

Bilateral Infarction in the Anterior Cerebral Artery Vascular Territory Due to an Unusual Anomaly of the Circle of Willis

Fons Borggreve, MD; Peter P. De Deyn, MD, PhD; Peter Mariën, MS;
Patrick Cras, PhD; Rudi A. Dierckx, MD

Background Bilateral infarction in the anterior cerebral artery vascular territory is rare and usually results from rupture of an aneurysm of the anterior communicating artery. In the case of an anomaly of the anterior part of the circle of Willis, thrombosis also may lead to bilateral infarction.

Case Description A 73-year-old right-handed man presented with a complete loss of communicative behavior and an almost complete quadriplegia. ^{99m}Tc-hexamethylpropyleneamine oxime single-photon emission-computed tomography (HMPAO SPECT) of the brain showed a “horseshoe” image, reflecting bilateral perfusion defects in areas supplied by the anterior cerebral arteries. Postmortem examination of the brain revealed an unusual anomaly of the circle of Willis in

which the postcommunicating part of the right anterior cerebral artery was absent.

Conclusions In our patient a unique developmental anomaly of the circle of Willis indirectly contributed to bilateral infarction in the territory of the anterior cerebral artery. The patient’s neuropsychological profile, dominated by akinetic mutism, was highly suggestive of involvement of the receptive language modalities. For the first time, ^{99m}Tc-HMPAO SPECT of the brain revealed a horseshoe image in bilateral infarction in the territory of the anterior cerebral artery. (*Stroke*. 1994;25:1279-1281.)

Key Words • cerebral arteries • cerebral infarction • circle of Willis • tomography, emission-computed

Bilateral infarction in the anterior cerebral artery vascular territory is very rare¹ and usually results from rupture of an aneurysm of the anterior communicating artery or from thrombosis of the precommunicating part of the anterior cerebral artery in combination with an agenesis of the contralateral part.² We present a patient in whom bilateral infarction occurred because of thrombosis of the postcommunicating part of the left anterior cerebral artery and in whom the postcommunicating part of the right anterior cerebral artery was absent.

Case Report

A 73-year-old right-handed man was admitted to the hospital after he was found at home sitting in a chair in a verbally unresponsive state and unable to move a single limb. According to his daughter, he had experienced several transient episodes of weakness alternating in both legs during the weeks before admission. On admission the patient looked around and frowned when he was spoken to. He did not react differently to verbal commands and made no attempts to speak or utter sounds. Neurological examination revealed flaccid right hemiplegia, flaccid plegia of the left leg, and paresis of the left arm with an intact grip function of the hand.

Received December 9, 1993; final revision received February 18, 1994; accepted March 10, 1994.

From the Departments of Neurology (F.B., P.P. De D., P.M.) and Nuclear Medicine (R.A.D.), General Hospital Middelheim, Antwerp, and the Departments of Neuropathology (P.C.) and Neurochemistry and Behavior (P.P. De D.), Born-Bunge Foundation, University of Antwerp (Belgium).

Reprint requests to P.P. De Deyn, MD, PhD, Department of Neurology, AZ Middelheim, Lindendreef 1, 2020 Antwerpen, Belgium.

Tendon reflexes were normal and symmetrical. A positive Hoffmann-Trömner sign was evident on the right side. Both plantar reflexes were in extension, and the snout reflex was present. With the exception of the appearance of a grasping reflex on the right side, the patient’s clinical situation remained unchanged during the following weeks.

Computed tomographic (CT) scan of the brain on the day of admission was normal. Examination of cerebrospinal fluid did not show cells; the protein level was slightly elevated at 72 mg%. The electroencephalographic registration showed bilateral, mostly sharp, 4- to 7-Hz theta waves of 30 to 50 μ V and disseminated polymorphic 1- to 3-Hz delta waves of 40 to 70 μ V. ^{99m}Tc-hexamethylpropyleneamine oxime single-photon emission-computed tomography (HMPAO SPECT) of the brain on day 2 of admission showed parasagittal hypoperfusion bilaterally in the frontoparietal regions, more pronounced on the left (Fig 1).

