NEWSLETTER
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in Neurotology (ARNO)
CYCLOPS™ Learning Lab
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DIZZCOVER

Systematic discussions on specific neurotology topics

INDEX

1. Issue In Focus
1.1 Central Vertigo: A brief writeup on this topic

2. Vertigo Grand Round
An online presentation of a neurotology case held on the last Friday of every month

2.1 Presentation of 3 cases of Dizziness with different Causes and therapeutic approach
2.2 Discussion Pearls
2.4 Link to the online VGR Video

3. Journal Scan
A brief review of current publications of relevance on the topic

4. History
An attempt to present events and people pictorially.

5. Gallery
Art of the Month

1. Issue in Focus:

1.1 Central Vertigo

Central vertigo occurs because of strokes, intracranial tumors, metabolic conditions, and paroxysmal or degenerative disorders. It mandates an early diagnosis and proper treatment, given its potential to worsen to life threatening levels with grave prognosis. Strokes are usually due to posterior cerebral circulation vascular compromise.

Dizziness or vertigo is the most common presenting symptom of posterior circulation strokes, which accounts for about 20% of all ischemic strokes. Patients with vertigo, due to strokes have a high chance of misdiagnosis in the emergency department despite modern imaging techniques.

To improve identification of vertigo due to strokes and an integrated approach using detailed neuro-otological examination such as HINTS plus, risk estimation strategy including ABCD2 Scoring system, and perfusion imaging is recommended.

Terms Explained

HINTS is an acronym for HI (Head Impulse Test), N (Nystagmus), TS (Test of Skew) PLUS (+/- auditory examination).

Head Impulse Test (HIT): This test assesses the vestibulo-ocular reflex (VOR), which is a reflex that helps to stabilize vision during head movements.

The examiner rapidly turns the patient's head to the side while the patient tries to keep their gaze fixed on a target. In peripheral vertigo, there will be a corrective saccade (a quick eye movement) to bring the eyes back onto the target. In central vertigo, there will be no corrective saccade or a corrective saccade that is delayed.

Nystagmus: Nystagmus that changes direction is more likely to be caused by central vertigo, while nystagmus that does not change direction is more likely to be caused by peripheral vertigo.

Test of Skew: The test of skew assesses for vertical misalignment of the pupils. This can be a sign of a brainstem stroke.

The examiner shines a light into the patient's eyes and observes the position of the corneal light reflex (the reflection of the light on the cornea). If the corneal light reflexes are at different levels, this is a positive test of skew.

Auditory assessments: Presence or absence of hearing loss is a significant indicator of peripheral causes of vertigo (though not invariable).

B) ABCD2 SCORING SYSTEM:

The ABCD2 score helps to predict stroke risk after a transient ischemic attack (TIA). Originally developed for TIA patients, ABCD2 is also found useful for those with acute vertigo/dizziness. This scoring system is based on five parameters (age, blood pressure, clinical features, duration of TIA, and presence of diabetes).

Table:

Risk Factor	Points	Score
Age >60 years	1	
Blood Pressure Initial Systolic BP >140 mm Hg OR Diastolic BP >90 mm Hg	 1 2	
Clinical features of TIA (choose one)* Unilateral weakness with or without speech impairment OR Speech impairment without unilateral weakness	 1 1	
Duration TIA duration >60 minutes TIA duration 10-59 minutes	 2 1	
Diabetes	1	
Total ABCD2 Score	0-7	

Score 1-3 (low): 2 day risk = 1.0%, 7 day risk = 1.2%
Score 4-5 (moderate): 2 day risk = 4.1%, 7 day risk = 5.9%
Score 6–7 (high): 2 day risk = 8.1%, 7 day risk = 11.7%

D) PERFUSION IMAGING

The several imaging modalities that can be used for perfusion imaging are

(a) Magnetic Resonance Perfusion Imaging (MRI)
(b) Computed Tomography Perfusion Imaging (CTP
(c) Single-Photon Emission Computed Tomography (SPECT)
(d) Positron Emission Tomography (PET)

VERTIGO Grand Rounds

2.1 Case Presentations

Case 1

A 21-year-old woman with a history of diminished right hearing, acute abrupt onset of vertigo, nausea, vomiting and with reduced right hearing.

