Asymptomatic aberrant carotid artery of the middle ear space

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Introduction

First reported in 1899 at a meeting of the Austrian Otologic Society, an aberrant internal carotid artery (ICA) occurs in less than 1% of the population.[1,2] It is defined as an ICA that courses laterally within temporal bone and passes through the middle ear cavity. Failure to recognize this anomaly during otologic surgery risks iatrogenic hemorrhage and devastating complications.

The clinical presentation is nonspecific. Hearing loss, the most common symptom, is present in about half of patients, but audiometry may or may not show a conductive hearing loss. About one third of patients report pulsatile tinnitus.[2] Otalgia, vertigo, aural fullness and headache appear less commonly. The presentation may be asymptomatic in up to 5% of patients.[2] In a review of 16 cases reported from 1982-2003, 3 cases were identified incidentally on imaging.[3]

In addition to an aberrant ICA, other vascular anomalies of the middle ear include a high riding jugular bulb, persistent stapedial artery, dehiscent carotid artery canal, and dehiscent high jugular bulb. One percent of the population has a dehiscent ICA canal, which creates the risk of an aberrant ICA, the most rare of the aforementioned.[4] A high jugular bulb is present in anywhere from 6-22% of the population, but a dehiscent high jugular bulb occurs in just 1%.[1,2] It is unknown how frequently these anomalies occur concordantly.

We present an unusual case of a concomitant aberrant internal carotid artery and dehiscent high jugular bulb, both identified incidentally on otologic examination. This rare combination elevates the rick of catastrophic bleeding if encountered during surgery.[5]

Case Report

A 15-year-old female was referred to our tertiary care pediatric otolaryngology clinic for evaluation of persistent middle ear fluid incidentally diagnosed by her pediatrician. She denied pain, tinnitus, and hearing loss and had no history of otitis media or otologic surgery.

Otoscopy showed no fluid in the middle ear, but did show a red pulsating retro-tympanic vascular mass located behind the inferior portion of the tympanic membrane (see Figure 1). A pure tone audiogram demonstrated mild conductive hearing loss at 250 Hz rising to within normal limits at 500 Hz and above.

High-resolution computed tomography (CT) of the temporal bones revealed a high riding jugular bulb with a dehiscent sigmoid plate resulting in protrusion of the jugular bulb into the mesotympanum reaching the tympanic membrane (see Figure 2). Computed tomography angiography (CTA) further delineated an aberrant carotid artery coursing through the middle ear, in addition to the high riding jugular bulb (see Figure 3 and 4). Imaging also demonstrated an absent foramen spinosum, but no evidence of a persistent stapedial artery (see Figure 5).

The patient was diagnosed with an aberrant carotid artery with a dehiscent high jugular bulb. She was counseled on the risk of middle ear surgery and external ear canal manipulation. She has been followed closely with serial audiometric and otoscopic examination.

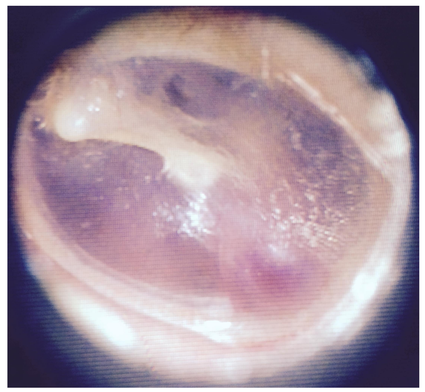

Figure 1. Otoscopic view showing a protruding retro-tympanic vascular mass (arrow) located behind the inferior portion of the tympanic membrane.

Disucssion

The An aberrant ICA is a rare anomaly, occurring in less than 1% of the population. The clinical presentation can include nonspecific symptoms including hearing loss and tinnitus, but it may also present asymptomatically.[6] Its presence on exam can be masked by otitis media with effusion, or, as in our case, it can be misinterpreted as an effusion. In some instances, an aberrant ICA may be identified only during surgery when life-threatening bleeding can occur. Every otolaryngologist should be aware of this anomaly so that it can be identified prior to middle ear surgery, preventing dramatic complications.

The vertical segment of the ICA, in its normal course, ascends to enter the petrous temporal bone medial to the styloid process and anterior to the jugular bulb and inferior tympanic cannaliculus.[7] Proximally along its vertical course, the artery travels within the carotid canal, anterior and inferior to the cochlea.[2] The thin bony plate of carotid canal, separating the ICA from the middle ear cavity, is less than 0.5mm thick.[7] Finally, the ICA turns anteromedially, traversing the foramen lacerum of the sphenoid bone and entering the middle cranial fossa.[2] Here, from the petrous portion of the ICA, the caroticotympanic artery branches off to supply the middle ear. The middle ear also receives blood supply from the inferior tympanic artery, a branch of the ascending pharyngeal artery. The inferior tympanic artery passes through the inferior tympanic canaliculus along with Jacobsens neve and anastomoses with the caroticotympanic artery.

