Clinical Reasoning: An Elderly Woman With a Jugular Bulb Anomaly and Acute Headache

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Section 1

History

A 68-year-old woman with a history of type II diabetes mellitus, hypertension, coronary artery disease (s/p coronary angioplasty 2 years before), and right hemiplegia secondary to left internal capsule infarct 1 year before developed central chest pain while on an evening walk. She took a tablet of glyceryl trinitrate (GTN) sublingually, which relieved her chest pain instantly. However, she developed an acute severe throbbing headache, which started behind her left ear and abruptly spread to her entire head. Within a few minutes, the headache peaked in intensity. The headache was associated with tinnitus in her left ear and a vertiginous sensation. She had no vomiting, conjunctival injection, lacrimation, photophobia, or phonophobia. Her headache did not improve with oral analgesics, and she had no history of migraine or other headache types. She reported episodic tinnitus and fullness in the left ear for 1 year. She was taking metoprolol, aspirin, atorvastatin, and telmisartan tablets. Before this episode, she had never taken sublingual GTN that was prescribed for chest pain. On examination, her pulse rate was 100 beats per minute and regular with palpable peripheral pulses. Her blood pressure was 140/90 mm Hg. She had a mild residual right facial palsy. The remainder of her clinical examination was normal.

Questions for Consideration:

1. What is the differential diagnosis of her neurologic symptoms?
2. What investigations will you consider in this patient?

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Section 2

Our patient complained of headache that peaked in severity within a few minutes. The International Classification of Headache Disorders 3rd edition (ICHD-3) defines a headache that peaks in intensity within a minute as thunderclap headache. However, because we could not verify the precise time course of her symptoms and this was her first lifetime episode, we considered her headache to be a thunderclap-like headache to look for ominous causes, including subarachnoid hemorrhage, reversible cerebral vasoconstriction syndrome (RCVS), pituitary apoplexy, cortical venous sinus thrombosis, posterior reversible encephalopathy syndrome, and unruptured aneurysms. With no underlying cause, thunderclap headache may be an idiopathic benign recurrent headache.[1]

Her headache precipitated after taking sublingual GTN. ICHD-3 classifies GTN-related headache as "nitric oxide donor-related headache."[2] The immediate GTN-related headache is a frontotemporal headache that starts within 1 hour of absorption of GTN and dissipates within 1 hour after the release of nitric oxide has ended. Posterior circulation stroke may also produce headache, vertigo, and tinnitus. However, such patients will usually have neurologic deficits. In addition, an acute attack of Meniere disease may produce acute vertigo and tinnitus, but symptoms do not include sudden headache.

A cranial CT scan did not show any evidence of subarachnoid hemorrhage. Lumbar puncture revealed normal CSF examination. MRI of the brain was normal. MR angiography revealed a normal circle of Willis without any evidence of aneurysm or RCVS. However, postcontrast MR venography showed a dilated left jugular bulb with normal dural sinus and deep cerebral veins.

Questions for Consideration:

1. What is the radiologic diagnosis of this patient?
2. How will you confirm the diagnosis?

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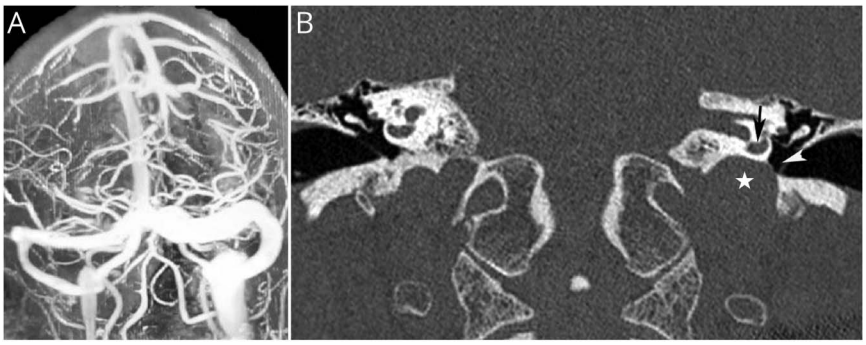
Section 3

The dilated left jugular bulb suggests a diagnosis of jugular bulb anomaly (JBA). JBA can be either a high-riding jugular bulb (HRJB) or a jugular bulb diverticulum.[3] HRJB is considered when the jugular bulb reaches the level of the internal acoustic meatus, whereas jugular bulb diverticulum is an outpouching arising from the dilated jugular bulb. JBA is an aneurysmal dilatation of the jugular bulb that may remain asymptomatic or present with symptoms secondary to pressure/dehiscence into the surrounding structures, including the vestibular aqueduct, semicircular canal, and the facial nerve. Our patient had a history of episodic tinnitus and fullness in the left ear. This time, she presented with headache accompanied by acute pulsatile tinnitus in the left ear and vertigo. To confirm the diagnosis, a high-resolution CT (HRCT) scan of the temporal bone was performed because CT is more sensitive to the diagnosis of JBA and its relationship with the structures of inner ear. HRCT of the temporal bone showed a HRJB with dehiscence into the middle ear (Figure).

