A Rete Mirabile of the Posterior Cerebral Artery

Embryonic and Phylogenetic Remnant of Choroidal Artery Origin

L. ZHENG, R. METCALFE*, JJ BHATTACHARYA

Departments of Neuroradiology and Neurology, Institute of Neurological Sciences, Southern General Hospital, Glasgow, UK*

Key words: rete mirabile, arteriovenous malformation, anatomic variants, anterior choroidal artery, posterior cerebral artery

Summary

We present the case of a 41-year-old man with a vascular network associated with the left posterior cerebral artery (PCA). Although initially considered to be a brain AVM, further investigation showed no arteriovenous shunting and a rete mirabile of the PCA was diagnosed. A well known but rare occurrence mainly in the anterior circulation, rete mirabile has not previously been described in the PCA.

The PCA is a composite vessel, developmentally. Its proximal portion derives from the posterior division of the ICA while its cortical territories are annexed from the anterior choroidal artery adjacent to the lateral geniculate body. We propose that this rete (or network) represents a persistence of the embryonic anastomosis between the AChA and the PCA.

Introduction

The rete mirabile of the brain holds occupies an almost legendary place in the history of neuroscience¹. Described by Galen, this supposed network separating the extracranial from the intracranial internal carotid artery was responsible for converting the 'vital spirit' into the 'animal spirit'. The presence of the rete in man was accepted in Western medicine until the renaissance when it was refuted by Vesalius. The rete mirabile is well recognized in oth-

er species² and indeed the rete at the skull base of the pig, supposedly involving the internal carotid artery but actually lying on the ascending pharyngeal artery, is familiar to interventional neuroradiologists as an in vivo model for testing embolic agents³. Intracranial arterial retes, or vascular networks, continue to excite interest as rare anatomic variants usually associated with segmental agenesis of a parent artery. We present a patient with an unusual vascular pattern of the posterior cerebral artery (PCA) that was initially suspected to be an arteriovenous malformation. Further investigation revealed this to be an arterial network, or rete mirabile, in the course of the PCA which to the best of our knowledge has not been described before. A plausible embryological reason for such a network at this specific site would help to confirm this as an anatomic variant rather than an acquired pathological defect. We seek here to offer such a phylogenetic and embryological explanation.

Case Report

A 41-year-old engineering manager presented with an episode of hemiparesis of the right leg and tingling sensation affecting the right side of his face and leg lasting 30 seconds. These were followed by more persistent tingling in the right foot and cheek. Neurological examination was normal. His symptoms were

Figure 2 Schematic illustration of the neural tube of early human embryo (Carnegie stage 6, 18-26mm) showing the limbic arterial arch around the telencephalic vesicle, formed by the ACA anteriorly and the PCom posteriorly. As the developing telencephalic vesicle (cerebral hemisphere) enlarges posteriorly, new sources of blood supply form anastomotic networks and subsequently replace the PCom cortical territory. These derive from the posterior choroidal artery and the PCA represented at this stage by the tectal arteries.

thought to be due to transient ischaemic attacks or migraines. His neck vessels were normal and there was no evidence of cerebral ischaemia. MRI and MRA did however show a small cluster of abnormal vessels arising from the posterior cerebral artery in the left ambient cistern (figure 1), suggestive of an AVM. Digital subtraction angiography was performed which again demonstrated the network of vessels arising from the PCA, but no draining vein was identified. Was this an AVM for which treatment would be considered or a benign anatomical variant which could be left alone? Superselective angiography under local anaesthetic was performed by means of a microcatheter in the left PCA which finally excluded the presence of any arteriovenous shunting

confirming the abnormality to be a purely arterial network. It was unlikely that the patient's symptoms were caused by this vascular anomaly, and they were attributed to migrainous equivalents for which prophylactic Propranolol was initiated, with no further episodes on follow-up.

