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Percheron Artery Syndrome: A Case Report in Pediatrics

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Abstract: ***The Percheron syndrome, first described by the French neurologist Gerard Percheron in 1973, consists of a synchronous bilateral paramedian thalamic infarction resulting in the occlusion of the corresponding artery. The classic triad includes gaze palsy, memory disturbances, and coma. Pediatric patients may present with seizures during the perinatal period. Magnetic resonance imaging is the preferred imaging technique for diagnosis. We present the case of a preschool patient, 4 years and 8 months old, who presents to the emergency department with altered consciousness and bilateral thalamic ischemic process.***

Keywords: Stroke, posterior cerebral artery infarction, thalamus, magnetic resonance imaging

1. Introduction

The Percheron syndrome, named in honor of the French neurologist Gerard Percheron, who first described it in 1973, is characterized by synchronous paramedian bilateral thalamic infarction resulting in occlusion of the corresponding artery, leading to a sudden sensory deterioration. Thalamic irrigation largely depends on the posterior communicating and posterior cerebral arteries, along with their branches, although multiple anatomical variants have been observed, such as the absence of polar branches in up to 30% of the population, which can be replaced by Percheron or paramedian arteries. Due to its rarity, Percheron syndrome is an uncommon disorder with few documented cases, which complicates its diagnosis (1). The occlusion resulting in Percheron artery infarctions only accounts for 0.3% of ischemic stroke cases (2). Cardioembolic origin the most common etiology of Percheron syndrome (3). Clinically, it manifests with consciousness alterations, including coma, with fluctuations, as well as disorders of language, mood, cranial nerves, movement, memory (amnesia), and sleep (1). The classic triad includes gaze palsy, memory impairments, and coma (3). In pediatric patients, symptoms may be similar to those in adults but may also include seizures during the perinatal period (4). Magnetic resonance imaging (MRI) is the preferred imaging technique for diagnosis. This work aims to present a case of bithalamic ischemic stroke due to the presence of an anatomical variant resulting in the common origin of the paramedian artery (of Percheron).

2. Case Report

Our case involves a 4-year and 8-month-old male patient who presented to the emergency department with a clinical picture evolving over approximately 24 hours, characterized by "weakness" that has intensified to the point of somnolence, associated with fever spikes and hyporexia. On the inicial physical examination, there was decreased level of consciousness with a Glasgow Coma Scale score of 13/15, with poor response to verbal stimuli. The patient's relevant medical history includes nephrotic syndrome diagnosed 5 months ago, treated with prednisolone with poor adherence, as well as an Epstein-Barr virus infection 2 months ago.

On physical examination, the patient was somnolent, with poor interaction with the examiner and exhibited ataxic gait. Further questioning revealed a history of head trauma 2 days prior. Initially, the patient was managed as moderate traumatic brain injury and possible viral encephalitis. Serial laboratory tests were ordered, and due to clinical deterioration, the patient was transferred to the pediatric intensive care unit.

Upon admission to the unit, the patient was stuporous, encephalopathic, with poorly responsive pupils. Cultures were obtained, including cerebrospinal fluid, and empirical antibiotic coverage with third-generation cephalosporin + glycopeptide was started. Acyclovir was added due to suspicion of viral etiology, possibly herpes virus. Initial laboratory results showed a positive procalcitonin level of 10.9 mg/dL, slight elevation in transaminases, and cerebrospinal fluid analysis revealed no abnormalities, pending culture and meningeal FilmArray. A head CT scan revealed poorly defined bilateral hypodense thalamic images, raising suspicion of either ischemic or infectious etiology.

The patient continued to deteriorate with fluctuating consciousness, ranging from lethargy to agitation, with preserved muscle strength, hyperreflexia in all four limbs, dysarthria, and reactive intermediate pupils. The report from the meningeal FilmArray was negative. An MRI was ordered for better visualization of the lesion observed on the CT scan. Due to the unusual course and negative infectious profile, suspicion of autoimmune encephalitis was considered, and markers for this were requested in the cerebrospinal fluid.

Two days later, an MRI was performed, revealing hypointense lesions on T1-weighted sequences and hyperintense lesions on FLAIR and T2-weighted sequences, symmetrically involving the bilateral thalamus, suggestive of a hypoxic ischemic process. Additionally, laboratory tests showed decreased antithrombin III levels, as well as low levels of TSH (0.91) and T4L (< 0.42), both below their respective reference values. Pediatric neurosurgery assessed the patient and recommended surgical treatment, suggesting a possible diagnosis of Percheron syndrome. Subsequently, pediatric neurology confirmed the diagnosis, attributing the underlying cause to the patient's hypercoagulable state due to nephrotic syndrome, along with central hypothyroidism caused by bilateral thalamic ischemia.

