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Aberrant Internal Carotid Artery: An Unusual Anatomic Variant

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Abstract

The aberrant internal carotid artery is a vascular anomaly of the temporal bone and occurs due to the agenesis of the first and second segments of the primitive internal carotid artery, resulting in dominance of a collateral pathway. The internal carotid artery provides the primitive brain its entire blood supply until the posterior circulation forms later in embryonic development. This condition is present in only 1% of the population and produces very subtle imaging findings potentially only discovered during middle ear surgery. During surgery, surgeons may misinterpret it for a glomus tumor or mass, and injury to the site can result in profuse bleeding and even death. Therefore, it is vital for physicians to have knowledge of this disease. Since radiologists are often the ones who diagnose this condition, we present a case report and detailed embryology and review anomalies of the internal carotid artery development.

Keywords: Aberrant, cerebral vasculature, internal carotid artery, radiology, vascular anomaly

Introduction

The aberrant internal carotid artery (ICA) is a vascular anomaly of the temporal bone and occurs due to the agenesis of the first and second segments of the primitive ICA, resulting in dominance of a collateral pathway.[1] The ICA provides the primitive brain its entire blood supply until the posterior circulation forms later in embryonic development.[2] This condition is present in only 1% of the population and produces very subtle imaging findings potentially only discovered during middle ear surgery.[3-6] During surgery, surgeons may misinterpret it for a glomus tumor or mass, and injury to the site can result in profuse bleeding and even death.[4,6] Therefore, it is vital for physicians to have knowledge of this disease. Since radiologists are often the ones who diagnose this condition, we present a case report and detailed embryology and review anomalies of ICA development. Consent from the patient was given to publish her medical information.

Case Presentation

A 37-year-old Hispanic female G9P9 with a past medical history of migraine headaches, hypertension, and an unspecified cardiac murmur presented to the otolaryngology office with a chief complaint of difficulty in hearing for the last 3 months. Three months prior, she noticed a sudden hearing loss particularly in her left ear. She denied any trauma or possible explainable cause of her hearing loss. In addition to decreased hearing, she also noted a ringing sensation in both her ears that has been persistent since onset. Additionally, she noted that she experienced vertigo when all of her symptoms first started but later resolved with right-sided Epley maneuver.

The patient had a past medical history of migraine headaches, hypertension, and a cardiac murmur. She was taking lisinopril 20 mg daily and over-the-counter non-steroidal anti-inflammatory drugs (NSAIDs) as needed for her headaches. The patient had a past surgical history of cholecystectomy and an allergy to penicillin. The patient denies drinking alcohol, current or historic tobacco use, or intravenous drug use. She is sexually active with her husband, and they are mutually monogamous. The patient does not have any family history of chronic medical problems.

Otoscopy revealed normal-appearing ear canals and tympanic membrane bilaterally. Tympanograms were "type A" normal bilaterally, suggesting an appropriate middle and external ear functioning. Audiogram revealed normal hearing in the right ear and a moderate-to-mild mixed hearing loss of the left ear. The speech reception threshold was in good agreement with the 3 frequency pure tone average. Speech discrimination score was termed "excellent" at 30 dB 5L bilaterally. The

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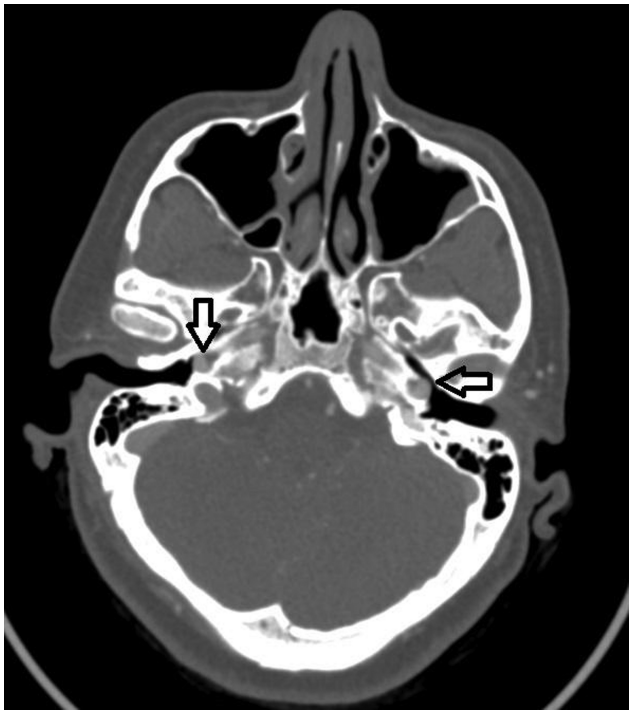

Figure 1. Computed tomography angiography (CTA) of the head demonstrating lateral displacement (arrows) of bilateral internal carotid arteries. There is no bone plate seen separating the internal carotid artery from the middle ear cavities.

patient's physical exam was consistent with otosclerosis but, in combination with vertigo, required further evaluation. A high-resolution computed tomography (CT) scan of the temporal bone was ordered. The result of the CT scan showed lateral displacement of the petrous segments of the bilateral ICAs with no obvious bony septum separating the carotids from the middle ear cavities (Figures 1, 2, 3, 4 and 5).

