“Duplicated” or “Multiple” Cervical Internal Carotid and Vertebral Arteries from Fenestration, Duplication and Vasa Vasorum to Segmental Rete

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Summary

The appearance of multiple cervical arteries may be due to a variety of anatomic situations. Arterial fenestrations and duplications have different anatomic origins, with distinct appearances on angiography. They are associated with incomplete segmental development.

The vasa vasorum of the internal carotid artery are rarely seen unless enlarged in pathological situations. They represent a peculiar segmental, in-situ, collateral circulation. Retes, on the other hand, correspond to a more complex reconstitution of an early segmental regression. Careful analysis of each of these features is required to choose the best appropriate terminology. The purpose of this paper is to report illustrative cases to enhance the distinctive features of each disposition.

Introduction

Arterial fenestration and duplication, enlarged vasa vasorum and arterial rete - can all give the appearance of multiple arteries (or lumens) along the course of cervical arteries. Duplication recruits two distinct arterial channels; it occurs when two embryologically separate vessels persist or remain unfused. Fenestration involves two lumens corresponding to a single

arterial segment or vessel. It can be related to a transarterial course of a peripheral nerve or a bony structure. Vasa vasorum is a normal arterial network in the adventitia of normal large arteries, which contributes to the nourishment of the arterial wall.

When enlarged, these arteries may appear as multiple channels along the path of the artery they follow. Retes, on the other hand, correspond to a more complex reconstitution of an early segmental regression. The vertebral artery (VA) and transcranial internal carotid artery (ICA) can harbour such variations. The purpose of this paper is to report illustrative cases to enhance the distinctive features of each disposition in both systems.

Illustrative Cases

Case 1 Fenestration of the VA (figure 1)

A 72-year-old woman presented with an intracerebral parenchymal haematoma for which diagnostic cerebral arteriography was performed. The angiogram revealed no significant vascular pathology, except for an unrelated fenestration involving the VA at the level of the third and fourth cervical spaces. In this case, the two separate lumens can be seen running along the path of two consecutive VA segments and remaining inside the vertebral canal.

Figure 1 Vertebral subtraction angiogram shows slit-like vertebral fenestration at the level of the third and fourth cervical spaces (arrows).

Case 2 Fenestration of the cervical ICA (figure 2).

Selective ICA arteriography performed in a 57-year-old patient (the clinical history was not available as the patient's file was from another

hospital and not available for review) revealed an arterial fenestration of the cervical segment. The corresponding source images of the patient's magnetic resonance angiogram (MRA) clearly showed two separate lumens within the same vascular structure.

Case 3 Duplication of the VA (figure 3)

A 51-year-old patient was diagnosed with a soft tissue arteriovenous malformation of the cervico-occipital region. The patient also had congenital block vertebrae (involving the first two cervical vertebrae) and occipitalisation of the first cervical vertebra. Arteriography was requested to assess for vascular compression of the VA within the vertebral canal during flexion and extension of the neck. There was duplication of the left VA at the craniocervical junction. In this case, one of the arterial channels leaves the vertebral canal to take a subarachnoid course within the spinal canal; the other artery remains extradural and follows the usual path of the VA within the vertebral canal. The intradural vessel at this level corresponds to the lateral spinal artery and accompanies cranially the spinal accessory nerve (XIth cranial nerve).

Case 4 Vasa vasorum of the cervical ICA (figure 4)

This 48-year-old patient presented with history of familial, multiple paraganglioma, including bilateral tympanic and vagal tumours. An-

Figure 2 A) Right internal carotid injection lateral projection and B) MRA source image at the corresponding level show fenestration of the right cervical ICA. The two lumens shown on angiography (arrow) are seen to lie within the same vascular structure (double arrow).

giography was performed for the purpose of pre-operative embolization of the tumours. There was a large vagal tumour supplied by an artery arising from the proximal intrapetrous segment of the ICA. This artery ran retrograde along with the ICA within the carotid canal, and caudally anastomosed with a branch arising from the ascending pharyngeal artery, which supplied the inferior aspect of the tumour. This artery running on the outer surface of the ICA is likely to represent an enlarged artery of the vasa vasorum. In this case, the artery was opacified from the site of origin of the caroticotympanic artery (embryonic hyostapedial artery remnant). Its enlargement was felt to be related to the angiogenic activity of the tumour and its effect on the wall of the cervical ICA.

