An Additional Variant of the Persistent Primitive Trigeminal Artery: Accessory Meningeal Artery – Antero-Superior Cerebellar Artery Anastomosis Associated with Moyamoya Disease

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Summary

Persistent embryological carotid-basilar anastomoses are rare. There has been no report on persistent anastomosis between the accessory meningeal artery and the antero-superior cerebellar artery. We describe a 3-year-old boy with moyamoya disease manifesting such a congenital vascular anomaly, namely, a large anastomosis between the accessory meningeal artery and the antero-superior cerebellar artery associated with marked hypoplasia of the basilar artery. This persistent vascular anomaly is considered embryologically as an additional variant of the persistent primitive trigeminal artery, i.e., a "*stapedo-trigemino-cerebellar*" variant.

Keywords: Accessory meningeal artery; moyamoya disease; persistent carotid-basilar anastomosis; primitive trigeminal artery.

Introduction

Persistent embryological carotid-basilar anastomoses are rare, but well-known congenital vascular anomalies, which include the persistent primitive trigeminal artery (PTA), otic (acoustic) artery, hypoglossal artery, and pro-atlantic intersegmental artery, although the otic artery has not been clearly demonstrated in the literature. The PTA is most frequently observed among them [13]. Several variants of the PTA have been reported: the middle meningeal artery (MMA) originating from the basilar artery [19], the ophthalmic artery originating from the basilar artery [18], and the cerebellar artery originating from the internal carotid artery [9, 20, 23]. Persistent anastomosis between the accessory meningeal artery (AMA) and the antero-superior cerebellar artery (ASCA) in association with hypoplasia of the basilar artery, however, has not been reported in the literature.

Moyamoya disease presents typical angiographic features: bilateral steno-occlusive changes at the terminal portion of the internal carotid arteries and fine collateral vessels known as moyamoya vessels in the basal ganglia. Only ten cases of moyamoya disease with persistent carotid-basilar anastomosis have been reported in the literature [2, 6–8, 15, 16, 21, 22]. We describe a patient with moyamoya disease manifesting persistent AMA-ASCA anastomosis and discuss the embryological interpretation and clinical importance of this vascular anomaly in association with moyamoya disease.

Case Report

Our patient, a boy, was born at the gestation period of 39 weeks by normal spontaneous delivery with a birth weight of 2,850 g. He had a port wine stain of the left face and naevus spilus on the trunk, but was otherwise healthy at birth. Development was normal until the child was 6 months old, when he developed sudden weakness of the right upper extremity. This subsided spontaneously in a week. One month later, he developed focal seizures and left hemiparesis. He was admitted to a hospital for evaluation. With a diagnosis of moyamoya disease, confirmed by cerebral angiography, encephaloduro-arterial synangiosis (EDAS) on the left side was performed. Six months after this operation, MMA-middle cerebral artery (MCA) anastomosis and EDAS on the right side were carried out. The patient did not have any repeat ischaemic episodes, but did often experience generalized seizures, even with the administration of anti-convulsant drugs.

The boy was admitted to our hospital for evaluation at the age of two. He was hemiparetic on the left and was not ambulatory. Facial port wine stain and trunk naevus spilus were consistent with a diagnosis of *phacomatosis pigmentovascularis* type IIIb [4]. Results of laboratory tests and a cerebrospinal fluid examination were normal. Magnetic resonance images disclosed diffuse atrophy of the right cerebral hemisphere with extensive ischaemic change and dilated
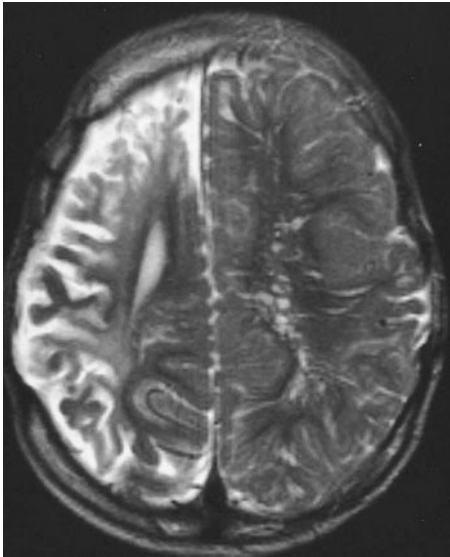
Fig. 1. T2-weighted fast spin-echo image (repetition time: 5000 msec, echo time: 120 msec, echo train length: 15, slice thickness: 6 mm, field of view: 20 cm × 20 cm, excitations: 2) shows marked atrophy of the right cerebral hemisphere with extensive ischaemic change as well as the dilated Virchow-Robin spaces in the left deep white matter

