INFANTILE BASAL GANGLIA STROKE DUE TO MINERALIZING ANGIOPATHY FOLLOWING MINOR TRAUMA - A CASE REPORT WITH RADIOLOGICAL REVIEW

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ABSTRACT

Basal ganglia stroke in infants is commonly associated with minor fall. Affected infant present with hemiparesis and facial paresis after injury. On neuroimaging small infarcts in basal ganglia are detected. We report a case of acute basal ganglia stroke after minor trauma in infant associated with mineralizationof lenticulostriate arteries detected on MDCT. It is a distinct clinico-radiological which needs further research for cause of mineralization of lenticulostriate arteries. Neurological outcome in most children is good.

Keywords: Pediatric stroke, minor trauma, lenticulostriate arteries, mineralizing angiopathy, juvenile head trauma syndrome, sonographic lenticulostriate vasculopathy.

Introduction

Stroke in children is considered a rare event. Reported incidence of combined ischemic and hemorrhagic pediatric stroke is 1.2 to 13 cases per 100,000 children below 18 years of age.[1]

Mortality rate of pediatric stroke is 0.6 per 100,000 dead per year.[2] It has high morbidity and long term outcome and is gaining more interest due to its heavy consequence and cause at both personal and social level. Half of surviving patients develop neurologic and cognitive impairment while more than quarter develops epilepsy.[3]

Basal ganglia stroke occurring in infant following minor fall is a known entity. Several case reports and small series in the last decade had shown its association. They have favorable prognosis on investigations, cause of stroke usually cannot be found out. Soon after injury, affected infant presents with hemiparesis with facial paresis and show small non-hemorrhagic infarct in basal ganglia on CT and/or MRI. Such infarcts show characteristic way of presentation and evolution of symptoms 1) affection in previously healthy infant of age 6 - 24 months. 2) Rapid onset and progression of neurological deficit (usually hemiparesis) minutes to hours following a minor fall. 3) Dystonia on affected side between day 2 and 4 after onset of illness which subside within 24 hours. 4) MDCT showing linear mineralization along course of bilateral lenticulostriate arteries. 5) Recurrence of stroke after minor trauma subsequently. 6) Good prognosis with good short term and long term neurodevelopment outcomes except in infarct in recurrent stroke.[4]

Clinical Report

A 1 year old child asymptomatic 3 days back, suddenly fell down while playing. This was followed by sudden onset weakness of left lower limb which gradually progressed to left upper limb. There was no history of loss of consciousness, seizures, vomiting and fever. There was no major illness in the past. No

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history of any congenital heart disease. He had full term normal vaginal delivery. Milestones were normal as per age. No history of consanguineous marriage. No family history of any neurological conditions. On CNS examination, the patient had left hemiparesis (3/5 power), increased tone in left upper limb and lower limb, deep tendon reflex in left upper and lower limb were brisk, left plantar reflex was extensor. Sensory examination and cranial nerves examination was normal. His laboratory investigation revealed mild anemia (Hb= 9 gm %), Microcytic hypochromic anemia. LFT, RFT and coagulation profile were normal.

MRI brain showed focal hyperintense lesion in dorsal portion of right lentiform nucleus and adjoining posterior limb of internal capsule extending to right corona radiata on T2WI (Fig.1), appearing hypointense on T1WI showing restricted diffusion on DWI