Multiple theories exist explaining the formation of an aberrant ICA. Historically, a persistent stapedial artery (PSA) was believed to be the cause. During embryologic developing, the stapedial artery serves to connect future branches of the external carotid artery (ECA) with the ICA, so persistence of these connections would create an aberrant ICA course.[2] Traction from the PSA, or its fibrous remnant, could pull the ICA into the tympanic cavity.[8] A PSA may be associated with an aberrant ICA in up to 50% of cases, but most studies report a lower incidence.[9] The association was likely present in our case due to the absent foramen spinosum, but the PSA could not be definitively diagnosed on our imaging studies (see Figure 5).

Despite the frequent co-occurrence of an aberrant ICA and PSA, an aberrant ICA is now most commonly thought to develop as a result of aplasia of the vertical ICA segment.[2,3] As a result, the anastomosis of the inferior tympanic artery to the caroticotympanic artery, which connects the ECA system to the ICA system, bypasses the missing ICA segment.[10] In fact, on CTA, the segmental ICA agenesis can be visualized as a stenotic ICA, and the collateral flow can visualized as a an enlarged tympanic canaliculus (see Figure 3).

The clinician should have a low threshold to obtain imaging to confirm a suspicion of an aberrant ICA. CT is generally sufficient, but must be obtained with intravenous contrast. In our case, the initial study without contrast identified the dehiscent jugular plate, but could not distinguish the high jugular bulb from the aberrant carotid artery. An enhancing intratympanic mass is considered one of four essential imaging characteristics on CT.[3] As discussed above, other essential characteristics include absence of the vertical segment of the ICA canal and an enlarged inferior tympanic canaliculus. Absence of bone covering the tympanic portion of the ICA is the final feature.[3] MRA can facilitate diagnosis in some cases. It will reveal a reduced diameter of the tympanic ICA, and, on frontal view, the vertical segment of the ICA is lateral to a line drawn vertically through the vestibule.[5] Conventional angiographagy is considered an invasive procedure, now used only for interventional purposes in the case of bleeding or aneurysm.

The presence of carotid canal dehiscence is significantly correlated with high jugular bulb presentation.[11] While a high jugular bulb may be present in up to 22% of the population, a dehiscent high jugular is only present in 1% of the population.[2] One prior case of an aberrant ICA in combination with a dehiscent jugular bulb is in the literature. The anomaly was identified after the patient presented with hearing loss and pulsatile tinnitus, unlike the asymptomatic presentation in our case.[5] It is unknown how frequently multiple great vessel anomalies occur concordantly and what the relationship between an aberrant ICA and a dehiscent high jugular bulb may be.

If bleeding from an aberrant ICA occurs during middle ear surgery, the surgeon must be prepared to control the hemorrhage to prevent further complications. In a review of 16 cases published from 1982 to 2003, 25% of aberrant ICAs were first identified when massive bleeding ensued.[3] The bleeding can be severe and result in neurologic sequelae including facial nerve paresis, deafness, hemiparesis, aphasia, and intractable vertigo.[2] The middle ear should be carefully packed with Surgicel or Gelfoam, and the nasopharynx may require packing if there is epistaxis. Over packing may damage the ICA and cause neurologic consequences.[2] A neurosurgery consult should be obtained, and an angiogram should be performed to assess the adequacy of cerebral blood flow provided by the contralateral side.[7] If bleeding persists despite conservative management, embolization using balloons or coils may be required.

Treatment of an aberrant ICA is controversial and most authors recommend conservative treatment when there are no bleeding complications.[3-5] Patients are instructed to avoid middle ear surgery and should be followed serially with exams and audiometric evaluation.

Conclusion

An aberrant ICA is a rare anomaly that all otolaryngologists should be familiar with, as middle ear surgery can result in severe bleeding and devastating complications. It may be present with non-specific symptoms including hearing loss and pulsatile tinnitus, or without symptomatology. A retrotympanic mass identified during otoscopy should raise suspicion and be further evaluated with contrasted high-resolution imaging, either CT or MRI. An aberrant ICA in combination with a dehiscent high jugular bulb further increases the risk of bleeding, and, in our experience, is best managed conservatively.

