A rare case of ischemic stroke following occlusion of the artery of Percheron

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A 57-year-old man was admitted to the ER after falling on the back of his head without any prodromi. No loss of consciousness was noted. The patient was responsive and able to walk into the ambulance without any help. Besides alcohol and nicotine abuse, medical history was blank.

At ICU admission physical examination showed a patient with impaired consciousness, a Glasgow Coma Scale of 8 and distinct anisocoria with an unresponsive mydriatic left pupil. Vestibulo-ocular reflexes were preserved. The man had appropriate responses to noiceptive stimuli and normoflexia was seen with down going planar reflexes with Babinski sign negative for both sides. Blood pressure was elevated but further cardiovascular, respiratory and abdominal examination was unremarkable. Apart from the elevated non-sober blood glucose level and hypercholesterolemia, all parameters ranged within normal limits.

Chest X-ray depicted an enlarged heart shadow with bilateral perihilar vascular consolidation. Cerebral computed tomography (CT) (Fig. A) showed no obvious brain lesions. Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) of the brain revealed T2-weighted hyperintense signals of the thalami and left mesencephalon (Fig. B), suggesting bilateral paramedian thalamic and left mesencephalic infarction. Diffusion weighted MRI also confirmed the suspected diffusion restriction in these regions (Fig. C), highly suggestive for an acute infarction in the artery of Percheron territory. Transcranial Doppler and 3D time-of-flight MR angiography (TOF-MRA) displayed lightgraded ostial occlusion of the internal carotid on both sides, without evidence for carotid or vertebral dissection.

The patient's condition improved gradually and after 12 days he was transferred for further revalidation. Both electrocardiogram and Holter monitoring demonstrated nothing abnormal. Echocardiography confirmed hypertrophy of the heart with a tricuspid insufficiency grade I and the patient was diagnosed with a hypertensive hypertrophic cardiomyopathy. No evidence was found suggesting a possible extracranial cause for embolism.

Antithrombotic and antihypertensive therapy was initiated at the time of admission, along with insulin to correct hyperglycemia, A statine was proposed because of the hypercholesterolemia. Cognitive revalidation was initiated after his transfer.

Comment

The artery of Percheron (AOP) is an uncommon anatomic variant supplying bilateral paramedian thalami and sometimes the rostral part of the mesencephalon. Occlusion of the AOP can cause typical neurological, ophthalmological and neuropsychological symptoms. The posterior cerebral artery is divided into four segments. The P1 segment extends from the junction between the basilar artery and the posterior communicating artery. It supplies the medial part of the thalamus on both sides and the rostral part of the mesencephalic midbrain. The artery of Percheron (AOP) is a variation of the P1 segment where a single prominent perforating branch supplies both thalami and the mesencephalic midbrain. The exact prevalence remains unknown. The anomaly has no significant difference in prevalence between males and females.

The AOP occlusion can result into three typical features. The neurological signs are most prevalent and have been reported in up to 92% of cases (consciousness impairment, focal neurological signs), ophthalmological signs were seen in 64% of cases (ocular motility, abnormal light reflexes, ptosis) and neuropsychological abnormalities up to 42% of cases (executive function disorders, memory deficit or language disorders). Although in most cases coma resolves rapidly, when present, the neuropsychological and ophthalmological signs may persist. In most cases occlusion is due to cardioembolism.

The diagnosis of an AOP occlusion is dependent on imaging studies, mainly MRI, as CT findings may appear normal early on. Though CT is easier to obtain, MRI remains essential when a typical thalamomesencephalic syndrome is suspected. In these cases, ischemic lesions of the paramedian areas of both thalami are seen, with possible rostral mesencephalic involvement. The AOP occlusion can also show a characteristic midbrain “V” sign, appearing as a high-intensity signal on diffusion-weighted and axial FLAIR images along the pial surface of the midbrain in the interpeduncular fossa, and present in 67% of patients. MRI coupled with 3D TOF-MRA of the carotid, vertebral and basilar arteries is preferred as the first choice investigation to rule out a basilar artery occlusion.

Other etiological investigations such as electrocardiography and Holter monitoring can be useful as they may reveal cardiac abnormality or arrhythmia associated with embolism.

The main differential diagnosis is dural venous sinus thrombosis as a subset of cerebral venous thrombosis. Other rarer causes of restricted thalamic diffusion include Creutzfeld-Jakob's disease, Wernicke encephalopathy and extrapontine myelinolysis.

ICU admission is definitely required in the management of patients with an AOP occlusion due to possible life-threatening complications (increased intracranial pressure, hyperglycemia, deep venous thrombosis, etc.).

Reference
