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A Rare Case Report: Stroke Infark In A 5 Years Old Children with Suspected Moya-Moya Disease

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Abstract

Ischemic stroke in children is extremely rare, with an incidence rate of approximately 1.0-2.0 per 100,000 children annually in Western countries. The highest incidence occurs in infants and children under 5 years, with a higher prevalence among males than females and among Black and Asian children than White children. Ischemic stroke is commonly associated with arteriopathy, particularly moyamoya disease. A 5-year-old male presented to the Emergency Department with weakness of the right limbs for 1 day, starting in the morning upon waking, accompanied by left-sided facial drooping and slurred speech. Vital signs were normal, general physical examination was unremarkable, with motor strength 3/5 in the right arm and 2/5 in the right leg. Laboratory results were normal, and CT scan revealed focal ischemia in the left lentiform nucleus. Acute stroke symptoms in children are similar to those in adults, commonly presenting with hemiparesis, hemifacial weakness, and speech or language disturbances. Pediatric ischemic stroke often has uncertain risk factors, including moyamoya disease, characterized by chronic, progressive stenosis of the distal internal carotid artery and proximal middle cerebral artery. Neuroimaging is crucial for diagnosis, with CT scans of the head being essential but limited in detecting acute ischemia, necessitating MRI. Conventional angiography remains the gold standard for cerebrovascular imaging, defining specific arteriopathy. This patient underwent a head CT scan revealing left lentiform nucleus ischemia and was referred to PON Hospital for MRI and further management. The diagnosis of stroke in children is primarily based on clinical presentation and radiological findings, allowing for targeted management given the heterogeneous etiology.

Keywords: Stroke, Infark, Children, Moya-moya Disease

PENDAHULUAN

Stroke is a major cause of neurological disability not only in adults but also in children. Although its incidence is lower than in the adult population, pediatric stroke remains a critical condition that is often underrecognized because its symptoms may mimic other neurological disorders. Pediatric stroke is defined as a clinical and radiological manifestation of cerebral infarction or hemorrhage, and therefore diagnosis requires confirmation through neuroimaging (Danu & Hidayah, 2021). Epidemiologically, the incidence of ischemic stroke in children is estimated at approximately 1–2 per 100,000 children per year, with the highest rates occurring in infants and children under 5 years old, and with a greater prevalence among males and among Asian and African ethnic groups (Golomb et al., 2009; Lehman et al., 2018). The middle cerebral artery is the vascular territory most frequently involved in childhood arterial ischemic stroke (AIS).

The etiology of AIS in children is highly diverse, including infection, coagulation disorders, trauma, and arteriopathies. Among the arteriopathies, moyamoya disease is one of the most significant causes, especially in Asian populations. Moyamoya disease is characterized by progressive stenosis of the terminal internal carotid artery and the proximal segments of the anterior and middle cerebral arteries, leading to the formation of abnormal collateral vessels that appear as a "puff of smoke" on angiography (Kuroda & Houkin, 2008; Scott & Smith, 2009). The disease exhibits two peak ages of onset, one of which occurs in children aged 5–9 years, and it tends to be progressive, with unilateral lesions potentially evolving into bilateral involvement within two years (Kelly et al., 2006).

Clinical manifestations of pediatric stroke commonly include hemiparesis, facial weakness, speech impairment, and other focal neurological deficits, as demonstrated in the present case. Neuroimaging plays a crucial role in early diagnosis; head CT is usually the initial modality used

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to exclude intracranial hemorrhage, although its sensitivity for detecting acute ischemia is lower than that of MRI (Rajani et al., 2018). In this case, the CT scan revealed a focal ischemic lesion in the left lentiform nucleus, as shown in Figure 1, presenting as a hypodense area consistent with an acute infarct. This finding is highly relevant because the basal ganglia are among the common infarct locations in pediatric arteriopathies, including moyamoya disease.

