Nervus Intermedius Neuralgia With Vestibular Paroxysmia -A Rare Combination of Nerve Compression Syndromes

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ABSTRACT

Introduction: Posterior cranial fossa nerve compressions in a rare combination are discussed here.

Case Reports: 51 year old male with intractable vertigo, vomiting and left ear ache, had left spontaneous nystagmus. Another 25 year old female had recurrent ear ache and vertigo. Both MRI Brain showed compression of VII/VIII nerve complex in the cistern. They were given Ox carbamazepine with supportive therapy tapered with no recurrence in past 1 year.

Discussion: The combination of the 7th and 8th cranial neuralgias presents with common symptomatology. The investigation of choice is HR T2 weighted MRI brain (CISS/FIESTA SPACE sequence), treated medically. Surgically, decompression or nerve sectioning are done.

Conclusion: Though rare due to different positions of the nerve entry/exit zones of these nerves, this combination of posterior cranial fossa nerve compression syndromes can coexist. Diagnosis can help in the proper management of such patients and even open the horizon for surgical options in recalcitrant cases.

Keywords: Vertigo, Neuralgia, Micro-Vascular, Carbamazepine

*See End Note for complete author details

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INTRODUCTION

Two of the common ENT symptomatology are vertigo and otalgia which represent the clinical presentation of a vast range of disorders. Here we report 2 cases who presented with similar complaints but was confirmed a rare diagnosis of a combination of posterior cranial fossa micro vascular compression syndromes.[1] Nervus intermedius neuralgia is one of the rare causes of facial neuralgias with only around 174 cases reported world-wide from 1932 to 2018.[2] This neuralgia with vestibular paroxysmia is even rarer compared to combinations with other cranial nerves like the trigeminal or facial motor nerve. The investigation of choice is high resolution T2 weighted MRI brain (CISS/FIESTA SPACE sequence) with 3D reconstruction.[3] It can be treated with anti-convulsant and anti-vertigo drugs.[2] Surgical micro vascular decompression or nerve sectioning may be required in recalcitrant cases.

CASE REPORT I

A 51 year old male presented with sudden spontaneous onset of intractable rotatory vertigo lasting few minutes recurring intermittently with more than 10 episodes that day, aggravated with head movements and associated with vomiting, tinnitus and paroxysms of deep seated sharp, shooting type left ear ache lasting few seconds in our casualty. No history of ear block, hearing deficits,

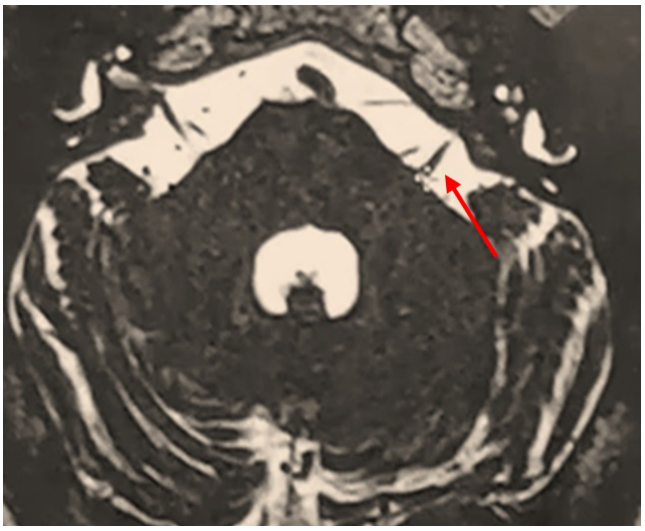

Figure 1. 3T HR Axial MRI-T2W 3D Sequence showing Vascular Looping in the 7th/8th Nerve Complex on Left.

ear discharge, head ache, facial deviation or limb weakness.

