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Case Report

Duplicated Cervical Internal Carotid Artery with Ipsilateral Persistent Stapedial Artery and Contralateral Aberrant Internal Carotid Artery: A Case Report

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Objective: Duplication of extracranial cerebral arteries is a rare anatomical variant, most commonly observed in the vertebral artery. However, duplication of the cervical internal carotid artery (ICA) is extremely rare. We present a unique case of a patient diagnosed with a right facial artery aneurysm, in whom a right duplicated ICA, ipsilateral persistent stapedial artery (PSA), and contralateral aberrant ICA were incidentally identified.

Case Presentation: A 56-year-old woman presented with a painless mass in the right lower jaw. Physical examination revealed a round, pulsatile mass. Contrast-enhanced CT demonstrated a 25 mm saccular aneurysm originating from the right facial artery, along with a right duplicated ICA and a left aberrant ICA. The lateral limb of the right duplicated ICA and the left aberrant ICA were seen traversing the tympanic cavity and entering the carotid canal. The absence of the right foramen spinosum indicated the presence of a PSA. Angiography confirmed the duplicated ICA, aberrant ICA, and PSA. The lateral limb of the right duplicated ICA gave rise to the occipital artery and a small branch, the PSA, which entered the middle cranial fossa. The facial artery aneurysm was successfully embolized. Follow-up MR angiography showed stable occlusion of the aneurysm, with the duplicated ICA giving off both the PSA from the lateral limb and a pharyngeal branch from the medial limb.

Conclusion: This case provides valuable insights into the development of the cervical arterial system during the fetal period, particularly the collateral pathways connecting the third arch to the dorsal aorta.

Keywords ▶ duplicated internal carotid artery, aberrant internal carotid artery, persistent stapedial artery, vascular embryology

Introduction

Duplication of the extracranial cerebral arteries is a rare anatomical variant, most of which are observed in the vertebral artery.[1)] Duplication of the cervical internal carotid artery (ICA) is extremely rare. Persistent stapedial artery (PSA) is also a rare anomaly, which is occasionally associated with an aberrant (intratympanic) ICA.

Herein, we present a rare case of duplication of the right ICA associated with ipsilateral PSA and contralateral aberrant ICA, and discuss embryological development of these rare anomalies.

Case Presentation

A 56-year-old woman presented with a painless mass in the right lower jaw, which was gradually growing for 6 months. She had no history of head trauma. Physical examination revealed a round pulsatile mass. Contrast-enhanced CT demonstrated a 25 mm saccular aneurysm originating from the right facial artery

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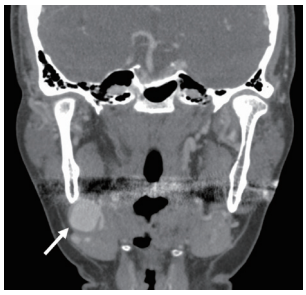

Fig. 1 Coronal section of contrast-enhanced CT. The coronal section of contrast-enhanced CT shows a saccular aneurysm (arrow) originating from the right facial artery in the submaxillary region.

