

A Persistent Pharyngohyostapedial Artery: Embryologic Implications

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Summary: A 3-year-old child was examined because of otorrhea. CT scans showed an unusual vessel, confirmed by angiography, related to a persistent pharyngohyostapedial artery. This embryonic persistent artery associated with the normal internal carotid artery would explain the "duplication" aspect of the internal carotid artery.

Anomalies of the internal carotid artery are rare, usually detected as an incidental finding. The case presented here illustrates the persistence of a pharyngohyostapedial artery associated with a normal internal carotid artery. Embryologic considerations are discussed.

Case Report

A 3-year-old girl underwent two tympanotomies on the left ear, both of which were heavily hemorrhagic. Immediately after the second tympanotomy, a high-resolution CT study was performed with 1-mm collimation and 1-mm-thick sections in the axial (Fig 1A-D) and coronal (Fig 1D) planes. Each scan was targeted for maximal bony detail with extended gray scale to 4000 HU.

The right temporal bone was normal. On the left side, in addition to the middle ear hemorrhage, the CT scans revealed an enlarged inferior tympanic canaliculus at the skull base, continuing with a bony canal that penetrated the floor of the tympanic cavity, ran along the promontory, and then joined the carotid canal at the junction of the vertical and horizontal segments.

The anterior tympanic segment of the facial nerve canal was enlarged, reaching the geniculate ganglion. Lateral to the geniculate ganglion, a specific opening was noted in the antero-superior surface of the petrous bone. The stapes superstructure was poorly defined, owing to the hemorrhage. The ipsilateral foramen spinosum could not be identified, but the carotid bony canal was normal. Although the CT findings indicated the persistence of a hyostapedial artery, they did not explain the enlarged inferior tympanic canaliculus.

Digital subtraction angiography (selective left carotid injection) (Fig 1E and F) showed a so-called "duplication" aspect of the left cervical internal carotid artery from the level of the bifurcation to the petrous segment. On the external carotid artery, there was neither a middle meningeal artery nor an ascending pharyngeal artery. The latter arose from the internal carotid artery, 1 cm after the bifurcation, and its enlarged inferior tympanic branch ran posteriorly and laterally to the internal carotid artery, following a parallel course. Both inferior

tympanic and internal carotid arteries were selectively catheterized. The inferior tympanic artery crossed the tympanic cavity, then joined the internal carotid artery at the junction of the vertical and horizontal segments. The stapedial artery was identified as originating from the distal inferior tympanic artery, and distally supplying the middle meningeal artery. The persistence of a pharyngohyostapedial artery was thus established. It was associated with a normal internal carotid artery, thereby simulating a duplication of the internal carotid artery.

Discussion

The embryologic development of the carotid system has been well known since the fundamental studies of Congdon in 1922 (1), Altmann in 1947 (2), and especially Padget in 1948 (3), as well as the more recent works of Steffen in 1968 (4) and Lasjaunias and Moret in 1978 (5). According to Congdon (1) and Altmann (2), during early embryologic development of the human embryo there is anterior enlargement of the truncus arteriosus, called the aortic sac (as opposed to the lower vertebrates, which have paired ventral aortas), from which six pair of aortic arches arise. Each pair related to the cranial nerve roots belongs to the corresponding pair of branchial arches, and connects to the homolateral dorsal aorta. The aortic arches develop in a cranio-caudal sequence, and are not present at the same time. The first undergoes involution about the time the fourth is complete, and the second disappears before the sixth (pulmonary) is finally developed (2). The first three aortic arches form the basis for the development of the carotid system. After involution, the first and second aortic arches are replaced by the primitive mandibular and hyoid arteries. The mandibular artery has no important role, and remains as the vidian or pterygoid artery (3).

Concurrently, Congdon (1), Padget (3), and Tandler (cited in [3]) noted a pair of symmetrical arterial sprouts, extending cranio-laterally from the aortic sac at the midline to the mandibular nerve root. These vascular structures, named ventral pharyngeal arteries, supply the first and second branchial arches and have an important role in the formation of the stapedial and external carotid arteries. They will be interrupted into distal and proximal portions in the region of the mandibular root of the fifth nerve.

