Mineralizing Angiopathy of Lenticulostriate Arteries with Infantile Basal Ganglia Infarct Following Minor Head Trauma: A Case Series

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Abstract

Acute arterial ischemic stroke after minor head trauma has been reported in the past, mostly in infants. Most of these affected children had basal ganglia infarct on imaging. Investigations for other etiologies of stroke were noncontributory in most of the cases. Thin-slice computed tomography scan may show mineralizing angiopathy of lenticulostriate arteries. We report a clinical series of four infants who presented with the classical features of this distinct clinico-radiological entity. Clinical characteristics and risk factors at the time of stroke were described in detail. The long-term outcome on standard antiplatelet therapy is reported. None of the children had stroke recurrence during follow-up. The current literature on this clinico-radiological syndrome is reviewed in detail. In the typical cases, extensive etiological workup may not be warranted.

Keywords: Basal ganglia infarcts, head trauma, lenticulostriate arteries, mineralizing angiopathy, stroke

Introduction

Acute arterial ischemic stroke (AIS) after minor head trauma has been reported in the past, mostly in infants.[1] Affected children present with hemiparesis with evidence of basal ganglia infarct on imaging. Investigations for etiologies of stroke were noncontributory in most of the cases. Mineralizing angiopathy of lenticulostriate arteries associated with basal ganglia stroke following minor trauma in infancy was described by Lingappa *et al.* as a distinct clinicopathologic syndrome.[2] Here, we are reporting a case series of four children who presented with this rare entity.

Case Report – Observations

Four infants who had the classical features were included in this cohort. All the children presented in the acute phase of stroke. The mean age at presentation was 11.7 months (range- 9 months–18 months), and male:female ratio was 1:1. The stroke followed the trauma by mean duration of 55 minutes (range-10 minutes to 90 minutes). Trauma was trivial in nature in all of them (fall from the bed, slipped over the floor). None of them had loss of consciousness or seizures after fall. Neurological examination revealed hemiparesis and two children had upper motor neuron facial weakness on the paretic side. General examination was normal except for the presence of anemia in two. Hemidystonia was noted in two children on the next day of admission which improved within 48 hours. Magnetic resonance imaging (MRI) with magnetic resonance angiogram (MRA) was done in all the children. MRI images showed acute infarct in the gangliocapsular region in all the children [Figure 1] and MRA was normal. All the secondary etiological workup was negative including prothrombotic workup. None of them had patent foramen ovale on conventional transthoracic echocardiography. Transesophageal echocardiogram was not done in any of them. The clinical presentations, investigations and follow up data are summarized in Table 1.

Computed tomography (CT) brain study showed basal ganglia calcification in all the four children and linear calcification in basal ganglia were demonstrated by oblique and coronal reconstruction in three of them [Figure 2]. All of them were treated with aspirin. The mean duration of follow-up was 43.5 months (interquantile range- 14–96 months). Three of them had full recovery in 1 week, 2 months, and 7 months, respectively. Case 4 had residual hemiparesis (4/5 power). None of them had recurrence of stroke during the follow-up period. Iron supplements were also given to those with iron deficiency.

Discussion

Acute ischemic strokes involving basal ganglia after minor head trauma constitutes <2% of all childhood ischemic strokes.[2] Even though this phenomenon has been reported for over a decade, the evolution of clinical manifestations,

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Table 1: Summary of the clinical presentations, investigation, and the follow-up data

Case number	Sex/age	History of fall	Onset of weakness after fall (minutes)	Neurological deficit	Other positive findings	BG calcification/ mineralization	Treatment given	Recovery	Recurrence	Neurodevelopmental outcome	Follow-up (months)
1	Male/10 months	Yes Fall from walker	10	Grade 2/5 power left upper and lower limb	Hb 7.62 gm/dl Serum iron 6.4 ug/dl TIBC 491 ug/dl	Bilateral More on the right side	Aspirin, iron supplements	2 months	Nil	Normal	22
2	Female/18 months	Fall following slippage on floor	60	Grade 2/5 power left upper and lower limb	Hb 9.55 gm/dl Serum iron 18 ug/dl Serum ferritin - 12 ng/dl	bilateral	Aspirin and iron supplements	1-2 weeks	Nil	Normal	14
3	Female/10 months	Fall while walking	60	Grade 3/5 right upper and lower limbs Right UMN facial weakness	WNL	Bilateral Linear calcification	Aspirin	2-3 months	Nil	Normal	42
4	Male/9 months	Fall from 2 feet height	90	Grade 2/5 power left upper and lower limb	WNL	Bilateral calcification	Aspirin	7 months	Nil	Residual weakness - 4/5 power	96