On examination he showed left sided horizontal nystagmus with positive head impulse test, Rhomberg's test and Unterberger test rotating to left side. Cerebellar signs were negative. The local ENT examination including palpation of styloid process and audiological evaluation was normal. His tinnitus was matched to 60 kHz at 1 kHz in left ear. MRI Brain showed prominent vessels arching over the cisternal segment of VII/VIII nerve complex on the left **(Figure 1).**

He had dramatic improvement with Ox carbazepine 300 mg daily along with other supportive IV medications including chlorproperazine and was discharged within 2 days on medications with no such further episodes. He was tapered on ox carbazepine dosage from 300 mg daily to 150 mg during this period and is off medications for past 2 months in this 1 year period.

CASE REPORT II

25 year old female presented to our OPD with bilateral deep seated stabbing type intermittent ear ache since 1 year lasing few seconds with some days having more than 10 such episodes to none on the other days. However these episodes affected her quality of life. It was also associated with mild imbalance during the episodes. No other ear symptoms, head ache noted. Her clinical examination was normal, but considering the intractable shooting nature of the pain, after ruling out ear, dental, TMJ pathologies and stylalgia (CBCT of styloid process was normal in length and angulation), an MRI Brain CISS sequence was done which showed compression of the VII/VIII nerve complex in the cistern bilaterally **(Figure 2,3,4).** She was treated with ox carbazepine 300 mg daily later tapered to 150 mg daily in the next 3 months and now in a stable dose without any remissions of pain or imbalance for past 1 year.

DISCUSSION

Neuro-vascular compression syndromes are diseases caused by abnormal contact between a vessel/vessel loops with the root entry/exit zone of a cranial nerve causing mechanical irritation of the nerve.[4] In the order of decreasing incidence, they are trigeminal neuralgia (5th), hemi facial spasm-HFS (7th), glossopharyngeal neuralgia (9th), their combinations like TN+HFS > HFS+GPN > TN+GPN. More rare ones are hemi laryngo-pharyngeal spasm-HeLPS (10th), nervus intermedius neuralgia (7th-sensory) and vestibular paroxysmia (8th).

Nervus intermedius neuralgia also known as geniculate ganglionitis is a very rare disease with only <174 cases reported between 1932 and 2018.[1] Some secondary causes of NIN include Herpes zoster (Ramsay Hunt syndrome), TMJ dysfunction, nasopharyngeal carcinoma, cerebello-pontine angle tumours, petrous bone osteoma, multiple sclerosis, neuro- borreliosis. The clinical diagnostic criteria of NIN[5] according to the ICHD-3rd edition include: (a) at least 3 attacks of u/l otalgia, (b) pain in auditory canal sometimes radiating to the parieto–occipital region(c) characteristic of pain –at least 3 out of the 4 needed: recurring in paroxysmal attacks lasting few seconds to minutes, severe intensity, shooting, stabbing, sharp in quality and precipitated by stimulation of a trigger in the posterior wall of EAM and/ or peri auricular region(d) no clinically evident neurological deficit (e) not better accounted for by another ICHD-3 diagnosis.

Vestibular paroxysmia or earlier known as disabling positional vertigo (DPV)[6] is diagnosed with Classification Committee of the Barany Society Diagnostic criteria (2016)[7] as definite VP when there is (a) at least 10 attacks of spontaneous spinning/non spinning vertigo (b) duration < 1 minute (c) stereotyped phenomenology in a particular patient (d) response to treatment with carbamazepine/ oxcarbazepine (e) not

