Case Report

The ‘8 of Willis’: large anterior communicating artery fenestration associated with posterior circulation anomalies

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Abstract

Introduction
Anomalies and variations of the anterior communicating artery are common. Fenestration has been reported as the most common variation of this artery. Dolichoectasia of intracranial arteries is a rare arteriopathy characterised by elongation and widening of the arteries, mostly involving vertebral and basilar arteries. This report discusses large anterior communicating artery fenestration associated with posterior circulation anomalies.

Case Report
We report here a rare case of large anterior communicating artery fenestration associated with mild dolichoectasia of the basilar artery and marked dolichoectasia of the left posterior inferior cerebellar artery, forming a large vascular loop at the area of cisterna magna, as well as hypoplasia of the left anterior inferior cerebellar artery. The study discusses the details of this case.

Conclusion
Dolichoectasia of intracranial arteries is rare and arteriopathy characterised by elongation and widening of the arteries and disturbance of the laminar blood flow, mostly involving vertebral and basilar arteries.

Introduction
The anterior communicating artery (ACoA) appears in the human embryo of 18 mm as a reticulated anastomosis between the two anterior cerebral arteries (ACAs). At the 24 mm crown-rump length stage, this network fuses to form a single ACoA.

During the foetal period, the artery acquires the same size as that of the ACA. Because of this growth in foetal life, the artery in adults is often of a large size, duplicated or plexiform but rarely absent. Padget noticed occurrence of anomalies in the ACoA to be fairly common and explained their presence on the basis of its embryonic development.

Congenital anomalies of the intracranial arteries predispose to the formation of saccular aneurysms due to an increased haemodynamic stress. Aneurysms of the ACA and ACoA are common, and their microvascular surgical management requires sound knowledge of the normal and variant vascular anatomy. Variations and anomalies of the ACA-ACoA-recurrent artery complex are commonly observed when associated with a symptomatic intracranial aneurysm and especially during ACoA aneurysm surgery. Regardless of whether a vascular anomaly has been identified preoperatively, ACoA aneurysm surgery should be undertaken with that possibility in mind.

The fenestration has been found to be the most common ACoA variation raising concern as this has been shown to compromise collateral flow and predispose to aneurysm formation. In contrast, fenestration of the internal carotid artery (ICA) is extremely rare and may be associated with aneurysms arising from the fenestrated segment.

Dolichoectasia of intracranial arteries is a rare arteriopathy characterised by elongation and widening of the arteries and disturbance of the laminar blood flow. It involves mostly the vertebral and basilar arteries. In advanced cases, formation of a fusiform aneurysm is possible.

Intracranial arterial dolichoectasia may be asymptomatic for a long time. However, in many cases, it leads to neurological symptoms associated with haemodynamic disturbance (due to unstable wall clots) and mass effect caused by the widened vessel.

The most frequently diagnosed complication of vertebrobasilar dolichoectasia (VBD) is the compression of structures adjacent to the vertebral and basilar arteries. A giant VBD with only slight compressive symptoms is unusual. Further, VBD can trigger various clinical symptoms such as posterior circulation stroke (including ischemia of cervical spine, cerebellum and cerebral trunk, as well as occipital lobe syndromes), cranial nerve palsies, subarachnoid or intracerebral haemorrhages and even symptoms of a neoplasm in the posterior fossa and in the cerebellopontine angle, but there is no effective treatment for their prevention.

The clinical symptom that should be stressed is headache, which precedes the occurrence of stroke for several days. Most arterial compressive lesions have been attributed to VBD and prior reports have concentrated mainly on the pressure effects of basilar artery ectasia. Much less is known about vertebral artery compression of the medulla.

We report here an asymptomatic case of a large ACoA fenestration associated with posterior circulation anomalies, including dolichoectasias. These Willis circle anomalies were found during routine dissection by chance. They were observed in a formalin-embalmed cadaver of a 75-year-old female who had died from heart disease. The details of this case as well as a relative literature-based discussion are presented below.

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Case report

Dissecting the Willis circle arteries of this case’s brain, it was observed that the ACoA was largely fenestrated (duplicated), forming a second smaller arterial circle just anterior and connected to the Willis circle. Given the fact that the ACoA belongs to the Willis circle, the appearance of this variation could be described as an ‘8 (eight)’ instead of the classical ‘circle’ of Willis (Figure 1). The maximum diameter of the ACoA fenestration was approximately 10 mm.