BG=Basal ganglia, Hb=Hemoglobin, TIBC=Total iron-binding capacity, UMN=Upper motor neuron, WNL=Within normal limits

radiological features, and the mechanism of stroke are still not delineated completely. A review of imaging features in many of these reports revealed documented hyperdense foci within the infarcts which have been variably interpreted as thrombus or tiny foci of hemorrhage.[1,3,4] The association of basal ganglia calcification with minor head trauma was suggested in 10 out of 16 infants in a report from China by Yang *et al.* in 2013.[5] Basal ganglia calcification was demonstrated on CT images, but the vascular nature of mineralization (linear mineralization) has not been documented in these series. The pattern of mineralization was similar to that described in sonographic lenticulostriate vasculopathy.[5] Mineralizing angiopathy of lenticulostriate arteries in children presenting as acute basal ganglia stroke was first described as a clinicoradiological syndrome by Lingappa *et al.* in 2013.[2] They reported 22 infants with this rare entity characterized by the occurrence of stroke in previously healthy children in the age group of 6 months to 24 months with rapid onset of hemiparesis within minutes to hours after minor trauma. Transient hemi dystonia was also noted on the affected side typically within 2 to 4 days of fall. Five infants experienced recurrence and nine had mild residual hemiparesis in the original series. None of the children in our case series had recurrences of stroke so far.

Several hypotheses have been put forward for basal ganglia stroke in association with minor head trauma in infants.[6,7] A proposed mechanism for post-traumatic arterial occlusion is that mechanical forces during head trauma causes rapid brain displacement, thus resulting in excessive stretching of arteries at the point of arterial tethering. In infancy, there is an acute angle between the middle cerebral artery and lenticulostriate arteries and it becomes more obtuse during a person's life time.[5] The anatomical peculiarity of lenticulostriate arteries as well as the relatively unmyelinated brain increases the risk of mechanical injury.[8] The incidence of minor trauma is more during infancy. The mineralizing angiopathy is known to regress over time; hence, infancy may represent an age for increased vulnerability for strokes due to this entity.

Lingappa *et al.*[2] has reviewed over 400 cases of brain CT performed in infants of the same age group for unrelated symptoms, and identified incidental lenticulostriate artery mineralization in only four individuals (1%). The cause for mineralization in affected children is usually unknown. Squier and Mack[9] proposed that mineralized vessels have defective physiological contractility leading to alteration in their barrier properties leading to the thrombotic event. They have suggested that there is evidence to implicate pericytes derived from mesoderm with osteogenic properties for the mineralization.

MRI most of the time may fail to identify mineralizing angiopathy of lenticulostriate arteries. When a gangliocapsular infarct is identified in an MRI in the age group of 6 months to 24 months, it is preferable to do a thin sliced multiplanar reconstruction CT to delineate linear calcification in lenticulostriate vessels. If there is a clear-cut evidence for mineralizing angiopathy of lenticulostriate arteries in the CT

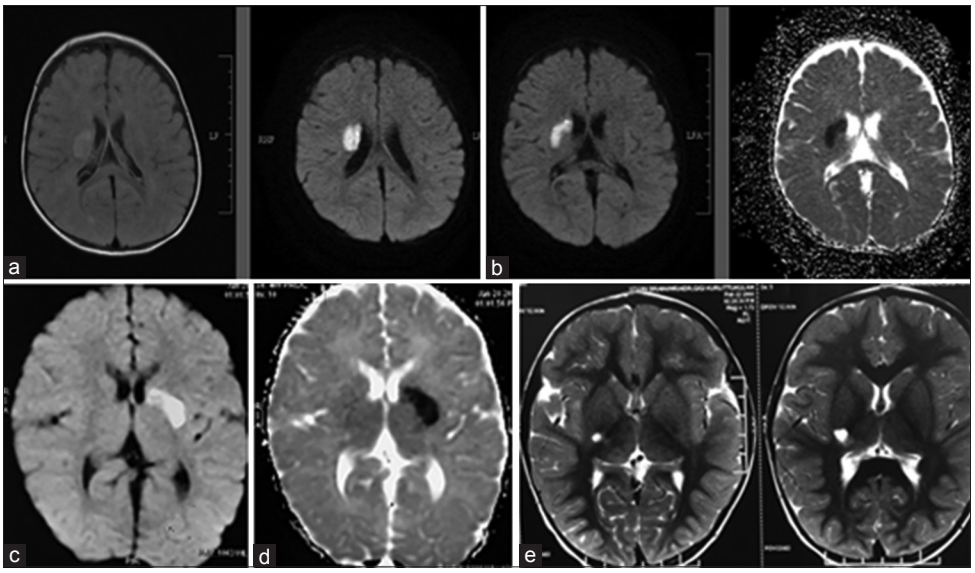

Figure 1: (a) (Patient1)-MRI Fluid attenuated inversion recovery(FLAIR) sequences showing hyperintensity and diffusion weighted images(DWI) showing diffusion restriction over right corona radiata (b) (Patient 2) showing diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) images showing diffusion restriction over right gangliocapsular area suggesting acute infarct. (c and d) (Patient 3) over the left gangliocapsular area and (e) (Patient 4) T2 images showing hyperintensities over the right gangliocapsular area

