CASE REPORT

Acute Infarction, Mineralizing Lenticulostriate Angiopathy (Case Report)

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ABSTRACT

A three-year-old boy presented with left hemiparesis after an alleged fall at home. It was associated with gait disturbance, saliva drooling, and slurred speech. Physical examination revealed normal vital signs, left hemiparesis, and tongue deviation to the right side. Blood gases showed respiratory acidosis while other laboratory investigations were normal. Non-contrast computed tomography (CT) brain, electrocardiography, and echocardiography revealed no abnormalities. T2-weighted and FLAIR magnetic resonance imaging (MRI) brain showed signal hyperintensity at the right putamen extending to the right corona radiata. There was restricted diffusion on diffusion-weighted images and signal hypointensity on apparent diffusion coefficient maps, suggestive of an acute infarct. The diagnosis was revised to mineralizing lenticulostriate angiopathy. The patient was started on oral aspirin, physiotherapy, and occupational therapy, which helped him make a full recovery.
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INTRODUCTION

Infarction of the basal ganglia after minor head injury is a well-described entity. "Mineralizing angiopathy" is a condition characterized by mineralization along the lenticulostriate vessels and it usually presents with acute stroke after minor head injury (1). The supratentorial compartment is relatively mobile compared to the fixed infratentorial compartment, predisposing it to greater shear forces between the perforating vessels and the brain parenchyma during trauma. Another postulate is the acute angulation of the lenticulostriate vessels, suggesting the propensity of the basal ganglia for injury. Children with mineralizing angiopathy usually have an excellent prognosis.

CASE REPORT

A three-year-and-seven-month-old Malay boy presented to the emergency department with left hemiparesis after an alleged fall at home. There were no eyewitnesses to establish the mechanism of the fall. After the fall, he complained of pain in the left side of his head. There were no external injuries. The patient was well until 15 minutes later when his mother noticed that he developed weakness in his left upper limb. He also developed an abnormal gait in which he dragged his left lower limb along and occasionally fell. During feeding, his mother noticed saliva drooling from the left angle of his mouth and that his tongue was shifted to the right side. The patient also had slurred speech. Otherwise, there was no loss of consciousness, altered sensorium, seizures, fever, or respiratory tract symptoms

At the emergency department, the patient's GCS score was 15/15, blood pressure was 107/53 mmHg, heart rate was 75 beats/minute, radial pulse was regular and strong, respiratory rate was 26 breaths/minute, and temperature was 37.3°C. He weighed 17.5 kg and height of 90 cm. The pupils were 3/3 and reactive bilaterally. There was no facial asymmetry, ptosis, drooling of saliva, or drooping of the angle of the mouth. Inspection of the oral cavity revealed that the tongue was deviated to the right side and the uvula was located centrally. The patient was able to protrude his tongue and the gag reflex was present. The left upper limb was partially flexed and pronated. The left upper and lower limbs had an MRC

grade of 1/5, poor tone, and 1+ reflexes. The Babinski sign was positive on the left side. Examination of the right side was normal. There were no fasciculations seen and no clonus elicited. Examination of other systems was normal

Full blood count and renal profile were normal while his venous blood gases revealed respiratory acidosis (pH: 7.25, PCO2: 52 mmHg, PO2: 29 mmHg, HCO3-: 22.8 mmol/L). He was provided with a high-flow oxygen mask at 10 L/minute and IV maintenance fluid therapy using normal saline with dextrose 5% at 55 mL/hour. A non-contrast CT brain revealed no tumour, bleeding, or oedema. The patient was given a provisional diagnosis of acute stroke with left hemiparesis complicated with bulbar palsy and gait disturbance.

The patient underwent an MRI brain under sedation the next day (Fig. 1,2). T2-weighted and FLAIR MRI images revealed a well-defined signal hyperintensity measuring 1.0 x 0.8 cm (AP x W) at the right putamen extending to the right corona radiata (T2-weighted sequences are used to show haemorrhage in various lesions, including vascular malformations). There was also restricted diffusion on diffusion-weighted images (DWI) and signal hypointensity on apparent diffusion coefficient (ADC) maps, suggestive of an acute infarct.

A small tubular void was noted at the right cerebellum, suggestive of a developmental venous anomaly. There were no other structural abnormalities. MRI angiography showed normal middle, anterior, and posterior cerebral artery signal flow. There were no signs of vascular malformations, aneurysms, or vasculitis, and no brain stem involvement. MRI findings show an acute infarct at the right putamen (lentiform nucleus) (Figure 3).

On the third day of admission, the patient was still experiencing weakness (MRC grade 2/5) in the left upper and lower limbs. He passed the formal swallowing test but was found to have mild dysarthria. Nonetheless, a nasogastric tube was inserted for feeding. The patient was referred for physiotherapy and occupational therapy. The case was reviewed by the Hospital Sultan Aminah paediatrics team who then suggested a revised diagnosis of mineralizing lenticulostriate angiopathy. The local team was advised to initiate oral prophylactic aspirin 75 mg (3.5 mg/kg) OD. The diagnosis was explained to the mother, and she was counselled regarding the importance of long-term follow-up, rehabilitation, fall prevention, and stroller use. She was also reassured that the patient had a good prognosis.

Muscle power in the left upper and lower limbs improved to an MRC grade of 5/5 over the next few days with the help of physiotherapy and occupational therapy. There were no choking episodes and his vital signs remained stable throughout the admission. Before the patient was discharged home, he was screened for thrombophilia, factor V Leiden, protein C and S abnormalities, and connective tissue disorders with serum C3 and C4, anti-nuclear antibodies, anti-double stranded DNA antibodies, and rheumatoid factor. The patient was discharged home with oral aspirin 75 mg OD. His screening results were reviewed during a follow-up visit at the paediatric neurology clinic. His serum C3 and C4 levels were within the normal ranges at 1.32 g/L and 0.22 g/L respectively. His rheumatoid factor was negative at <8 IU/mL.

