Normal variations of cervical-petrosal Internal Carotid Artery; Imaging and Embryologic consideration

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Learning objectives

1. To describe the embryological development of cervical-petrosal internal carotid artery (ICA).

2. To demonstrate anatomical variations of cervical-petrosal ICA and discuss the malformation mechanism of those anomalies.

3. To demonstrate the importance of the knowledge of these anatomical variations for diagnosis and surgical/interventional procedures.

Background

The development of neurointervention technique has attributed to the growth of the number of cerebral angiography. Occasionally, we encounter the cases with anatomical variations of internal carotid artery (ICA). There are several anatomical variations of ICA which requires special attention and/or can cause unexpected complications in diagnostic angiography and neurointervention. We illustrate the normal embryological development of carotid artery and demonstrate cases with anatomical variations of cervical-petrosal ICA. We also discuss about embryological malformation mechanism of these cases.
Normal embryological development of carotid artery

Cervical-petrosal ICA

In the human embryo, six paired aortic arches arise from aortic sac and terminate in the ipsilateral dorsal aorta (1). The cervical carotid arteries develop by complicated processes of regression, and the communication within the vascular network consists of the ventral aorta, dorsal aorta, aortic arches, and intersegmental arteries (caroticoisular anastomosis) (Fig. 1, 2). The first aortic arch is completed by day 24 but regresses as the second aortic arch forms on day 26. The third and fourth arches form on day 28; the second arch degenerates as the sixth arch forms on day 29. The dorsal aortic side of first aortic arch becomes the mandibular artery. After second aortic arch regresses, dorsal aortic side of second aortic arch forms the hyoid artery and ventral side forms the ventral pharyngeal artery (VPA) (Fig. 2-B,C). The VPA, the origin of which migrates to third aortic arch, is based on the primitive external carotid artery (ECA) (Fig. 2-D). A section of the dorsal aorta between the third and the fourth aortic arch, so-called ductus caroticus, regresses subsequently. The common carotid artery (CCA) is derived from the proximal part of the third aortic arches. Cervical ICA is derived from the distal part of the third aortic arches and the embryonic dorsal aorta. (2).

External carotid artery

The External carotid artery system, in terms of embryology, can be classified into following three groups; internal maxillary artery system, derived from the embryologic hyostapedial artery (second primitive aortic arch derivative), thyroid-facial-lingular artery system, which all come from regressed ventral aorta (VPA), and ascending paryngeal-occipital artery system, which are formed through caroticoisular anastomoses.

The hyoid artery system, which is a remnant of the distal part of the second aortic arch, gives rise to the stapedial artery, passing through the stapes. The stapedial artery bifurcates into superior (supraorbital) division and inferior (maxillomandibular) division. The supraorbital division becomes the middle meningeal artery afterward, and maxillomandibular division further becomes infraorbital artery and inferior alveolar artery (2). The primitive ECA, which is derived from the VPA, forms its definitive branches, including the internal maxillary artery, facial artery and lingual artery. The anastomosis of the VPA to the stapedial artery (maxillomandibular division) forms the ECA while the proximal stapedial artery regresses (Fig. 2). The ascending pharyngeal artery basically develops from the primitive hypoglossal artery. The occipital artery is also a remnant.
of the embryologic carotid-vertebrobasilar anastomosis, namely: type I and type II proatlantal intersegmental arteries. Because the both ascending pharyngeal and occipital artery can originate from either external carotid or internal carotid artery in normal development, there are potential communications between the VPA (ECA) and the distal portion of the IIIrd aortic arch or dorsal aorta (ICA) via the primitive ascending pharyngeal/hypoglossal artery and the primitive occipital/proatlantal artery (Fig. 3).

Review Cases of anatomical variations of cervical-petrosal ICA

1. Primitive cervical carotico-basilar/vertebral anastomosis

A. Persistent primitive proatlantal artery (Fig.4)

B. Persistent primitive hypoglossal artery (Fig.5)

**Persistent primitive proatlantal artery (PPPA):** a persistent of the suboccipital caroticobasilar anastomosis between the cervical carotid and vertebral artery

Type 1 PPPA- the first cervical segmental artery (the proatlantal intersegmental artery)

Type 2 PPPA- the second cervical segmental artery (the first cervical intersegmental artery)

**Persistent primitive hypoglossal artery (PPHA):** a persistent of the caroticobasilar anastomosis between the cervical carotid and basilar artery courses through the hypoglossal canal.