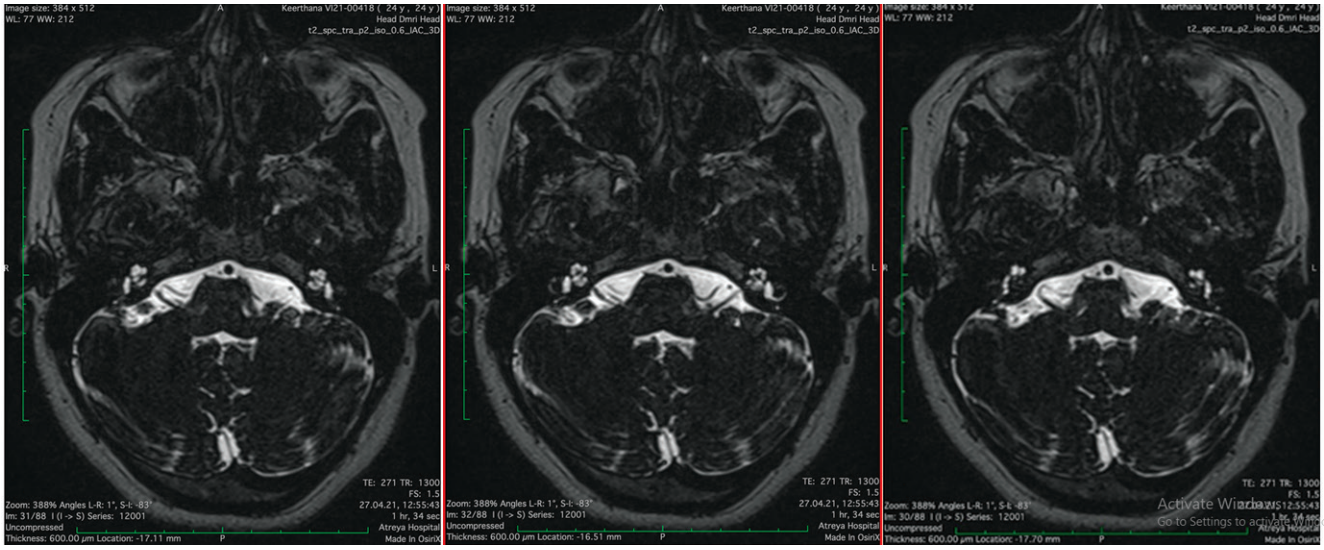

Figure 2,3,4. 3T HR Axial MRI 3D CISS Sequence-Subsequent 3 Sections Showing Multiple Vascular Looping in the Bilateral 7th/8th Nerve Complex more on the Right Side

better accounted for by another diagnosis. Diagnosis of probable VP include (a) at least 5 attacks (b) <5 minutes (c) spontaneous / provoked by head movements (d) stereotyped phenomenology in a particular patient (e) not better accounted for by another diagnosis.

Pathogenesis of NVCS is due to the constant pressure by a vascular segment or loop, mostly a branch of AICA/ PICA on the 7th/ 8th cranial nerve complex causing de-myelination of these nerves leading to ectopic generation of nerve impulses. These leads to ephaptic discharges and transmission of impulses along the compressed nerve giving rise to sensory/ motor effects.[8] The prominent vessels arching over the cistern segments of the 7th and 8th cranial nerves may be preferentially compressing the nervus intermedius fibres along with the vestibular component of the vestibulo-cochlear nerve giving rise to this symptomatology. Type 1 compression involves a non looping vessel passing superior to the nerve complex. Type 2 includes a loop of vessel passing just outside the internal auditory meatus compressing it. Type 3 refers to a vessel loop extending into the IAC compressing the nerve complex.[9]

The site of pathology is the root entry/exit zone of the nerve also called transitional zone/ Obersteiner-Redlich zone[1] which is closer to the brain stem in case of nervus intermedius while the 8th nerve has a long and distant transitional zone. This explains the rarity of this combination and also explains why nervus intermedius is less involved in NVCS compared to other posterior cranial fossa nerves. However the variability and tortuousness in the course of AICA observed by anatomists makes any combination of NVCS possible.