Discussion

A rete mirabile may be defined as a vascular network interrupting the continuity of a vessel, just as the great Niger River in Mali breaks into the myriad channels of its inland delta before reforming a single channel below Timbuktu⁴. In humans there are no such normally occurring arterial networks in the brain. The long

Figure 3 Example of a persistent cortical territory of the AChA preventing safe embolization of an AVM fed by the AChA. A) Contrast-enhanced CT brain scan showing a left periventricular haematoma and the AVM nidus (arrow) in the ambient cistern. B) Carotid angiogram in early arterial phase demonstrates the nidus supplied by the AChA (arrow) and shows the anterior choroidal artery continuing on to supply the occipital lobe (arrowheads).

held and erroneous belief in the miraculous network, or rete mirabile, proclaimed by Galen¹ and even traceable back to Aristotle, most likely results from observations in oxen and other animals, which unlike human cadavers, were then available for anatomic dissection. Vesalius

was among the first to describe the integrity of the internal carotid artery from its extracranial to its intracranial portions and refute a thousand years of anatomical dogma.

Pathological networks, however, are well recognized occurring most prominently in moyamoya disease⁵. In this situation a collateral network (involving perforating arteries) bypasses an acquired stenosis distally reforming distally the main arterial trunk. A similar pattern may be encountered in proliferative angiopathy, a type of diffuse brain AVM. In both conditions, the arterial stenosis causes local ischaemia evoking an angiogenic response which may result in sprouting or non-sprouting angiogenesis.

Our patient was striking for the normality of his brain and other blood vessels with no suggestion of ischaemic lesions. The absence of a draining vein also excludes arteriovenous shunting. These observations make an anatomical variation the most likely cause of the network of blood vessels. Variants do however follow a logic imposed by embryology and evolution. Thus fenestration of a vessel occurs when an anatomical structure 'pierces' the course of a developing artery, with both lumens corresponding to a single arterial path. Conversely, duplication of a vessel indicates persistence of two distinct anatomic channels as seen when incomplete fusion of the longitudinal neural arteries results in basilar artery duplications [6]. In our case, the posterior cerebral artery, rather than simply dividing into two channels becomes more irregular and reveals a more complex network. Is there a developmental explanation for such an appearance at this particular site in the ambient cistern adjacent to the lateral geniculate body? Is its appearance mere serendipity or testament to hundreds of millions of years of evolution of the vertebrate hemisphere?

To seek a plausible explanation we must return to the early embryo. At the start of the 5th fifthweek of human development (4-5 mm embryo) the primitive internal carotid arteries form from the 3rd third aortic arches. At the base of the optic vesicle, the carotid bifurcates into an anterior division (AD), the future anterior cerebral artery from which will arise the middle cerebral artery will arise, and a posterior division (PD), the future posterior communicating artery (PComA)⁷. This important division is replicated across the vertebrate phylum

Figure 4 Schematic illustration of the relationship between the anterior choroidal (large arrows) and posterior cerebral arteries (small arrows) seen from above. A) The occipital cortical territory originally belongs to the AChA. Anastomoses develop between AChA and developing PCA and posterior choroidal arteries. B) Normal adult configuration. The occipital artery territory has been completely annexed by the PCA (arrows). C) In our patient annexation of the cortical territory has successfully occurred but the anastomotic network has not fully regressed leaving a rete mirabile, shown here in simplified form (arrow).

and represents the true ICA bifurcation, in adult humans as in other vertebrates at the level of the PComA. Indeed the same pattern is apparent in fish whose evolutionary lineage diverged from ours almost 400 million years ago⁸. The AD curves round the optic, and then the telencephalic vesicles as the anterior cerebral artery (ACA). The posterior division initially only supplies the mesencephalon and tectum but during the fifth week anastomoses with the longitudinal neural arteries to form the PComA. By week six, the AD of the ICA forms an arterial ring, the so-called limbic loop, around the telencephalic vesicle (figure 2). Moffat showed this pattern to be present in the rat as well as human embryos⁹.