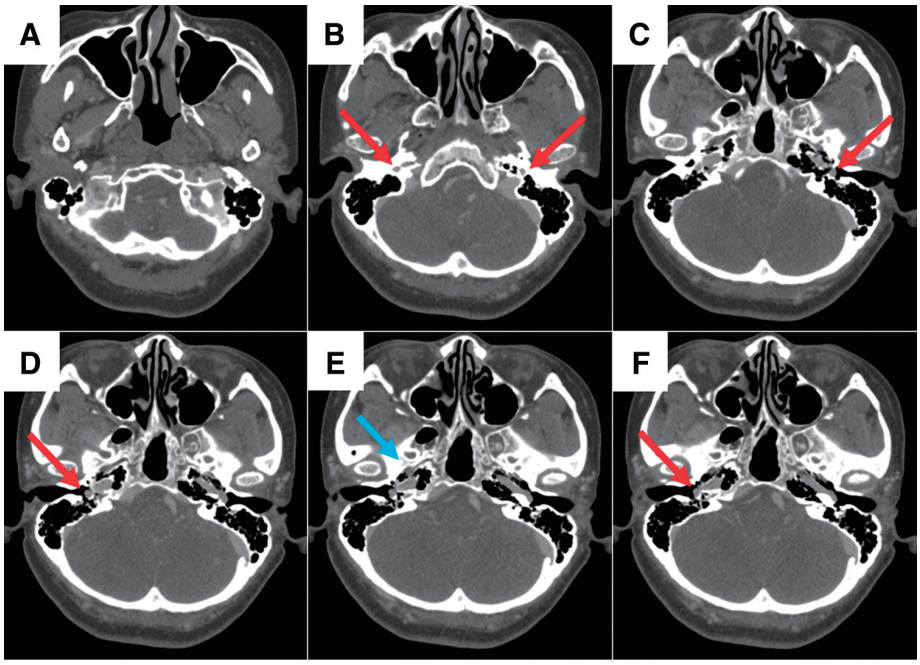

Fig. 2 Axial section of contrast-enhanced CT (**A–F**, caudal to cranial). In slice (**B**), the lateral limb of the right duplicated ICA and the left aberrant ICA are seen ascending to traverse the tympanic cavity through a dilated inferior tympanic canaliculus (Jacobson's canal) (red arrow). Slice (**C**) shows intratympanic aberrant ICA (**C**, **D**, **F**, red arrow). Slice (**E**) demonstrates an absent right foramen spinosum, indicating the presence of a PSA (blue arrow). In slice (**F**), the lateral limb of the right duplicated ICA and the left aberrant ICA traverse the tympanic cavity and enter the proximal portion of the carotid canal within the petrous bone. ICA, internal carotid artery; PSA, persistent stapedial artery

(**Fig. 1**). CT also showed a right duplicated ICA from the carotid bifurcation to the petrous portion and left aberrant ICA. The lateral limb of the right duplicated ICA and the left aberrant ICA ascended to traverse the tympanic cavity through the dilated inferior tympanic canaliculus (Jacobson's canal), entering the proximal portion of the carotid canal within the petrous bone (**Fig. 2**). The right foramen spinosum was not identified (**Fig. 2E**). Angiography also showed a right duplicated ICA and a left aberrant ICA exhibiting a vertical bend at the level of the carotid canal (**Fig. 3A** and **3C**). The lateral limb of the right duplicated ICA gave off the occipital artery posteriorly at its vertical portion and a small branch superiorly at the intratympanic segment (**Fig. 3B**). The small branch, representing PSA, entered the middle cranial fossa to become contiguous with the intracranial middle meningeal artery (**Fig. 3A**). The medial limb ran a parapharyngeal course and gave off pharyngeal branches of the ascending pharyngeal artery (**Fig. 3B**). The other branches of the external carotid artery (ECA) showed a normal branching pattern. The facial artery aneurysm was subsequently embolized with coils. The patient was discharged uneventfully. Follow-up MRA after aneurysm embolization demonstrated stable occlusion of the aneurysm. It also showed the duplicated right ICA giving off the PSA from the lateral limb and the pharyngeal branch of the ascending pharyngeal artery from the medial limb (**Figs. 4** and **5**). The course of the stapedial artery observed on MRA was inferred to pass between the anterior and posterior crura of the stapes based on comparison with the tympanic structures on previously acquired CT images.

Discussion

In the human embryo, carotid arteries, the aortic arch, and major thoracic arteries derive from transient blood vessels termed pharyngeal arch arteries (PAAs) that connect the aortic sinus to the dorsal aorta.[2,3] The paired PAAs evolve from a vascular plexus in the second heart field within the anterior lateral plate mesoderm under genetic regulation such as Hox genes and several molecular signal pathways.[4–6] The first and second arches regress early, and the remnant vascular plexus becomes the precursor to branches of the ECA, and distal parts of the first and second arches that connect to the dorsal aorta become the premandibular artery (future vidian artery)