The investigation of choice is High resolution 3D T2W MRI BRAIN-CISS/FIESTA sequence with 3D TOF angiography or a 3D T1W Gadolinium enhanced MRI with multi planar oblique reconstruction of above sequences in combination.[9] Recent imaging modalities like DTI-Diffusion tensor imaging with tractography can pin point the site and extend of nerve compression due to the vessel segment. Potential bio markers like COMT enzyme has been found useful in case of trigemenial neuralgia.[3]

Treatment includes medical anti-epileptic drugs with carbamazepine as the 1st choice.[10] A dose of 150 mg to 300 mg daily is recommended for the membrane stabilizing effect along with reducing the ectopic discharges along the nerve. Others like ox carbazepine, gabapentin, baclofen, amitriptyline have also been tried.[11] Symptomatic management with anti-vertigo drugs like prochlorperazine maleate 5-10 mg BD/ SOS, cinnarazine (20-75 mg BD) with dimenhydrinate (40-50 mg BD) are given based on the level of difficulty as in our case 1 who presented with acute onset severe intractable vertigo controlled with IV drugs. In our case study, both patients were weaned off medications in 3-6 months with a symptom free period of 8-12 weeks due to the membrane stabilising effects.

In recalcitrant cases, surgical management is advised. Sub occipital retro sigmoid craniotomy approach with or without removal of posterior wall of IAC is carried out based on extend of nerve–vessel segment involved. Micro vascular decompression is then carried out with a Teflon felt padding kept in between the vessel loop and the nerve segment to reduce the compression on the nerve. In rare cases, neurotomy is carried out.

CONCLUSION

NVCS of the nervus intermedius and vestibular nerve though rare, has a very common clinical presentation. A high degree of suspicion is necessary to think of rare causes of vertigo and otalgia when the symptomatology cannot be attributed to any one common cause alone. MRI brain can always hide surprises in the most common clinical presentations. A team of neuro otologists can well diagnose and treat such cases. Surgical management is reserved for recalcitrant cases with majority managed on anti-epileptics and other symptomatic drugs with long symptom free intervals.

END NOTE

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REFERENCES

1. Baldauf J, Rosenstengel C, Schroeder HWS. Nerve Compression Syndromes in the Posterior Cranial Fossa. Dtsch Arztebl Int. 2019 Jan 25;116(4):54–60.
2. Figueiredo R, Vazquez-Delgado E, Okeson JP, Gay-Escoda C. Nervus intermedius neuralgia: a case report. Cranio. 2007 Jul;25(3):213–7.
3. Donahue JH, Ornan DA, Mukherjee S. Imaging of Vascular Compression Syndromes. Radiol Clin North Am. 2017 Jan;55(1):123–38.
4. Langner S, Schroeder HW, Hosten N, Kirsch M. [Diagnosing neurovascular compression syndromes]. Rofo. 2012 Mar;184(3):220–8. [Pubmed] https://pubmed.ncbi.nlm.nih.gov/22179769/
5. Robblee J. A pain in the ear: Two case reports of nervus intermedius neuralgia and narrative review. Headache. 2021 Mar;61(3):414–21.
6. Brandt T, Dieterich M. VIIIth nerve vascular compression syndrome: vestibular paroxysmia. Baillieres Clin Neurol. 1994 Nov;3(3):565–75.
7. Strupp M, Lopez-Escamez JA, Kim JS, Straumann D, Jen JC, Carey J, et al. Vestibular paroxysmia: Diagnostic criteria. J Vestib Res. 2016;26(5–6):409–15.
8. Brandt T, Strupp M, Dieterich M. Vestibular paroxysmia: a treatable neurovascular cross-compression syndrome. J Neurol. 2016 Apr;263 Suppl 1:S90-96.
9. Haller S, Etienne L, Kövari E, Varoquaux AD, Urbach H, Becker M. Imaging of Neurovascular Compression Syndromes: Trigeminal Neuralgia, Hemifacial Spasm, Vestibular Paroxysmia, and Glossopharyngeal Neuralgia. AJNR Am J Neuroradiol. 2016 Aug;37(8):1384–92.
10. Holste KG, Hardaway FA, Raslan AM, Burchiel KJ. Pain-free and pain-controlled survival after sectioning the nervus intermedius in nervus intermedius neuralgia: a single-institution review. J Neurosurg. 2018 Aug 17;131(2):352–9.