Mineralizing angiopathy with basal ganglia stroke in an infant

Puneet Jain[1,2], Praveen Kishore[1], Jasjit Singh Bhasin[1], Subhash Chand Arya[1]

[1]Department of Pediatrics, [2]Division of Pediatric Neurology, BL Kapur Super Speciality Hospital, New Delhi, India

Abstract

Basal ganglia stroke is known following trivial head trauma. Recently a distinct clinic-radiological entity termed 'mineralizing angiopathy' was described. We report an infant who developed basal ganglia stroke following trivial fall. His clinic-radiological features are described.

Key Words

Intracranial calcification, mineralizing angiopathy, stroke, trauma

For correspondence:
Dr. Puneet Jain, Department of Pediatrics, Division of Pediatric Neurology, BLK Super Speciality Hospital, New Delhi - 110 005, India.
E-mail: puneet_mpa@yahoo.com

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Case Report

A developmentally normal, 12-months-old boy presented with paucity of movements of right upper and lower limbs noted for 7 days, following a fall from the bed. There was no loss of consciousness, seizures, and bleeding following the fall. The right-sided weakness was slowly improving over the last 7 days. There were no previous similar episodes and family history was not significant.

Examination revealed irritability, absence of neurocutaneous features, and normal fundus. There were poor antigravity movements of the right upper and lower limbs with decreased tone and depressed muscle stretch reflexes on the right side. He could not stand with support, but could sit unsupported with asymmetrical lateral propping response. There was no cranial palsy. Neuroimaging is shown in Figures 1 and 2. Further investigations showed microcytic hypochromic anemia (hemoglobin 9 g/dL), normal protein C and S and anti-thrombin III levels, negative lupus anticoagulant and anticardiolipin antibodies, normal homocysteine levels, and normal lipid profile. Arterial blood gas, lactate, ammonia, plasma tandem mass spectroscopy, and urinary gas chromatography-mass spectroscopy were normal. Serum calcium, phosphorus, alkaline phosphatase, and intact parathyroid hormone were also normal. Human immunodeficiency virus (HIV) and Cytomegalovirus (CMV) serologies were negative. The echocardiography was normal. The clinic-radiological presentation may be consistent with mineralizing angiopathy with basal ganglia stroke.

He was started on 3 mg/kg/day of oral aspirin, iron supplements, and physical rehabilitation. He showed gradual improvement. He could walk independently at 21 months of age with mild dragging of the right foot. The power of the right limbs had improved to around 4/5, but preferred to use left hand. The tone and reflexes were normal.

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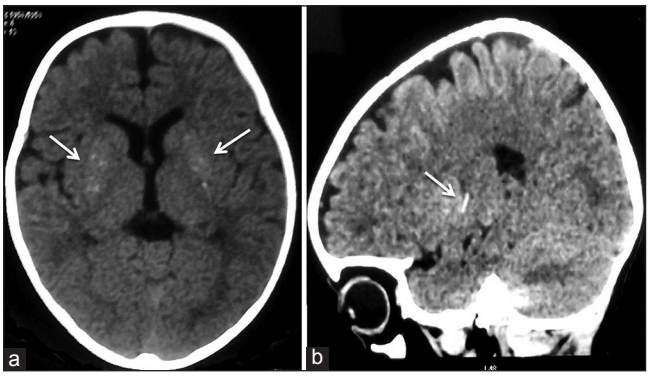

Figure 1: Neuroimaging of the child (a) Axial CThead image shows bilateral punctate lentiform nuclei calcifications (b) The sagittal image shows mineralization of the lenticulostriate artery (arrow). CT = Computed tomography

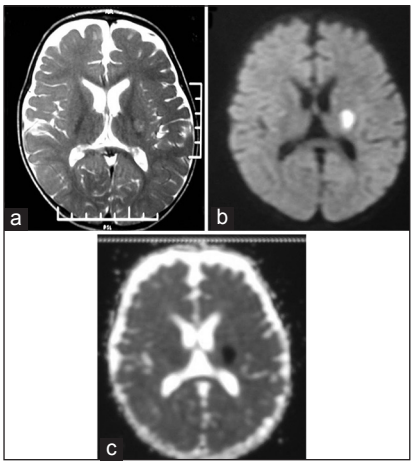

Figure 2: (a) T2-weighted axial MRI image shows left lentiform hyperintensity which showed restricted diffusion on diffusion weighted images (b) and hypointensity on ADC maps (c) GRE sequences and MR angiography were normal (not shown). MRI = Magnetic resonance imaging, ADC = apparent diffusion coefficient

Discussion

Basal ganglia stroke following trivial head trauma has been described previously.[1,2] The proposed mechanisms include transient arterial spasm, mechanical disruption of the flow in the perforating arteries, and intimal trauma with subsequent thrombosis. The acute angle at which perforating arteries arise from the middle cerebral artery in children predispose these vessels to stretching, and distorting forces even after trivial head injury.[3] Further, due to elasticity of the pediatric skull, the shearing forces are stronger.

A detailed history of any traumatic events preceding the stroke must be recorded. Further, as the pediatric strokes have multifactorial etiology, work up for other causes is also mandatory.

Yang *et al*.,[4] reported basal ganglia calcifications in 10 out of 16 infants with stroke following trivial trauma and identified as the potential risk factor. Similar observations have been made previously.[5-7] None of these studies had demonstrated calcified lenticulostriate vessels. The exact mechanism by which calcification was contributing to the development of stroke was largely unknown.

Recently, Lingappa *et al*.,[8] described a distinct clinic-radiological entity of mineralizing angiopathy. The classical phenotype consisted of a previously healthy 6-24-months-old infant with basal ganglia stroke following trivial trauma, with or without transient hemidystonia, linear mineralization along lenticulostriate arteries, and good short-term neurodevelopmental outcome. This case had similar presentation except for the absence of dystonia.

They proposed this entity as the severe and persisting form of sonographic lenticulostriate vasculopathy. This entity may be seen in 0.1% of all live born neonates and tend to regress with time. It is easily picked up by ultrasonography, but not by computed tomography (CT) or magnetic resonance imaging (MRI). The underlying lenticulostriate vasculopathy may predispose the infant to the vascular effects of trauma. The trauma-related stress across the mineralized lenticulostriate vessels may then cause thrombosis with subsequent stroke. This predisposition appears to be age related. Majority of the affected children reported by Lingappa *et al*.,[8] were less than 18 months.

The reported case was an infant who developed basal ganglia stroke following trivial trauma with calcified lenticulostriate artery with good short-term outcome. His clinic-radiological presentation was thus consistent with mineralizing angiopathy with basal ganglia stroke.

Mineralizing angiopathy may be an underrecognized cause of pediatric stroke. Thin-section spiral CT with multiplanar reconstructions may be prudent in children developing focal deficits following trivial trauma. Further research is required to explore the mechanisms underlying mineralizing angiopathy and long-term prognosis of such patients.

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