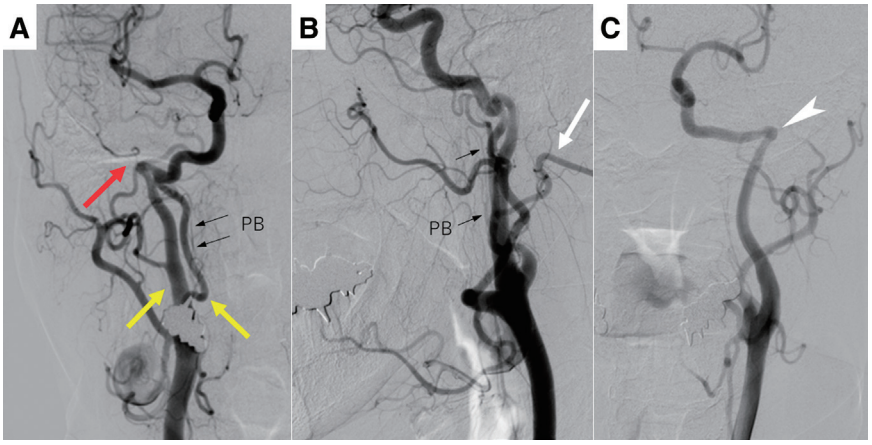

Fig. 3 Angiography. (**A**) The DSA frontal view of the right CCA shows a duplicated ICA (yellow arrow) and a PSA (red arrow). The PSA enters the middle cranial fossa and continues as the intracranial middle meningeal artery. The medial limb runs a parapharyngeal course and gives off the PB of the ascending pharyngeal artery (**A**, **B**, black arrow). (**B**) DSA lateral view of the right CCA shows the lateral limb of the right duplicated ICA, giving off the occipital artery posteriorly at its vertical portion (white arrow). (**C**) DSA frontal view of the left CCA shows an aberrant ICA exhibiting a vertical bend at the level of the carotid canal (arrowhead). CCA, common carotid artery; ICA, internal carotid artery; PB, pharyngeal branch; PSA, persistent stapedial artery

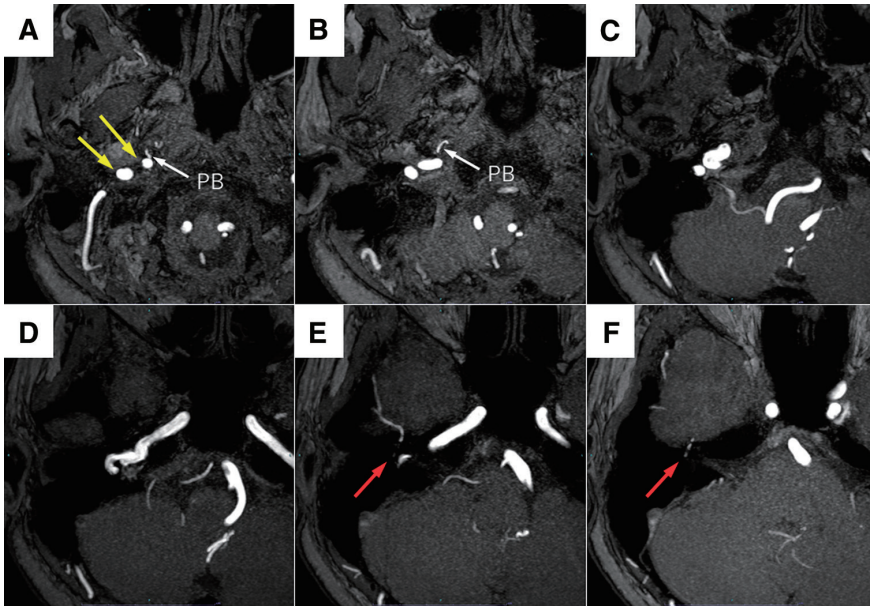

Fig. 4 Axial section of slab MIP MRA (**A–F**, caudal to cranial). Axial images of slab MIP MRA show the duplicated ICA (yellow arrow), the PSA (red arrow) originating from the lateral limb of the right ICA via the carotico-tympanic artery, and PB (white arrow) of the ascending pharyngeal artery. ICA, internal carotid artery; MIP, maximum intensity projection; PB, pharyngeal branch; PSA, persistent stapedial artery

and the hyoid artery (future carotico-tympanic artery).[3] The third arches and the paired dorsal aorta extending from the third arch remain to be the common carotid artery and the ICA. The dorsal aorta between the third and fourth arches (carotid duct) regresses, and the fourth arches become the brachiocephalic artery on the right side and the aortic arch on the left sides, respectively.

Different types of ICA duplication or fenestration have been reported in the literature. Among these, the majority of fenestrations might represent pseudo-fenestrations caused by arterial dissection rather than a developmental anomaly.[7] There are 2 types of cervical duplicated ICA. One is the duplication of ICA with the 2 limbs running parallel in the normal ICA course, which is probably due

