Redka anatomska variacija Willisovega kroga pri bolniku z ishemično možgansko kapjo

A rare anatomical variation of the circle of Willis in a patient with ischemic stroke

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Izvleček

Namen: *Cerebralni arterijski krog zagotavlja kolateralni obtok v možganih in ščiti možgane pred ishemijo, saj v primeru zmanjšanega ali oslabljenega pretoka krvi vzdržuje perfuzijo tkiva po njegovih vejah. Čeprav so številne naključne anatomske variacije v Willisovem krogu (CoW) manj pomembne, pa nekatere med njimi povečujejo tveganje za ishemične dogodke, anevrizme in aterosklerozo ter imajo pomembno vlogo pri načrtovanju tako endovaskularnega kot kirurškega zdravljenja.*

Predstavitev primera: *Poročamo o primeru redke arterijske variacije CoW, ki je bila diagnosticirana z računalniško tomografsko angiografijo pri bolniku z ishemično možgansko kapjo, ki je v literaturi redko opisana. Ugotovili smo aplazijo segmenta A1 leve sprednje možganske arterije in ipsilateralno*

Abstract

Purpose: *The cerebral arterial circuit provides collateral circulation to the brain and protects the brain from ischemia, by sustaining tissue perfusion in case of reduced or impaired blood flow through its branches. Although many random anatomical variations in the circle of Willis (CoW) are of minor importance, some increase the risk of ischemic events, aneurysm, and atherosclerosis, and play an important role in endovascular and surgical treatment planning.*

Case presentation: *We report a rare arterial variation of the CoW diagnosed by computed tomography angiography in a patient with ischemic stroke. We found aplasia of the A1 segment of the left anterior cerebral artery and ipsilateral hypoplasia of the P1 segment of the left posterior cerebral artery.*

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hipoplazijo segmenta P1 leve zadnje možganske arterije. ***Zaključek:*** *Za nevrokirurge, nevrologe in nevroradiologe je podrobno poznavanje številnih anatomskih variacij v arterijski oskrbi možganov ključnega pomena za postavitev pravilne diagnoze in izbiro ustreznega zdravljenja. Tip CoW prav tako vpliva na resnost in izid možganske kapi, pri čemer je nepopoln Willisov krog povezan s težjimi stanji (višji rezultati po National Institutes of Health Stroke Scale, hujši nevrološki izpadi in slabši nevrološki izidi) in slabšo prognozo.*

Conclusion: *For neurosurgeons, neurologists, and neuroradiologists, detailed knowledge of the numerous anatomical variations in the arterial supply to the brain is crucial to make a correct diagnosis and choose the appropriate treatment. The type of CoW variation influences stroke severity and outcome, with an incomplete CoW related to more severe conditions (higher National Institutes of Health Stroke Scale scores, more severe neurological deficits, and worse neurological outcomes) and poor prognosis.*

INTRODUCTION

Reduced blood flow to the brain activates compensatory mechanisms via the collateral supply of the circle of Willis (CoW), which protects the brain from ischemia and infarction (1). Variations in CoW lead to changes in cerebral hemodynamics and are associated with increased risk of stroke, aneurysm rupture, white matter disease, migraine, atherosclerosis, and vasospasm after subarachnoid hemorrhage. CoW variations determine the type and method of treatment, may affect the success of the procedure due to potential complications, and may influence the safety and outcome of treatment (2). Here, we present a case of a patient with ischemic stroke with a rare variation of the CoW and address the role of CoW variations as important risk factors for stroke.

CASE PRESENTATION

A 71-year-old right-handed man was brought to the emergency department with suspicion of ischemic stroke, as he suddenly developed right-sided hemiplegia and central facial palsy, dysarthria, and diplopia when looking to the right, homonymous hemianopia, and the condition progressed. Glasgow Coma Scale (GCS) was 15/15, and he was conscious and oriented. Tendon reflexes on the right side were less responsive and right-sided extensor plantar reflex was present. The electrocardiogram recorded a normal sinus rhythm. His past medical history included hypertension, hyperlipidemia, benign prostatic hyperplasia, symptomatic epileptic seizures, alcohol dependence syndrome, and toxic liver damage. He had been a smoker for 30 years. The patient had no history of ischemic stroke or transient ischemic attack, head injury, or trauma. His family history of stroke was negative.

Initial computed tomography and computed tomography angiography revealed left parieto-occipital subacute ischemia, with reduced perfusion time, volume, and flow. A 90% occlusion of the left internal carotid artery (ICA) was found. The left vertebral artery (VA) was subtotally narrowed in the V4 segment. The A1 segment of the left anterior cerebral artery (ACA) was absent (Figures 1 and 2). The P1 segment of the left posterior cerebral artery (PCA) was hypoplastic (Figures 1 and 2). The PCA was filled distally via a strong left posterior communicating artery (PCoA).