Case 5 Rete mirabile formation in segmental ICA agenesis (figure 5)

A 30-year-old man presented with an unruptured basilar tip giant aneurysm for which diagnostic cerebral arteriography followed by endovascular embolization of the aneurysm was performed. Left ICA angiography revealed segmental agenesis of the clinoid and terminal segments of the ICA. There was reconstitution of distal flow with typical rete mirabile formation, an arterial network at the skull base.

Discussion

A) Arterial fenestration and duplication

There have been many reports in the literature of arterial fenestration and duplication in the head and neck region. These may represent true duplications or merely be arterial slits or fenestrations, often described with incorrect terminology.

Arterial duplication recruits two different channels, occurring when two embryonic vessels persist or remain unfused¹. As shown in our third illustrative case (figure 3), in duplication of the VA, one of the vessels leaves the vertebral canal to enter the spinal canal hence following an intradural path, while the other vessel which arises from the original segmental artery remains within the vertebral canal. Likewise in duplication of the cervical ICA, which starts just distal to the origin of the artery and extends to the petrous portion, this involves two different embryonic vessels. The “accessory

Figure 3 3D vertebral angiogram shows duplication of the vertebral artery at the craniocervical junction. One of the vessels takes a subarachnoid course (double arrow), the other remaining extradural (arrow).

channel” corresponds to the tympanic branch of the ascending pharyngeal artery, which ascends in the same cervical space as the normal ICA. This “accessory channel” then enters the tympanic cavity and anastomoses with the hyoid remnant of the intrapetrous ICA. In such duplication, the medial ramus corresponds to the ICA itself, while the lateral ramus corresponds to the tympanic branch entering the tympanic cavity.

Fenestration, on the other hand, involves two lumens corresponding to a single path or vessel. This split of a single arterial lumen may be complete or incomplete, usually involving a single or few consecutive segments of the artery. The two lumens are seen to run in the same space together (case 2); fenestration may sometimes be barely visible (case 1). Vertebral artery fenestrations, however, should be differentiated from so-called basilar and anterior

Figure 4 A) Axial T2 weighted MRI reveals large right-sided vagal tumour (arrows) supplied by artery shown on B) 3D angiography, arising from the proximal petrous segment of the ICA.

spinal artery fenestrations: although basilar and anterior spinal artery fenestrations appear as slits within a supposed single vessel, these correspond to unfused segments of the paired ventral longitudinal arteries. Conversely, VA fenest-

trations arise as a developmental variant between the intersegmental arterial channels, which contribute to the formation of the artery.

Enlarged random anastomoses may also produce false arterial duplications: for example at

Figure 5 Left internal carotid subtraction angiogram A) lateral and B) frontal projections. There is segmental agenesis of the clinoid and terminal portions of the ICA with rete mirabile reconstitution of the distal flow.

Figure 6 A) Right ICA angiogram shows an enlarged fusion type of ACA at the anterior communicating artery complex (arrow), with associated infra-optic course of the right A1 segment. B) Right ICA angiogram shows an accessory MCA, often called the "duplicated" MCA, which corresponds to an enlarged Heubner artery sharing the cortical territory. C) Left VA angiogram shows a short left PcomA with early branching into P1 and P2, giving "pseudo-duplicated" appearance (arrow). D) Enlarged perforator anastomosis (arrow) at the proximal M1 segment shown on right ICA angiogram and E) similar disposition of perforator anastomosis at the basilar tip.

Figure 7 A) Common carotid angiogram shows carotid branch of the ascending pharyngeal artery within the carotid canal, anastomosing with the recurrent artery of the foramen lacerum (arrows). B) Schematic representation.

the circle of Willis where perforators anastomose prior to entering the brain substance (basilar tip, A1 or M1 segments) (figure 6D,E).