The posterior part of this loop represents the earliest appearance of the anterior choroidal

artery (AChA). Together the ACA and AChA form the primitive arterial supply to the whole of the telencephalon prior to enlargement of the MCA (a branch of the ACA). Thus the AChA supplies a large area of the posterome-

dial cortex in addition to the choroid plexus and diencephalon. As the cerebral hemispheres enlarge posteriorly, further sources of arterial supply are tapped. A network of anastomoses develops between the AChA and the posterior choroidal artery (PChA) and future PCA, a branch of the PD at the diencephalic level, close to the lateral geniculate body¹⁰. As the vertebral arteries form and fuse with the basilar artery, there is a reversal of the original caudal flow in the BA. At the same time there is shrinkage of the PComAs and a progressive transfer of the cortical territory of the AChA to the PChA and developing PCA. The AChA thus comes to supply mainly the choroid plexus

with a small residual cortical supply mainly to the mesial temporal lobe¹¹.

Incomplete transfer of territories from the AChA to the PCA may leave the AChA with a cortical supply to the occipital lobe (figure 3). Persistence of anastomoses around the lateral geniculate body provides a collateral pathway from the AChA if the proximal PCA is occluded (figure 4).

A more complete persistence of the anastomotic network with failure of the PCA to establish a dominant channel could be expected to leave a rete mirabile in the course of the PCA adjacent to the lateral geniculate body as in our case.

References

- 1 De Gutierrez-Mahoney CG, Schechter MM: The myth of the rete mirabile in man. *Neuroradiology* 1972; 4: 141-158, 1972.
- 2 Kimani JK, Opole IO: The structural organization and adrenergic innervation of the carotid arterial system of the giraffe (*Giraffa camelopardalis*). *Anat Rec* 230: 369-377, 1991.
- 3 Klisch J, Requejo F et Al: The two-in-one model. A new variation of the arteriovenous malformation model in swine. *Neuroradiology* 43: 393-397, 2001.
- 4 Niger River inland delta. 14° 52' North, 4°20' West. <http://earth.google.com/intl/en/>.
- 5 Okayasu I, Kajita A, Yamamoto M: Abnormal intracranial vascular networks ('moyamoya' disease), possibly due to occlusion of bilateral internal carotid arteries –a case report with histometrical analysis. *Acta Pathol Jpn* 26: 105-114, 1976.
- 6 Lasjaunias P, Berenstein A, terBrugge KG: Surgical Neuroangiography. Vol 1 Chapter 3. Springer-Verlag. Berlin Heidelberg, 2001.
- 7 Padget DH: The development of the cranial arteries in the human embryo. *Contributions to embryology* 212: 205-261, 1948.
- 8 Allman J: *Evolving Brains*. Scientific American Library. New York, 1993.
- 9 Moffat DB: The development of the posterior cerebral artery. *J Anat* 95: 485-495, 1961.
- 10 Abbie AA: The morphology of forebrain arteries, with especial reference to the evolution of the basal ganglia. *J Anat* 68: 433-470, 1933.
- 11 Rhoton AL Jr, Fujii K, Fradd B: Microsurgical anatomy of the anterior choroidal artery. *Surg Neurol* 12: 171-187, 1979.

Dr JJ Bhattacharya
Dept of Neuroradiology
Institute of Neurological Sciences
Southern General Hospital
1345 Govan Road
Glasgow G51 4TF, UK
E-mail:
j.j.Bhattacharya@clinmed.gla.ac.uk

EDITORIAL COMMENT

The manuscript provided by Zheng et Al provides a useful and interesting assessment of a much debated topic, the existence of retia networks in human. The discussion of the well explained embryological-phylogenetic background is informative and much amplified by the inclusion of appealing sketches that are valuable adjuncts to understanding the topic. The only minor reservation to this otherwise excellent contribution is whether the authors should label this “abnormal” network as a rete. The attribution of a rete mirabile at this level has not previously been mentioned in the literature and might better be considered using the terminology of an arterio-arterial network. Since this nomenclature has been utilized by the authors in their introduction, it might be more functional to maintain this term in the final diagnosis.

Given that the phylogeny and embryology of the intracerebral vessels suggest a mechanism related to transposed distributions or “abnormal” collateral networks, it might be informative if this background were employed to further explain the patient reported by the authors.

