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Case Report

A Highly Uncommon Variation of "Top of the Basilar" Syndrome

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Abstract

Stroke is a leading cause of death and acquired disability worldwide. Top of the Basilar Syndrome (TOBS), is a relatively uncommon subtype of stroke, and it has a wide spectrum of clinical presentations (abnormalities of alertness, inversion of the sleep–wake cycle, and abnormal oculomotor or pupillomotor functions) due to anatomic variations of posterior circulation, raising diagnostic challenges. In this case study, A 78-year-old man was admitted to our hospital with sudden-onset hemi paralysis and stupor followed by central facial palsy, dysphasia and dysarthria. He also revealed a blepharoptosis of the right eyelid, impairment of abduction on left side. He appeared hypersomnic, exhibited a reversed sleep–wake cycle, and was disoriented in space and time. It was also noticed that, he also had a personality change. Cerebral Magnetic Resonance Imaging (MRI) showed a bilateral thalamic stroke with extension to the right midbrain, temporal and occipital lobes. This unique case represents a highly uncommon variant of the "top of the basilar" syndrome because of a special anatomic arterial variation named Artery of Percheron (AOP).

Keywords: Artery of Percheron; Bilateral thalamic infarction; Top of the basilar syndrome

Introduction

Background

Stroke is one of the most common causes of death and acquired disability in the world. In China, stroke136.64 /100,000 has exceeded malignant tumor (135.88/100,000) and become the first cause of death. The incidence rate of stroke is approximately 205-584/100,000, the morbidity rate is approximately 400-700/100,000. Approximately two thirds of the 6000,000 people who survive a stroke are left with severe disabilities, which place a heavy economic burden on our society [1,2].

Bilateral thalamic infarction, a relatively uncommon subtype of stroke, combined with infarctions in the posterior circulation territories, results in a constellation of symptoms and signs that is generally referred to as "top of the basilar" syndrome [3,4]. Here, a highly uncommon variation of TOBS is discussed with the aim of increasing diagnostic awareness.

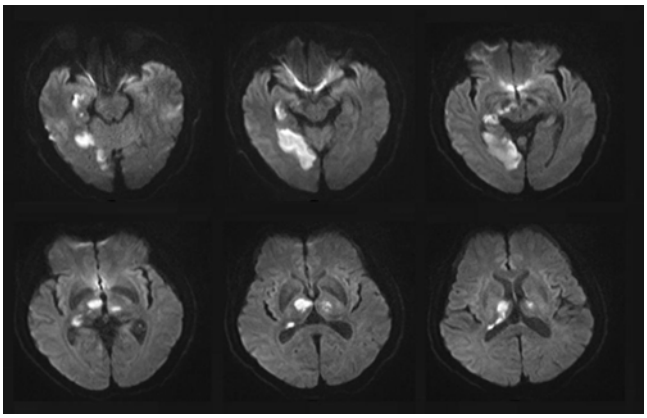

Figure 1: DWI shows infarctions of bilateral thalami, right midbrain, medial right temporal and occipital lobes.

Case Report

A 78-year-old man with an unremarkable medical history experienced sudden weakness of his left leg and simultaneously had difficulty lifting his left arm when he was jogging in the early morning. His gait was unsteady, and he deviated to the left. He also felt dizzy and experienced an overwhelming feeling of drowsiness. He then quickly fell asleep after he was sent home by his neighbors. Approximately seven hours later, his family came home from work and found that he was not very responsive. He was disoriented, his speech was slurred, and the left side of his mouth was drooping when he was finally admitted to the emergency department of our hospital.

Clinical findings

On clinical examination, he was in a stupor. His Glasgow coma score was 12/15; he was able to obey simple commands, but he was disoriented in time and place. His blood pressure was 150/102 mmHg, his pulse rate was 62 beats per minute, respiratory rate was 20 per minute, and his SpO_2 was 94%. An emergency cerebral Computed Tomography (CT) scan without contrast was performed at hospital admission and ruled out an intracerebral hemorrhage. No low-density focus was responsible for his staggered symptoms and signs.