The pre-medullary, intracranial “duplication” of the VA at the vertebrobasilar junction actually represents an enlarged pre-medullary bridge. This is neither a fenestration, nor is it a duplication involving the persistence of two different embryonic vessels, but an enlarged pre-fusion type of anastomosis. A similar disposition is seen at the anterior communicating artery complex (figure 6A).

Posterior communicating artery (PcomA) duplication corresponds either to an enlarged anterior choroidal artery retaining its cortical territory or to a short PcomA with an early branching into P1 and P2. (figure 6C) Similarly, the so-called duplicated middle cerebral artery (MCA) or accessory MCA corresponds to shared cortical supply with the Heubner artery² (figure 6B).

B) Vasa vasorum

Normal large cervical arteries have an extensive network of vasa vasorum in the adventitia, which arise from branch points of parent arteries³. Nourishment of arteries is accomplished by simple diffusion from the vascular lumen and from this network of vessels. The vasa va-

sorum can increase blood flow to the artery wall by dilatation of the existing arteries or by formation of new vessels (neovascularisation)⁴. Likewise, there is a network of vasa vasorum around the ICA. This has been described in cases of ICA occlusion or proximal surgical ligation where the enlarged vasa vasorum arteries form a single or multiple channels within the carotid canal, to vascularise and finally reconstitute the distal artery⁵. Atheromatous disease is known to stimulate the development of vasa vasorum extending to the ICA anterior division and basilar tip. In our illustrative case 4 (figure 4), the artery supplying the tumour is probably an example of a single enlarged artery of the vasa vasorum triggered by the tumoral angiogenetic activity. The various embryonic arteries related to each ICA segment contribute to the supply of the vasa vasorum, to finally constitute the continuous network running along the wall of the cervical and transcranial ICA. The junction between two consecutive ICA segments also provides branches involved in the vasa vasorum supply and is anastomosed with an ascending pharyngeal branch:

- The recurrent artery of the foramen lacerum 2 arises from the lateral surface of the vertical portion of the cavernous carotid siphon (junction between segments 4 and 5), and descends into the foramen lacerum. It supplies

the pericarotid autonomic plexus and is likely to contribute to the vasa vasorum. Within the carotid canal, it anastomoses with the carotid branch arising from the superior pharyngeal branch of the ascending pharyngeal artery (figure 7).

- The mandibular artery, which is the remnant of the first aortic arch, arises from the petrous ICA (junction between segments three and four). It anastomoses with the superior pharyngeal and carotid branches of the ascending pharyngeal artery⁶.

- The caroticotympanic artery (junction between segments two and three) anastomoses with another carotid contributor of the ascending pharyngeal artery entering the carotid canal (figure 4).

C) The rete

Arterial fenestrations and duplications in humans are anomalies, albeit pointing to an incomplete maturation process of the arterial wall, as in the association with intracranial berry aneurysms⁷. On the other hand, in some species, the persistence of multiple vascular channels is a normal occurrence. In the pangolin, for example, persistence of many channels within the path of the cervical VA gives rise to an arterial network (the cervical vertebral rete)⁸. The rete mirabile, on the other hand, is a distinct entity also present as a normal vascular network in some vertebrates. This rich arterial network has, however, been de-

scribed in the literature in association with segmental regression of the carotid artery in man⁹. The rete mirabile is typically located at the skull base, has contributions from external carotid branches, and reconstitutes the ICA distal to the missing segment(s). (Illustrative case 5) The appearance is that of an arterial network at the skull base, rather than multiple parallel vascular channels along the path of an embryonic artery as seen in duplications and fenestrations. This rete mirabile is a response to the early regression of the ICA.

Conclusions

The appearance of multiple cervical arteries may be due to a variety of anatomic situations. Arterial fenestrations and duplications have different anatomic origins, with distinct appearances on angiography. They are associated with incomplete segmental development. The vasa vasorum of the ICA are rarely seen unless enlarged in pathological situations. They represent a peculiar segmental, in-situ, collateral circulation. Finally, retes correspond to a more complex, although primitive appearing, reconstitution of an early segmental regression. Careful analysis of each of these features is needed in order to choose the most appropriate terminology.

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