At the end of the branchial phase, the hyoid artery, the dorsal remnant of the second aortic arch, gives off an important collateral branch near its origin from the internal carotid, called the stapedial

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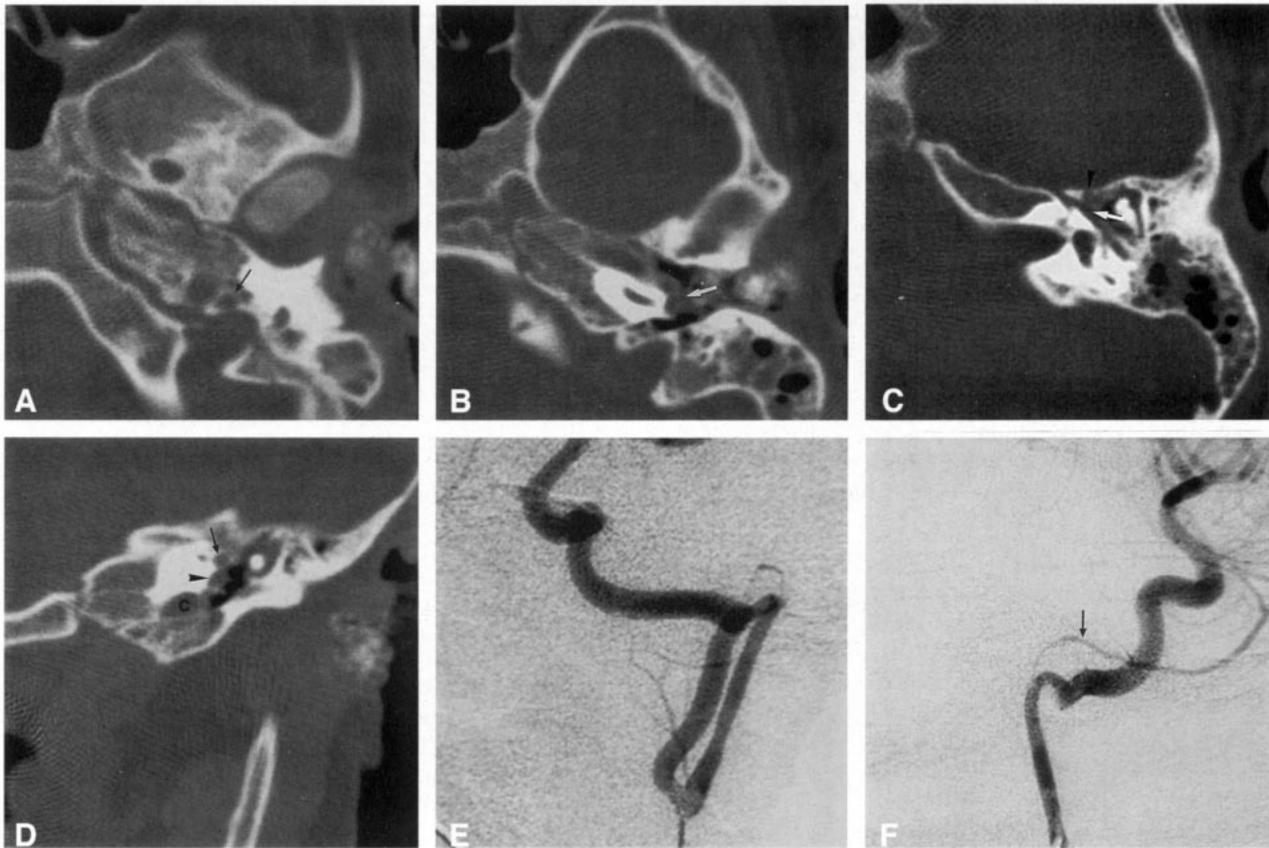


FIG 1. 3-year-old girl with a persistent pharyngohyostapedial artery detected after second tympanotomy of the left ear.