Furthermore, examination of the rest of the cerebral vessels revealed mild dolichoectasia of the basilar artery (mainly dilated and curved to the left side at the pontomedullary junction) (Figure 2) and marked dolichoectasia of the left posterior inferior cerebellar artery (PICA), forming a large vascular loop at the area of the cisterna magna (Figures 3 and 4). Hypoplasia of the left anterior inferior cerebellar artery (AICA) (Figure 4) was another interesting coexisting finding. Bilaterally, the inferior cerebellar arteries of larger diameter (i.e. right AICA and left PICA) were distributed to the cerebellar tonsil.

Discussion

Series

Anterior circulation

Kwak et al.14 investigated arterial anomalies accompanying 296 cases of a single ACoA aneurysm and the etiological significance of these anomalies in the development of cerebral aneurysms. Either the ACoA fenestration or the presence of more than two ACoAs was observed in 17 cases (5.7%). As the anomaly was duplicated in some of these cases, the total number of cases with ACoA anomalies resulted to be 26 cases (8.8%) out of 296 cases investigated. This occurrence rate was not higher than those observed in other previously reported cerebral aneurysmal cases and control cases14.

Ogawa et al.6 reported operations performed on 206 patients with ACoA aneurysms using a bifrontal craniotomy and an interhemispheric approach. A total of 44 (21.4%) of these patients had vascular anomalies in the vicinity of the ACoA; these included a median artery of the corpus callosum (MACC) in 27 cases (13.1%), duplication of the ACoA in 20 cases (9.7%) and duplication of the A1 segment of the ACA in one case (0.5%). A retrospective study of the angiograms indicated that diagnosis of the A1 or ACoA duplication was not possible. The majority of the cases of ACoA aneurysms with MACC (81.5%) showed trifurcation of the ACoA, A2 and MACC6.

Saidi et al.3 evaluated variations of the ACA and ACoA. Thirty-six cadaveric brains were studied by gross dissection for the pattern of arterial blood supply. Unique variations observed include an accessory ACA from the ACoA. Variations of the ACoA were also observed including fenestration (26%) and duplication (13%)3.

Kapoor et al.1 studied variations of the Willis circle using brains from 1000 medicolegal autopsy subjects of varying ages (Indian population). In 54.8% of specimens, there were variations in the Willis circle. The circle was deficient in 32 specimens (3.2%). The ACoA was absent in 1.8% cases, duplicate in 10%, triplicate in 1.2% and plexiform in 0.4%. Seventy-four brains (7.4%) had multiple variations. Intracranial saccular aneurysm
was present in 10 specimens (1%). Persistence of some embryonic vessel that normally disappear, disappearance of vessels that would normally persist or sprouting of new vessels due to haemodynamic and genetic factors, are the usual causes for such anomalies.

Among the 100 cases (10%) of duplicated ACoA that these authors found, 75 were males and 25 females. In 44 (31 males, 13 females) specimens, there were two parallel communicating arteries. The distances between the two vessels varied from 0 mm to 5 mm. One of the vessels was hypoplastic in some of these cases. In another 27 instances (21 males, 6 females), such double ACoAs were joined together by an anteroposterior vessel. In a further 29 specimens, the two ACoAs were not parallel with each other. The communication was Y-shaped (forked) in 16 subjects (11 males, 5 females) and V-shaped in 13 specimens (8 males, 5 females). The apex could be on either side. They also reported literature data suggesting that the frequency of ACoA duplication varies from 3.2% to 10.8%. Various authors found the combination of two or more variations in a single brain, for example hypoplasia of the posterior communicating artery (PCoA) was frequently associated with embryonic derivation of the posterior cerebral artery or with duplication of the ACoA.

Gasca-González et al.15 studied the recurrent artery of Heubner (RAH), dissecting 15 human brains from the Mexican population and reported the presence, length, branches, course and RAH-ACoA complex variants. The RAH was found in 93% of hemispheres and duplicated in 39% of hemispheres. The RAH was duplicated in at least one hemisphere in 46.6% of brains. In 53.3% of brains, some variant of the ACoA complex was found15.