Virchow-Robin spaces predominantly in the deep white matter of the left hemisphere (Fig. 1).

A right internal carotid arteriogram showed marked stenosis at the terminal portion of the carotid artery. The MCA and the anterior cerebral artery were partially visualized. Moyamoya vessels in the basal ganglia were also shown. The right ophthalmic artery was not demonstrated (Fig. 2). A left internal carotid arteriogram showed occlusion of the left internal carotid artery at the level of the ophthalmic artery. Through the collateral from the ethomoidal moyamoya vessels, the basal moyamoya vessels were visualized. The left MCA was not visualized (Fig. 3). A right external carotid arteriogram revealed a persistent large right AMA, anastomosing with the right ASCA (Fig. 4). The right AMA entered the middle fossa and coursed to the posterior fossa near the trigeminal ganglion. Since the basilar artery was markedly hypoplastic, the right AMA supplied the right ASCA and both posterior cerebral arteries (PCAs). There was no distinct left ASCA, but the small arteries originating from the proximal left PCA and the left anterior and posterior inferior cerebellar arteries seemed to supply the territory of the left ASCA through a collateral. Interestingly, the parieto-occipital and calcarine branches of the left PCA were supplied by the right anastomotic collicular artery coursing behind the brain stem. The inferior temporal branches of the left PCA, which had a connection with the distal basilar artery, was predominantly supplied by the right AMA-ASCA anastomosis. The parietal branch of the right MMA supplied the MCA territory to some extent through the surgically established anastomosis. The lacrimal branch of the right MMA supplied the orbit including the retinal circulation, which coursed through the superior orbital fissure. A left external carotid angiogram showed the left MCA territory supplied through the EDAS. A right vertebral angiogram showed solely the right posterior inferior cerebellar artery. A left vertebral angiogram showed the left anterior and posterior inferior cerebellar arteries and the markedly hypoplastic basilar artery, which joined the inferior temporal branches of the left

Fig. 2. Right internal carotid injection (lateral view) demonstrates the steno-occlusive change at the terminal portion of the internal carotid artery with development of the basal moyamoya vessels (arrowhead). There is no ophthalmic artery originating from the carotid artery. Through the moyamoya vessels, the right middle cerebral artery (arrow) is partially visualized. Contrast reflux into the external carotid system is faintly visualized

