of Meniere attacks

- Pathology is hydrops

- Elevated endolymph pressure ruptures membranes, contaminating endolymph with perilymph

- Elevated endolymph pressure directly affects the vestibular sensors in some other fashion
Problems with these models

- Rupture is sudden: why do attacks seem to come on more gradually?
- Ruptures heal slowly: why do attacks end in a few hours?
- How does extra endolymph pressure make hair cells or end organs malfunction during attacks?
- Why does permanent damage gradually develop?
Problem with hydrops

- Everyone with MD has hydrops
- Most people with hydrops do not have MD
- Hydrops is found in:
  - 16% of children
  - 8-9% of adults
- 27 million people in the US
- Hydrops is VERY COMMON; MD is rare
Common explanation

- Hydrops is a red herring
  - End stage pathology common to many disorders
  - Epiphenomenon of unclear relationship
Alternative model: migraine/vasospasm

- **1908** “Meniere’s disease is simply migraine”

- **1929** Meniere syndrome is due to vasospasm
  - Lermoyez M: La Vertige qui fait entendre (angiospasme labyrinthique). Annales des maladies des oreilles et du larynx 1929;48:575-583

- **1943** Meniere attack is aural migraine (vasospasm)

- **1991** 94% of basilar migraine patients had Meniere symptoms

- **1996** Meniere syndrome in a migraine kindred

- **2007** Migraine is a risk factor for Meniere disease
Problem with vasospasm:

Why just one ear?

- Most Meniere cases have repeated attacks in the same ear resulting in unilateral deafness

- Vascular disorders are systemic: bilateral

- Meniere’s syndrome can be bilateral, but not often
  - Bilateral hydrops is found on autopsy in these
  - Most hydrops is unilateral (30/39 or 76%)
Problem with vasospasm

**Why hydrops?**

- Hydrops on autopsy is found in association with h/o Meniere attacks during life

- All MD cases have hydrops

- Can vasospasm cause hydrops? How?
Problem with vasospasm

- Meniere attacks often occur in people without a personal or family history of migraine

- Meniere attacks are associated with a large number of vascular risk factors not related to migraine
Common risk factors for branch retinal artery occlusion & Meniere attacks

- Atherosclerosis
- Trauma
- Infection: syphilis, lyme, HIV
- Sickle cell disease
- Hormonal: Pregnancy, oral contraceptives
- Vasospasm: Migraine
- Clotting disorders: homocystinuria, protein C deficiency
- Embolization
- Vasculitis
  - Systemic lupus erythematosus
  - Sarcoid
  - Churg-Strauss syndrome
  - Granulomatous angiitis
  - Giant cell arteritis
  - Polyarteritis nodosa
  - Wegener's granulomatosis
  - Behcet's disease
  - Rheumatoid arthritis
  - Inflammatory bowel disease
  - Susac syndrome
Our model: Ischemia

- The mechanism of the Meniere attack is acute partial ischemia of the sensory tissues of the inner ear.

- This can be spotty, affecting some sensors and not others

- After re-perfusion, patchy areas of permanent tissue death may be found

- With repetition, these dead areas become confluent, resulting in near complete loss of aural function
Basic hemodynamics of ischemia:

Perfusion pressure in tissues \( P_p = P_a - P_v \)

Arterial pressure = inflow

Venous pressure = outflow resistance

**Starling resistor** = anything externally compressing capillary bed that exceeds venous outflow pressure

\( P_p = P_a - P_{sr} \)
Perfusion pressure (brain)

- Inflowing arterial pressure
  - $CPP = MAP - ICP$ (if ICP is higher than JVP)

- Outflowing venous pressure
  - $CPP = MAP - JVP$ (if JVP is higher than ICP)

MAP: mean arterial pressure
JVP: jugular venous pressure
ICP: intracranial pressure
\[ CPP = MAP - ICP \]