On Examination: left beating nystagmus, head thrust positive to left, progressive right facial nerve palsy collapsed in the emergency room, ventilated and after improvement was assessed.

Imaging (CT): Aneurysm rupture in the right anterior inferior cerebellar Artery (AICA) region resulting in hemorrhage into the intra-axial middle fossa. She underwent embolization treatment in the intensive care unit (ICU) and was discharged.

Diagnosis: Right AICA Aneurysm, severe ataxia (loss of muscular coordination), acute unilateral vestibular loss and dizziness, unilateral hearing loss, unilateral tinnitus and facial nerve palsy.

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VNG Findings on first referral to Audiovestibular medicine department :

Without fixation on primary Gaze: The patient had bilateral direction changing gaze evoked nystagmus. When she looked towards the right, it is beating towards the right. But when she looks towards the left it is changing its direction to beat to the left. There was a clear upbeat nystagmus consistent with cerebellar lesion. Head shaking provoked left beating nystagmus consistent with peripheral disorder. Hence patient presents with central and peripheral signs not in keeping with the original presentation.

The MRI contrast of brain confirmed that she had cerebellar infarct. It is a recognized complication of embolization procedure.

She was rehabilitated as follows:

Auditory rehab- CROS HA, vestibular rehab, neuro rehab and cognitive rehab resulting in near complete recovery and return to normal life

VNG findings after rehabilitation sessions shows much improved bilateral gaze evoked nystagmus. There was minimal residual vertical nystagmus beats and improvement in gaze control.

Discussion:

There is a probable Brain Stem Infarct. As ICA also supplies to the PONS, she also had a pontine infarct causing the additional gaze evoked nystagmus with changing direction while looking towards the left suggesting brain stem infarct and Bruns nystagmus where there is a combination of gaze evoked as well as vestibular sort of nystagmus. In right, it is coarser and slower but in left the jerks are faster suggesting a central involvement. It also had consistent upbeats. During headshaking, vestibular asymmetry also comes in play. There is also left beating nystagmus even at rest. So, a left beating and up beating and a direction changing gaze evoked nystagmus is observed. It was broadly classified into cerebellar eye movement sign. Therefore, it could be cerebellar or brain stem lesion.

In labyrinthitis, nystagmus is unidirectional, in this case gaze evoked nystagmus with changing direction suggesting brain stem infarct and Bruns nystagmus.

Cerebellar infarcts are a complication of embolization of AICA/PICA/SCA though it may also arise due to spasm of the artery due to the bleed.

It is recommended that an MRI be done as the first line cerebral imaging preferred over a CT scan, but even the MRI cannot rule out a central cause of vertigo and it will need to be repeated after 72 hrs, especially if the HINTS test suggest central vertigo.

Case 2

A 40-year-old female with a complaint of continuous headache, dizziness and imbalance with nausea and diplopia for the last 6 months. She is on medication for temporal lobe epilepsy. She also had treatment for , postnatal depression stress and anemia.

For several weeks, the patient experienced growing symptoms of nausea and dizziness alongside Headaches, exhaustion, nausea, vertigo, and diplopia. She also had phonophobia and photophobia. She experienced fullness in both ears and tinnitus especially in her right ear. She had co-ordination difficulties while using pen, combing hair, typing, and speech production errors.

Previously diagnosed to have vestibular migraine and treated with amityptyline.

On Examination:

Bilateral gaze evoked down beating nystagmus with fixation, left eye skew deviation, direction changing nystagmus with torsional component on positional tests, intention tremor on the left hand, gait abnormalities which were attributed to be functional in nature.

Videooculography Findings:

Downbeating nystagmus in all horizontal and vertical planes with and without optic fixation. Second degree right beating nystagmus without optic fixation. High frequency head shaking test showed downbeat nystagmus with torsional component immediately following the stoppage of head shaking. This then converted into a second-degree spontaneous right beating nystagmus. No Vibration induced nystagmus.