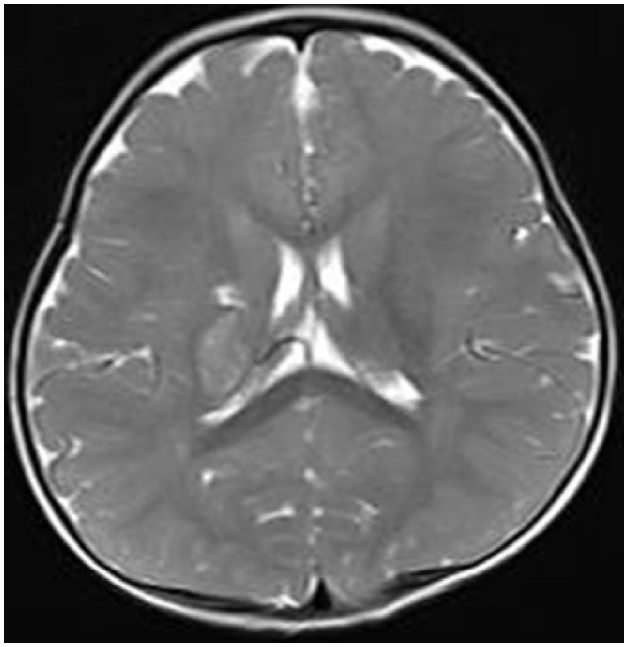


Figure 1: MRI: Axial T2WI shows hyperintense nonhemorrhagic infarct in dorsal portion in right lentiform nucleus and adjoining posterior limb of internal capsule.

with low ADC values (Fig. 2A & B). Findings were suggestive of acute non hemorrhagic infarct. MR angiography was normal. Non contrast CT scan showed hypodense lesion in dorsal portion of right lentiform nucleus and adjoining posterior limb of internal capsule extending to right corona radiata suggestive of non hemorrhagic infarct (Fig. 3B & C).

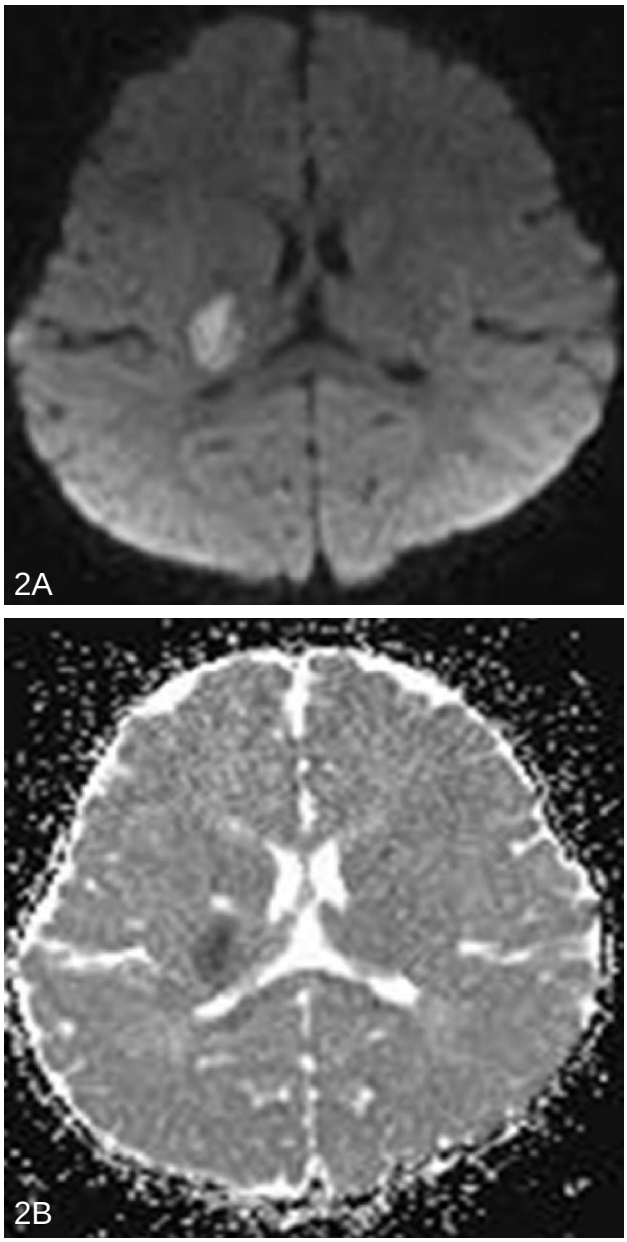


Figure 2A-B: MRI: DWI (A) and ADC (B) show restricted diffusion with low ADC values suggestive of acute nonhemorraghic infarct in dorsal portion in right lentiform nucleus and adjoining posterior limb of internal capsule.

