An Orbital Arteriovenous Malformation in a Patient with Origin of the Ophthalmic Artery from the Basilar Artery

M. Schumacher and A. K. Wakhloo

Summary: An abnormal origin of the ophthalmic artery from the basilar artery, found in conjunction with an orbital arteriovenous malformation, is described. The successful treatment of the arteriovenous malformation by embolization through the ophthalmic artery is also reported.

Index terms: Arteries, abnormalities and anomalies; Arteries, anatomy; Arteries, basilar; Arteries, ophthalmic; Arteriovenous malformations, embolization; Growth and development; Interventional neuroradiology, in infants and children.

Arteriovenous malformations (AVMs) of the eyelid and periorbital region are generally supplied by branches of the ophthalmic artery (1, 2). Technical problems and the risk of neuroophthalmological damage make therapeutic embolization through the ophthalmic artery a hazardous procedure. Therefore it is necessary to decide whether embolization or surgery is less risky (3). In the case described here, treatment was further complicated by the fact that the ophthalmic artery, which supplied the lesion, arose from the basilar artery. This variation, its abnormal development, and the endovascular treatment are described.

Case Report

A 3-year-old girl (Fig 1) presented with a huge AVM extending over the right upper eyelid and supraorbital region of the forehead. This malformation had been treated in Vietnam with conventional radiotherapy, and had resulted in blindness and scarring. She was admitted to our hospital for surgical treatment of the malformation and plastic skin reconstructive surgery. A computed tomography scan disclosed the persistence of parts of the AVM in the upper lid and near the lacrimal gland on the right side (Fig 2).

Angiography revealed that the main arterial supply of the lesion was derived from the frontal branch of the superficial temporal, the anterior deep temporal, and the orbital branches of the infraorbital arteries. As a first step, these vessels were embolized preoperatively with Ivalon. Computed tomography revealed a larger AVM than had been expected. A transbrachial retrograde angiogram displayed an artery entering the orbit through the optic canal to supply the AVM, which originated from the basilar artery proximal to its superior cerebellar branch. Subsequent selective injection of the vertebrobasilar system during external compression of the right internal carotid artery
Fig. 3. A, Left vertebral angiogram made during compression of the right internal carotid artery. The abnormal ophthalmic artery and a well-developed posterior communicating artery can be seen.
B, C, Late phase of the vertebral angiogram showing the abnormal ophthalmic artery and the orbital AVM.

Fig. 4. Supraselective angiography of the abnormal ophthalmic artery. Note origin of the middle meningeal artery from the ophthalmic artery (long arrow), and a normally developed retinal artery originating in a typical fashion from the middle segment of the ophthalmic artery (short arrow).

Fig. 5. Postembolization angiogram with intranidal placement of the N-butyl cyanoacrylate (arrows). Displayed a normal posterior communicating artery. It also confirmed that no communication existed between the abnormal ophthalmic artery and the internal carotid artery (Fig 3A). At a later phase of the angiogram, branches from the ophthalmic artery filling the AVM were demonstrated.

Supraselective angiography with the tip of the catheter in the middle segment of the ophthalmic artery revealed two vascular details: a middle meningeal arising from the ophthalmic artery and the origin of the central retinal artery. The tip of the catheter was advanced distal to the origin of the central retinal artery, and the AVM was embolized with 0.4 mL N-butyl cyanoacrylate in a 50% mixture with lipiodol (Fig 5). After this procedure the child’s progress was satisfactory, and plastic surgery could be undertaken.

Discussion
The origin of the ophthalmic artery from the vertebrobasilar system is a variant that requires embryologic explanation. As is recognized for other primitive arteries, early caroticobasilar connections can persist as abnormal variants between the two systems (4). Among these, primitive trigeminal, otic, and hypoglossal arteries should be included, as well as the proatlantic artery. These primitive arteries have the following characteristics in common: they are constantly found to connect the vertebral or basilar artery with the carotid and can be satisfactorily explained embryologically as collaterals between the two primitive carotids. The development and regression of these vessels have been described in detail by Padget (5). The vascular variant we found cannot be fully explained within his scheme of embryologic development for three reasons: 1) the internal carotid artery is in no way involved, 2) the anlage of the definitive ophthalmic artery appears significantly later than the appearance and regression of the primitive arteries described above, and 3) the ophthalmic artery is not formed from one primitive trunk, but from several single vessels, the development of which has been divided into stages by Padget (5).