Fig. 3. Left internal carotid injection (lateral view) shows occlusion of the left internal carotid artery at the ophthalmic artery (arrow). Through the collateral from the ethomoidal moyamoya vessels, the basal moyamoya vessels are visualized
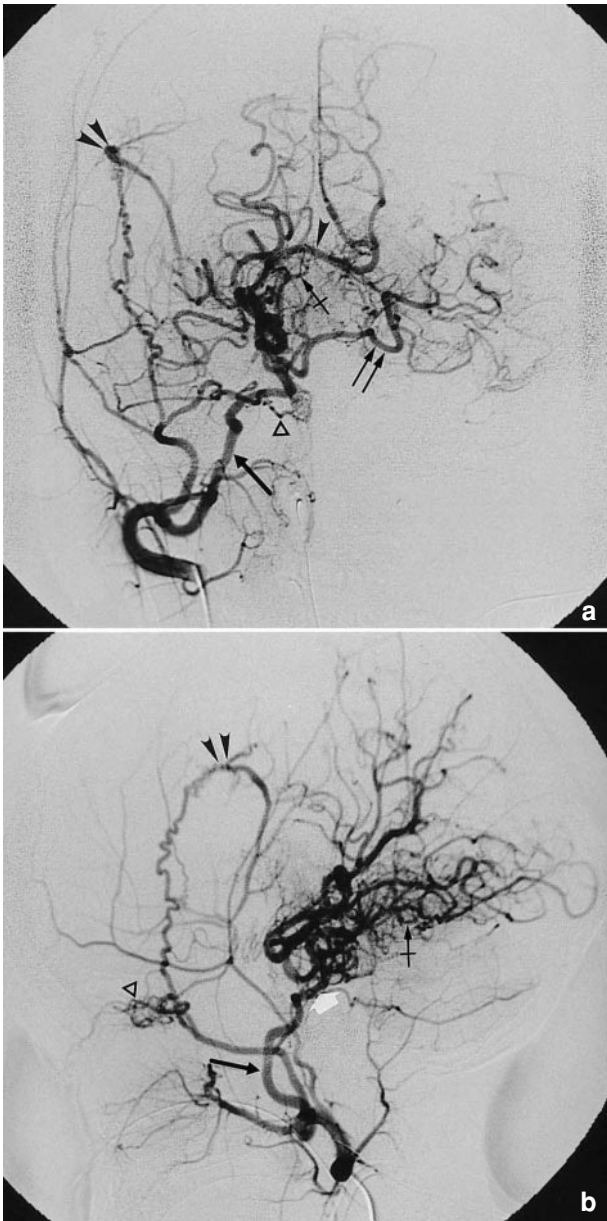
Fig. 4. The right external carotid injection (a) AP view (b) lateral view demonstrates the large anomalous accessory meningeal artery (single black arrow) coursing to the posterior fossa anastomosing with the right antero-superior cerebellar artery (crossed arrow). Single white arrow indicates the persistent trigeminal artery. The inferior temporal branches of the left posterior cerebral artery (*PCA*) (double arrows) and the right PCA are supplied by the above-mentioned anastomosis. The parieto-occipital and calcarine branches of the left PCA are supplied through the anastomotic collicular artery (single arrowhead) coursing behind the brain stem. The right middle meningeal artery anastomoses with the cortical branch of the middle cerebral artery (double arowheads). The orbital branch of the right middle meningeal artery (open triangle) supplies the orbit including the retinal circulation
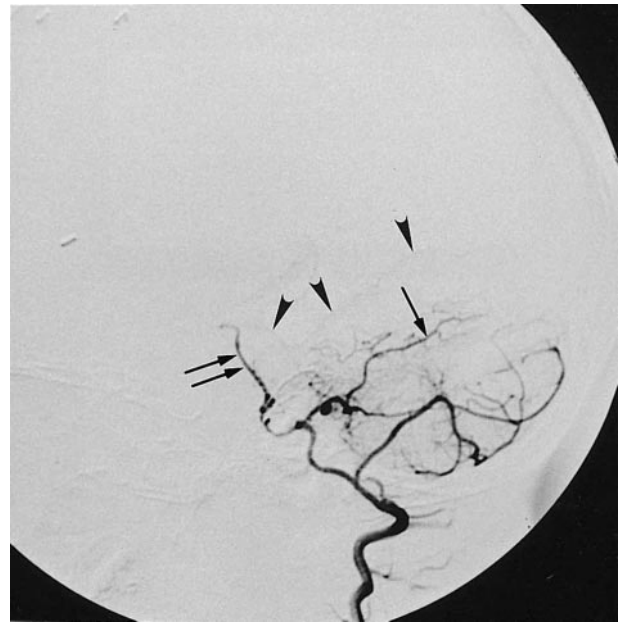