The patient was soon transferred to our neurology department. A neurological examination was performed. A physical examination of cranial nerve function revealed blepharoptosis of the right eyelid, impairment of abduction on the left side, left central facial palsy, dysarthria, and dysphagia. The muscle strength of his left extremities was scored at 2/5. Oppenheimer and Babinski signs were present

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on the left. No sensory deficits were present. A cardiopulmonary examination was unremarkable.

Results of Auxiliary Examinations

Extensive laboratory tests, including a complete blood count, biochemical analyses and fibrinolysis and cardiac troponin measurements, were normal. Holter Electrocardiograph monitoring revealed atrial fibrillation and arrhythmias. Transthoracic echocardiography was unremarkable, and transesophageal echocardiography was contraindicated by persistent arousal disorder. Duplex sonography of the carotid and vertebral arteries revealed light stenosis due to the presence of extensive atheromatous plaques. A chest CT scan revealed mild pneumonia. A cerebral MRI performed on the third day after admission revealed a bilateral thalamic stroke with extension to the right midbrain and the right temporal and occipital lobes (Figure 1). Because the patient became agitated and uncooperative during the latter part of the MRI scan, we were unable to perform cerebral MRA.

Treatment and clinical course

Anti-platelet aggregation therapy was initiated immediately on the day of admission. The patient's left hemiparesis improved within 24 hours, as did his left-side abduction deficit, left central facial palsy, dysarthria and dysphagia. However, he was found to be in a coma on the morning of the second day, so he was transferred to our Neurological Intensive Care Unit (NICU) for further intensive medical care. Heparin therapy had to be stopped after two days' administration because of urethral injury and persistent hematuresis as complications of urinary catheterization. The patient improved quickly with appropriate treatment. In the NICU, he appeared hypersomnic, exhibited a reversed sleep–wake cycle, and was disoriented in space and time. It was also noticed that he had undergone a personality change. He became frustrated and was apt to cry. He underwent an MRA of the head again 20 days later (Figure 2), which revealed no stenosis or occlusion of the basilar and bilateral posterior cerebral arteries. One month later, he was discharged home with a persistent personality change and right-sided ptosis.

Discussion

TOBS is usually characterized by abnormalities of alertness, inversion of the sleep–wake cycle, and abnormal oculomotor or pupillomotor functions [5,6]. Altered mental status ranging from drowsiness or confusion to hyper somnolence or coma is observed with lesions of the bilateral paramedian thalamus [7]. Involvement of the reticular activating system can cause alterations of the sleep–wake cycle. The presence of ptosis and INO usually suggest involvement of the mesencephalon [8]. Hemiparalysis affecting the left extremities together with stupor is rarely observed as the initial symptoms of TOBS. Coma, seizure and INO have also been reported as initial symptoms [9-11], which raises diagnostic challenges. We are unable to explain the deficit in left-side abduction observed in our patient.

The patient's pyramidal symptoms improved quickly, in contrast with his other symptoms of bilateral thalamic infarction, such as his reduced level of consciousness, which continued to progress after admission. To achieve a thorough understanding of the staggered symptoms and signs observed in this patient, it is necessary to review the blood supply of the bilateral thalami

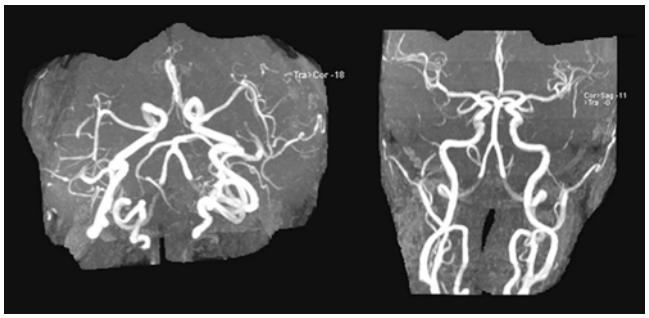

Figure 2: MRA on 20th day shows patent posterior circulation.