Extrinsic pressure on vascular bed

**STARLING RESISTOR**

- Brain: ICP=CSF pressure
- Eye: intraocular pressure
- Ear: intra aural fluid pressure
The ischemic threshold

$$CPP = MAP - ICP$$

- Average CPP is 85mmHg
- Ischemia usually ensues when CPP drops below 50mmHg
- There is about a 35mmHg “buffer” against ischemia
- Anything that reduces MAP by 35mmHg or raises ICP/resistor pressure by 35 mmHg, or both in combination, can cause acute ischemia
Vascular disease impacts

- MAP can drop very low with vascular disorders
  - Tight stenotic lesions
  - Clot in vessel
  - Vasospasm
  - Rigid arterial walls

- This can drop the ischemic buffer to the 1-2 mmHg range

- Even a small increase in resistor pressure can cause ischemia if this occurs
Our model: Vascular disease

- Vascular disease lowers MAP so ischemic risk is elevated

- Intermittent vasospasm (migraine) could explain the self-limited Meniere attack, but it would not always return to the same ear

- Other vascular disorders are not intermittent on the time scale of a Meniere attack nor are they focal

- Therefore, vascular factors alone cannot explain MD

- We need a Starling resistor that is focal to the ear
Our model: Hydrops

- Hydrops is focal to the Meniere ear
  - Found in 100% of cases
    - Rauch SD 1989
    - Merchant et al 2005

- This suggests that a resistor lies within the hydropic ear

- High resistor pressure in ear + intermittent vasospasm in migraine = ischemic Meniere attack
Our model: Intermittent resistor

- Hydrops in the face of fixed vascular disease cannot act as a continuous resistor
  - Continuous resistor = infarct, not spells

- Therefore the resistor associated with hydrops must be intermittent

- Therefore pressure in the hydropic ear must vary, giving rise to intermittent spells
What pressures vary in the hydropic ear?

■ Absolute atmospheric pressure
  ■ MD patients have dizziness with weather changes
  ■ Meniere attacks can be terminated by lowering atmospheric pressure in a chamber

■ Response to head position
  ■ MD patients like to rest with head propped up during spells
  ■ Nystagmus in hydropic animal models occurs only when head is inverted

■ Response to hydration
  ■ MD responds to salt intake

■ Therefore, the hydropic ear must be able to transmit pressure changes to the vascular bed to a greater degree than normal ears.
Range of external pressure variation

- **Atmospheric pressure**
  - Weather: varies by 1-2 mmHg (max: 10 mmHg)
  - Elevation: decreases by 130 mmHg per mile

- **CSF pressure**
  - Varies from <0 (upright) to 25 mmHg (supine)
An attack will occur when:

- There is reduced MAP due to one or more vascular disorders, particularly if vasospasm is present

  AND

- Interstitial tissue pressure is transiently elevated in the hydropic ear due to position, hydration, or atmospheric pressure change
Classic Meniere attack

- Starts with a stuffy, full feeling in one ear
  - Associated with hearing loss; pressure building in ear
- Ear seems deadened to sound
  - Insufficient perfusion of cochlea
- A low, roaring tinnitus develops
  - Corresponds to hearing loss in low tones
  - Hydrops is more common and severe in apex of cochlea
- Over minutes to hours, a vertigo spell begins
  - Insufficient perfusion of labyrinth
- The affected ear eventually dies in a progressive fashion
  - Patchy ischemia gradually results in confluent damage
Treatment
Aggressively reduce vascular risk on multiple fronts

- Treat vasospasm
  - Migraine prophylactic meds
  - Magnesium
  - Trigger avoidance
  - Aspirin

- Prevent elevated aural pressures
  - Sodium restriction
  - Diuretics
  - Elevate head at night
  - Decompression procedures?
Treatment: new thoughts

Acute attacks: neuroprotection

- Calcium channel blockers
  - Nimodipine

- IV Magnesium
  - Calcium channel/NMDA blocker

- Other possibilities
  - Steroids?
  - Local hypothermia?

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