Less common vestibular disorders presenting with funny turns

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Making the diagnosis

HISTORY TAKING

Clinical examination

Vestibular testing
Clinical presentation

One acute episode - recurrent episodes

Auditory symptoms - no auditory symptoms

Spontaneous episodes – triggering factors

Isolated vestibular symptoms – associated symptoms
Recurrent episodes of vertigo induced by changes in intracranial or middle ear pressure

- Coughing, sneezing, load noises
- Defect by the labyrinthine canal causing audiovestibular symptoms

- Perilymphatic fistula
- Superior semicircular canal dehiscence
Perilymphatic fistula

- Leakage of perilymph; usually round or oval window
- Chronic otitis media, cholesteatoma, otosclerosis or temporal bone fractures
- Positive Hennebert’s sign/fistula test – pressure sensitivity
- Symptoms  - episodic vertigo/chronic disequilibrium
  - hearing loss (sudden, fluctuating, progressive)
  - tinnitus/aural pressure
Hennebert’s test

- Positive result indicating perilymphatic fistula
  - when positive pressure - nystagmus towards the ipsilateral ear
  - when negative pressure – nystagmus towards the contralateral ear
Continue perilymphatic fistula

- No available diagnostic tests
- Erosion, related to cholesteatoma/otosclerosis visible on high resolution CT
- If disabling symptoms – surgical exploration with repair with graft

www.otosclerose.nl
Superior semicircular canal dehiscence

- Episodic vertigo and oscillopsia
- Hypersensitivity to bone-conducted sounds – autophony
- Pseudoconductive hearing loss – better hearing thresholds on bone conduction to low frequencies
- Mild low-frequency hearing loss
Pure tone audiogram

Right ear-HL

Left ear-HL

From http://synapse.koreamed.org/
Continue superior semicircular canal dehiscence

- Often bilateral
- ? Congenital or developmental
- High resolution CT - dehiscence of bone overlying the superior semicircular canal
- Abnormally low vestibular evoked myogenic potential (VEMP) threshold
- With debilitating symptoms - canal plugging
  - resurfacing
Superior semicircular canal dehiscence

www.dbi.udel.edu

otosurgery.org
From http://www.dizziness-and-balance.com/
Fig. 1. Hennebert’s sign associated with left superior semicircular canal dehiscence syndrome as seen under video-oculography goggles. Positive pressure in the left ear causes a conjugate vertical-torsional ocular deviation where the eyes rotate up and away from the left ear (solid arrows). This reverses with negative pressure (dotted arrows). These eye movements align in the plane of the left superior semicircular canal.

Shuman et al., Laryngoscope 122: February 2012
Vestibular Schwannoma (Acoustic Neuroma)

- A slow growing benign tumour of the vestibular branch of the vestibulocochlear nerve that arises in the auditory canal
- Progresses to cause ipsilateral tinnitus and sensorineural hearing loss as the VIIIth CN is compressed
- Dizziness and imbalance
- Large tumours may give ipsilateral cerebellar signs
  - Ataxia
  - Nystagmus
Acoustic neuroma on vestibulocochlear nerve puts pressure on facial nerve
Continue Vestibular Schwannoma

Treatment - monitoring
  - surgical removal
  - radiosurgery ("gamma knife")
Arnold-Chiari Malformation

- Dizziness/disequilibrium exacerbated by neck extension
- Headache – back of the head worsening with coughing/sneezing
- Downbeat nystagmus
- Cerebellum herniates through the foramen magnum
- Congenital – small posterior fossa or malformation brain/spine (estimated 1/1000 births)
- Injury
- Increased pressure (tumour/hydrocephalus)
Arnold-Chiari Malformation
Continue Arnold-Chiari malformation

• Diagnosis - MRI findings
• Management - wait and watch
  - surgical (posterior fossa decompression, shunting)
Familial periodic ataxia/vertigo

• Periodic ataxia – recurrent episodes of poor coordination and balance
• Additional associated symptoms – vertigo, dysarthria, muscle weakness
• Syndrome varies considerably from family to family - suggest separate disorders
• Mainly autosomal dominant inheritance
Episodic Ataxia Type 2

- Recurrent disabling attacks of imbalance, vertigo and ataxia (minutes to hours)
- Provoked physical exertion or emotional stress
- Central ocular motor disturbances >90% (particularly downbeat nystagmus)
- Cerebellar dyscoordination of limbs, dysarthria, paraesthesia
- Migraine headaches (>50%)

Continue episodic ataxia type 2

- Autosomal dominant (onset from early childhood to sixth decade), caused by mutations in the calcium channel gene CACNA1A
- MRI - cerebellar atrophy
- Treatment acetazolamide (250-1000mg per day) – prevents or attenuates the attacks in 50-75% but effect only transient. (4-aminopyridine).
Making the diagnosis

- Auditory symptoms/ear infections
- Triggering factors
- Family history
- Associated symptoms
Thank you!