In addition, multiple linear hyperdensecalcific densities (70- 90 HU) were noted in bilateral lentiform nuclei (Fig. 3A - C). On sagittal and coronal reformatted images, these calcifications were linear along course of lenticulostriate arteries (Fig. 4A- D). Number of small mineralized vessels ranged from 3-5 on each side. Stroke workup was done. 2D echo was normal. Sickling test was negative. Antiphospholipid antibody (APL) - IgG was 8.64 U/ml (negative < 12), Antiphos-

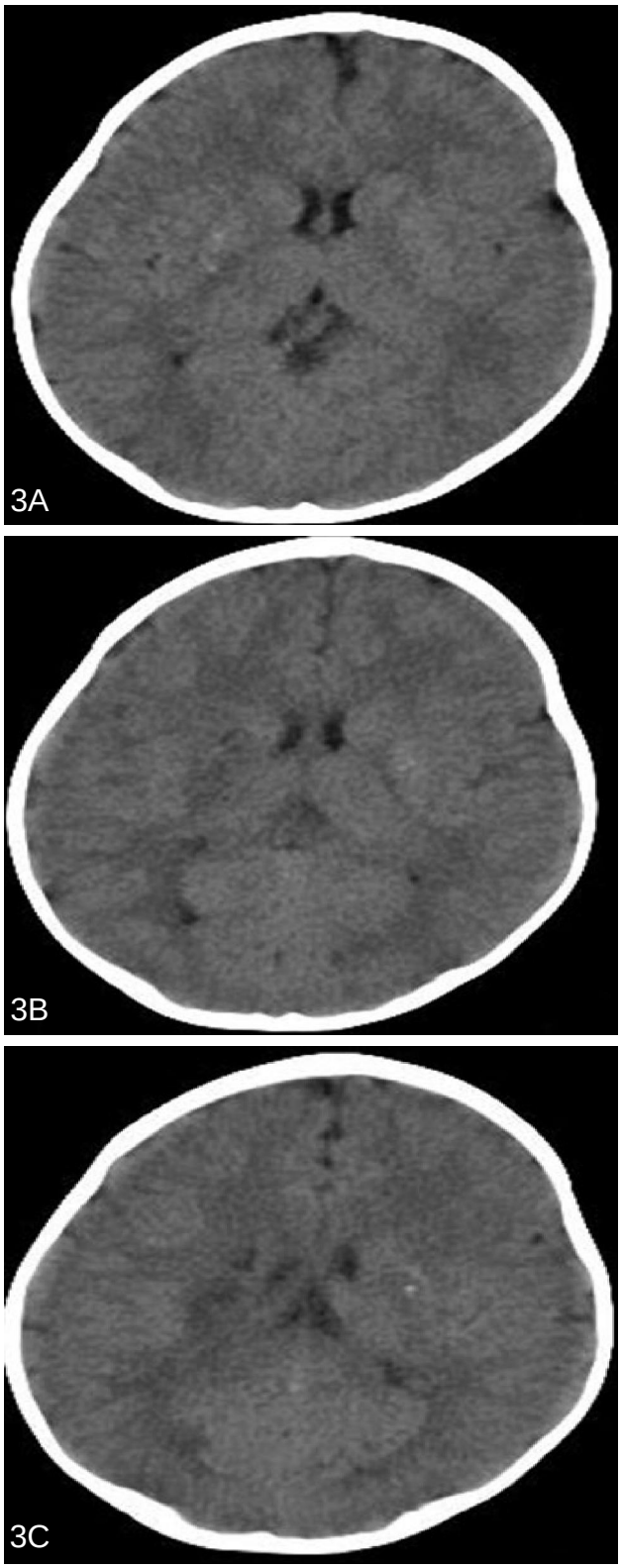
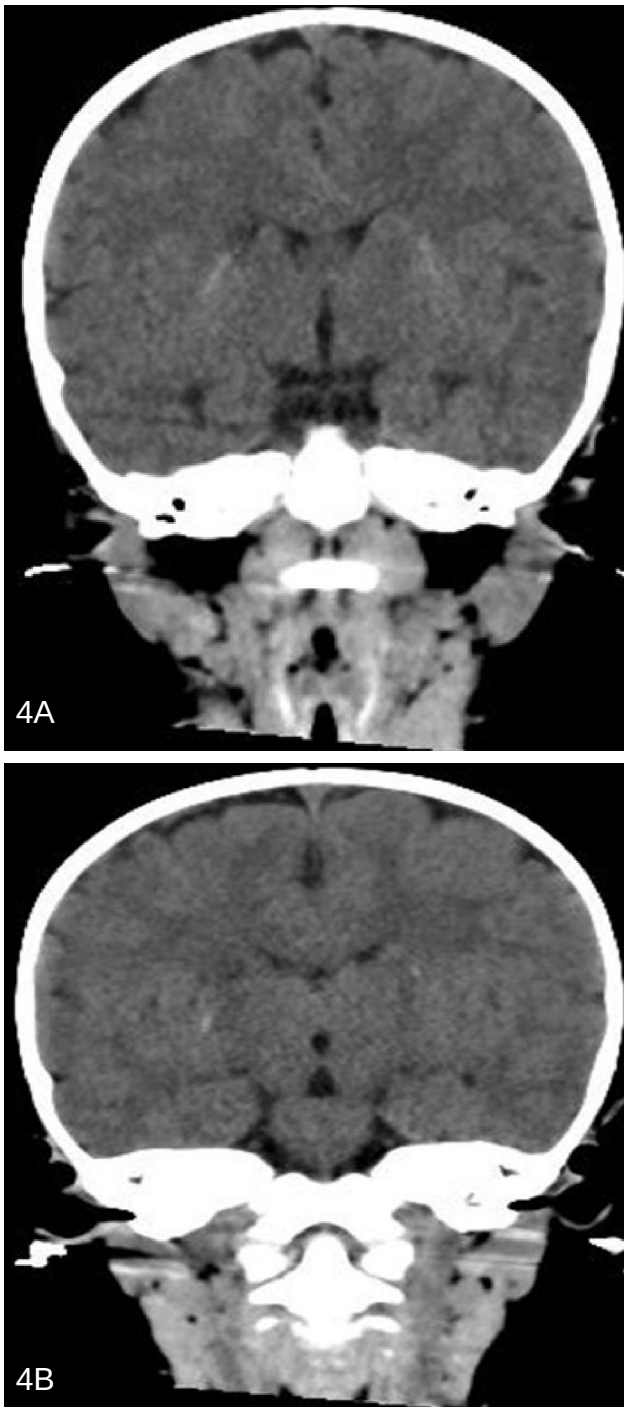
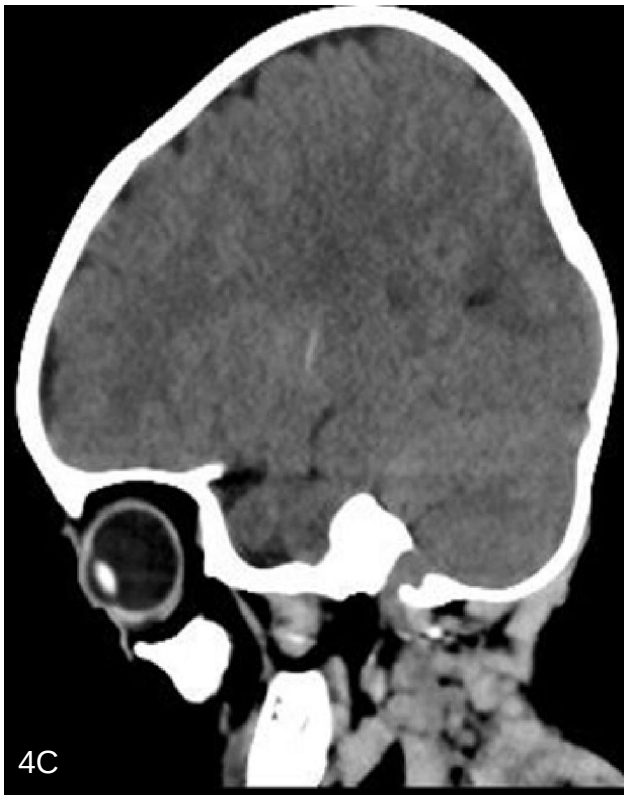


