Fetal and Primitive Type of Circle of Willis with Unilateral Trifurcation of Internal Carotid Artery

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Abstract

Circle of Willis is a network of arteries in the base of the brain between the internal carotid artery and the vertebral artery to establish the collateral circulation between these two major arteries. Anomalies in this network may lead to serious clinical conditions like stroke by impairing the vital blood circulation. Normally, the internal carotid artery communicates with the posterior cerebral artery from the basilar artery by a thin posterior communicating artery. The present case however shows a rarity where both anterior cerebral arteries were arising from the left internal carotid artery and the right anterior cerebral artery had a small communication from the right internal carotid artery. In addition, both posterior communicating arteries were large and continued as posterior cerebral arteries with small contribution from the basilar artery. The knowledge of such variations may be useful for neurosurgeons and radiologists during the diagnosis and to plan the treatment regime.

Key Words: Circle of Willis, variation, fetal, cerebral flow, aneurysms

(Rec.Date: Feb 10, 2014 Accept Date: Mar 31, 2014)
Introduction

Cerebral circulation receives 15-20% of the cardiac output which is closely regulated to maintain perfusion in response to metabolic and physiological demands. The main cerebral distribution center for blood flow, the Circle of Willis (Circulus arteriosus), is a ring-like network of collateral vessels. The blood is delivered to the brain through the two internal carotid arteries (ICA) and the two vertebral arteries (VA) that join intracranially to form the basilar artery (BA). Each of the internal carotid arteries branches to form the middle (MCA) and anterior cerebral arteries (ACA), which supply blood to the front and the sides of the brain including the frontal, temporal, and parietal lobes of cerebrum. The basilar artery bifurcates into the right and left posterior cerebral arteries (PCA), which perfuse the back of the brain including the occipital lobe, cerebellum and the brain stem. The ring is completed by communicating arteries that connect the posterior cerebral arteries with ICA via posterior communicating arteries (PCoA) and two anterior cerebral arteries via the anterior communicating artery (ACoA) [1].

The anastomosis thus provided by this vascular ring is of great significance when one of the major arteries supplying the brain becomes occluded. It has been found that in more than 50% of healthy brains and in more than 80% of dysfunctional brains, the Circle of Willis contains at least one artery that is absent or underdeveloped [2]. The most common topological variations include missing communicating vessels, fused vessels, string-like vessels, and presence of extra vessels [3]. These topological variations may affect the ability to maintain cerebral perfusion, which may increase the risk of stroke and transient ischemic attack in patients with atherosclerosis [4]. Existence of a relationship between the variations in the circle of Willis and the sites of aneurysms of arteries also postulated earlier [5]. Aneurysms usually tend to occur in the branches or at the bifurcations of cerebral arteries. [6]. The pattern of the circle therefore becomes important in determining the adequacy of the brain circulation in operations for cerebral aneurysms and also in ligation of the internal carotid artery. Therefore, a detailed knowledge of the variations in the circle of Willis is valuable to the radiologists and neurologists. The present case reports a unique combination of two variations, i.e., fetal trifurcation of ICA (bilateral) & anterior trifurcation of ICA on left. Even though these variations were separately reported in the previous studies, combination of the
same if occurs may be very grave and may affect the blood flow which may increase the risk of stroke and transient ischemic attack.

**Case Report**

During routine dissection classes for medical students in the department of Anatomy, Kasturba Medical College, Manipal, the following variation was encountered. In a 70 year old female cadaver, bilateral fetal trifurcation of ICA and anterior trifurcation of ICA on the left was observed.

The left ICA was trifurcating into a large antero-median branch (AmA), middle cerebral (MCA) and a large posterior communicating (PCoA) arteries. The PCoA was found to continue as PCA after joining with a left small terminal branch of the BA. The large antero-median branch then divided into left and right anterior cerebral arteries (ACA). Between right and left ACA, we found an anterior communicating artery (ACoA) which was short but with similar caliber of ACA.

The right ICA on the other hand provided a thin antero-medial branch, large MCA and PCoA. The right AmA was very small in caliber and was joining the right ACA which in turn was a branch of the left ICA. The large PCoA then continued as PCA after receiving a small right terminal branch from the BA (Figure 1).

The basilar artery was formed by fusion of large right and small left vertebral arteries. The vertebral arteries were asymmetrical, the left being hypoplastic in nature. The terminal branches of the basilar artery further bifurcated into two small branches on both sides. One branch joined the PCA while the other continued as the superior cerebellar artery (SCA) bilaterally. Additionally a large right SCA and a small left SCA was also provided from the basilar artery prior to its termination.
Figure 1: Left internal carotid artery (ICA) was dividing into a large antero-medial branch, middle cerebral artery (MCA) and large posterior communicating artery (PCoA). The large left antero-medial branch (Ama) then divided into right and left anterior cerebral arteries (ACA). The right ICA was dividing into small communicating branch (Ama) to right ACA, large MCA and PCoA. The large PCoA from the ICA of both side continued as PCA after receiving a small terminal branch from the basilar artery (BA). The terminal branches of the basilar artery further bifurcated into two small branches on both sides. One branch joined the PCA while the other continued as the superior cerebellar artery (SCA) bilaterally. Additionally a large right SCA and a small left SCA was also provided from the basilar artery prior to its termination.
Discussion