A diagnosis of bilateral infarction in the territory of the anterior cerebral artery was confirmed by a control CT scan and magnetic resonance imaging of the brain on day 3 of admission. Although an anomaly of the vessels was suspected, arteriography could not be performed because the patient’s clinical condition continued to worsen, leading to death 4 weeks after admission.

Postmortem investigation revealed an unusual anomaly of the circle of Willis (Fig 2). The posterior part of the circle was entirely normal. The anterior part, however, showed the following anomalies: the left anterior cerebral artery had a greater diameter compared with the right (0.3 versus 0.15 cm, respectively). The postcommunicating part of the right anterior cerebral artery was not developed, and only a small orbital branch was

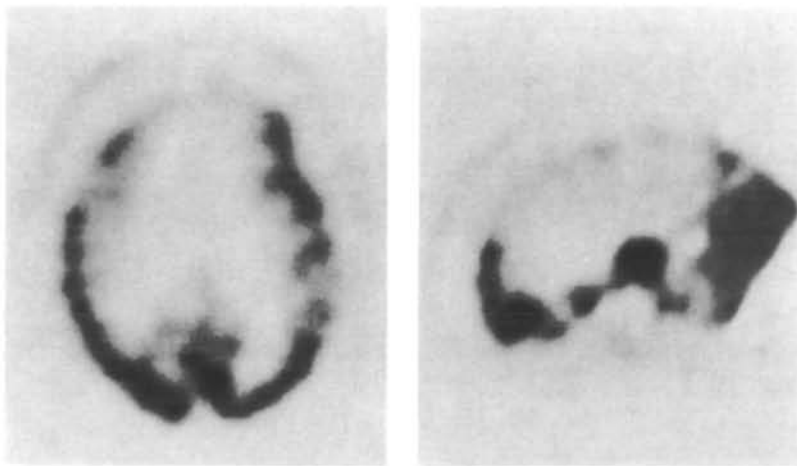


FIG 1. Left, Oblique single-photon emission-computed tomography (SPECT) slice shows bilateral frontoparietal hypoperfusion, located just above the basal ganglia. Orientation is similar to computed tomographic scan: left=right side, top=frontal side. Right, Left parasagittal slice illustrates lateral extension of the hypoperfusion. Left=frontal region. ^{99m}Tc -hexamethylpropyleneamine oxime brain SPECT was performed using a three-head SPECT system (Triad 88, Trionix) equipped with ultra-high-resolution parallel-hole collimators.

found. The diameter of the postcommunicating part of the left anterior cerebral artery was 0.4 cm and split into two arteries that supplied both frontal lobes. A complete occlusion by thrombus of the left anterior cerebral artery was located approximately 2 cm distal to the anterior communicating artery. The extent of the cerebral infarction followed the boundaries of the anterior cerebral artery territory, as described below.

Discussion

The anterior cerebral artery supplies the medial surface as well as the adjacent rim of the lateral convexity of the entire frontal and parietal lobes. It also contributes the anterior limb of the internal capsule, the inferior half of the head of the caudate nucleus, portions of the inferior putamen and globus pallidus, parts of the hypothalamus, the anterior column of the fornix, and the anterior part of the corpus callosum.^{2,3} Infarction in the territory of the anterior cerebral artery is quite rare and is responsible for 0.6% to 3% of all cerebrovascular accidents. According to Bogousslavsky and Regli,¹ unilateral infarction in this area usually is a result of embolism from the heart or the internal carotid

artery (63%). Other causes are contralateral or ipsilateral occlusions of the internal carotid artery, distal extensions of a thrombosis of the carotid artery, and local thrombosis caused by vasculitis.⁴ Occlusion of the stem of the anterior cerebral artery proximal to its connection with the anterior communicating artery is usually well tolerated because adequate collateral flow is provided via the anterior communicating artery from the anterior cerebral artery on the opposite side. When the occlusion is localized distal to the anterior communicating artery, neurological symptoms appear.² Symptomatology (including hemiparesis, hemihypoesthesia, mutism at onset, transcortical motor aphasia, conflictual tasks impairment, mood disturbances, and, far less frequently, incontinence, grasp reflex, hemineglect, acute confusional state, and unilateral left apraxia) correlates well with the topography and size of the infarct.⁵

