Treatment of Vestibulocochlear Nerve Hyperactivity with Carbamazepine

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Case Report

A 63-year old female presented with a chief complaint of intermittent vertigo proceeded by a unilateral high pitched right sided tinnitus. Her symptoms began six years earlier, as a crackling isolated to the right ear, like “firecrackers at a distance”, followed immediately by a left beating nystagmus and a sensation of vertigo. The episodes last on average 15-20 seconds with spontaneous resolution. The attacks have gradually increased in frequency, and in the past year they recurred in episodic bouts of two minute intervals. The spells are secondarily elicited with loud noise and with Valsalva maneuvers. Over this time frame there was no associated subjective hearing loss or aural fullness.

Exam was unremarkable. Audiogram demonstrated normal hearing bilaterally. However, acoustic reflexes could not be obtained due to inability to obtain hermetic seals. Additional studies were performed below.

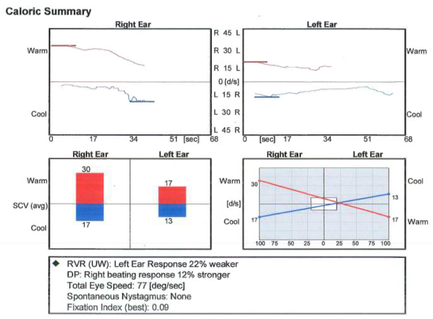

Figure 1- Patient’s caloric response. Note the relative weakness in the left ear (22%) despite the right sided symptoms.

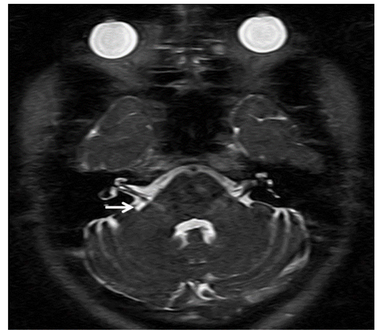

Figure 2- Note the loop of the anterior inferior cerebellar artery (AICA) (arrow) sitting at the porous acousticus in contact with the cochleovestibular nerve. There was bilateral compression of CN VIII on FIESTA MRI.

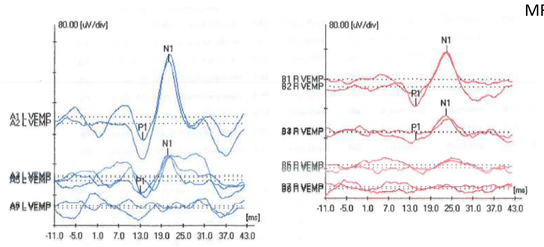

Figure 3- Bilateral vestibular evoked myogenic potential (VEMP) responses with thresholds at 85dB on the left and 75dB on the right. Normal P1 and N1 latencies bilaterally. However, there is a decreased amplitude response of 30.59% on the right compared to the left.

On assessment, vestibular testing elicited only a left sided caloric weakness that was within the accepted normal range (Fig 1). Rotational chair testing corroborated the unilateral peripheral weakness. There was no evidence of conductive or sensorineural hearing loss with type A tympanograms bilaterally. Slight VEMP amplitude asymmetry, with no evidence of diminished thresholds (Fig 3). FIESTA MRI revealed bilateral compression CN VIII bilaterally at the porous acousticus (Fig 2).

Case Resolution

The unilateral peripheral weakness on the left side from the VNG confounded the case. However, because the patient’s symptoms were right-sided, additional investigation with a VEMP and MRI further clarified the patient’s symptoms. The history of unilateral staccato tinnitus with vestibular involvement, evidence of AICA and CNVIII contact, and the paucity of congruent findings on physical exam or audiologic/vestibular assessment was consistent with many cases of CN VIII NVC described in the literature. Our patient presented with a mixed diagnostic picture with symptoms of a hyperactive right vestibulocochlear nerve with possible early development of peripheral vestibular weakness on the left secondary to bilateral CN VIII neurovascular compression. An initial trial of carbamazepine 50 mg daily provided partial relief of symptoms. An increase to 50 mg twice daily led to complete symptom resolution with no associated side effects.