A–C, Axial CT scans from the skull base to the superior surface of the petrous bone. Enlarged inferior tympanic canaliculus is interposed between the carotid and jugular foramina (arrow, A). No ipsilateral foramen spinosum is identified. The bony canal along the promontory (arrow, B), where the hyoid artery crosses, has an appearance similar to a pair of glasses. The anterior tympanic segment of the facial nerve canal is enlarged (arrow, C), and a special opening is seen at the anterosuperior surface of the petrous bone lateral to the geniculate ganglion (arrowhead, C).

D, Coronal CT scan shows enlargement of the anterior tympanic segment of the facial nerve canal (arrow) as compared with the labyrinthine segment. The bony canal containing the hyoid artery is enclosed in the promontory (arrowhead). C = carotid canal.

E, Digital subtraction angiogram, oblique view, shows “duplication” aspect of the left internal carotid artery. The inferior tympanic artery courses parallel laterally and posteriorly to the internal carotid artery. Note the other branches of the ascending pharyngeal artery.

F, Lateral subtraction angiogram of selective inferior tympanic artery from which the stapedia artery arises, further supplying the middle meningeal artery (arrow).

artery (normally persistent in some species but temporary in the human embryo), which courses anteriorly through the stapes primordium and continues between the chorda tympani and the facial nerve. At the height of its development (embryos of 16 to 18 mm), the stapedia artery has two major divisions (3):

- The dorsal or supraorbital division courses laterally to the gasserian ganglion and advances far into the primitive orbit, accompanying the ophthalmic root of the fifth cranial nerve. The primitive ophthalmic artery retains the orbital end of the supraorbital division to form the lacrimal artery. The remainder of the supraorbital division is incorporated into the middle meningeal artery.

- The stapedia artery annexes the distal end of the ventral pharyngeal artery at the mandibular nerve root, thereby forming the primary (ventral) maxillomandibular division, which continues the course of the main trunk of the artery and termi-

nates into separate maxillary and mandibular arteries. The proximal end of the ventral pharyngeal artery carries the anlage of the primitive external carotid artery.

At the 20- to 24-mm stage of embryologic development, the stapedia artery is interrupted in the area of the stapes. Its maxillomandibular division becomes secondary, linked to the external carotid artery, to form the middle meningeal artery, a collateral of the internal maxillary artery. The distal remnant of the stapedia artery beyond the stapes remains as the superior tympanic artery, a branch of the middle meningeal artery. The remnant of the hyoid artery is the caroticotympanic artery, a branch of the intrapetrous carotid artery, which accompanies the inferior tympanic nerve and anastomoses with the inferior tympanic artery, a branch of the ascending pharyngeal artery (5).

The different stages of development of the internal and external carotid arteries explain the oc-

currence of rare persistent embryonic arteries, such as, in our case, the hyostapedial and pharyngo-hyostapedial arteries.

Since the first human case was reported by Hyrtl in 1836 (in [2]), many authors have reported descriptions of the persistent stapedial artery's course through the middle ear, or more precisely, for Padgett (3), the persistent hyostapedial artery (2, 4, 6). According to Altmann (2), the vessel originating from the internal carotid artery penetrates the floor of the tympanic cavity and runs upward and backward along the promontory, enclosed in a bony canal. Leaving the promontory, it goes upward between the crura of the stapes and then enters the facial canal, accompanying the facial nerve. Behind the geniculate ganglion, the artery leaves the facial canal through a special opening and proceeds between the dura and inner surface of the middle fossa, forward and upward to terminate in the middle meningeal artery.

The persistent pharyngostapedial artery has been previously reported by Moret (6), but the persistent pharyngo-hyostapedial artery, in fact, has never been clearly established without segmental agenesis of the internal carotid artery. In the latter case, the well-known course of the aberrant internal carotid artery (flux) through the middle ear corresponds to the anastomoses in the inferior tympanic canaliculus between the inferior tympanic artery and the hyoid artery, which can be associated or not with stapedial artery persistence. There is neither an ascending segment of the intrapetrous carotid artery nor a consequently ascending segment of the bony carotid canal. For unknown reasons, the blood flow regresses in the cervical carotid artery and goes preferentially through the inferior tympanic artery and then the hyoid artery, to finally join the horizontal portion of the carotid artery (5, 6).