VNG Findings:
Saccadic smooth pursuit at all frequencies, but with otherwise accurate saccades. Spontaneous first-degree nystagmus to the right without optic fixation. Sinusoidal rotation showed symmetrical responses with normal gain and phase lead at most frequencies. VOR suppression was within normal limits.
Impulsive rotation showed a significant right directional preponderance of 26.2%.
Subjective visual vertical and horizontal were within acceptable limits with high variation.

Immediate Management:
Repositioning manoeuvres, Imaging, blood tests including autoimmune, infectious and paraneoplastic screen, neuro-vestibular physiotherapy, Restart Amitriptyline (although resistant to medications)

Imaging:
MRI Head, IAM and MRI Head with gadolinium, DaT scan, HIV, syphilis, Lyme and paraneoplastic markers, PET scan

Findings:
Hb 64.0 g/L with microcytosis, ABR – normal, GAD antibody positive (greater than 50,000)

Treatment:
She was later treated with high dose of steroids, plasma exchange, and immunoglobulin resulting in reduced frequency of seizures, improved gait and eye improved movement.

Discussion: Autoimmune disorders are extremely rare. A high suspicion index is required in cases with atypical findings. The case presented illustrates the importance of testing for autoimmune conditions.

Disclaimer: Above case presentation is very brief summary. For detailed discussion please watch Vertigo grand round video for which link was given below.

VERTIGO Grand Rounds

2.2 Discussion Pearls

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Bruns Nystagmus: Bruns nystagmus is an uncommon type of involuntary eye movement (nystagmus) that occurs in both eyes (bilateral) and is a potential indicator of tumors in the cerebellopontine angle, which is the area where the brainstem meets the cerebellum.

Bruns nystagmus is characterized by a combination of two different types of nystagmus:

- **Slow, large-amplitude nystagmus (gaze-paretic nystagmus):** This occurs when the person looks towards the side of the lesion.
- **Fast, small-amplitude nystagmus (vestibular nystagmus):** This occurs when the person looks away from the side of the lesion. Coil embolization is a minimally invasive procedure used to treat aneurysms. While it offers benefits over surgery, there are potential complications associated with the procedure itself, like the following:

General Embolization Complications:

- **Dissection of proximal vessels:** This occurs when the catheter used during embolization damages the lining of a blood vessel, leading to clotting or blockage or spasm and distal ischemia and even infarcts.
- **Non-target embolization:** The embolic material used to block the clot can migrate to unintended locations, causing complications in other parts of the brain. I do not think this happens with coiling, but with glue embolization.
- **Hemorrhage:** Bleeding in the brain can occur during the procedure or in the following days.
- **Stroke worsening:** If there is already spasm and ischemia, embolization procedure itself can worsen it.

Head shaking Nystagmus:

Head-shaking may be a useful bedside maneuver in diagnosing AICA infarction since it can detect central as well as peripheral pathology.

Head-shaking nystagmus (HSN) typically beats to the intact side in unilateral peripheral vestibular lesions. In contrast, patterns and mechanisms of HSN are varied in central vestibular disorders. In general, central patterns of HSN include unusually strong HSN elicited by weak head-shaking, intense HSN in patients without caloric paresis, ipsilesional HSN, and perverted HSN (i.e., vertical or torsional nystagmus developing in response to horizontal head-shaking).

VERTIGO Grand Rounds

2.3 Online VGR Video

Scan the below QR Code

to watch the complete Vertigo Grand Round Session

POSTURA DR **Static Posturography / Stabilometry**

Postura - DR comes with dozens of calibrated exercise protocols as well as interactive games to rehabilitate every aspect of balance. Patient's progress can be measured from one visit to another.

3. Journal Scan

1. Posterior inferior cerebellar artery aneurysm presenting with severe vertigo and altered sensorium. Vijayashree S. et al., (2024) DOI: 10.7759/cureus.60869

Editorial Comment:

A study reported a rare case of a 31-year-old woman experiencing vertigo, headache and confusion due to a ruptured posterior inferior cerebellar artery aneurysm. Diagnosed through MRI and angiography, she underwent successful endovascular coiling intervention. This study highlights the importance of prompt diagnosis and treatment for this uncommon but serious condition. While endovascular coiling appears to be a promising treatment approach, further long-term studies are needed for validating various treatment approaches.