The anatomy of the circle of Willis is highly variable, particularly with regard to the anastomoses in posterior circulation [7]. During development, ICAs are formed between 28–30 days, and the BA is formed between 31–36 days, when the longitudinal neural arteries combine [8]. In embryos of 52 days of age, completely formed circle of Willis appears with all slender segments having an identical caliber. As the embryo grows, the dominant fetal-type feeding of the PCA from the ICA via the PCoA changes towards a normal adult configuration where the feeding of the PCAs takes place from the vertebro-basilar system. Caliber of the PCoAs regress as the vertebro-basilar system develops and starts to feed the PCA resulting in a normal adult-type circle of Willis. In some cases, this change is incomplete and leads to persisting fetal-type feeding of the PCA from the ICA [9] as seen in the present case, the fetal type of feeding with large anterior, middle cerebral arteries and persistent large PCoA which continued as the PCA on both side with small contribution from the basilar system.

Earlier, it has been shown that, out of the variations found in the arteries participating in the circle of Willis, most of the abnormalities were in the posterior communicating or posterior cerebral arteries [10]. In a study using multi detector CT angiography, it was shown that in Chinese population, 79% of the cases demonstrated a complete anterior circle of Willis, and 31% showed a complete posterior circle of Willis and 27% of the participants showed a complete circle of Willis [11]. Present case even though showing the complete circle of Willis, the distribution pattern ICA and BA systems are very different from other complete circles.

A comparative study of the circle of Willis in man, cow, sheep, goat, and pig revealed that the PCoA are larger in the ruminants and pigs indicating the large quantity of blood flowing from the ICA through the PCoAs to the posterior aspect of the cerebrum [12]. Therefore, the carotid arteries are the main arteries supplying the blood to the brain in these animals. In man, the smaller caliber of the posterior communicating artery indicates less flow of blood from ICA to the posterior part of the cerebrum. The reason could be due to the additional contribution of blood to the posterior aspect of the brain by the vertebro-basilar system which is a recent development in human. As PCoAs are the new link and being a weak point, several changes happening here, leads to more variations in the posterior half of the circle as seen in
the present case. The present case also shows a primitive type of circle of Willis with the PCoA of a larger caliber as compared to the PCA.

The embryonic origin of the posterior cerebral artery from the internal carotid is a fairly common anomaly and was found in 10% of the circles in a study by Iqbal [6]. Such a vessel was connected to the basilar artery by a small communicating type of vessel. Several other studies have also recorded the incidence of the embryonic origin of the PCA [2,13].

Accessory vessels may also be present in the circle of Willis in the form of duplications/triplications. Studies have reported duplications in the anterior portion of the circle i.e., in the ACoA and ACA [6,14]. The disappearance of the vessels that normally persist or the persistence of the vessels that normally disappear or formation of new vessels also occurs due to hemodynamic factors during early development [9]. However, in most of the arterial variations, the normal brain function may not be affected due to the collateral circulation and compensation from the arteries of the other side. But, these variations gain importance in abnormal conditions like ischemia, aneurysm and stroke.

Interestingly, in the present case, a large antero-medial branch from the left ICA was giving rise to both right and left ACA with a very small and short communication from the right ICA to the right ACA. In such condition, the occlusion of left ICA may lead to severe ischemia to the both the cerebral hemisphere as about three fourth of the brain will be deprived of the blood supply, as the left PCA, MCA, ACA & right ACA all are derived from left ICA and the resultant clinical consequences would be severe.

Under normal conditions, blood flow in the communicating arteries is negligible. However, if a subject has an atypical Circle of Willis, e.g., missing one of the main arteries or communicating arteries or under pathological conditions such as complete or partial occlusion of one of the cerebral or carotid vessels, the flow can be redirected to perfuse deprived areas [15]. Such topological variations in the arterial circle may affect the ability to maintain flow through arterioles, which may increase the risk of stroke and transient ischemic attack in patients with atherosclerosis [4].

These clinical scenarios typically occur in older patients, who have a limited ability to compensate to acute changes in blood flow and thus are at greater risk for developing an acute ischemia (stroke) or chronic hypofusion. The significance of these problems cannot be
underestimated since stroke ranks third among leading causes of death and is the leading cause of disability in older adults [4]. The variations in the Circle of Willis are of clinical importance since they may have important influences on symptomatology, clinical examinations and investigations. The present case was a unique combination of variations with left ICA dominance indicating the fetal type of circulation. Thus, occlusion of left ICA may have significant impact on the cerebral arterial supply. Therefore, the knowledge of such variations in the formation of circle of Willis resulting from a number of developmental errors may be useful for neurosurgeons, radiologists during diagnosis and to plan the treatment regime.

Conflict of Interest
The authors declare no conflicts of interest.

References


