Semicircular Canal Dehiscence from a High Riding Jugular Bulb Diverticulum Causing Tumarkin Crisis: A Case Report

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Abstract

Objectives: To describe the clinical features and audiovestibular testing of Tumarkin crisis secondary to posterior semicircular canal dehiscence from high riding jugular bulb diverticulum.

Study Design: Case report and literature review

Methods: Retrospective review of a single case of Tumarkin crisis presenting to Neurotology Clinic at UCLA Medical Center, Los Angeles, CA. Quantitative audiologic and vestibular function testing, relevant imaging, neurologic history, and examination were performed.

Results: We present a 78 year old female with a 10 year history of Tumarkin crisis (sudden drop-attacks) lateralizing to the right. Vestibular Evoked Myogenic Potential showed mild weakness on the right, while audiogram demonstrated an asymmetric right-sided mixed hearing loss. Delayed intravenous contrast-enhanced 3D FLAIR MRI showed no endolymphatic hydrops; however, CT scan revealed a high riding jugular bulb diverticulum (JBD) on the right side with osseous dehiscence of the posterior semicircular canal. To our knowledge this is the first reported case of non-Meniere's Tumarkin crisis secondary to high riding JBD.

Conclusion: High riding JBD should be considered in patients with Tumarkin crisis that do not meet diagnostic criteria for Meniere's disease.

Introduction

In 1936, Tumarkin described patients with Meniere's disease that experienced sudden falls and violent vertigo without loss of consciousness or neurologic symptoms.[1] Tumarkin crisis have since been hypothesized to result from mechanical stimulation to the otoliths, producing a burst of neural signaling from the vestibular system to the vestibulospinal pathways. Tumarkin crisis have been demonstrated in patients with and without Meniere's disease.

The apex of the jugular bulb (JB) lies below the tympanic cavity and is described as "high-riding" (rising above the lower margin of the round window) in up to 24% of cases and, more rarely, may form a diverticulum (JBD).[2] A high riding JBD can cause semicircular canal (SC) dehiscence which, in turn, may produce conductive hearing loss, autophony, chronic imbalance, and pressure or noise induced vertigo or nystagmus. Due to the significant variability of presenting symptoms, canal dehiscence has been called "a great otologic mimicker" and represents a challenging diagnosis.[3]

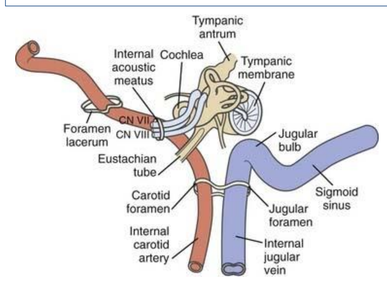

Figure 1. Diagram displaying the close association of the vestibular system and the jugular bulb.

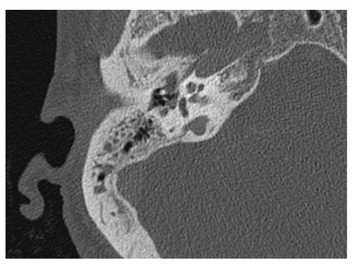

Figure 2. Axial CT of right temporal bone demonstrating a high riding jugular bulb bordering the vestibular aqueduct

Case Report

A 78 year old woman with a 10 year history of Tumarkin crisis presented to our institution for assessment. She first experienced slight positional vertigo while doing her hair. These spells increased in frequency and, in the 6 months prior to her presentation, she had experienced four falls. First: she was shopping and fell in the hallway, breaking her shoulder. Second: she was seated in her endocrinologist's office, started walking to the water cooler, then fell and hit her head on the counter. Third: she fell while walking up concrete stairs. Fourth: at a thanksgiving party, she stood from a seated position and fell. All falls lateralized towards the right. She felt as though she was being pushed over during the spell, but denied loss of consciousness or feeling abnormal prior to it. She denied worsening of the symptoms with Valsalva or bending over. She had progressive, episodic tinnitus and progressively worse hearing in her right ear. Prior treatments included a PE tube, without improvement of symptoms. Medical history was significant for childhood migraines (resolved with botox therapy), 15 pack-year smoking history (ceased), Ehlers-Danlos syndrome (III), diabetes with peripheral neuropathy, hypothyroidism, and hyperlipidemia.

Vestibular evoked myogenic potentials (VEMPs) showed decreased response on the right (0.5) and normal response on the left (0.7). Audiogram revealed an asymmetric right-sided mixed hearing loss. Electronystagmography, caloric testing, and quantitative rotational testing were all within normal limits. Delayed intravenous contrast-enhanced 3D FLAIR MRI showed no endolymphatic hydrops. CT scan revealed a high riding JBD on the right side with a foci of osseous dehiscence into the middle ear cavity and posterior SC (Figure 4). The diverticulum extends superiorly to the vestibular aqueduct, laterally to the mastoid facial canal, and superolaterally to the inferior posterior SC.