The development of the definitive ophthalmic artery is not only complicated but also late in
time. In its earlier stages it is like that of the rabbit, which has been well described by Fuchs (6). Whereas the primitive caroticobasilar arteries have regressed at the latest by the 12 mm stage, development of the ophthalmic artery continues almost up to the 40 mm stage, the hyaloid artery being always present as its terminal branch. If one attempts to bring the development of the ophthalmic artery and its supraorbital branches into a definitive scheme, the detailed description by Padget recognizes six different stages (5).

At the 5 mm stage, there are branches of the primitive maxillary artery, a primitive dorsal ophthalmic artery, and a primitive hyaloid artery. With the development of primitive dorsal and ventral ophthalmic arteries, all these vessels end at about the 9-mm stage in a plexus. They give rise to a stapedial branch at about 14 mm, which will in turn give rise at 18 mm to a definitive stapedial artery. This vessel is of decisive importance for the development of the blood supply to the orbit. It is also of decisive importance that by this stage the trigeminal artery has completely regressed. These facts are, in our opinion, proof that the present variant is not dependent upon the embryonic trigeminal artery alone. This is also confirmed by the normal internal carotid, which is not connected with the variant vessel, and which cannot therefore be explained as a "trigeminal type" of vessel (Fig 6B).

The 18-mm stage is decisive for further development because it is here that a definitive stapedial artery has appeared, and a permanent ophthalmic trunk developed out of the dorsal primitive ophthalmic artery. At this stage also the dorsal ophthalmic trunk gives rise to the first ocular branches. At 20 mm a critical union of the trunk of the ophthalmic artery with the stapedial artery takes place. By the mature stage of the 40 mm embryo the original stapedial artery has regressed, with only a supraorbital division remaining that gives rise to the lacrimal and middle meningeal arteries. In our own case it was possible to demonstrate the presence of meningeal branches of the ophthalmic artery, which verifies the embryologic significance of the stapedial artery. Because there is at no stage of its development any direct connection between the stapedial artery and the vertebrobasilar system, it is necessary to postulate a connection that has persisted. A completely unambiguous embryologic explanation does not seem to be possible, and in any case there is the possibility of an abnormal embryonic connection having already been pres-
ent between the primitive trigeminal and the stapedial arteries. This would account for a "stapedotrigeminal" variant, which can also lead to an origin of the ophthalmic artery proximal to the anterior superior cerebellar artery.

An unusual occipital-basilar anastomosis has been reported by Tsai et al (7). They interpreted it as a collateral pathway caused by a congenital absence of both vertebral arteries. Because of an underlying atresia followed by a true collateralization, their case is different from ours, in which all supraaortal vessels were patent. Therefore, a demand for collateralization did not exist. The anatomy of the abnormal orbital-basilar anastomosis in our case also differs from the carotid-superior cerebellar artery anastomosis described by Teal et al (8), insofar as there was no connection with the carotid artery.

The cause of the persisting abnormal embryonic connection described here is unknown. It at least takes over the function of replacing the normal ophthalmic artery because this was not present, as can be seen from the angiogram of the internal carotid artery. This condition corresponds to the other commonly seen persisting embryonic connections which are frequently associated with incomplete development of the circle of Willis (5, 9). The same significance may be attributed to a remaining AVM (10–13), which in our case remained open. It could possibly be this condition that supported the persistence of the embryonic orbital vessel.

Acknowledgment

We thank Prof P. Stoeter of the Department of Neuroradiology, University of Mainz, for his help in explaining the development of the embryonic vascular system.

References