DISCUSSION

The CoW is complete in less than half of the population due to developmental anomalies. Twice as many complete circles are in the anterior half, whereas only one-third are complete in the posterior half of the circle (3). Aplasia of the A1 segment is found in 1% of people (1). Thus, both A2 segments are supplied via the available A1 segment, which covers the entire ACA territory (4). Variation is crucial between intravascular

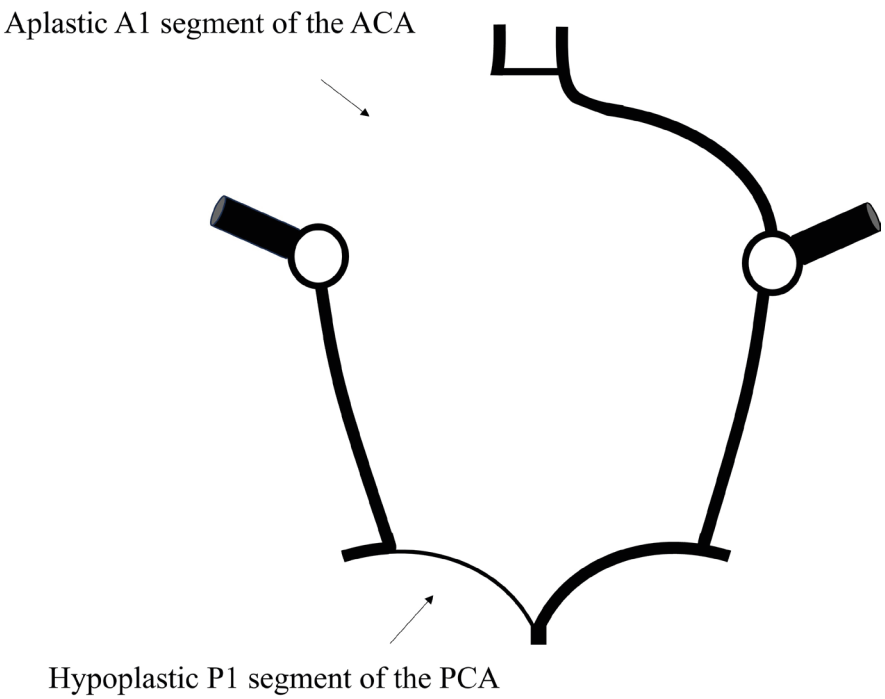

Figure 1. *Scheme of anatomical variation of the Willis polygon with absent A1 segment of the left ACA and hypoplastic P1 segment of the ipsilateral PCA. ACA – anterior cerebral artery; PCA – posterior cerebral artery.*

interventions in the anterior communicating artery territory or ischemic events, as it increases the risk and extent of brain ischemia in the frontal lobe (5).

Two-thirds of cases have more common anatomical variants of CoW in the posterior region (4). When the P1 segment is hypoplastic, the main blood supply to the occipital lobe comes from the ICA, and the PCA is filled distally via a stronger PCoA, as in our patient. The variation is found unilaterally in 11%–29% and bilaterally in 1%–9% (6).

Studies on CoW variations are numerous and vary widely depending on age, sex, health status, side and number of vessels affected, location within the CoW, and method for data collection (4). Despite the high degree of CoW variability described and classified into 22 different types by Riggs, Lazorthes, and Krabbe–Hartkamp (2), to our knowledge, there is no research on the incidence of simultaneous A1 aplasia and ipsilateral P1 hypoplasia in the literature. He et al. (7) found a similar case with left A1 aplasia but contralateral hypoplasia of the P1 segment in a patient with hypertension, who complained of long-term dizziness, fatigue, depression, insomnia, and memory loss. Sonobe et al. (8) reported a case of A1 aplasia and bilateral P1 hypoplasia in an infant who suffered a massive infarction during extracorporeal life support because of a rare anatomic variation. An incomplete anterior part of the CoW combined with an incomplete posterior part of the CoW results in anterior circulation stroke (4).

The effectiveness of compensation of the CoW depends on the presence of its vessels and their size. A complete CoW has an effective intracranial collateral circulation and a greater compensatory capacity in cerebral ischemia, but variations in the CoW cause changes in hemodynamics (4). In the case of ICA occlusion and A1 hypoplasia or aplasia, collateral supply to the ACA and middle cerebral artery occurs through reverse flow in the PCoA ipsilateral to ICA occlusion. When the P1 segment is hypoplastic, the main blood supply to the occipital lobe comes from the ICA. In P1 hypoplasia, flow through the dilated PCoA is preserved in 5–20% of cases, as in our patient (9). When large artery stenosis or occlusion is present in patients with variations in the posterior portion of the CoW, the brain tissue is vulnerable to hypoxicity because of inadequate compensation by collateral blood flow. Variations, incompleteness, and hypoplasia of any part of the CoW are 1.4 times more likely to cause ischemic stroke compared with patients with normal anatomy. Poor CoW integrity is associated with frequent stroke recurrence in patients with severe intracranial atherosclerotic stenosis and poor prognosis in acute ischemic stroke. Recent studies suggest that CoW variants affect the development and progression of intracranial atherosclerosis (10).