The patient was started on comprehensive physical therapy, showing significant improvement. Nephrotic syndrome control was achieved, along with adherence to corticosteroid therapy. The patient was discharged in good condition, with improved gait, and prescribed oral corticosteroids and Warfarin as an anticoagulant. A follow-up evaluation at 2 months post-event revealed normalization of TSH and T4L levels.

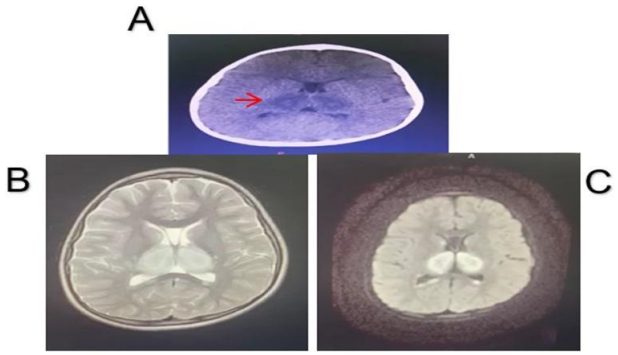

Figure 1: Image of the patient with Percheron syndrome (PS). A. CT scan image showing bilateral thalamic hypodensities. B. Brain MRI axial T2-weighted sequence image demonstrating hyperintense lesions in bilateral thalamic regions. C. Brain MRI FLAIR sequence image depicting bilateral thalamic hyperintense areas, indicative of hypoxic ischemic process

3. Discussion

Percheron artery syndrome presents as a variant of the paramedian artery or basilar communicating artery. It was described by Percheron in the 1960s, who observed different forms of its origin, including a symmetric, bilateral origin, and an asymmetric origin, where the right and left thalamic arteries arise from a common communicating artery (5).

The thalamus is irrigated by several arteries originating at the base of the skull, including the anterior communicating artery and the posterior cerebral artery. The term Percheron artery or Percheron paramedian artery refers to the first portion of the posterior cerebral artery coursing between the upper end of the basilar artery and the exit of the posterior communicating artery; its occlusion leads to bilateral thalamic infarction resulting in diverse symptomatology (5).

Percheron syndrome, also known as bilateral thalamic infarction, is a very rare entity, accounting for 0.1 to 0.3% of all ischemic strokes, which poses a diagnostic challenge for healthcare personnel and requires a high clinical suspicion for early diagnosis (6).

The most common etiologies are cardiogenic embolism and small vessel arterial disease. In some patients, the presence of a patent foramen ovale is associated with the occurrence of embolisms and infarctions in median territories (7).

The typical clinical presentation consists of a classical triad, including altered consciousness, cognitive impairment, and vertical gaze palsy. Additionally, hemiplegia, cerebellar ataxia, and oculomotor deficits may be present (8).

The diagnosis of Percheron syndrome is made through MRI, with special emphasis on diffusion-weighted sequences. The MRI typically shows restricted diffusion in the bilateral paramedian thalamic region, with or without involvement of the rostral midbrain, corresponding to the vascular territory of the Percheron artery. The characteristic lesions are described as V-shaped hyperintensity on FLAIR and DWI sequences along the pial surface of the midbrain next to the interpeduncular fossa (7).

In the presented case, poorly defined bilateral thalamic hypodense lesions were seen on plain cranial CT, with hyperintense areas on T2 and FLAIR sequences suggestive of bilateral thalamic hypoxic ischemic process on brain MRI.
Initial treatment in the hyperacute stage involves the use of intravenous or intra-arterial thrombolytics in the adult population, but it is not approved for pediatric patients. Additionally, endovascular treatment with thrombectomy can be attempted within the first 6 hours of symptom onset. Beyond this time window, anticoagulant therapy is indicated (7), as was performed in this case.

Our patient had a satisfactory clinical evolution, achieving control of the underlying condition through corticosteroid use, which reduces the procoagulant state; additionally, the patient received physical and speech therapy with an appropriate response. In conclusion, Percheron syndrome is a very rare entity in the general population, even more so in the pediatric population, so recognizing the classical symptoms would lead to a higher clinical suspicion and thus a timelier diagnosis.

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