Agenesis and hypoplasia of the internal carotid artery are rare congenital anomalies, occurring in less than 0.01% of the population [1]. “Agenesis” can be defined as total failure of an organ to develop embryologically; “aplasia,” as lack of development despite the presence of a precursor; and “hypoplasia,” as incomplete development of the organ [2]. These terms are used interchangeably despite their different definitions, and the term “absent internal carotid artery” is preferred for simplification.

The ophthalmic artery as the first major branch of the internal carotid artery typically originates under the lateral aspect of the optic nerve and passes through the optic canal. Abnormal origins of the ophthalmic artery are rare and vary depending on the fetal anastomoses established by the artery with the adjacent vessels. We report an unusual case of unilateral internal carotid artery absence associated with anomalous origin of the ophthalmic artery from the ipsilateral posterior communicating artery.

Case History

A 38-year-old woman with acute right-sided headaches but no neurologic deficit underwent duplex sonography, which raised the suspicion of an acute ipsilateral internal carotid artery occlusion. To exclude an acute dissection, we performed cerebral angiography, which surprisingly revealed an absent right internal carotid artery with an anomalous origin of the ipsilateral ophthalmic artery from the posterior communicating artery (Fig. 1). The right middle cerebral artery was directly supplied by the posterior circulation through a hypertrophied posterior communicating artery, whereas the right anterior cerebral artery was supplied through a patent anterior communicating artery. Except for a direct origin of the left vertebral artery from the aortic arch, the remaining extra- and intracranial vasculature was otherwise normal.

Discussion

Absence of internal carotid artery can be observed as a single anatomic variant or in association with complex vascular malformations such as in patients with PHACE syndrome [3]. It may be asymptomatic if sufficient collateral circulation exists but may also be associated with transient ischemic attacks, hemiplegia, and intracranial hemorrhage [1]. On the basis of his observations, Lie [2] described two basic patterns of collateral circulation in association with absence of the internal carotid artery. The fetal type, seen in nearly all cases, develops during an early embryogenic stage. The anterior cerebral artery is supplied by the contralateral internal carotid artery via the anterior communicating artery, whereas the middle cerebral artery is supplied by an enlarged posterior communicating artery as in our patient. The second, the adult type, is believed to occur later in embryogenesis and after completion of the circle of Willis. Given et al. [4] recently described six possible patterns of collateral circulation, surprisingly without including the ophthalmic artery; little is known about its anomalous origin and collateral flow to the optic organ in case of internal carotid artery absence.

According to Padget [5], the internal carotid artery originates from the dorsal aorta and the third aortic arch at approximately the 3-mm embryonic stage, but complete development does not occur earlier than the 16–18 mm stage (40 days). So far, there is no exact explanation for developmental anomalies of the internal carotid artery, but all variations are thought to occur because of insults to the developing embryo. Keen [6] suggested that...
mechanical insults to the developing embryo such as excessive folding of the embryo to one side, pressure effects, or restriction by amniotic bands may cause unilateral absence of internal carotid artery.

The ophthalmic artery has a complex embryogenesis, which is closely related to the development of the internal carotid artery. As described by Padget [5], the primitive internal carotid artery in a 4-mm embryo gives rise to its cranial and caudal branches and supplies the orbit with two main primitive arteries. The dorsal ophthalmic artery arising from the future internal carotid artery siphon reaches the orbit through the superior orbital fissure. The ventral ophthalmic artery, arising at that time from the anterior cerebral artery, reaches the orbit through the optic canal and supplies the optic tract [7]. By the time the embryo reaches 18 mm, the dorsal ophthalmic artery and the ventral ophthalmic artery develop an anastomosis with each other and eventually terminate in a plexus that supplies the optic cup. At the same time, the ventral ophthalmic artery forms an anastomosis with the internal carotid artery and its proximal segment regresses. The stem of the permanent ophthalmic artery, also called primitive ophthalmic artery [7], is first identified arising from the supracavernous internal carotid artery and reaches its

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Fig. 1.—38-year-old woman with acute headaches. 
A, Arteriogram shows aortic arch, left anterior oblique projection, with origin of common carotid artery (single long arrow) from right subclavian artery (brachiocephalic trunk) giving rise to external carotid artery only. Double arrow indicates left vertebral artery; short arrow indicates left vertebral artery directly arising from arch. 
B, Right external arteriogram, lateral view, shows no anastomosis to internal carotid artery territory. Particularly, middle meningeal artery does not supply ophthalmic artery. 
C and D, Arteriograms, right vertebral artery injection, lateral (C) and anteroposterior (D) views, show origin of right ophthalmic artery (arrows) from posterior communicating artery. Arrowheads (C) indicate choroidal blush.