Fig. 5. The left vertebral injection (lateral view) shows the left anterior inferior cerebellar artery (single arrow) and left posterior inferior cerebellar artery. Through the markedly hypoplastic basilar artery (double arrows), the inferior temporal branch of the left PCA (arrowheads) is faintly visualized
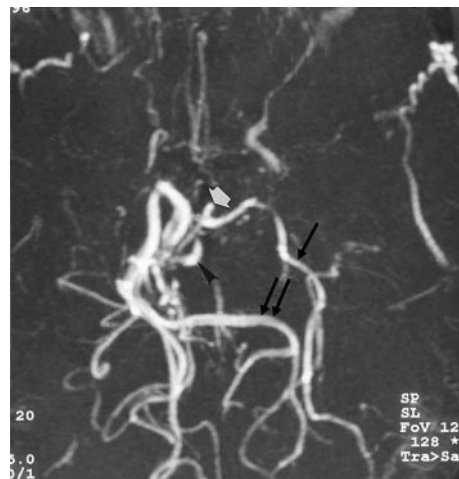

Fig. 6. Magnetic resonance angiography (axial view) shows the temporal branch (single black arrow) and parieto-occipital branch (double arrows) of the left PCA originating from the right accessory meningeal artery (arrowhead). Single white arrow indicates the basilar artery

PCA (Fig. 5). Except for the surgically established extracranial to intracranial anastomoses, the above-mentioned vascular anomalies were observed in the preoperative cut-film angiography at the age of 8 months. The axial and lateral views of the magnetic resonance angiography helped to understand the complex anomalous vasculature (Figs. 6 and 7).
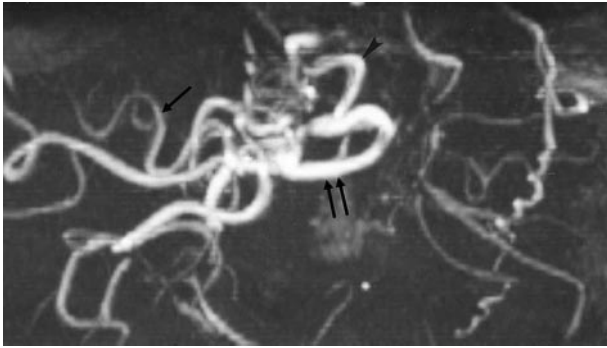
Fig. 7. Magnetic resonance angiography (lateral view) of the right half vasculature including the midline structure shows the right accessory meningeal artery (arrowhead), which supplies the right antero-superior cerebellar artery (single arrow) and the right PCA (double arrows)
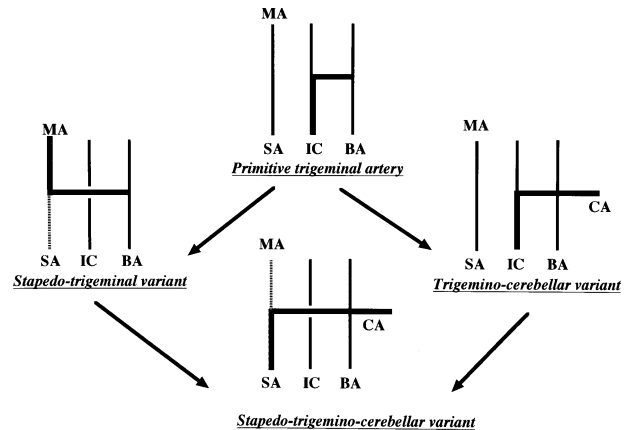

Fig. 8. Diagrammatic explanation of the variants of the persistent primitive trigeminal artery. Abbreviations: *BA* basilar artery; *CA* cerebellar artery; *IC* internal carotid artery; *MA* meningeal artery; *SA* stapedial artery

Discussion

The AMA represents the persistent portions of the medial limb of the primitive internal maxillary artery [1], which derives from the primitive stapedial artery [10]. The AMA usually has intracranial and extracranial branches [1, 3]. The intracranial branch courses into the intracranial cavity through the foramen ovale or the foramen of Vesalius (sphenoidal emissary foramen), and gives blood supply to the trigeminal ganglion and adjacent dura [1]. This intracranial branch may anastomose with the branch from the carotid siphon. In case of acquired occlusion or congenital agenesis of the internal carotid artery, this anastomosis may play an important role as the blood supply from the external carotid system to the internal carotid system [3].

