Weber's syndrome revealing a Percheron artery infarction: a case report

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Abstract

Percheron's artery (PA) occlusion is an uncommon type of stroke involving paramedian thalamus and/or midbrain infarction. It accounts for 4-18% of all thalamic infarcts and 0.1-2% of all strokes. Its clinical manifestations are variable and its mode of presentation as Weber's syndrome is exceptional due to the unusual clinical presentatio

Introduction:

The artery of Percheron (AP) is an anatomical variant of the arteries of the posterior circulation of the brain. Occlusion of this artery will result in bilateral paramedian thalamic infarction with or without midbrain involvement (1) . The clinical manifestations of ischemic stroke in this area can be summarised as a triad: altered consciousness, paralysis of verticality of gaze and memory disorders (2). They are rarely associated with pyramidal signs or cranial nerve damage and the brain scan is usually normal (3). This often leads to misdiagnosis in our low-income countries where MRI (Magnetic Resonance Imaging) is still not widely available. We report a case of Weber's syndrome secondary to an occlusion of Percheron's artery.

Clinical case

This is the case of a 50 years old hypertensive for 5 years on amlodipine 10 mg. She was admitted with consciousness disturbance with a right hemiplegia. The exact time of beginning of the symptoms was unknown. The patient was found unconscious. On admission, the neurological examination showed a Glasgow scale at 9/15, a right hemiplegia with Babinky sign, associated with anesthesia and a left third nerve palsy (Figure1) and the initial NIHSS (National Institutes of Health Stroke Scale) score was 20. Blood pressure was 145/90 mmHg and capillary blood glucose 1.15g/l. The brain CT showed a bilateral thalamic and mesencephalic infarction (figure 2 and 3), the MRI was not done because it was not available. CT angiogram of supra aortic arteries showed an occlusion of the V4 portion of the left vertebral artery (figure 4). Selective cerebral angiography to make the diagnosis was not performed. The electrocardiogram and echocardiogram were normal but Holter-electrocardiogram wasn't done. The blood count, urea, creatinine, lipid profile and blood glucose were normal. She was treated with 100 mg of aspirin, statins. The evolution was marked by the occurrence of an aspiration pneumopathy at day 4 of hospitalization. This was successfully treated with amoxicillin and clavulanic acid. At 10 days of hospitalisation, the consciousness problems had regressed. On discharge, she retained hemiplegia, motor aphasia and third nerve damage. Her NIHSS score was 16. However, follow-up was not performed because the patient was lost to follow-up.

Discussion:

Cerebral infarcts affecting both paramedian thalami are unusual and may raise suspicion of occlusion of a single arterial trunk known as the artery of Percheron (4,5) . This is the third anatomical arterial variant (Type IIb) which vascularises the thalami and/or the midbrain (3) (see table 1). These structures are responsible for the regulation of the sleep-wake cycle. Therefore, thalamic infarcts ca cause paresthesias or numbness, speech and cognition disturbance, memory impairment, and stupor (6). Our patient presented a motor deficit associated with consciousness discturbance like stupor. This anatomical variant occurs in 33% of the population (7) and infarcts of this artery account for 4-18% of all thalamic infarcts and 0.1-2% of all strokes (8). Macedo et al. who had one of the most representative series found a frequency of 0.17%(9). The classic clinical presentation of an infarction of Percheron's artery is a triad of altered consciousness, paralysis of verticality of gaze and memory disorders (2) . The association of these signs with Weber's syndrome in our patient constitutes one of the main particularities of our clinical case. Indeed, Weber's syndrome is part of the midbrain syndrome and is characterised by a contralateral hemiplegia associated with an attack on the homolateral common oculomotor nerve. Its incidence is unknown and it rarely occurs in isolation (10,11). Mesencephalic and Percheron's artery infarction with Weber's syndrome is a rare and difficult clinical finding (12). In our patient, the common oculomotor nerve damage was not accompanied by pupillary damage. The most plausible hypothesis would be an absence of damage to the nuclei of the superior mesencephalon, responsible for pupillary innervation. Thus, a careful clinical examination allows the diagnosis to be suspected. Brain CT allowed to exclude the infarction involving occipital lobe (or lobes), that could suggest basilar artery occlusion, but it did revealed symmetrical ischemic lesions in both thalami and anteromedial midbrain, which were consistent with clinical presentation (13)(6). Carotid and basilar Doppler ultrasound was irrelevant. CT angiography was performed (arterial and venous phases), which revealed no signs of arterial or venous thrombosis. Although it also did not show evidence for artery of Percheron (AOP) occlusion. It is not unusual as artery of Percheron is rarely visible on angio-MRI or angio-CT or conventional angiography. In our patient the CT angiogram showed an vertebral occlusion. Our patient did not benefit from intravenous thrombolysis because it was not available in our region. Finally, the treatment of Percheron's artery infarction must be oriented according to the underlying pathological process. It can range from antiaggregation treatment to anticoagulation depending on the aetiology (14). The long-term evolution had not been done in our patient who was lost to follow-up. We could not determine if she presented persistent vigilance disorders or dementia as reported by Macedo et al. In his series of 8 patients, he found hypersomnia in 3 patients. (9)

Conclusion

The combination of consciousness disorders, oculomotor impairment and hemiplegia should lead to the suspicion of brain stem damage and to emergency imaging, in particular MRI, which is the gold standard. If the latter is not available, a brain CT associated with CT angiogram of supra-aortic trunks can be used for investigation. If the imaging reveals lesions in the thalami, an infarction of Percheron's artery should be considered. Although Weber's syndrome associated with Percheron's artery infarction is rare, the management is no different from other types of stroke and requires the same etiological approach. However, follow-up is important in order to determine the evolution, which is variable depending on the patient.

Table 1: Anatomical variations in the vascularity of the thalamus

Anatomical description	**Anatomical description**
I	Normal anatomy of the paramedian artery: the left and right paramedian arteries arise from t
IIA	Variant of the paramedian artery anatomy: both paramedian arteries arise from the left and ri
IIB	Anatomy of Percheron's artery: a branch of the right or left PCA supplies the two thalami
III	Variant of the paramedian artery anatomy: the two paramedian arteries originate from an arte

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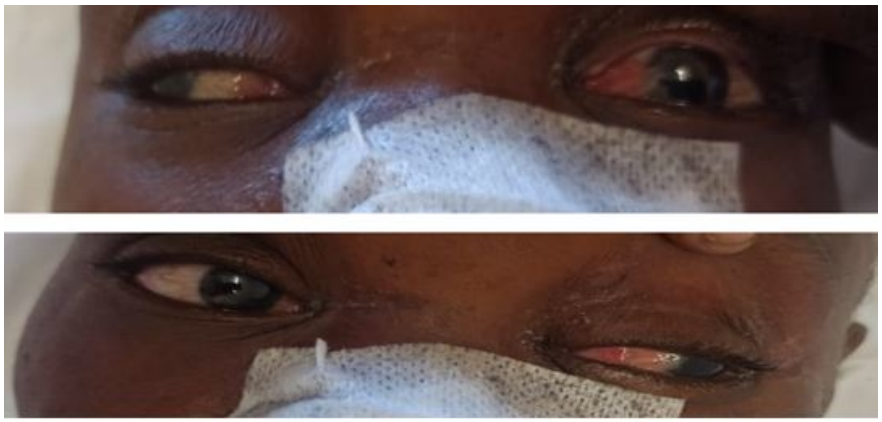

A: there is a lack of adduction of the left eye during the conjugate deviation of the eyes to the right

B: conjugate deviation of the eyes to the left