Discussion

The ICA consists of a cervical segment which starts at the common carotid artery bifurcation and ends at the skull base when the artery enters the petrous temporal bone.[2] The second segment, the short vertical petrous segment, ends at the origin of the carotid–tympanic artery.[2] The horizontal petrous, the third segment, ends at the skull base at the foramen lacerum.[2] Next, the ascending cavernous, the fourth segment, enters the cavernous sinus and ends at the meningohypophyseal trunk.[2] The fifth segment, otherwise known as the horizontal cavernous, starts from the inferolateral trunk in the cavernous sinus. The clinoidal segment, the sixth segment, is distal to the inferolateral trunk and proximal to the ophthalmic artery.[2] Lastly, the supraclinoid segment, the seventh segment, is in between the

Main Points

- Presentation of an aberrant internal carotid artery (ICA) can vary and be as subtle as changes in hearing.
- A high clinical suspicion should be kept in order to avoid biopsy and potential fatal bleeding.
- Management of an aberrant ICA is controversial, but extreme caution should be taken as there is an elevated risk of complications.

Figure 2. Normal CTA demonstrating the normal course of the internal carotid artery with the bone plate separating the middle ear cavity.

ophthalmic artery and the ICA bifurcation into the anterior and middle cerebral artery.[2]

The medial wall of the middle ear artery is supplied by the caroticotympanic branches from the ICA and the internal tympanic artery via the ascending pharyngeal artery.[7] In normal development, the ICA forms within 24 days of embryonic development.[2] It forms from the third branchial arch artery and the distal segments of the paired dorsal aortae.[2,8] The ICA develops as 7 separate segments (Figures 6 and 7).[2,9]

When agenesis of the cervical and petrous segments occurs in the early embryological period, the blood supply of brain

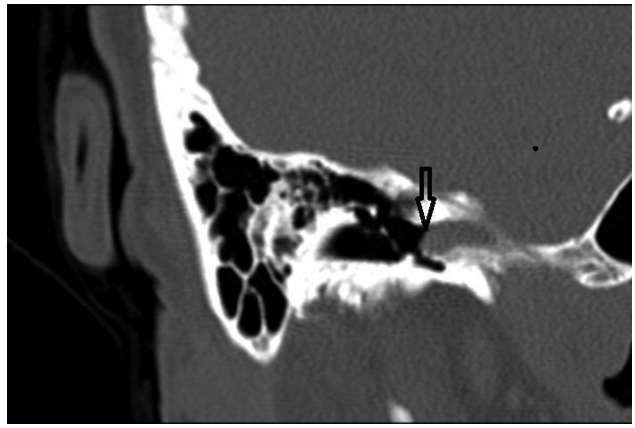

Figure 3. Coronal view of a high-resolution computed tomography of temporal bone demonstrating laterally displaced internal carotid artery (junction of vertical and horizontal petrous segment) without any bone plate separating from the middle ear cavity.

Figure 4. High-resolution computed tomography of temporal bone demonstrating laterally displaced internal carotid artery (vertical petrous segment of right internal carotid artery) segment along the medial wall of middle ear cavity.

is reconstituted via a collateral pathway from the ascending pharyngeal artery to inferior tympanic artery to the carotid tympanic artery to the horizontal segment of ICA (Figure 8).[2,10] This anatomical variant is called an aberrant ICA. Sometimes, this is associated with a persistent stapedial artery and can be bilateral.[2,8]

When this occurs, symptoms are often present, including pulsatile tinnitus, a pulsatile retrotympanic mass, conductive hearing loss, ear pain, and aural fullness. On otoscopic examination, a dark red or bluish swelling may appear in the middle ear area. The clinical differential diagnosis includes an aberrant ICA, persistent stapedial artery, jugular diverticulum glomus tumor, facial nerve schwannoma, and cholesterol granuloma.

Figure 5. Normal high-resolution computed tomography of the temporal bone demonstrating normal course of the internal carotid artery.

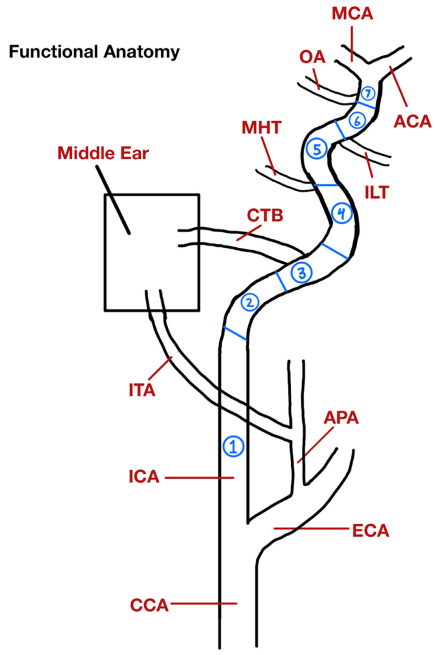

Figure 6. Common carotid artery (CCA), external carotid artery (ECA), internal carotid artery (ICA), ascending pharyngeal artery (APA), inferior tympanic artery (ITA), caroticotympanic branch (CTB), meningohypophyseal trunk (MHT), inferolateral trunk (ILT), ophthalmic artery (OA), anterior carotid artery (ACA), and middle carotid artery (MCA). Internal carotid segments: 1, cervical; 2, vertical petrous; 3, horizontal petrous; 4, ascending cavernous; 5, horizontal cavernous; 6, clinoidal; and 7, supraclinoidal.