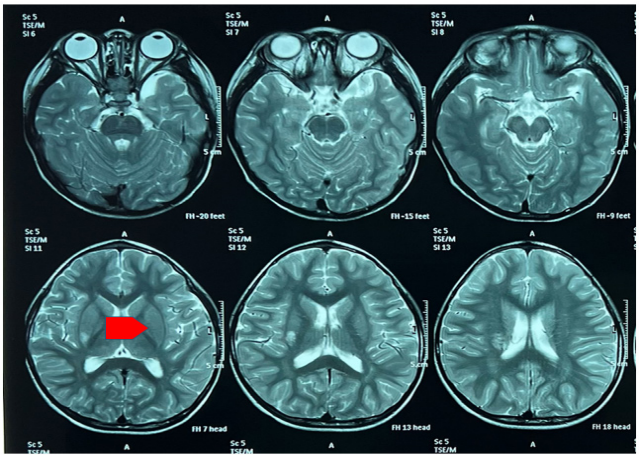

Figure 1: MRI brain images showing lesions at the putamen area (indicated by the red arrow).

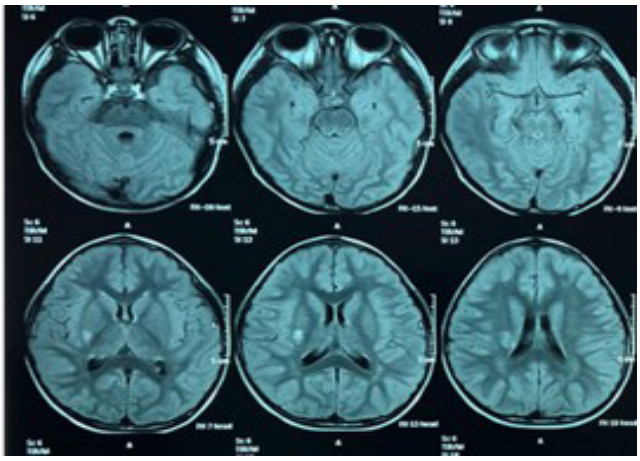

Figure 2: FLAIR sequence.

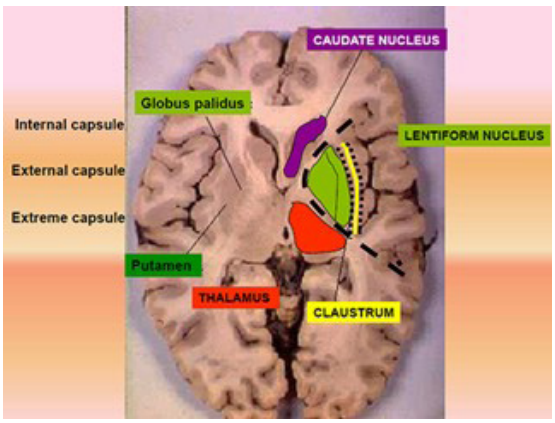

Figure 3: Diagram of the lentiform nucleus (putamen and globus pallidus).

DISCUSSION

Lingappa et al. 2014, described mineralizing angiopathy as a clinical-radiological stroke syndrome in children. It is characterized by cerebral parenchymal calcifications along the lenticulostriate vessels. Lacunar infarcts are diagnosed based on clinical presentation and radiologic imaging (1). Patients usually present with hemiparesis and facial paresis minutes to hours after a minor head injury. Neuroimaging would reveal basal ganglia infarction and linear calcifications along the lenticulostriate vessels. Mineralizing angiopathy is also characterized by a favourable prognosis with minimal risk of recurrence (1).

Lacunar stroke or lacunar cerebral infarct is the most common type of ischaemic stroke and results from occlusion of small penetrating arteries. During cadaver dissections of stroke patients, Charles Miller Fisher observed empty spaces, known now as "lacunae" in deep brain structures with occlusion of small penetrating arteries. Five classical lacunar stroke syndromes have been described by Giacomozzi S. et al. 2020 (2):
1. Pure motor stroke (most common: 33 – 50%)
2. Ataxic hemiparesis (second most common)
3. Dysarthria-clumsy hand syndrome
4. Pure sensory stroke
5. Mixed sensorimotor stroke

Pathophysiology

According to Koffler et al. 2019 (3), lacunes result from occlusion of penetrating arteries which originate directly from the constituents of the circle of Willis, the cerebellar arteries, and/or the basilar artery. Lesions associated with lacunes are also observed in the deep nuclei of the brain (putamen: 37%, thalamus: 14%, caudate: 10%), the pons (16%), or the posterior limb of the internal capsule (10%). These lesions are less common in other brain regions such as the cerebellum, the cerebral white matter, and the anterior limb of the internal capsule.

Atheroma formation is the most common mechanism of arterial occlusion. Atheromas can be found in either the main arteries (luminal atheroma) or the origins of the penetrating arteries (junctional atheroma). or due to artery stenosis. Alternatively, hypoperfusion may also be caused by penetrating artery stenosis. When no evidence of small vessel disease could be found in histologic examinations, an embolic cause is assumed, either originating from another artery or the heart. Reports suggest that 25% of patients with clinically and/or radiologically defined lacunar stroke have a cardiac cause for the infarct (4).

CONCLUSION

Mineralizing angiopathy is an underrecognized cause of stroke in children. This case report demonstrates the characteristic development of focal neurological deficits after a minor head injury in a child. Further research is required to establish the pathophysiology of mineralizing angiopathy.

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