The PTA runs along the trigeminal nerve and anastomoses with the internal carotid artery at the C5 segment and the basilar artery at the point between the ASCAs and the anterior inferior cerebellar arteries. According to Padget [17], the PTA is first observed in the 3-mm embryo. At the 4-mm stage, this artery communicates with the primitive internal carotid artery. The PTA also communicates with a fragment of the longitudinal neural artery. The basilar artery is formed by the union of the paired longitudinal neural arteries at 7- to 12-mm stage. The PTA begins to involute and then disappears at the 14-mm stage. The incidence of persistent PTA is estimated at 0.1–0.2% [13].

Two modifications in the development of the PTA are observed in our patient (Fig. 8). One modification is that the PTA takes over the territory of the intra-

cranial branch of the primitive stapedial artery (the "*stapedo-trigeminal*" variant). An example of this vascular anomaly is the MMA originating from the basilar artery [19]. Another is the ophthalmic artery originating from the basilar artery, in which the primitive dorsal ophthalmic artery is annexed by the "*stapedo-trigeminal*" variant (the "*ophthalmo-sta-pedo-trigeminal*" variant) [18]. The other modification is that the PTA takes over the territory of the ASCA. An example of this anomaly is the cerebellar arteries originating from the C5 segment of the internal carotid artery (the "*trigemino-cerebellar*" variant) [9, 20, 23]. Haughton, *et al.* [5] postulated that this anomaly is the result of a persistent PTA associated with an incomplete fusion of the longitudinal neural arteries. In some cases with this anomaly, there is a communication between the cerebellar artery and the basilar artery. The incidence of this anomaly is reported at 0.18% [20]. These two modifications in the development of the PTA resulted in the persistent anastomosis between the AMA and the ASCA (the "*stapedo-trigemino-cerebellar*" variant). Haemodynamic balance in the early stage of development between the large anomalous AMA-ASCA anastomosis and the vertebrobasilar system might have caused regression of the basilar artery between the ASCA and the inferior cerebellar arteries.

Embryologically, the posterior communicating artery and the P1 portion of the PCA (diencephalo-mesencephalic territory) belong to the caudal division of the internal carotid artery. The PCA gathers the telencephalic supply by distal annexation of the

territory of the anterior choroidal artery (temporo-occipito-parietal territory) [12]. Inferior temporal branches of the PCA may have a separate origin from the parieto-occipital and calcarine branches of the PCA [14]. The collicular artery in our patient supplied the parieto-occipital and calcarine territories of the contralateral PCA through the midline anastomosis. This midline collateral of the collicular artery is the remnant of the tectal embryonic network [12]. We postulate that the collicular artery becomes the anastomotic vessel to maintain the haemodynamic balance in association with regression of the proximal segment of the left PCA.

The right ophthalmic artery in our patient originates from the right MMA, reaching the orbit through the superior orbital fissure. This anomaly is caused by the anastomosis between the primitive ophthalmic artery and the orbital branch of the primitive stapedial artery. Regression of the proximal segment of the primitive ophthalmic artery causes the ophthalmic artery to originate from the MMA [11].

Bilateral steno-occlusive changes of the terminal portions of the internal carotid arteries and moyamoya vessels are consistent with the angiographic criteria for moyamoya disease. Our diagnosis is based on these angiographic criteria, however, the systemic disease and congenital anomaly are exclusion criteria for moyamoya disease which is proposed by Research Committee on Spontaneous Occlusion of the Circle of Willis sponsored by Japanese Ministry of Health and Welfare in 1988. Therefore, since our patient had congenital vascular anomalies and phacomatosis pigmentovascularis, the diagnosis is not moyamoya disease, notwithstanding the presence of typical angiographic features, if we use the guidelines of the above-mentioned committee. The aetiology of moyamoya disease is unknown. Although there are congenital and acquired theories regarding its origin, it is conceivable that congenital factors play a role to some extent in the aetiology of moyamoya disease in our patient. It is unclear if the vascular anomalies found in our patient are associated with moyamoya disease or are just chance occurrences.

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Comments

This paper reports a variant, combining multiple embryonic vessels including the trigeminal system; it is indeed a very unusual variant.

This case is well documented and the embryological discussion is correct. It is infomative for our readers and a helpful reminder of the potential anastomoses which may occur with the caudal ICA.

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