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Cochleovestibular Neurovascular Compression

- Defined as compression of cranial nerve VIII anywhere from the cerbellopontine angle to within the internal auditory canal by aberrant vasculature. This source of ectopic excitation yields hyperactive stimulation in the absence of efferent input[1].
- First noted as a contributory etiology of cochleovestibular dysfunction in 1975 by Jannetta et al., and may represent up to 4% of all clinical vestibular complaints in adults and children[2,3,4].
- Longstanding precedent in the otolaryngology literature of utilizing microvascular decompression for CN VIII neurovascular compression (NVC).

Diagnostic Criteria

The continued controversy over vascular compression as a disease entity stems from an un-elucidated pathophysiology and a lack of consistent clinical findings to foster wide spread acceptance of diagnostic guidelines.

Unique Clinical Presentations of Vestibulocochlear Hyperactivity Secondary to Compression:
Patients typically present in middle age, with increasing frequency of spells with occasional neighboring ipsilateral cranial nerve neurovascular compression[5,6].

Paroxysmal Vertigo[5]
Spontaneous recurrent attacks of vertigo lasting seconds to minutes
- 69%- rotatory vertigo / 25%- to-and-fro sensation
- 50%- unprovoked / 22%- regularly provoked
  - Accompanying symptoms
    - 75%- unsteady gait
    - 41%- nausea / vomiting
    - 28%- unilateral tinnitus

Typewriter Tinnitus[6,7]
Intermittent paroxysmal spells of unilateral tinnitus lasting only seconds
- Accompanying symptoms
  - Hearing loss at the tinnitus frequency
  - Associated ipsilateral compressive symptoms of adjacent cranial nerves

Clinical Workup:
- Audiometric Workup
  - Stratifying for time and severity of symptoms, SNHL correlates with a longer duration and greater burden of disease[8].
  - Hearing loss often reported as high frequency[9].
- Vestibular Studies
  - Variable statistically significant abnormal vestibular deficiencies across the literature (caloric deficiency[10,11], reduced VEMP response[7]).
  - Independent of abnormal finding, noted worsening of ENG exam with time[5].
- Auditory Brainstem Response
  - Temporally progressive lengthening of inter-peak latencies ipsilateral to the compressive etiology with contralateral compensation, paralleling symptom progression[13].
- Imaging
  - Contact of CN VIII and AICA is a normal anatomic variant in up to 34% of population[14].
  - Patients with pulsatile or intermittent tinnitus are nearly 80 times more likely to have a vascular loop in contact with CN VIII than patients with non-pulsatile tinnitus[14].
  - Patients with a unilateral sensorineural hearing loss have an increased odds ratio of nearly 2 (95% CI 1.5-2.63) of having an ipsilateral vascular compression[14].

Treatment Options

Microvascular Decompression[15]

Symptoms	Subjective Symptom Resolution (meta-analysis of 19 studies)
Tinnitus	27.8-100%
Vertigo	75-100%

Carbamazepine Therapy

Presenting Syndrome	Subjective Symptom Resolution

Paroxysmal Vertigo[5]	• Decreased the attack *frequency* by 90%,
	• Decreased attack duration by 79%
	• Decreased attack *intensity* by 69%
	  • Carbamazepine average dose requirement is 568 ± 200.9 mg/day
	  • Oxcarbazepine average dose requirement is 870 ± 429.5 mg/day

Typewriter Tinnitus[6,16,17,18]	Five case series (13 patients) with 100% initial response rate • One patient reported resistant to maximal medical therapy with long term therapy • Tinnitus secondary to NVC, as previously published and including our case study, requires low dose carbamazepine with an average maintenance dose of 353.8 ± 194.1 mg

Discussion

- Diagnosis of CN VIII neurovascular compression as the etiology of unilateral symptoms remains largely based on clinical history, vestibular testing and imaging, while ruling out competing pathologies.
- Carbamazepine therapy represents an efficacious and safe alternative intervention for non surgical candidates.