Although bilateral infarction in the territory of the anterior cerebral artery has been reported to be very rare, no data regarding the incidence are available. In his study of 1490 cases, Bogousslavsky described only two patients with bilateral infarction. Bilateral infarc-

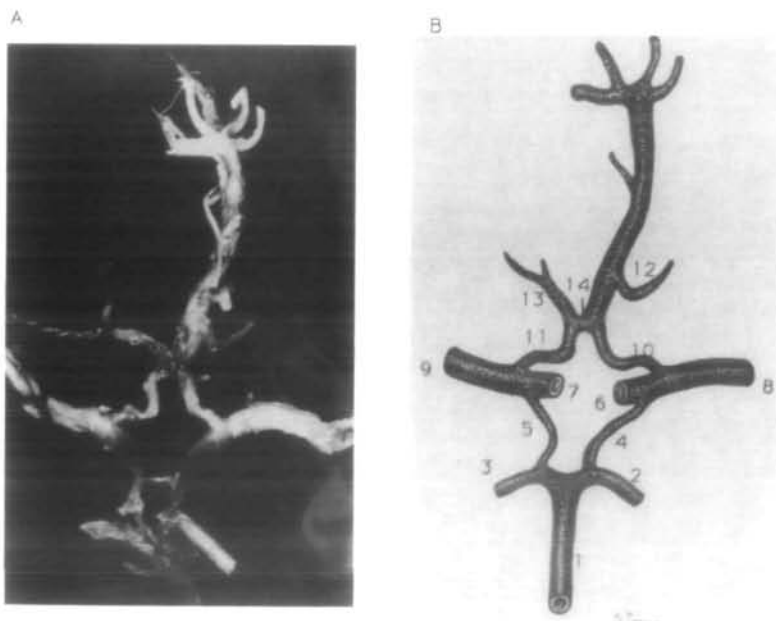


FIG 2. Photomicrograph (A) and schematic representation (B) of the circle of Willis, in which the postcommunicating part of the right anterior cerebral artery was not developed. 1 indicates basilar artery; 2 and 3, left and right posterior cerebral artery, respectively; 4 and 5, left and right posterior communicating artery, respectively; 6 and 7, left and right internal carotid artery, respectively; 8 and 9, left and right middle cerebral artery, respectively; 10 and 11, left and right anterior cerebral artery (precommunicating part), respectively; 12, left anterior cerebral artery (distal part); 13, small orbital branch replacing distal part of right anterior cerebral artery; and 14, communicating anterior cerebral artery.

tions are usually due to embolism or spasm that complicates subarachnoid hemorrhage from an aneurysm of the anterior communicating artery. Another cause is thrombosis of the proximal part of one anterior cerebral artery when the contralateral proximal branch is rudimentary or absent. Bilateral infarction resulting from cardiac or carotid embolism is exceptional.² Furthermore, the symptoms that accompany bilateral infarction of the anterior cerebral artery territory differ from those caused by unilateral infarction, particularly the profound mental changes, which vary from abulia to akinetic mutism.^{3,6} Bilateral infarction may even mimic a basilar artery occlusion.⁷

Bilateral infarction in our patient resulted from thrombosis of the distal part of the left anterior cerebral artery, which supplied both anterior cerebral artery vascular territories. This congenital anatomic anomaly of the circle of Willis, in which the postcommunicating part of the right anterior cerebral artery was not developed, is a unique finding: none of the large anatomic studies that involved the circle of Willis revealed the variant that occurred in our patient.⁸⁻¹⁵