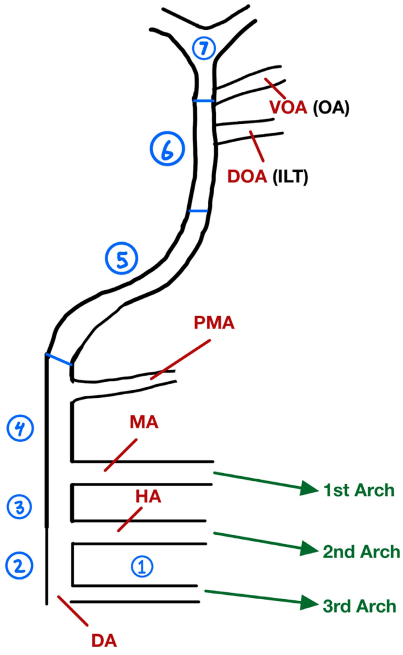

Figure 7. Dorsal aortic arch (DA), hyoid arch (HA), mandibular arch (MA), primitive maxillary artery (PMA), dorsal ophthalmic artery (DOA), inferolateral trunk (ILT), ventral ophthalmic artery (VOA), and ophthalmic artery (OA). Internal carotid segments: 1, cervical; 2, vertical petrous; 3, horizontal petrous; 4, ascending cavernous; 5, horizontal cavernous; 6, clinoidal; and 7, supraclinoidal.

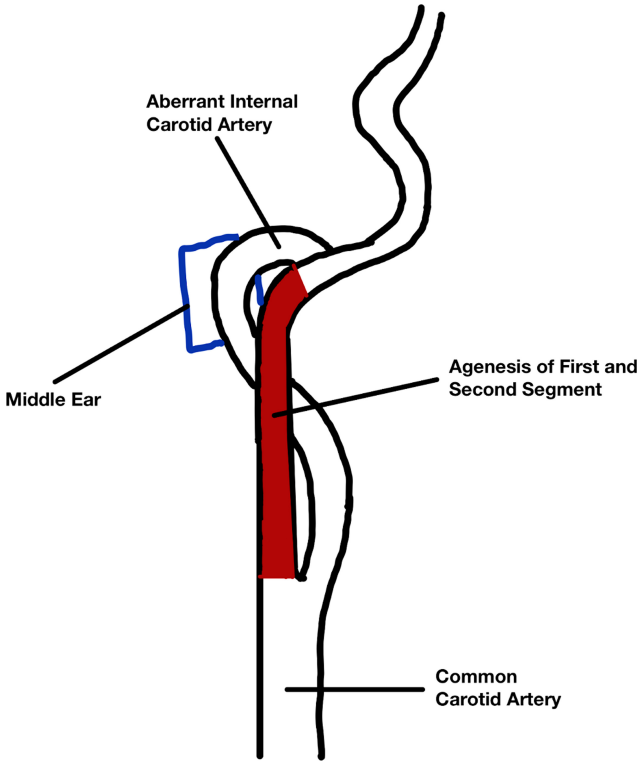

Figure 8. Graphic demonstrating agenesis of the first and second embryological segments of the internal carotid artery (ICA). The aberrant ICA formed from an enlarged inferior tympanic artery, the carotid tympanic artery, which continues as horizontal petrous segment.

High-resolution CT scan or CTA is needed to differentiate vascular lesions from the neoplasm since any surgical intervention of vascular lesions may cause catastrophic bleeding.[11] Specific imaging findings of aberrant ICA include soft tissue mass in the middle ear, a reduced caliber aberrant ICA entering the middle ear through the inferior tympanic canal, sharp turn anteriorly as it continues with the horizontal portion of petrous segment of the ICA, absence of the proximal portion of the carotid canal and absence of bone covering between ICA and middle ear cavity, and hearing loss due to the artery which lies beneath the incudostapedial joint.

If an aberrant ICA is present, management is controversial.[4] Surgical correction is mostly avoided since manipulation could result in stroke, Horner's syndrome,[4] paralysis of the facial nerve, and deafness.[12] Nevertheless, treatment techniques such as placing silicone sheets or bone and fascial grafts in between the aberrant ICA and the ossicular chain have been proposed.[13,14]

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

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Declaration of Interests: The authors have no conflict of interest to declare.

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