A COMPLEX ABNORMALITY OF THE CIRCLE OF WILLIS. CASE PRESENTATION

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ABSTRACT

Objective. We report a case of a patient with medium ACI stenosis and border zone infarct that occurred under normal hemodynamic conditions (normal BP, sinus rhytm), as normal supply mechanisms through the circle of Willis were absent. The circle of Willis was practically absent: the right ICA ramified in right MCA and a single ACA. Left ICA ramified in MCA and PCA. Right PCA was a branch of BA.

Case presentation. A 75 year old patient was admitted in emergency with dressing apraxia. CT scan and MRI disclosed a 5/4 cm infarction in the temporo-parieto-occipital junction on the right side. Doppler ultrasonography of the precerebral vessels revealed medium stenosis of right ICA. 3D TOF MRA reconstruction disclosed a complex abnormality of the circle of Willis: absence of A1 and P1 on the left side, absence of PCoA on the right side, a single and fenestrated ACA originating in right ICA. The circle was practically absent; there was no possibility of circulatory supply. The territories of distribution of the precerebral vessels suffered severe modifications: right ICA ramified in MCA and a single ACA, left ICA ramified in MCA and PCA, right PCA was a branch of basilar artery. The infarction occurred between the boundaries of the right ICA (MCA respectively) and the posterior territory (PCA respectively).

Conclusion. We report a complex and extremely rare abnormality of the circle of Willis that became clinically manifest because of a severe hemodynamic compromise.

Key words: circle of Willis abnormalities, border zone infarction, carotid stenosis

OBJECTIVE

Border zone infarction, due to a hemodynamic mechanism, is frequent in severe stenosis/occlusion of cerebral vessels. We report a case of a patient with medium ACI stenosis and border zone infarction under normal hemodynamic conditions (normal BP, sinus rhytm), as the normal supply mechanisms through the circle of Willis were absent. The circle of Willis was practically absent: the right ICA ramified in right MCA and a single ACA. Left ICA ramified in MCA and PCA. Right PCA was a branch of BA.

CASE PRESENTATION

A right handed and hypertensive 75 year old patient, blind because of a posttraumatic retinal detachment, treated by antihypertensive medication, presented for impairment of dressing ability in the last week. That inability was initially ignored both by the patient and by the family and was thought to be explained by the vision impairment (although the patient could dress himself previously and perform other household activities too by the time when his visual acuity had already been impaired).

On physical examination BP was 180/90 mm Hg, unremarkable for the rest.

Neurological examination: the patient was alert, oriented in space and time, with no motor deficit, deep tendon reflexes were symmetrical, brisk, the palmomental reflex was bilateral present, and the plantar response was bilateral flexion. The right eye was with amaurosis and outwards deviated. The patient could perceive visual stimuli with the left eye and could identify them in the internal inferior quadrant. The patient presented digital agnosia and dressing apraxia (he kept turning his clothing inside out on all sides and then in the opposite way too and was unable to put it on, although he recognized it well).

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Doppler ultrasonography of precerebral vessels revealed medium stenosis of right ICA.

CT scan disclosed a 5/4 cm hypodensity at the junction between T-P-O lobes on the right side, with no mass effect. Normally sized and positioned ventricular system. The radiological report was available only, unlike the slides that were not.

All laboratory findings were normal.

MRI after 1 week disclosed the same area of infarction described on the CT scan (Fig. 1).

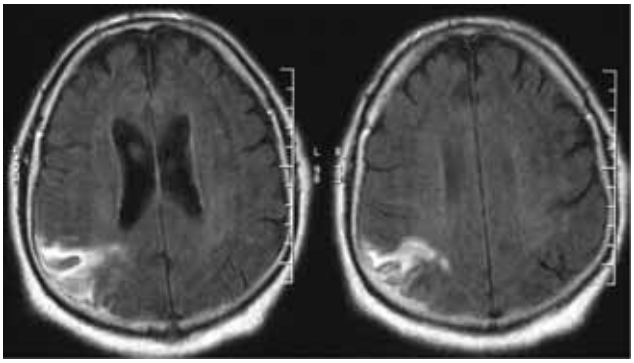

Figure 1

3D TOF MRA with vascular reconstruction revealed multiple abnormalities of cerebral vessels (Fig. 2, Fig. 3):

- Absence of A1 on the left side
- Single ACA originating in the right ICA, having a fenestration area at the usual level of AComA
- Absence of P1 on the left side, PCA being a branch of left ICA
- Absence of PComA on the right side

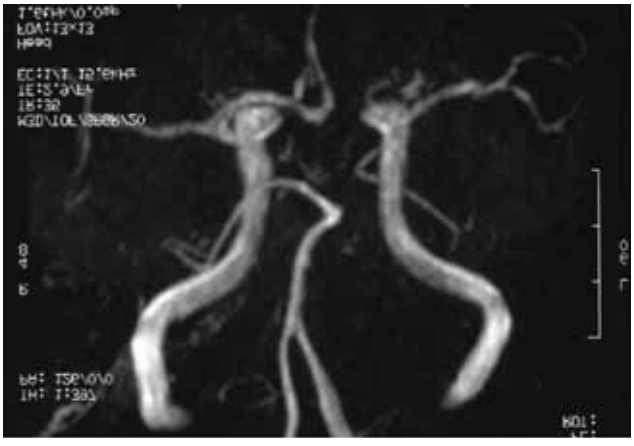

Figure 2

Antiplatelet and neurotrophic medication was administered, while BP values maintained normal without antihypertensive treatment. The evolution was good, although dressing inability persisted.