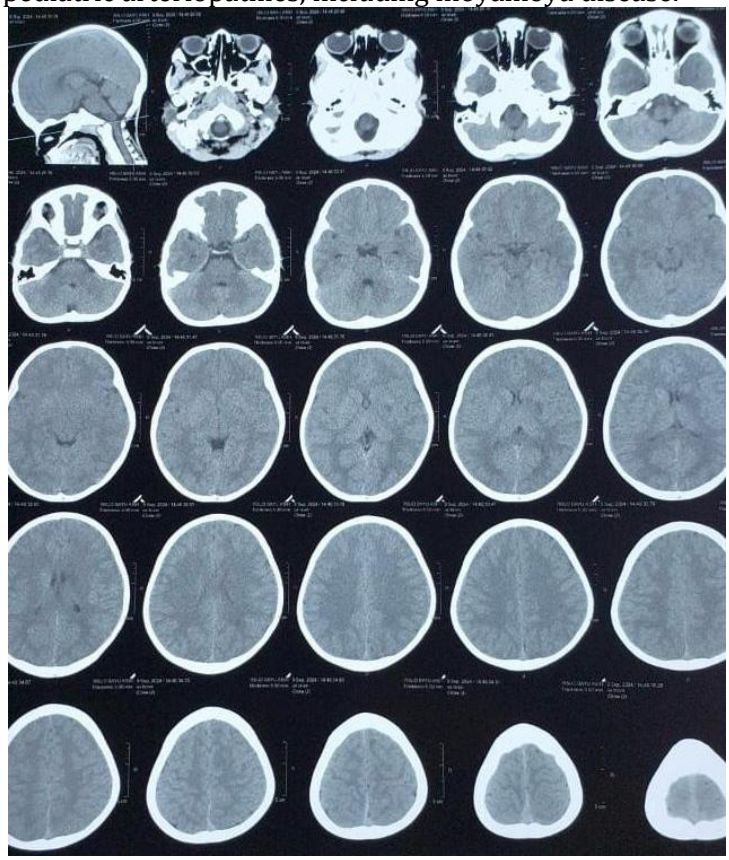

Figure 1. Head CT scan of a 5-year-old boy showing focal ischemia in the left lentiform nucleus

Identifying stroke in children is crucial because more than 50% of survivors are at risk of developing long-term disabilities, cognitive impairment, or epilepsy (Ferriero et al., 2019). Therefore, further evaluation using MRI and angiography is essential to determine the underlying etiology, particularly when arteriopathies such as moyamoya disease are suspected. The case of this 5-year-old boy underscores the importance of early detection and a multidisciplinary approach in the management of pediatric stroke, given its heterogeneous etiology and the potential for significant long-term neurological complications.

METHOD

The research method used in this case report employed a clinical descriptive approach aimed at providing a comprehensive overview of the presentation, radiological findings, and management process of a 5-year-old boy with suspected ischemic stroke due to an underlying arteriopathy, including the possibility of moyamoya disease. Data were obtained through direct observation of the patient's condition in the Emergency Department and inpatient ward, supported by neurological physical examinations, assessment of vital signs, and evaluation of motor function using a standardized muscle strength grading scale. Additional diagnostic investigations included basic laboratory tests—such as hematology, electrolytes, and inflammatory markers—to rule out infectious or metabolic etiologies that may mimic pediatric stroke. Initial neuroimaging with a head CT scan was performed to identify potential cerebral infarction or intracranial hemorrhage, in accordance with Ferriero et al. (2019), given that CT scanning is a rapidly accessible modality crucial for early identification. The CT findings showing

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an ischemic lesion in the left lentiform nucleus served as the basis for referral to a tertiary center for further diagnostic evaluation using MRI and angiography, consistent with AHA/ASA guidelines emphasizing advanced neuroimaging when pediatric arteriopathy is suspected. Additionally, a systematic review of medical history was conducted through interviews with the patient's parents and examination of medical records to identify risk factors, prior infections, or previous neurological symptoms. Data analysis was carried out qualitatively by comparing clinical and radiological findings with current literature on pediatric stroke and moyamoya disease to ensure diagnostic accuracy and guide case management. This approach allowed for a thorough understanding of the disease progression and informed subsequent clinical decision-making.