Questions for Consideration:

1. What is the relationship between JBA and headache?
2. How would you treat this patient?

Figure Left-Sided High-Riding Jugular Bulb



(A) Magnetic resonance venography showed a dilated left jugular bulb with prominent left transverse sinus and internal jugular vein. (B) CT of the temporal bones also showed a dilated left jugular bulb (white star) in close proximity to the inferior turn of cochlea (black arrow) that dehisced into the left middle ear (white arrowhead).

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Section 4

JBA can produce migraine-like headaches and intractable headaches.[4,5] Our patient had headache immediately after the use of sublingual GTN. GTN produces acute, bilateral, frontoparietal severe throbbing headache due to dilatation of blood vessels. The initiation of headache from left posterior aural location and associated left-sided tinnitus and vertigo suggested an acute dilatation of the left HRJB due to GTN. The long-standing left ear tinnitus is also consistent with left HRJB. GTN produces both arterial and venular vasodilatation by the nitric oxide-cyclic guanosine monophosphate pathway. Sublingual GTN-related headache is transient because intracranial arteries are rich in sympathetic vasomotor fibers that promptly normalize after GTN-related vasodilatation. Unlike arteries, veins have poorly innervated tunica media.[6] The abnormally dilated veins such as HRJB will have hypertrophy of the intimal layer and thinning of the elastic lamina and smooth muscle layer.[7] Therefore, sublingual GTN may have a prolonged dilatation effect on HRJB producing an acute-onset but prolonged headache that explains the longer headache duration in our patient.

After her headache resolved over 24 hours, she remained asymptomatic except for episodic left ear tinnitus. She was offered a surgical treatment of HRJB, but she chose to continue with conservative treatment.

Discussion

Asymptomatic anatomic variations of the jugular bulb are not uncommon. JBA is usually discovered incidentally and can be seen on up to 8% of all CT scans.[3] Anomalies include jugular bulb diverticula, high-riding jugular bulb, or an unusual lateral location of the jugular bulb. JBA usually remains clinically silent in most patients, although some people may complain of aural fullness, unilateral hearing loss, tinnitus, and vertigo.[3-5] Otalgia, migraine-like or thunderclap headaches, and cranial nerve involvement are rare manifestations of JBA.

ICHD-3 does not define diagnostic criteria for headaches attributable to HRJB. However, the headache characteristics and associated features in our patient shared similar characteristics to headaches attributable to unruptured saccular aneurysm as defined by ICHD-3.[2] Headaches in unruptured aneurysms occur because of sudden dilatation of the aneurysm. The ICHD-3 diagnostic features of unruptured aneurysms include radiologic evidence of aneurysm, sudden or thunderclap headache, headache occurring in temporal relation to other symptoms of aneurysm (related to pressure effect of aneurysm, like 3rd nerve palsy in posterior communicating artery aneurysm), and relief from headache after aneurysm treatment.[2,8] As seen with unruptured aneurysms, acute dilatation of HRJB by GTN possibly produced the sudden headache in our patient. Her ipsilateral headache was associated with accompanying symptoms secondary to pressure on the surrounding structures in the form of acute left-sided tinnitus and vertigo, and her headache improved spontaneously after the effect of GTN was over.

GTN-related headache phenotypes can range from migraine without aura to episodic tension-type headaches in a patient with known chronic tension headaches, or they can produce a new cluster episode in a patient with cluster headache.[2] The clinical features related to headache in our patient are difficult to explain by GTN alone. GTN produces bilateral frontotemporal pulsatile headache. In our patient, headaches originated from the left retroaural area and became holocranial. The accompanying left-sided tinnitus and vertigo with headache are not seen with GTN headache, which typically subsides after the release of nitric oxide. Thus, GTN does not fully explain the headache that lasted more than 24 hours but may have unmasked the quasi-symptomatic HRJB in our patient. The cause-to-effect relationship remains correlative.

HRJB can erode superiorly into the vestibular aqueduct, posterior semicircular canal, and middle ear and posteriorly into the facial nerve. Most commonly, it erodes into the vestibular aqueduct and produces tinnitus, dizziness, hearing loss, and rarely endolymphatic hydrops. It can also remain clinically silent. The second most commonly involved structure is the facial nerve, which can produce recurrent facial palsy or hemifacial spasm. Dehiscence into the posterior semicircular or internal auditory canal is less common but frequently symptomatic, leading to sensorineural hearing loss. Middle ear dehiscence can produce tinnitus and conductive hearing loss.[9]

A recent study[10] classified these anomalies based on their relationship with posterior semicircular canal, internal auditory canal, and presence or absence of dehiscence. In our patient, the jugular bulb was situated below the inferior margin of the posterior semicircular canal and dehisced into the middle ear, and therefore, it can be categorized as a type 2b.

HRJB with persistent aural symptoms or recurrent facial palsy may be treated by endovascular approach using an occluding stent device or by surgical repositioning of the HRJB.

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