Comparative anatomic and embryologic studies have shown that the internal carotid artery is consistently developed in mammalia, while in other species a secondary obliteration of the vessel (complete or only partial) occurs during embryonic or early postembryonic development. Also in human development, aplasia or hypoplasia of the internal carotid artery would be related to secondary atrophy of the originally normally developed internal carotid artery (Tandler in [1], Wolff in [4]).

An explanation of the noteworthy aspects of our observation of a persistent pharyngo-hyostapedial artery requires an examination of two additional changes in normal embryologic development: first, the persistence of a pharyngo-hyoid artery, with or without a stapedial artery, and second, the direct emergence of the ascending pharyngeal artery from the internal carotid artery. The ascending pharyngeal artery arises more often from the external carotid artery (proximal or distal branches), but its internal carotid origin is nevertheless well known. These conditions would explain the so-called "fenestration" or "duplication" aspects of the internal carotid artery (Fig 2), first reported by Killien in

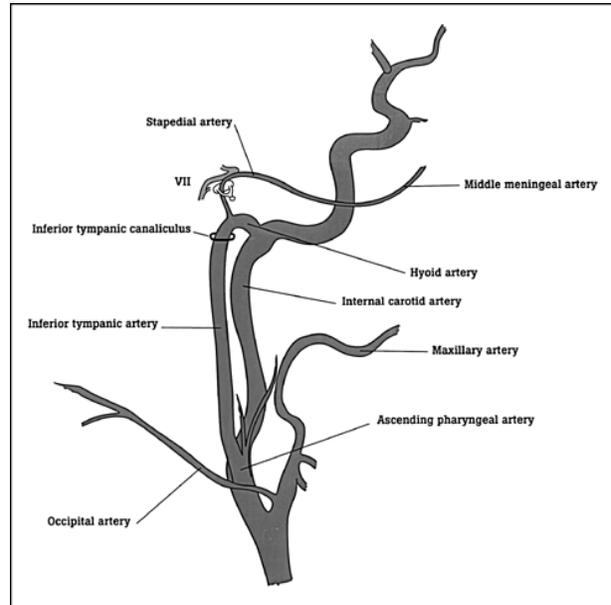


FIG 2. Diagram of persistent pharyngo-hyostapedial artery.

1980 (7, 8), as well as in more recent publications (9–11), for which neither a satisfactory embryologic hypothesis has been made nor anatomic descriptions given. Moreover, fenestration does not exist in other species as a normal and usual variant (8). There is actually neither duplication nor fenestration of the internal carotid artery, but the presence of two different vessels, as suggested by Koenigsberg et al (10): one is the true internal carotid artery and the other is the inferior tympanic artery, a collateral of the ascending pharyngeal artery, which emerges directly from the internal carotid artery, connects to the persistent hyoid artery, and finally joins the internal carotid artery at its junction of the vertical and horizontal portions.

The anastomoses between the inferior tympanic artery and the hyoid artery in the inferior tympanic canaliculus would be preserved because of the direct emergence of the ascending pharyngeal artery from the internal carotid artery, with a consequently enlarged inferior tympanic artery related to increased arterial blood flow. Nevertheless, whether persistence of both the inferior tympanic and internal carotid arteries occurs in this particular variation remains unclear, probably because of equal blood flow through both vessels.

Conclusion

As has been previously reported for other vascular anomalies of the middle ear, it appears clear that basic embryologic studies, however old, will continue to play a major role in the explanation of arterial anomalies. High-resolution CT of the petrous bone is an excellent diagnostic method to illustrate arterial vascular anomalies of the middle ear, which must be detected before surgery, even though they are rarely encountered, because of the

risk of profuse bleeding. In this case, the specific course of the persistent hyostapedial artery was the same as previously reported in conventional anatomic descriptions, and the persistent pharyngo-hyostapedial artery, confirmed by angiography, could have been suspected because of the enlarged inferior tympanic canaliculus.

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