Embryology

Primitive carotid-vertebral and carotid-basilar anastomoses are formed at early stage during human embryogenesis at approximately 24 days. These ventro-dorsal anastomoses includes a cranial extension of the primitive internal carotid artery (posterior communicating artery), trigeminal artery, otic artery, hypoglossal artery, and cervical segmental arteries from cephalic to caudal direction. In the 5- to 6-mm (28-30 days) embryo, these anastomotic channels provide the proximal supply to the longitudinal neural arteries (Fig.6). Involution of these anastomotic channels starts when the embryo is 7 to 12 mm (3). There are two types in the persistent primitive proatlantal artery of type1 and type2. Type 1 is corresponding to the first cervical segmental artery, and type 2 is corresponding to the second cervical segmental artery.
Imaging findings and Clinical significance

Type 1 PPPA originates from the internal carotid artery or the external carotid artery at the proximal portion, and ascends posterolaterally. It runs without passing through the transverse foramen of the cervical vertebrae, and joins the intracranial vertebral artery after running through the foramen magnum. Type2 PPPA arising from the ECA ascends more laterally than the type1. It runs through the transverse foramen of C1 vertebra and then joins the extracranial vertebral artery below the C1 level. Fifty-seven percent of the described proatlantal arteries are of the type 1, 38% are type 2, and 5% arise from the common carotid artery (4). The PPHA arises from cervical ICA, and ascends parallel to the cervical ICA. It curves posteromedially, entering intracranial space through the hypoglossal canal, and joins the caudal portion of the basilar artery. Hypoplasia of the ipsilateral (or bilateral) vertebral artery is frequently seen in the cases of these carotidobasilar(vertebral) anastomoses.

The particular course of each types of anastomosis would be an essential point in differentiation of these three persistent of primitive arteries. CT or MR images can exactly demonstrate the particular course of these arteries with a relationship of the surrounding structures, e.g. hypoglossal artery passing through the hypoglossal canal (5,6).

The carotid bifurcation is the most common site of atheromatous plaques that can cause ischemia in both carotid and vertebral/basilar territories. The embolic sources separated from carotid bifurcation can reach to the vertebral/basilar artery through the PPPA/PPHA and result in neurological symptoms in both anterior and posterior cerebral territories (3).

PPHA may be associated with an anomalous structure of the vessel wall and exposes the basilar trunk to an unusual hemodynamic stress, predisposing to the onset of aneurysm with possible hemorrhagic consequences (7). However, the PPPA is thought not to be associated with the cerebral aneurysms.

2. Aberrant ICA (intratympanic ICA) (Fig.7, 8)

Intratympanic ICA: the ICA running unusual tortuous course through the middle ear

Embryology

The following genetic mechanism of this anomaly has been supposed by Lasjaunias et al. (8, 9). They hypothesized the alternate blood flow theory. If the third primitive aortic arch involutes abnormally, the ICA can be supplied via the persistence of the VPA system. As a consequence, an anomalous course develops with blood flowing via the VPA and the enlarged inferior tympanic artery with retrograde flow through the caroticotympanic vessels into the horizontal segment of the ICA (VPA-hyoid
anastomosis) (Fig.9). Therefore, the aberrant intratympanic ICA is frequently associated with persistent stapedial artery.

**Imaging findings and Clinical significance**

Aberrant intratympanic ICA takes an aberrant course which runs adjacent to jugular bulb, and passes through middle ear. It is associated with the absence of the bone plate between the carotid canal and tympanic cavity.

Aberrant intratympanic ICA can be associated with pulsatile tinnitus, conductive hearing loss, and a pulsatile retrotympanic mass (10). Otologists should be aware the intratympanic ICA to avoid surgical injury of the ICA due to misdiagnosis. Although otoscopic findings of this variation may mimic glomus tumor, vascular malformations or high jugular bulb, High resolution CT of the temporal bone and MR images can clearly demonstrate the characteristic course of the intratympanic ICA with enlargement of inferior tympanic canal, and absence of bone covering the tympanic portion of the ICA. Endovascular embolization has been applied for acute bleeding due to iatrogenic injury of the intretympanic ICA during middle ear surgery.