(Fig. 1 continues on next page)
Agenesis of Internal Carotid Artery

definite origin from the future C2 or C3 segment (according to Fischer’s [8] classification) by considerable caudal migration. The dorsal ophthalmic artery starts to regress and will be finally reduced to the anteromedial branch of the inferolateral trunk. (In this stage, the stapedial artery originating from the primitive hyoid branch of internal carotid artery gives off two branches: the maxillomandibular and the supraorbital arteries. When the embryo reaches 20 mm, the supraorbital branch of the stapedial artery forms an anastomosis with the permanent ophthalmic artery. Later, at the 40-mm stage, the anastomosis is complete and the adult configuration of the ophthalmic artery is identifiable.

Anomalous origins of the ophthalmic artery from the posterior communicating artery are very rare with four such variants reported so far [6, 9–11], of which the only one proven by cerebral angiography is published in the non-English literature [11]. The most commonly reported variants are the origins from the middle meningeal [12] and the anterior cerebral [7] arteries. Other possible origins are the accessory meningeal artery, the basilar artery [7], and possibly the middle cerebral, the anterior deep temporal, and the external carotid arteries [12].

The first observation of a posterior communicating artery origin was made by Fisher [9] in 1914. He described a bilateral absence of both internal carotid arteries in a cadaver and showed the origin of the ophthalmic artery from the posterior communicating artery. Keen [6] described agenesis of both internal carotid arteries in a 55-year-old man with a marked enlargement of both the vertebral and the basilar arteries. Each posterior cerebral artery gave rise to an enlarged posterior communicating artery, which itself gave off small ophthalmic arteries on either side. Hills and Sament [10] reviewed the findings in a 10-week-old infant who died because of severe cardiac anomalies. Both internal carotid arteries were absent at the base of the brain. The posterior communicating artery gave off ophthalmic arteries and continued forward to bifurcate into the anterior and middle cerebral arteries.

So far, the only radiographically visualized case of unilateral internal carotid artery absence and posterior communicating artery origin of ophthalmic artery has been reported by Nakata and Iwata [11] in a 41-year-old woman with sudden onset of headache and fever. Cerebral angiography revealed complete absence of the left internal carotid artery and an enlarged left posterior communicating artery, which gave off the left ophthalmic artery. The embryologic explanation for an origin from the posterior communicating artery remains speculative but probably needs involvement of internal carotid artery as a coexisting factor. The type of collateral flow pattern suggests an interruption before the circle of Willis is completed (24-mm stage). Probably, the ventral ophthalmal artery had stopped migrating caudally at the segment where the posterior communicating artery joins the M1 segment of the middle cerebral artery (usually the terminal internal carotid artery segment).

Otherwise, as discussed in the case of Nakata and Iwata [11], a part of the posterior communicating artery in our patient may actually belong to the internal carotid artery, and the dominant collateral flow through the posterior communicating artery plays a major role in determining it as an origin for the ophthalmic artery.

Knowledge of anatomic variants in the cerebral circulation such as internal carotid artery absence and their association with anomalous origins of the ophthalmic artery may be helpful in avoiding misinterpretation of clinical and imaging patterns in ischemic stroke. In patients undergoing carotid endarterectomy, unilateral supply of brain territories can become crucial. Furthermore, it may also help to differentiate a questionable internal carotid artery dissection or occlusion from a simple anatomic variation without clinical consequence.

Conclusion

Association of an absent internal carotid artery with a posterior communicating artery origin of the ophthalmic artery is extremely rare. We provide the first angiographic documentation in the English literature and discuss related embryologic theories.

References


Fig. 1. (continued)—38-year-old woman with acute headache. E, Schematic drawing shows anatomic disposition in presented case. MCA = middle cerebral artery, ICA = internal carotid artery, PcomA = posterior communicating artery, MMA = middle meningeal artery, OA = ophthalmic artery, ECA = external carotid artery, and IMA = internal maxillary artery.
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