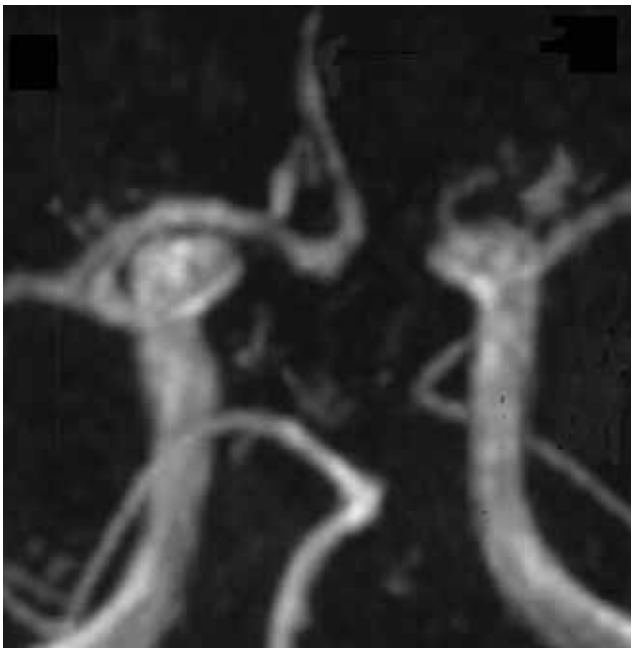

Figure 3

DISCUSSION

The circle of Willis was practically absent in the presented case: the right ICA ramified in right MCA and a single ACA. Left ICA ramified in MCA and PCA. Right PCA was a branch of BA (Fig. 4).

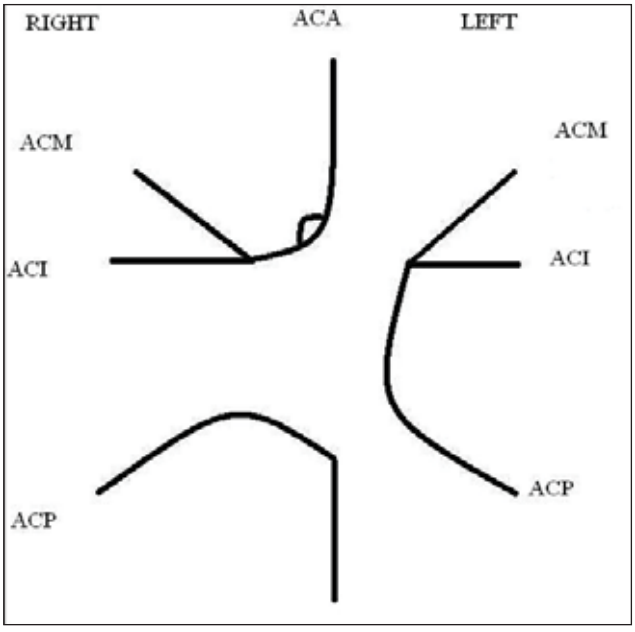

Figure 4

Cerebral vessels had important changes of the territories of distribution comparatively to the common type. (Fig. 5) The circle of Willis could not offer any possibility of supply between those territories.

The infarction was situated between right ICA territory (its branch MCA, respectively) and the posterior territory (the right PCA that originated in the BA) (Fig. 4).

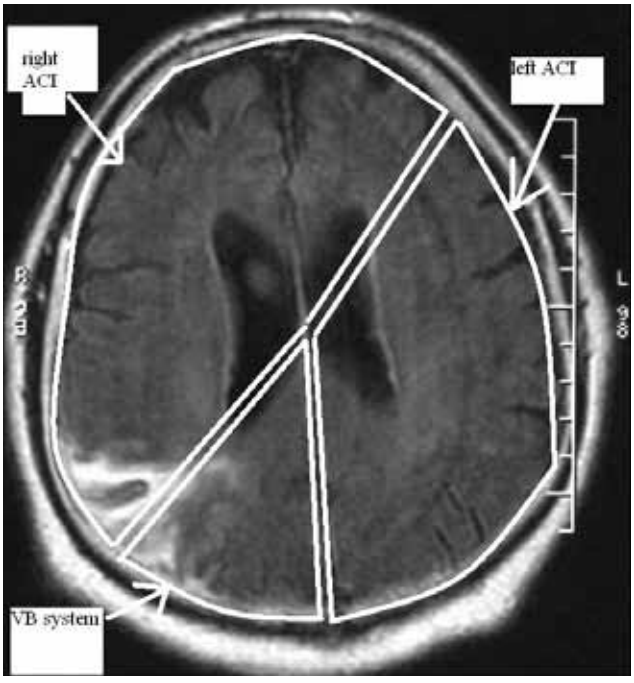

Figure 5

That complex abnormality of cerebral vessels takes place in the early stages of development, between the 30th-45th days. In the 30th day of development, inside the 4-6 mm embryo, the third pair of the bronchial arches and a part of the dorsal aorta forms together the distal portion of the ICA which gives 2 branches: the cranial branch (that divides into ACA, MCA, and ACorA) and a caudal branch that divides into PComA, PCA, PCorA and Superior Cerebellar arteries.