Posterior circulation

Milandre et al.16 reported 23 patients (16 males, 7 females, mean age of 62 years) with symptomatic VBD, during a 13-year period. Arterial hypertension was noted in 20 cases and associated aortic ectasia in four cases. The malformation was identified in all patients on computed tomography (CT) completed by angiography in 19 cases and magnetic resonance imaging (MRI) in seven cases. Autopsy was performed in five cases. Fourteen subjects (group 1) presented with a vascular event (ischemic in 13) affecting the brainstem and/or cerebellum. Nine other patients (group 2) had a chronic symptomatology resulting from compression of the cranial nerves, central nervous system and/or the cerebrospinal fluid pathway. Two patients died of stroke within the first month (rupture of the ectasia in one and occlusion in the other). The 21 survivors were followed for a mean period of 45.3 months. Eight patients had a stroke, with a significantly higher incidence in group 1 than in group 2. Ten patients (five in each group) developed progressive dementia possibly resulting from multiple cerebral infarction, hypertensive leucoencephalopathy and/or hydrocephalus. Except for the incidence of stroke, inaugural manifestations (stroke vs nervous compression) did not seem to influence the long-term prognosis16.

Savitz et al.13 studied nine symptomatic patients (four men, five women, age range: 32–79 years) with medulla compression from the vertebral artery and found that compression most commonly occurs at the ventrolateral surface. The clinical features can be transient or permanent and are predominantly motor and cerebellar or vestibular, but a poor correlation exists between the clinical findings and the severity or extent of impingement. The vertebral arteries were angulated, tortuous or dilated but not necessarily dolichoectatic to cause obvious indentation. Seven patients were treated with antiplatelets and anticoagulants or analgesics, whereas two underwent microvascular decompression, resulting in temporary or no relief. One surgical patient developed cranial nerve complications. Among the medically treated patients, none had progression of deficits, and those with single episodes had no recurrence of symptoms13.

Krisht et al.17 reported their experience using the pretemporal, transzygomatic, transcavernous approach with 50 complex basilar aneurysms. Complexity criteria in the 50 aneurysms included large or giant size in 27 patients, wide dysmorphic base in 18 patients, low bifurcation in 21 patients, posteriorly projecting dome in 11 patients and dolichoectasia of the apex in three patients. Twenty-five patients presented with subarachnoid haemorrhage (14 men, 36 women, age range: 32–76 years). The authors supported microsurgery as a safe and more durable treatment option for the management of complex basilar apex aneurysms that tend to have a higher rate of failure with endovascular therapy17.
Wu et al.\textsuperscript{11} aimed to validate the feasibility of coil-assisted stent reconstruction in the vascular lumen for the treatment of VBD and to evaluate its long-term effectiveness in preventing ischemic events. They found that endovascular reconstruction with coil-assisted stent placement or stent placement alone in the vascular lumen for the treatment of VBD is technically feasible and can prevent ischemic events in the territory of stented vessels compared with the natural course, though further studies in larger samples are needed\textsuperscript{11}.

**Cases**

**Anterior circulation**

Gurdal et al.\textsuperscript{18} examined 30 cadavers and reported two unusual variations of the ACoA. In the first case, ACoA was duplicated with a fenestrated ACA. In the second case, an oblique ACoA was present. Further, two branches of the oblique ACoA were joined to the right ACA. According to the authors, during neurosurgical exposure of the region for different purposes, knowledge of the vascular variations will increase the success of the procedure\textsuperscript{18}.

Dey and Awad\textsuperscript{7} reported two cases of ICA fenestration with successful surgical clipping of associated saccular aneurysms. In both instances, the fenestration involved the supracranial ICA and the aneurysm arose from the duplicated segment proximal to the origin of the PCoA. The aneurysms were more proximal than typical PCoA aneurysms and fenestration was suspected by rotational three-dimensional angiography and confirmed at surgery\textsuperscript{7}.

Furthermore, Osonuga et al.\textsuperscript{19} reported a case of ACoA duplication with no associated vascular variations or aneurysms in other major blood vessels. One ACoA was shorter than the other with external diameters of 1.0 mm and 1.4 mm, respectively. A quadrangular opening was formed between them\textsuperscript{19}.

**Posterior circulation**

Davous et al.\textsuperscript{20} reported a 54-year-old man who was affected by three successive infarctions in the vertebro-basilar territory. These infarctions were related to a dolichoectatic basilar artery. Deafness occurred first on the left side and then, after the third infarction, on the right side. The authors underlined that deafness can be observed after a pontine infarction in the territory of the AICA. A dolichoectatic basilar artery can be the source of thrombotic or embolic strokes. Their prevention by anti-aggregant or anticoagulant therapy is suggested\textsuperscript{20}.