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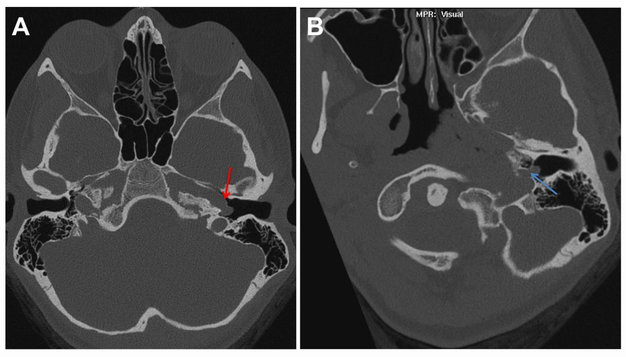

Figure 2. (a) Axial unenhanced CT through level of left external auditory canal demonstrates an abnormal tubular structure running along the medial aspect of the left middle ear, suspicious for aberrant ICA (red arrow). (b) Axial reformatted bone CT demonstrates the jugular bulb and soft tissue density mass within the inferior left middle ear cavity contiguous through the apparent dehiscent jugular plate (blue arrow) making it difficult to differentiate from aberrant ICA without intravenous contrast.

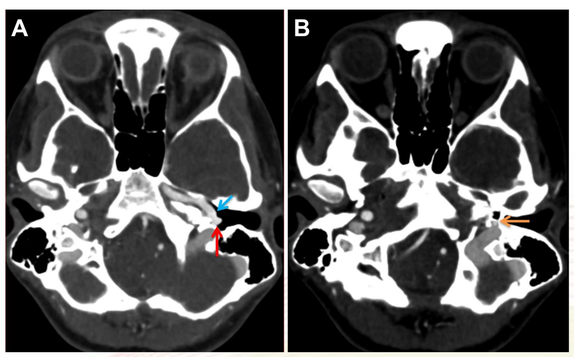

Figure 3. Typical case of aberrant internal carotid artery seen on CTA. Axial CTA image through the middle ear (a) shows the looping aberrant internal carotid in the middle ear (red arrow). Notice it rejoining the normal horizontal segment of the ICA with caliber change (blue arrow) from small proximal to large distal. More inferior axial bone CT image (b) reveals the expected enlarged inferior tympanic canaliculus with stenosed ICA(orange arrow).

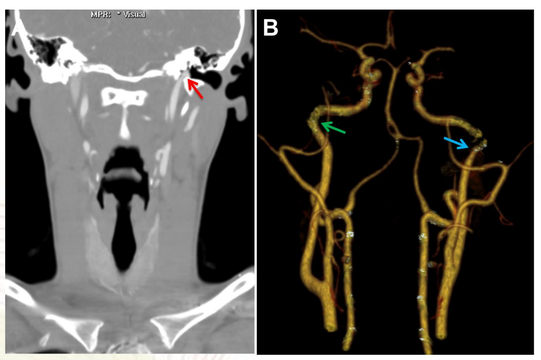

Figure 4. Typical CTA case showing left aberrant ICA. Coronal CTA image (a) through the enlarged inferior tympanic canaliculus (red arrow) reveals the stenosed artery within this structure. CTA reprojection image (b) reveals the left ICA with significant irregularity and stenosis visible (blue arrow). This alone should allow the radiologist to make the diagnosis of left aberrant ICA. The right ICA is normal (green arrow). Frontal CTA reprojection demonstrates the angulated aberrant ICA (blue arrow) compared to the smooth opposite right ICA (green arrow).

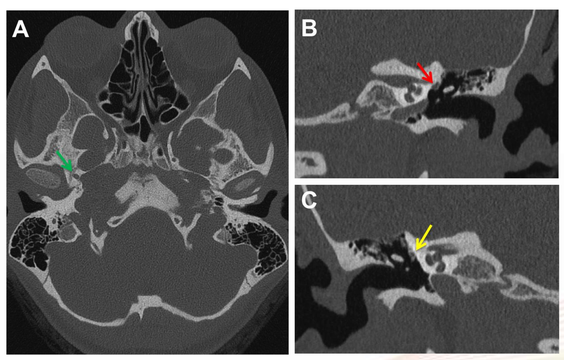

Figure 5. a) Axial CT scan shows a normal right foramen spinosum (green arrow) and absence of the left foramen spinosum. b) Coronal CT scan through left middle ear shows prominent soft tissue at the tympanic segment of the facial nerve likely representing presence of a stapedial artery (red arrow). c) Coronal CT scan through right middle ear shows the tympanic facial nerve is normal in size (yellow arrow). These two findings are diagnostic of an associated persistent stapedial artery. However, there is not enough vascular detail to directly visualize a persistent stapedial artery in this case.