Superior neural arteries will form together the vertebral arteries that will join on the median line and form the basilar trunk.

The circle of Willis can be identified in the 45th day of development, and the entire arterial system has the typical adult pattern in the 30-40 mm embryo of the 52 nod day (stage IV according to Padget, stage 20 according to Carnegie) (1,2,3,4,5)

Both initially and including after the anastomoses between the 2 systems, the flow goes from the carotid towards the basilar system, but the communicating arteries become functional later on only when pressure becomes unequal inside the 2 arterial systems as it may be due to pathological or physiological causes.

The ideal, perfectly symmetrical circle is relatively rare. Since Willis (6) described the anatomical structures of that amazing anastomotic system and intuited its functional role, numerous variants and abnormalities have been described by dissection studies, microsurgery, and imagistic studies (angiograms, angio CT scans, MRA).

Circle of Willis anomalies mainly is the rule than the exception. (1)

Despite this rule, numerous variants have been described by several authors. Kleis, in 1941, cited by (2, 3), reported 12 variants for the circle of Willis. They consisted in asymmetrical and incomplete variants, some structures being duplicated or absent, other structures being fused together.

Lazorthes and Gouanzesin 1968, cited by (2,3) described 25 types of the circle according to the diameter of its arteries. Other criteria there may also be added, such as different other size, the absence of certain communicating arteries or the presence of multiple variants for each of them. A very great number of possibilities arise, resulting in the statement that every individual has his own vascular design, in which only the general principles of organisation are common.

Numerous anatomical studies report multiple variants and anomalies of the Circle of Willis. (7)

In 1988 SenGupta RA and Mc Allister VL, cited by (5), proposed 8 types of classification, all the vessels being present: normal or hypoplasic.

Various and numerous studies of the entire circle performed by different modalities (anatomical, Doppler, angioCT, MRA) reported variants corresponding to the above mentioned descriptions and other new variants too (8, 13). There may also be added numerous studies and case reports upon certain segments of the circle.

In our presented case, 3D TOF MRA reconstruction image arguments that the circle did practically not exist. The patient presented a complex abnormality of development that involved the branches of all main arteries that would have normally form the circle: there was no PComA on the right side, there was no ACA on the left side, the basilar artery did not communicate with PCA, and PCA was a branch of ICA. Besides, the single ACA displayed a segmental dedublation, fenestration respectively, at its origin.

All these segments embryological developed between day 30 and day 45.

Multiple mechanisms explain these abnormalities: vessel fusion of normal separated vessels, lack of fusion and persistence of some independent vessels, persistence of vessels that should disappear, disappearance of vessels that should persist, incomplete development, and unusual paths in the complete development of a vessel.

Numerous factors, hard to identify in the most cases, may trigger these mechanisms. We can speculate that PComA did not arise from right ICA , and P1 of PCA did not arise from BT. Azygos ACA may result from the fusion of the 2 ACA, when segmental dedublation persists in the place where there should be AcomA, followed by the disappearance of A1 on the left side. Or, ACA can develop only on the right side, and the segmental dedublation might be the right AComA which remains just a curl, because left ACA does not exist.

Absence of these segments is essential in the hemodynamic mechanisms. (14,15) Arterial distribution territories are severely modified (16,17). Each precerebral vessel supplies strictly one cerebral area, and there is no compensation possibility between them, as it is shown in Fig.4

Absence of PComA was essential for generating the cerebral lesion. PComA is the essential arterial structure providing circulatory compensation both in carotid lesions and in vertebrobasilar system lesions too. (14,15,19,20,21,22)

A watershed (border-zone) infarct is the consequence of hemodynamic compromise (so-called low-flow infarcts): severe stenosis, arterial occlusion or severe homodynamic disturbances (hypotension, arrhythmia, dehydration, a.s.o.) (23).

Small/medium stenosis in normal hemodynamic conditions (normal BP, sinus rhythm), may result in a border zone infarction in cases where normal supply mechanisms through the circle of Willis are absent.

CONCLUSION

We reported a complex and extremely rare abnormality of the Circle of Willis that resulted in severe impairment of the compensatory mechanisms of the Circle of Willis, becoming clinically manifest therefore.

Abreviations: ICA – internal carotid artery, ACA-anterior cerebral artery, MCA – middle cerebral artery, AComA – anterior communicating artery, PComA – posterior communicating artery, PCA – posterior cerebral artery, BA – basilar artery, A1 – ACA segment of circle of Willis, P1 – ACP segment of circle of Willis, ACorA – anterior choroidal artery, PCorA – posterior choroidal artery

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