The clinical symptomatology in our patient, which consisted of quadriplegia that particularly affected the lower limbs, incontinence, a grasping phenomenon, and a complete loss of communicative abilities, entirely fit the classic symptoms of anterior cerebral artery occlusion. The complete absence of verbal output could be differentiated from speechlessness in "locked-in" syndrome since any attempt to develop an alternative, nonverbal communication strategy based on the few voluntary movements the patient could still perform remained unsuccessful. Because no consistent facial responses or directional eye movements could be elicited, evaluation of receptive language modalities was impossible to exclude a supplementary aphasic disorder. However, the neuropsychological profile, dominated by the functional ablation of expressive language, clearly corresponded to a diagnosis of akinetic mutism.¹⁶ This diagnosis was supported by ^{99m}Tc-HMPAO SPECT, which revealed bilateral hypoperfusion in the territory of the anterior cerebral artery.⁶

From the point of view of functional neuroimaging, the "horseshoe" image shown on ^{99m}Tc-HMPAO SPECT of the brain, performed on day 2 of admission, greatly contributed to the diagnosis of bilateral infarction in the territory of the anterior cerebral artery.

Because unilateral anterior cerebral infarctions are known to involve strips of the superior frontal cortex along the hemispheric fissure on SPECT images,³ bilateral involvement may be suspected to exhibit a horseshoe image, as it did in our patient. This primary horseshoe hypoperfusion image on SPECT, following the boundaries of the anterior cerebral artery territory, has never been described before and seems to be inherently connected to bilateral infarction in this area.

Acknowledgments

We would like to thank Professors B. Hillen and J.J. Martin for their comments and S. Joacim for skillfully preparing the figure of the anomaly of the circle of Willis.

References

1. Bogousslavsky J, Regli F. Anterior cerebral artery territory infarction in the Lausanne Stroke Registry. *Arch Neurol.* 1990;47:144-150.
2. Ross Russell RW. *Vascular Disease of the Central Nervous System.* Edinburgh, Scotland: Churchill Livingstone, Inc; 1983:170-173.
3. Wood JH. *Cerebral Blood Flow: Physiological and Clinical Aspects.* New York, NY: McGraw-Hill Book Co; 1987:24-25, 97-99, 247-252.
4. Gacs G, Fox AJ, Barnett HJM, Vinuela F. Occurrence and mechanisms of occlusion of the anterior cerebral artery. *Stroke.* 1983;14:952-959.
5. Vanderwegen L, Van den Bergh R. Infarct in het gebied van de arteria cerebri anterior. *Tijdschr Geneesk.* 1992;48:299-302.
6. Freedom FR. Akinetic mutism and bilateral anterior cerebral artery occlusion. *J Neurol Neurosurg Psychiatry.* 1971;34:693-698.
7. Ferbert A, Thron A. Bilateral anterior cerebral artery territory infarction in the differential diagnosis of basilar artery occlusion. *J Neurol.* 1992;239:162-164.
8. Fawcett E, Blachford JV. The circle of Willis: an examination of 700 specimens. *J Anat.* 1905;40:63-70.
9. Von Mitterwallner F. Variationsstatistische Untersuchungen an den basalen Hirngefäßen. *Acta Anat.* 1955;24:51-86.
10. Baptista AG. Studies on the arteries of the brain, II: the anterior cerebral artery: some anatomic features and their clinical implications. *Neurology.* 1963;13:825-835.
11. Tulleken CAF. A study of the anatomy of the anterior communicating artery with the aid of the operating microscope. *Clin Neurol Neurosurg.* 1977;80:170-173.
12. Perlmutter D, Rhoton AL. Microsurgical anatomy of the distal anterior cerebral artery. *J Neurosurg.* 1978;49:204-228.
13. Hillen B. The variability of the circle of Willis: univariate and bivariate analysis. *Acta Morphol Neerl Scand.* 1986;24:87-101.
14. Hillen B. The variability of the circulus arteriosus (Willisii): order or anarchy? *Acta Anat.* 1987;129:74-80.
15. Nathal E, Yasui N, Sampei T, Suzuki A. Intraoperative anatomical studies in patients with aneurysms of the anterior communicating artery complex. *J Neurosurg.* 1992;76:629-634.
16. Lebrun Y. *Mutism.* London, England: Whurr Publishers Ltd; 1990:88-94.