2. Cerebellar aneurysm with acute cerebellar infarction causing episodic vertigo. Qinghua Z. et al., (2024) DOI: https://doi.org/10.1007/978-981-99-6995-1_19

Editorial Comment:

A 31-year-old man with sudden vertigo underwent workup for common inner ear causes, which were found negative highlighting the challenges in diagnosing the causes of vertigo, especially in younger patients. While initial workup excluded common inner ear etiologies, further investigation revealed a rare cause - a cerebellar infarction and a coinciding left posterior inferior cerebellar artery aneurysm by MRI and CTA. This case emphasizes the importance of maintaining a broad differential diagnosis for vertigo and considering uncommon causes like cerebellar stroke and aneurysms, even in young adults without traditional risk factors.

3. Treatment strategies for saccular anterior inferior cerebellar artery aneurysms: a systematic review. Badary A. et al., (2024) DOI: 10.1007/s10143-024-02338-8

Editorial Comment:

This systematic review analyses AICA aneurysms (over 140 patients), a rare but serious stroke condition. It highlights a diverse profile where 83% patients lacked identifiable risk factors. This emphasized considering AICA aneurysms even without typical risk factors. Treatment modalities have also been discussed. While microsurgery remains common, endovascular procedures showed promise with similar effectiveness and low mortality rates. Importantly, the study stresses tailoring treatment to each patient based on their unique presentation. Overall, this review provides a valuable roadmap for clinicians navigating the complexities of AICA aneurysms.

4. Head-shaking aids in the diagnosis of acute audiovestibular loss due to anterior inferior cerebellar artery infarction. Young EH. et al., (2013) DOI: 10.1159/000345643

Editorial Comment:

This study investigated the characteristics of head-shaking nystagmus (HSN) and its usefulness in diagnosing stroke of the anterior inferior cerebellar artery (AICA) in patients with acute audio-vestibular loss. The majority (83%) of patients with AICA infarction showed HSN. In most cases, nystagmus moved in the direction opposite the side of the stroke (contra-lesional HSN). However, 50% of the patients also displayed atypical HSN patterns. The presence of central HSN, gaze-evoked nystagmus, and a normal head impulse test were highly specific for AICA infarction. This study also reported one case where central HSN was the only indicator of stroke in a patient with isolated hearing and balance problems. The damage to flocculus was linked to perverted central HSN. Therefore, a careful evaluation is important for HSN in AICA infarction (typical and atypical) in patients with sudden hearing and balance loss.

4. History

As early as 460-370 BC, **Hippocrates** described stroke symptoms as "apoplexy," focusing on paralysis and convulsions. Treatments were rudimentary, often involving enemas, bloodletting, and balancing diet and exercise.

Johann Jacob Wepfer (1658), a physician practicing in Schaffhausen, Switzerland was the first to hypothesize that the effects of a stroke were caused by bleeding in the brain. He also mentioned that these symptoms could be caused by a blockage of one of the main arteries that supply blood to the brain. From postmortem studies, he provided information on the carotid and vertebral arteries that supply the brain with blood. He has been hailed as the author of the "classic" modern treatise on apoplexy because he demonstrated that it resulted from brain hemorrage or occlusive diseases of the vessels. The original text demonstrates that Wepfer's medical thinking is deeply rooted in pre-modern concepts, e.g. Galenic theories of brain function and iatrochemical concepts of disease. He published a classic treatise on strokes, titled Historiae apoplecticorum.

Antonio Egas Moniz (1927), Portuguese neurologist and the developer of cerebral angiography used X-ray technology to produce the first image of the cerebral arteries in a living person.

Désiré Collen (1970), a Belgian molecular biologist marked the starting point for the development of a medicinal treatment for ischemic stroke. In his search for a way to dissolve blood clots, Collen developed a substance known as alteplase recombinant tissue plasminogen activator (rt-PA), which could dismantle the underlying framework of the clot (or “thrombus”) and therefore break it down. Since the mid-1990s, this drug has been used for the acute treatment of stroke in lysis therapy. He published 172 review articles and 39 issued US patents and has more than 70,000 citations.

5. Gallery - Art of the month

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