3. **Aberrant ICA (retropharyngeal/lateral pharyngeal ICA) (Fig.10)**

Retropharyngeal/lateral pharyngeal ICA: the ICA running unusual, tortuous and medial course in the posterior pharyngeal wall

**Embryology**

There are the two hypotheses about the etiology of aberrant retropharyngeal/lateral pharyngeal ICA. One is by tortuosity and atherosclerotic changes in old and hypertensive patients. The other is the congenital cause; embryonic ICA uncoils as the dorsal aortic root descends into the chest and finally assumes a straight course in the neck (2, 11). It is postulated that failure to fully uncoil or migrate is due to a greater relative growth of the dorsal aortic arch resulting in a congenitally tortuous carotid vessel, usually where it crosses the glossoopharyngeal nerve. The exact cause is unclear, however, abnormal development of cervical carotid artery from the third aortic arch to CCA or ICA would cause of retropharyngeal/lateral pharyngeal ICA for young patients.

**Imaging findings and Clinical significance**

Retropharyngeal/lateral pharyngeal ICA runs the anomalous course located in the midline of the posterior pharyngeal wall. Although majority of this variation are asymptomatic, It
can present as oropharyngeal mass on oral/pharyngeal examination. This variation can cause fatal hemorrhage during oropharyngeal surgery (e.g. tonsillotomy). Therefore, it should be remained in the case of oropharyngeal diseases.

4# Non-bifurcating carotid artery (Fig.11,12,13)

Non-bifurcating carotid artery: the carotid artery ascending without forming a bifurcation of the ICA and ECA

Embryology

Non-bifurcating carotid artery is an extremely rare anatomical variation of cervical carotid artery (12, 13, 14, 15, 16, 17). It can be classified into two types according to its course at the proximal portion. One runs into carotid canal in straight course (Fig.11). The other shows significant tortuosity of the carotid artery at the C1 level, and is sometime associated with an arterial stump (Fig.12, 13).

Two hypotheses have been developed to explain the causes such anomalies (12,13,14,18). One is agenesis of the main trunk of the ECA with regression failure of the hyoid art

When the non-bifurcating carotid artery is caused by agenesis of the common stem of the ECA with regression failure of the hyoid artery, the hyoid-stapedial artery system would be the main route supplying most parts of the ECA territory (Fig.14). Parts of the ascending pharyngeal artery (APA) and occipital artery (OA) arise from the remnant of the hypoglossal artery and the proatlantal arteries, respectively, both of which form caroticobasilar anastomoses. According to Lasjaunias et al. (19), the OA can arise from primitive ICA and ECA and vertebral artery, and sites of regression determine the origin of the OA. The origin of the APA is determined in the same way (20). Furthermore, a communicating artery should exist between the proximal portion of the APA and the OA because they often originate from a common trunk (Fig.14).

It is speculated that an arterial stump and tortuosity of the carotid artery are contributed to an abnormal regression of 3rd aortic arch. But this developmental mechanism is different from that of the aberrant intratympanic ICA associated with the abnormal regression of the 2nd aortic arch.

As previously described, it is considered that the ICA is formed via communicating arteries of hypoglossal/proatrantal system with abnormal regression of 3rd aortic arch (Fig.15). However, the developmental mechanism of the ECA has not been completely explained, in case which the carotid artery ascends up straightly, the agenesis of the ECA could be owed to this anomaly.
Imaging findings and Clinical significance

As described before, there are two types of non-bifurcating carotid artery. One shows a looping at the cervical portion with or without stump-like findings at the loop, and the other shows straight course. Several cerebrovascular lesions that coexist with non-bifurcating carotid artery have been reported. Among them, Rodríguez H et al. have suggested that steno-occlusive changes in the carotid artery are often associated with non-bifurcating carotid artery, in which both the mechanisms of atherosclerosis and congenital hypoplasia could be included (15). They also have speculated that the hemodynamic alterations, which are caused by the absence of a carotid bifurcation, could be responsible for the predilection of plaque formation in the carotid artery (12, 14).

In the neurointerventional procedures through the non-bifurcating carotid artery, endovascular approach to the target arteries could be difficult in the cases that have significant looping of the carotid artery (Fig.12, 13). The evaluation of origin of branches of the external carotid artery is necessary for superselective catheterization.