There is considerable evidence attesting to the fact that during the early stages of fetal development, AchA provides the main blood supply to the postero-medial aspects of the developing cortex. Thus, under circumstances where the telencephalic branches of the embryonic PCA fail to take over this vascular territory, cortical branches of the AchA may persist as the dominant supply to the temporal, parietal and occipital lobes. This phenomenon has led to the observation that under such circumstances a spectrum of anomalies may exist, that range from a single cortical branch originating from AchA to a complete replacement of the PCA by AchA branches. Takahashi when describing the variety of phenotypes of AchA hyperplasia was in reality delineating the different PCA territories that may be taken over by AchA and subcategorizing the variations that lie between the two extremes. (Takahashi S. et Al, 90, Abrahams J. et Al, 99, Lasjaunias P. et Al 01).

In some cases that have been reported, a persistent communication between PCA and AchA has also been demonstrated (Patient No 4 in (Abrahams J. et Al 99).

In general, anastomoses are located at the Pcom, interpenducular, choroid plexus, temporal lobe and lateral geniculate body. (Takahashi et Al 90, Lasjaunias et Al 01).

There seems to have been no evident trigger from the unremarkable- in relevance to ischemic lesions- brain parenchyma that might allow for the postulate that this network could be an acquired pathological defect.

Since the PCA had successfully achieved annexation of its cortical territory, it seems unlikely that a segmental agenesis or regression might be responsible for the development of a possible “rete”.

More generally, the concept of the variability in men is based on phylogenetic steps that represent a series of discrete the milestones in embryonic development and can globally be viewed as a path in regards to individual human disposition. The functionality of a partic-

ular or specific disposition includes several patterns compatible with a normal brain activity. In this capacity the vascular system can be considered as a provider of blood and therefore the final possible permutations and commutations of the arterial trees reflect the multiple options which are all compatible with normal cortical function/perfusion.

The embryology of the vascular supply to the limbic system is in this matter, exceptionally illustrative. All the possible steps between AChA supply, as seen in the opossum for example, to the PCA predominance, in man (in equilibrium with ACA contribution), are demonstrable. Retes per se are different in that they represent dispositions which are often erroneously considered as an “archaic” arrangement, yet are evident in such advanced species as dolphins where they are particularly numerous. In such a situation, a rete may be considered as representative of a “modern” vascular architecture adapted to exigencies of the pressure flow variables encountered in the new environmental conditions faced by a “mammal returning to the sea”.

If this proposition is considered viable it would be instructive to rigorously analyze all networks in order to differentiate what might represent a ‘secondary collateral circulation’ leading to the “take over” of supply from one source to another, in contrast to a ‘true rete’ which most likely corresponds to a stable (non-triggered) anatomic option. Given the information presented, it is plausible that the PCA-AChA most likely corresponds to the former interpretation whereas the rete ophthalmicus, mirabilis or vertebralis rather to the second one. Intra cavernous anastomoses variations represent the most typical difference between them as illustrated by: the classic disposition of the ILT and ICA-ECA anastomoses in the greatest majority of human dispositions the cavernous ECA-ICA collateral circulation when ECA is recruited and triggered to “re supply” an occluded ICA the true rete mirabile as encountered in sheep, swine or pig, when associated with an absent (regressed) ICA.

Dr E. Psatha MD

Universitätsspital Zurich

Prof. P. Lasjaunias MD PhD

Centre Hospitalier de Bicêtre

- 1 Suga T, Kawata Y, Sakamoto K: “AChA: Angiographic Analysis of variations and anomalies” Am J Neuroradiol 11: 719-729, 1990,
- 2 Abrahams J, Hurst R, Bagley L, Zager E: “AChA supply to the PCA distribution: Embryological basis and clinical Implications” Neurosurgery 44(6) 1999.
- 3 Lasjaunias P, Berenstein A, terBrugge K: e al Surgical Neuroangiography. Vol 1, 2nd edit. Springer Verlag 2001.