Muñoz et al.\textsuperscript{21} reported a 52-year-old man with rapid jerking movements of his head to the right and clonic involuntary twitches involving his left eyelid and cheek. Electromyographic recordings were consistent with left sternocleidomastoid and facial asynchronous myoclonus. MRI showed the presence of a dolichoectatic left vertebral artery displacing the medulla. The authors proposed that direct compression of the XI\textsuperscript{th} and VII\textsuperscript{th} cranial nerves by the dolichoectatic left vertebral artery might be the mechanism responsible for myoclonus in their patient\textsuperscript{21}.

Campos et al.\textsuperscript{9} reported a 48-year-old woman who presented a two-month history of continuous buzz and a slight right-sided hearing loss that was followed by a cerebellar ischemic stroke. Brain CT and MRI revealed a marked compression of the brainstem due to an ectatic, tortuous and partially thrombosed basilar artery. The largest cross-sectional diameter of the basilar artery was 18 mm. They underlined the fact that an unusual giant VBD caused an impressive brainstem compression with displacement of important structures in an oligosymptomatic patient. Diagnosis was made only after the occurrence of a stroke. According to the authors, the presence of significant atherosclerotic changes and the large basilar artery diameter may indicate a poor outcome\textsuperscript{9}.

Interestingly, Tsutsumi et al.\textsuperscript{22} reported a 71-year-old female with basilar and bilateral carotid dolichoectasia manifesting as dysarthria and hemisensory disturbance, which resolved spontaneously within a day. She suffered a brainstem infarction (infarct in the perfusion area of the superior cerebellar artery) 28 months later, manifesting as drowsiness, dysarthria and right hemiparesis. Her consciousness level progressively deteriorated to stupor and she died of acute respiratory failure on the seventh day. Autopsy demonstrated a tear in the lateral wall of the broad-based aneurysm on the ectatic basilar artery and a diffused subarachnoid haemorrhage. The authors concluded that VBD is a dynamic vasculopathy that may rapidly progress in the affected basilar artery following an indolent clinical course. The prognosis for patients with VBD may depend mainly on the pathological changes in the basilar artery\textsuperscript{22}.

Finally, Baran et al.\textsuperscript{8} reported a case of a 64-year-old female with hypertension who was admitted to the hospital with severe non-systemic vertigo and dysarthria, which had lasted for a couple of weeks. Imaging of the brain revealed dolichoectasia of the Willis circle arteries (ICA, vertebral artery, basilar artery, middle cerebral artery) and she died of acute respiratory failure on the seventh day. Autopsy demonstrated a tear in the lateral wall of the broad-based aneurysm on the ectatic basilar artery and a diffused subarachnoid haemorrhage. The authors concluded that VBD is a dynamic vasculopathy that may rapidly progress in the affected basilar artery following an indolent clinical course. The prognosis for patients with VBD may depend mainly on the pathological changes in the basilar artery\textsuperscript{22}.

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**Conclusion**
Anomalies of the ACoA are common and fenestration has been reported as its most common variation. Dolichoectasia of intracranial arteries is a rare arteriopathy characterised by elongation and widening of the arteries and disturbance of the laminar blood flow, mostly involving vertebral and basilar arteries. However, a case combining large ACoA fenestration with mild basilar artery and marked left PICA dolichoectasia, as well as left AICA hypoplasia, is quite unusual. Persistence of some embryonic vessels that normally disappear; disappearance of vessels that would normally persist or sprouting of new vessels due to haemodynamic and genetic factors, are probably the usual causes of Willis circle anomalies. Knowledge of such variations and anomalies (together with their potential clinical manifestations) is of paramount importance, primarily for neurosurgeons and neuroradiologists.

**Abbreviations list**
ACA, anterior cerebral artery; ACoA, anterior communicating artery; AICA, anterior inferior cerebellar artery; CT, computed tomography; ICA, internal carotid artery; MACC, median artery of the corpus callosum; MRI, magnetic resonance imaging; PCoA, posterior communicating artery; PICA, posterior inferior cerebellar artery; RAH, recurrent artery of Heubner; VBD, vertebrobasilar dolichoectasia.

**References**