RESULTS AND DISCUSS

Acute stroke in children presents with clinical manifestations similar to those seen in adults, most commonly hemiparesis, hemifacial weakness, and speech or language disturbances, as demonstrated in this case. Pediatric arterial ischemic stroke, however, often occurs without a clearly identified risk factor, and arteriopathies—particularly moyamoya disease—represent one of the most important underlying etiologies in children (Golomb et al., 2009). Moyamoya disease, derived from the Japanese term meaning "puff of smoke," refers to the angiographic appearance of fragile collateral vessels that form in response to progressive stenosis of the distal internal carotid artery (ICA) and the proximal segments of the anterior and middle cerebral arteries (Kuroda & Houkin, 2008; Scott & Smith, 2009). The pattern of vascular involvement varies depending on the site of occlusion and includes proximal M1 occlusion with extensive MCA infarction, distal M1 occlusion affecting the basal ganglia, M2 branch occlusion involving frontal, parietal, or temporal regions, and lenticulostriate artery involvement affecting the basal ganglia and deep white matter. Such distinct vascular patterns play an important role in predicting the etiology of ischemic stroke and frequently indicate an underlying arteriopathy (Kelly et al., 2006).

The global incidence of moyamoya disease ranges from 0.086 per 100,000 individuals in the United States to approximately 0.54 per 100,000 worldwide, with substantially higher prevalence in East Asia—particularly Japan and South Korea—and a notable predominance in females (Kuroda & Houkin, 2008). In children, unilateral involvement occurs in approximately 18% of cases but may progress to bilateral disease within two years (Kelly et al., 2006). Pediatric moyamoya most commonly manifests with cerebral ischemia (approximately 80% of cases), while hemorrhagic events are less frequent (Scott & Smith, 2009). Children typically present with MCA territory symptoms such as hemiparesis, aphasia, sensory loss, and visual field deficits.

Neuroimaging plays a critical role in establishing the diagnosis of ischemic stroke. A head CT scan is usually the first imaging modality performed to exclude intracranial hemorrhage, although its sensitivity for detecting acute ischemia in children is limited, ranging from 16% to 56% (Rajani et al., 2018). MRI, particularly diffusion-weighted imaging (DWI), provides earlier and more accurate detection of acute infarction, while perfusion-weighted imaging (PWI) evaluates cerebral hemodynamics. Magnetic resonance angiography aids in identifying stenosis or abnormal collateral vessels. However, conventional cerebral angiography remains the gold standard for diagnosing moyamoya disease due to its superior ability to delineate vascular architecture and identify arteriopathy when CT and MRA findings are inconclusive (Scott & Smith, 2009).

Management of pediatric ischemic stroke differs substantially from adult protocols because randomized clinical trials in children are limited. Evidence does not strongly support the routine use of antiplatelet or anticoagulant therapy in the acute phase; however, the American Heart Association recommends either low-molecular-weight heparin or low-dose aspirin (1 mg/kg/day) until a definitive etiology is established (Ferriero et al., 2019). A prospective study demonstrated no significant difference in preventing recurrent stroke between these two therapies, emphasizing the need for individualized treatment decisions. Children with presumed ischemic stroke should therefore be managed in specialized pediatric centers with expertise in neurovascular disorders. Revascularization surgery—either direct, indirect, or combined

techniques—remains the definitive treatment for moyamoya disease, aiming to improve cerebral perfusion and reduce recurrent ischemic events (Kuroda & Houkin, 2008).

Long-term outcomes highlight the importance of early diagnosis and intervention. More than 50% of children who survive a stroke develop moderate to severe neurological sequelae or epilepsy, underscoring the critical role of rehabilitation targeting motor, speech, cognitive, behavioral, and psychosocial domains (Ferriero et al., 2019). Although pediatric stroke is relatively rare, it is clinically important because of its distinct risk factors, broader differential diagnoses, and potentially severe complications. Early recognition by clinicians is essential, as pediatric stroke carries a mortality rate of 5–10% and is among the top ten leading causes of death in children (Golomb et al., 2009; Ferriero et al., 2019).

CONCLUSION

Stroke in children is characterized by clinical and radiological manifestations, accompanied by radiological evidence of cerebral infarction or hemorrhage. The etiology of pediatric stroke is diverse, necessitating comprehensive diagnostic procedures to identify underlying causes. Treatment strategies are tailored to the specific etiology and individual child's condition. Prompt recognition of childhood stroke is crucial, enabling the implementation of targeted and neuroprotective therapies.

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