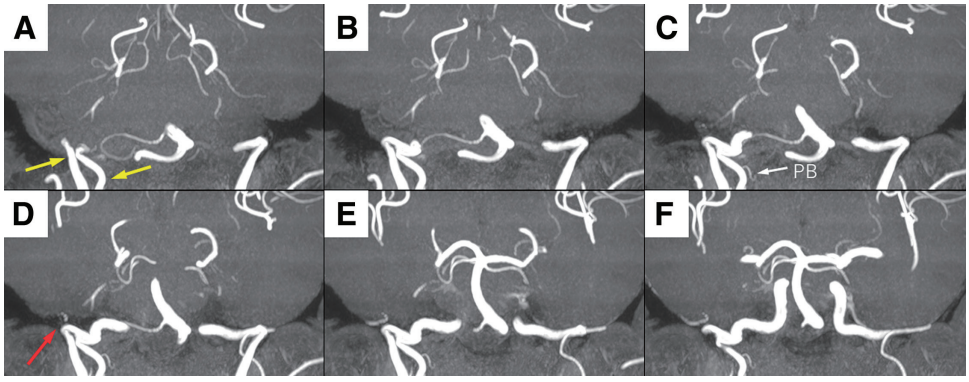

Fig. 5 Coronal section of slab MIP MRA (**A–F**, posterior to anterior). Coronal images of slab MIP MRA show the duplicated ICA (yellow arrow), the PSA (red arrow) originating from the lateral limb of the right ICA via the carotico-tympanic artery, and the PB (white arrow) originating from the medial limb. ICA, internal carotid artery; MIP, maximum intensity projection; PB, pharyngeal branch; PSA, persistent stapedial artery

to incomplete fusion of the third PAA from the primitive vascular plexus.[8,9] Our case is the other type, which is a duplicated ICA with an aberrant course. To the best of our knowledge, 3 cases of bilateral aberrant ICA with duplication have been reported.[10–12] It is thought to be caused by hypoplasia or interruption of the third PAA or its junction between the third arch and the dorsal aorta with the development of collateral via the ascending pharyngeal arterial branch.[11] This alternate circulation makes the external carotid system connect to the internal carotid system. The inferior tympanic branch of the ascending pharyngeal artery anastomoses with the carotico-tympanic artery (hyoid artery remnant) of the ICA. The stapedial artery further branches into the internal maxillary and middle meningeal arteries. The proximal portion of the hyostapedial trunk generates the carotico-tympanic artery (external artery branch). Normally, as the ECA develops, the stapedial artery regresses, although it persists in some cases, such as aberrant ICA. This complex developmental pattern can result in the simultaneous presence of an intratympanic aberrant ICA and a PSA, as both anomalies stem from the same embryological deviation from typical vascular development. The intratympanic aberrant ICA corresponds to the lateral limb of the duplicated ICA. Regarding the medial limb, our case showed the medial limb running a parapharyngeal course and giving off the pharyngeal branches (originally from the ascending pharyngeal artery). That may suggest the medial limb can be formed by the development of another collateral pathway to the ICA through the pharyngeal and carotid branches of the ascending pharyngeal artery. The anomalies involving duplication of the ICAs may be associated with abnormalities in the regulation of Hox genes or angiogenic factors during the embryonic period.

In our case, the lateral limb of the right duplicated ICA and the left ICA showed the intratympanic aberrant ICA course. During otolaryngological examination, the intratympanic aberrant ICA is often identified as a reddish mass in the middle ear space.[12] It may cause pulsatile tinnitus, conductive hearing loss, and aural fullness. However, some cases may remain asymptomatic.[13] The diagnosis of the aberrant ICA is important because otologic intervention may result in severe iatrogenic complications.[14,15] CT is useful for the diagnosis of the intratympanic aberrant ICA, typically identified by a thin or absent bony covering over the tympanic portion of the ICA and the lack of a vertical segment in the carotid canal.[16] PSA is often associated with an aberrant ICA, and it may also present symptoms similar to those of an aberrant ICA.[17] Thiers et al. summarized the image findings of a PSA: (1) a small canaliculus leaving the carotid canal; (2) a linear structure crossing the middle ear over the promontory; (3) an enlarged facial nerve canal or a separate canal parallel to the facial nerve; and (4) absence of the foramen spinosum.[18] In our case, we could confirm only the absence of the foramen spinosum by CT. Regarding the delineation of blood vessels, MRI may allow for better visualization of vascular continuity and could be useful in understanding vascular anomalies. While reports of bleeding during middle ear surgery are also present in PSA cases, no neurological complications have been reported in cases of intraoperative resection.[19] A single case report described preoperative embolization

of PSA to prevent intraoperative bleeding, which resulted in no complications.[20)]

Conclusion

We present a rare case of duplicated cervical ICA with ipsilateral PSA and contralateral aberrant ICA. The present case is of great interest for understanding the development of the cervical arterial system during the fetal period, particularly the collateral pathways connecting the third arch to the dorsal aorta.

Disclosure Statement

The authors declare that they have no conflicts of interest.

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