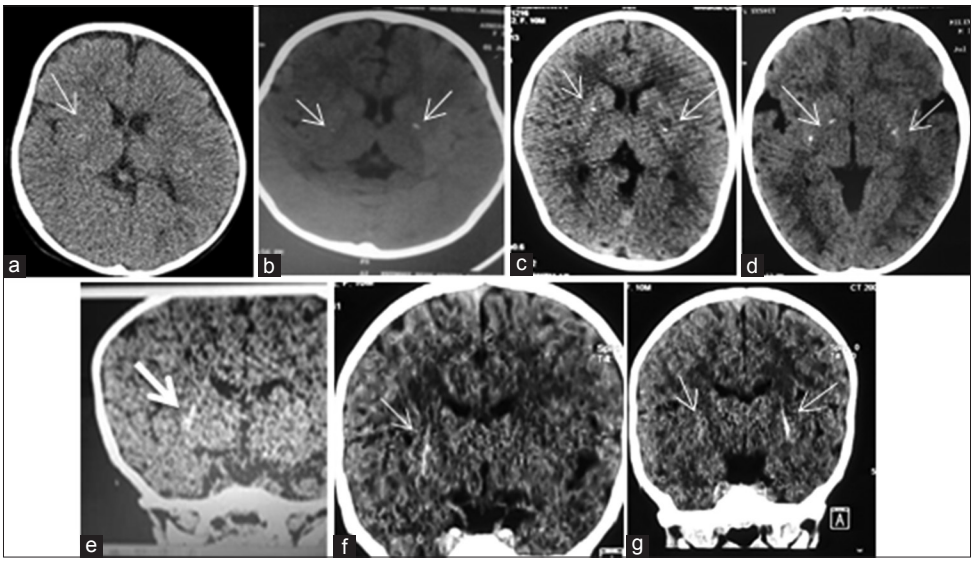

Figure 2: (a-d) Plain computed tomography images of patient 1, 2, 3, and 4, respectively, showing bilateral basal ganglia calcification (marked by white arrow). (e-g) Reconstructed computed tomography images of patient 1, 2 and 3 showing linear calcification suggesting mineralizing angiopathy of lenticulostriate arteries (marked by white arrow)

scan, the expensive battery of further etiological investigations may be avoided. This clinicopathological entity has a very favorable prognosis with standard antithrombotic therapy.

The role of aspirin as well as the duration of treatment in this condition is not certain at present. Lingappa *et al.* had initially used aspirin for all the children, adding cilostazol after the recurrence of stroke in one of their patients.[2] Yang *et al.*, however, continued on conservative management without antiplatelet agent or anticoagulants.[5] In their series of 16 children with acute stroke after head trauma, one patient had stroke recurrence. Most of the other previously reported cases had a relatively good prognosis, recovering from the symptoms without any residual neurological deficits. However, some patients had recurrent strokes and developed persistent residual weakness along with dysarthria.[2,5] In this series, all the children received long-term aspirin and none had a recurrence of stroke. Only one child had residual weakness, others showed full recovery on long-term follow-up. However, the numbers are too small to make firm conclusions. Multicentric cohorts with a larger number of patients and longer follow-up periods may be one way forward to clearly delineate the recurrence risk and the role of long-term antiplatelet agents in this syndrome.

Conclusion

Mineralizing angiopathy of lenticulostriate arteries presenting as infantile basal ganglia stroke after minor trauma is a distinct clinico-pathological entity with a good long-term outcome. Thin-sliced multiplanar reconstruction CT is the investigation of choice to pick up the linear calcification of these vessels. The pathophysiology of this new disorder is not very evident at this point and might require further research. This case series illustrates the salient clinical features of this rare association. Treating physicians, as well as radiologists, should be aware of this rare entity to judiciously plan the investigations and management options in a potential case.

What is known?

- Infantile basal ganglia stroke after minor trauma associated with mineralizing angiopathy of lenticulostriate arteries is a recently described clinico-radiological entity.

What is new?

- This case series adds to the literature on the long-term outcome of infantile basal ganglia stroke associated with mineralizing angiopathy of lenticulostriate arteries
- Recurrence for stroke is found to be very low on long-term follow-up
- In the typical case, extensive secondary etiological workup may not be warranted.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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