Images for this section:
**Fig. 1:** Schematic view of primitive six paired aortic arches system

**Fig. 2:** Schematic drawing of development of cervical carotid artery system
Fig. 3: Schematic drawing of development of external carotid artery system. The Primitive ECA communicates with dorsal aorta via the hyoidal anastomosis, and with 3rd aortic arch via the hypoglossal or proatlantal anastomosis. These anastomoses regress, and ECA develops as a common stem.
Fig. 4: Persistent primitive proatlantal artery. A 71-year-old man present with transient disorientation. MR angiography (A-C; source image, D; maximum intensity projection) shows left proatlantal artery, which originates from the cervical portion of the ICA and anastomoses the vertebral artery through the foramen magnum (A-C; arrow D; arrowhead).
Fig. 5: Persistent primitive hypoglossal artery. A 63-year-old man present with sudden headache and disturbance of consciousness. CT scans showed a subarachnoid hemorrhage. (A, B) Right carotid angiography shows left persistent primitive hypoglossal artery (arrows) and the ruptured basilar-tip aneurysm (arrowhead). (C) Post contrast axial CT image and (D) 3D reconstructed CT angiography images reveal persistent hypoglossal artery running through the hypoglossal canal (C; arrowhead, D; blackarrow). Transcatheter embolization was performed via carotid basilar anastomosis with femoral arterial approach. The aneurysm was successfully packed with coils (D; blackarrowheads).
Fig. 6: Schematic demonstration of hypoglossal artery and proatlantal artery

Fig. 7: Otoscopic findings obtained from 71-year-old female with mild hearing loss. (A) Right tympanic membrane is normal. (B) Left tympanic membrane is reddish and bulging.
**Fig. 8:** (A), (B) axial view and (C) coronal view of post contrast CT show that abnormal course of the left internal carotid artery which protruding into the middle ear. (A, B; arrow C; arrowhead). (D) Frontal view of left carotid angiography reveals an unusual course of petrous ICA, which runs with forming convex shape to lateral side (white arrow).
Fig. 9: Schematic demonstration of developmental mechanism of intratympanic ICA
Fig. 10: Aberrant retropharyngeal/lateral pharyngeal ICA. A 57-year-old woman with a meningioma. Preoperative axial postcontrast CT scan shows bilateral internal carotid artery running along the posterior pharyngeal wall near the midline (A, B; arrows). The common carotid angiography reveals an unusual course of the cervical ICA, which runs with forming convex shape to medial side (C, D; arrowhead). Right ascending pharyngeal artery originates from right cervical ICA (C; arrow).
Fig. 11: Non-bifurcating carotid artery which ascends up straightly. A 71-year-old man present with chronic aortic dissection at the thoracic aorta. Magnetic resonance (MR) angiography for preoperative screening reveal a dural AVF with cortical venous drainage at the right transverse sinus. Right carotid angiography in the (A) anteroposterior view and (B) lateral view show the non-bifurcating carotid artery. Several branches, such as the internal maxillary artery, occipital artery, lingual artery, facial artery and internal maxillary artery, originate independently from the single carotid artery. (C) 3D image of the right common carotid angiography shows the dural AVF (asterisk) and clearly demonstrates the separate origin of the main branches of the external carotid artery from the carotid artery.
Fig. 12: Non-bifurcating carotid artery with an arterial stump of the carotid artery. A 39-year-old man with a tumor of maxillary sinus. The carotid angiography in the (A) anteroposterior view and (B) lateral view show the non-bifurcating carotid artery. There are the arterial stump in location which the primary ICA originates (arrow). (by courtesy of Dr.Y.Akiyama, department of Neurosurgery, Tenri Hospital, Tenri, Nara, Japan)
**Fig. 13:** Non-bifurcating carotid artery with the tortuosity of the carotid artery. A 79-year-old man underwent carotid artery stenting. Contralateral carotid angiography in the (A) anteroposterior view and (B) lateral view shows the non-bifurcating carotid artery with an arterial stump (arrow). The carotid artery runs with configuration projecting caudal (arrowhead). (by courtesy of Dr. A. Hyodo, department of Neurosurgery, Dokkyo Medical University Koshigaya Hospital, Koshigaya, Saitama, Japan)

**Fig. 14:** Schematic view of maldevelopment in the non-bifurcation carotid artery
Fig. 15: Schematic demonstration of developmental mechanism of the non-bifurcating carotid artery via the communicating arteries of hypoglossal/proatrantal system with abnormal regression of 3rd aortic arch.
Conclusion

Recent developments in imaging modalities allow us to obtain detailed structures of the carotid artery system. As a result, the anatomic variations of the ICA could be often observed in clinical practice. Several cerebrovascular lesions that coexist with anatomic variations of the ICA have been reported. The variations of cervical-petrosal ICA can be attributed to a maldevelopment of the vascular network during embryogenesis. The knowledge of these variations from the point of embryology is essential for the diagnosis and surgical/interventional procedure in Head and Neck.

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References