The CoW is strongly associated with cerebral infarction in patients with severe stenosis of the ICA or vertebrobasilar artery (10), as in our case, as the

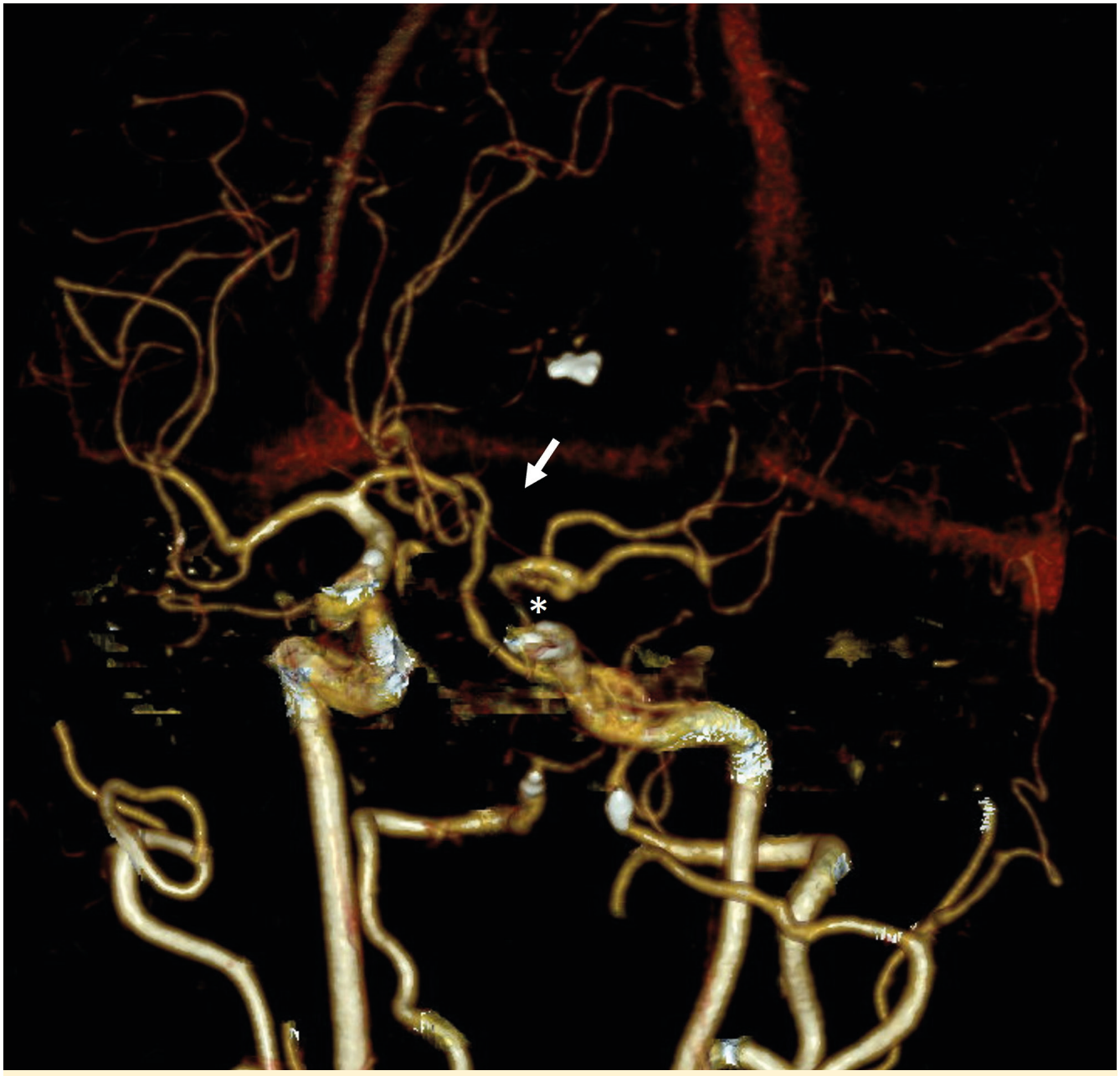

Figure 2. *3D computed tomography angiography reconstruction of the cerebral arteries – anterolateral view. Absence of the proximal segment (A1) of the left ACA (arrow) and hypoplastic proximal segment (P1) of the left PCA (asterisk). ACA – anterior cerebral artery; PCA – posterior cerebral artery.*

patient had 90% occlusion of the left ICA and subtotal stenosis of the V4 segment of the left VA. Henderson and Eliasziw (11) found that the rate of hemispheric cerebral infarction or transient cerebral ischemic attack was significantly lower in patients with >70% ICA stenosis in the perioperative or long-term treatment period when the CoW developed normally.

CONCLUSIONS

Understanding the anatomical variations in the arterial supply to the brain is important for planning surgical or interventional treatment of ischemic attacks and strokes, and has a significant impact on disease outcome and patient recovery.

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