[12,13]. There are notable variations and overlaps in the blood supply of the bilateral thalami. The thalamic blood supply is divided into anterior, paramedian, inferolateral, and posterior vascular territories according to the arteries supplying these regions; these arteries arise from several sources. The anterior territory is supplied by the thalamotuberal artery or polar artery, which arises from the posterior cerebral communicating artery (PcomA). The paramedian territory is supplied by the thalamoperforating arteries, also called the paramedian arteries, which arise from the P1 segment of the Posterior Cerebral Arteries (PCAs). The inferolateral territory is supplied by the thalamogeniculate arteries, and the posterior territory is supplied by the posterior choroidal arteries, both of which arise from the P2 segment of the PCAs. In some cases, paramedian arteries also supply the anterior territories and the even midbrain due to anatomical variations in the paramedian arterial supply [14].

Percheron [12] categorized arteries supplying the bilateral paramedian territories into three types. Type I arteries are pairs of arteries that arise from the bilateral P1 segments of the PCAs. Type II arteries are pairs of perforating arteries that arise from the P1 segment of an ipsilateral PCA. Type II arteries are further subdivided into 2 subtypes according to whether both paramedian arteries originate in a common trunk or not. Type IIa arteries originate in a single arterial trunk arising from the proximal PCA that irrigates both paramedian thalamic regions; this single arterial trunk is known as an artery of Percheron and accounts for approximately 30% of variants. The Type III variant is rare; this type of artery bridges both PCAs with an arcade of perforating vessels. Occlusion of an AOP always leads to infarction of the bilateral paramedian thalamus, including anterior territories, with (57%) or without (43%) midbrain involvement [7].

In this case, with evidence of bilateral thalamic stroke with extension to the right midbrain and to the right temporal and occipital lobes and with the patient's constellation of symptoms and signs, TOBS was diagnosed. Caplan3 described "top of the basilar syndrome" in 1980. "Top of the basilar" refers to a vascular structure network, centered at the bifurcation of the basilar artery and with a diameter of approximately 2 centimeters, which consists of the distal basilar artery, proximal bilateral posterior cerebral arteries and superior cerebella arteries. An occlusion of the top of the basilar network results in simultaneous or asynchronous disturbances of the blood supplies to the territories served by these five arteries and can affect the thalamus, midbrain, pons, superior cerebellum, and the medial temporal and occipital lobes [4].

DWI of our patient showed that the primary focus of the infarction

was in the territory of the right PCA, indicating the presence of a proximal occlusion of right PCA. It is difficult to explain the existence of a small area of infarction of the left paramedical thalamus together with an occlusion of the right PCA in this patient. We hypothesize that that it may be due to the presence of an AOP. It seems likely that occlusion of the right PCA combined with the presence of an AOP arising from the right side, rather than synchronous occlusions of the right PCA and the left posterior thalamoperforating artery, was responsible for the infarctions observed in this patient. We further suggest that infarction of the AOP causes a subtype of TOBS. An AOP is rarely observed in the clinic; only several cases in which an AOP was visible on imaging have been reported in the literature [15,16]. Lazzaro et al. found that the presence of the "V" sign, which is a hyper intense signal on FLAIR and DWI sequences, together with midbrain involvement, exhibited a sensitivity of 67% in cases of occlusion of the AOP and supported this diagnosis [7].

To be able to develop appropriate therapies for TOBS that address the underlying etiologies of this disorder, further investigation of the pathogenesis of this disorder is needed. In several retrospective studies, hypertension (34.2%), atrial fibrillation (16.7%), diabetes mellitus (16.6%) and dyslipidemia (15.8%) have been described as risk factors for TOBS. The most common etiology of TOBS is cardioembolism (20.8%), and undetermined etiologies account for 69.2% of cases10. In this case, atrial fibrillation is considered a possible cause. The patient was discharged home with anticoagulation therapy.

Conclusion

TOBS has a wide spectrum of clinical presentations due to anatomic variations in the posterior circulation. Hemi paralysis together with stupor is rarely observed as the initial symptoms of TOBS. It is important for clinicians to notice that symptoms and signs are staggered in this disorder. It is quite possible that occlusion of the proximal right PCA, together with the presence of an AOP, a special branch of the PCA, led to the distinctive imaging pattern observed in this patient. We further suggest that infarction of the AOP causes a subtype of TOBS.

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