A rare case of stroke- mineralizing angiopathy

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Abstract
Mineralising angiopathy is a condition characterized by parenchymal cerebral calcifications, and is usually seen in children as a complication of cranial radiotherapy and chemotherapy [1-2]. Basal ganglia stroke is known following trivial head trauma. Recently a distinct clinic-radiological entity termed ‘mineralizing angiopathy’ was described. His clinic-radiological features are described. We report an infant who developed left sided hemiparesis following trivial fall. It is a rare entity.

Keywords: Intracranial calcification, mineralizing angiopathy, stroke, trauma

Introduction
Mineralising angiopathy is a condition characterized by parenchymal cerebral calcifications, and is usually seen in children as a complication of cranial radiotherapy and chemotherapy. Basal ganglia stroke has been reported to occur in infants in association with minor falls. Affected infants present with facial paresis and hemiparesis soon after the injury and small infarcts in the basal ganglia are visible on neuro imaging. This condition has been reported to have a favourable prognosis. Basal ganglia stroke following trivial head trauma has been described previously [3, 4]. 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. [5] Further, due to elasticity of the pediatric skull, the shearing forces are stronger. 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. 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. Pathologically, lenticulostriate vasculopathy is characterized by thickened hypercellular vessel walls with intramural and perivascular mineralization. Although lenticulostriate vasculopathy is easily visualized on neurosonography, the vascular changes are generally not picked up on CT or MRI.

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.

Case Report
A developmentally normal, 1 year old male child presented with left sided hemiplegia following a minor fall while playing. There was no loss of consciousness, seizures, and bleeding following the fall but the child was very irritable. There were no history of similar episodes in the past and family history was not significant. Examination revealed irritability, absence of neurocutaneous features, and normal fundus. There were poor antigravity movements of the left upper and lower limbs with decreased tone and absent reflexes, both cortical and non-cortical sensations were intact. He could not stand with support. There were no signs of meningitis or raised intra-cranial pressures. There was no cranial nerve palsy. On the same day he had three episodes of left sided focal convulsions and was started on anti-epileptic treatment.
Further investigations showed normocytic normochromic in hemogram (hemoglobin 10.4 g/dL), homocysteine levels, protein C and S levels were normal, negative lupus anticoagulant and anticardiolipin antibodies, 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. All the other causes of infantile hemiplegia were ruled out.

**Neuroimaging**

CT Scan Brain was done which revealed acute infarct in right putamen and corona radiata with calcification of lenticulostriate artery with mild cerebral edema.

MRI Brain reported acute non-haemorrhagic infarct in dorsal aspect of putamen, posterior limb of internal capsule, body of caudate nucleus and corona radiata on right side (lateral lenticulostriate arterial territory on right side) with calcification in lateral lenticulostriate artery suggesting mineralizing angiopathy.

MR angiography and venography were normal.
He was started on oral aspirin at 3mg/kg/day, anti-epileptics (oral leviteracetam), multivitamins, and physiotherapy. He showed gradual improvement over next six days and was able to stand without support. He could walk independently at 13 months of age with normal gait. The power of the left limbs had improved with normal tone and reflexes.

Discussion
The estimated incidence of ischemic stroke in children older than 28 days of life is variable [6-8] but, according to a large prospective, population study, it averages 13/100,000 for all strokes, 7.9/100,000 for ischemic strokes, and 5.1 for hemorrhagic strokes.

Acute basal ganglia stroke after minor trauma associated with mineralization of lenticulostriate arteries in infants is a distinct clinicoradiological entity. Investigations for prothrombotic states and vasculopathies are normal. Although neurological outcomes in most children are good, trauma is a risk factor for recurrence of stroke.

Recently, Lingappa et al., [9] described a distinct clinicoradiological 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. [9] were less than 18 months.

Reference