The patient was scheduled for right vestibular neurectomy. Post-operative care will include physical therapy to improve equilibrium.

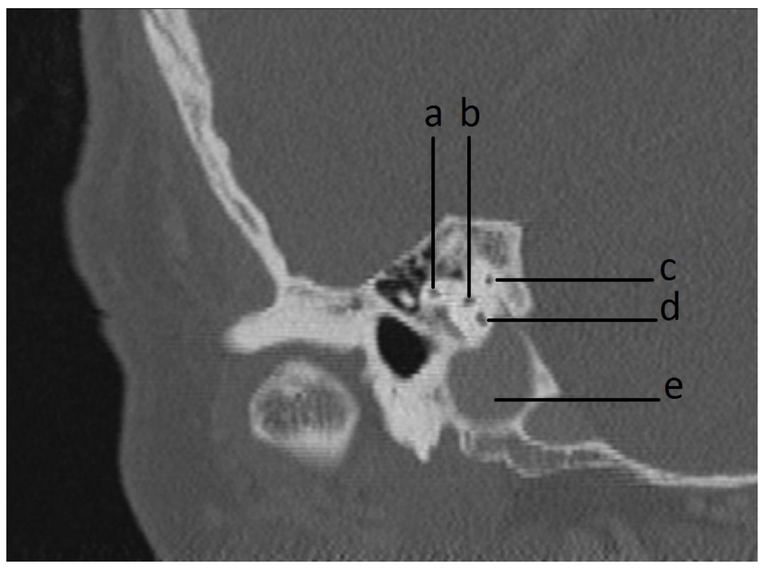

Figure 3. Oblique CT, right temporal bone. (a,b) lateral and medial aspect of lateral SC, respectively, (c,d) superior and inferior aspects of the posterior SC, respectively, (e) jugular bulb

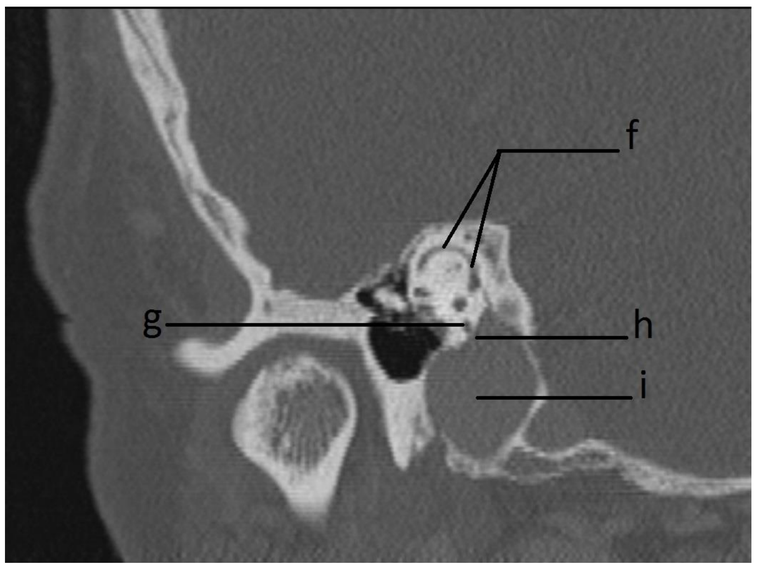

Figure 4. Oblique CT, right temporal bone. (f) superior SC, (g) inferior aspect of the posterior SC, (h) osseous dehiscence of the jugular bulb into the inferior aspect of the posterior SC, (i) jugular bulb

Discussion

In this case report, we demonstrate how anatomic abnormalities of the jugular bulb can cause SC dehiscence and provoke vestibular disturbances. Though the correlation has been well demonstrated,[3-5] the exact pathophysiology of the disease process is still unknown. Hypothesis include venous hypertension and turbulent venous flow.[4] It would be reasonable to consider that vascular disease can predispose towards the development of jugular bulb abnormalities. In the patient presented, multiple vasculopathologic factors including age, smoking history, diabetes, hypothyroidism, Ehlers-Danlos syndrome, and hyperlipidemia, may have contributed to her condition.

If symptoms are disruptive and disabling to a patient with JBD, surgical management may be pursued. Ligation or embolization of the jugular vein, surgical lowering of the jugular bulb through a transmastoid approach, resurfacing of dehiscence, SC plugging, and endovascular obliteration of diverticuli with coils have been reported with varying degrees of success. Our planned approach is vestibular neurectomy to definitively disrupt the pathological neural signaling from the vestibular periphery that is provoking her attacks.

Conclusions

High riding JBD should be considered in patients with Tumarkin crisis that do not meet diagnostic criteria for Meniere's disease. CT, Valsalva maneuvers, VEMP and auditory testing are useful diagnostic modalities for symptomatic JBD. Though the pathophysiology of JBD is not definitively known, the mechanical nature of this particular etiology of Tumarkin crisis allows for management with a variety of surgical techniques.

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