Figure 3A-C: Axial plain CT shows hypodense area of non-hemorrhagic infarct in dorsal portion in right lentiform nucleus and adjoining posterior limb of internal capsule and multiple punctate areas of calcific foci in bilateral basal ganglia.

pholipid antibody (APL) - IgM was 4.63 U/ml (negative < 12),homocystenine was 25.6 micro mole/L (negative < 30). Patient was put on aspirin 5mg/kg/day and physical rehabilitation. After 15 days of treatment patient was able to stand with support and grasp object with left hand and there was improvement in power (3/5). A diagnosis of mineralizing angiopathy of arteries leading to stroke was made.





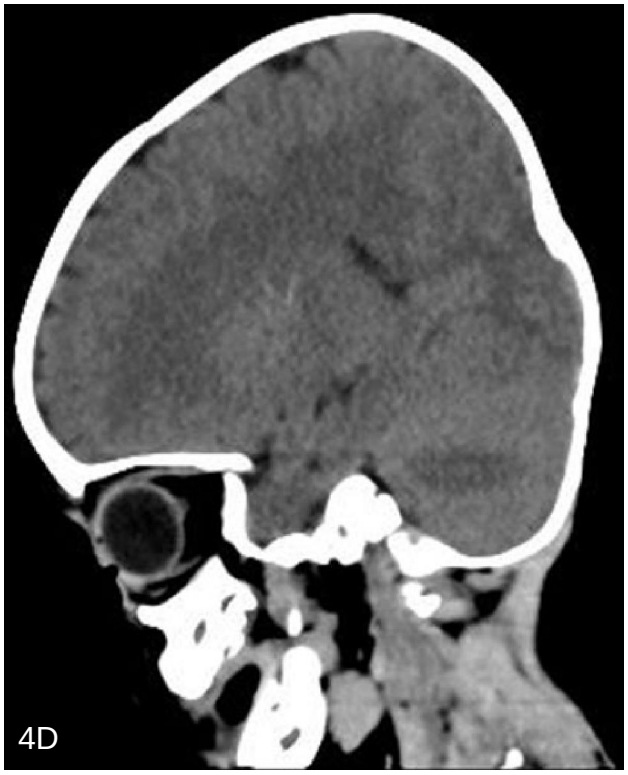


Figure 4A-D: CT: Coronal and Sagittal reformatted images show linear distribution of calcification along course of lenticulostriate arteries.

Discussion

Acute basal ganglia infarct following minor trauma have been reported in children below age of 18 months and constitute less than 2% of all childhood ischemic stroke. Evolution of clinical manifestation, mechanism of stroke and radiological features are still unclear.[5] Neuroimaging in these cases show hyperdense foci within this infarct which were interpreted as thrombus or tiny foci of hemorrhage.[6] Study by Yang et al in infarct with stroke after minor trauma showed basal ganglia calcification on axial CT images in 10 out of 16 infants. However vascular mineralization (linear mineralization) was not documented in this series. These cases probably represent previously unrecognized cases of same entity. With advent of MDCT, it is possible to demonstrate linear distribution of mineralization along lenticulostriate arteries by performing coronal and sagittal reformatted images.[7] Cause of mineralization of lenticulostriate arteries is not yet clear. Yang et al reported association between post traumatic basal ganglia infarction and positive serological testing for CMV, echovirus, Epstein-Barr virus and mycoplasma.[7] There was coexistence of basal ganglia mineralization with only CMV seropositivity. Echovirus, Epstein-Barr virus and mycoplasma were detected as co-infection with CMV and not independently associated with basal ganglia mineralization. As mineralization is generally a result of long standing pathology, it is not yet clear whether acute CMV infection (as demonstrated by CMV DNA in urine) can cause mineralization. Hence further research is needed. Distribution of linear areas of mineralization in basal ganglia region is similar to that described in sonographic lenticulostriate vasculopathy (SLV). This occurs in 0.4% of all live born neonates and 1.9% - 5.8% of ill neonates.[8] It occurs in association with variety of congenital and acquired disorders and usually regresses with over time. Pathologically lenticulostriate vasculopathy is characterized by thickening of hypercellular vessel wall with intramural and perivascular mineralization. These changes are easily visualized on neurosonography and are not often picked up on CT/MRI.[8,9] Mineralizing angiopathy most probably represent a more severe and persistent form of lenticulostriate mineralization which is extensive enough to be detected on CT.

There may be possible association between SLV and mineralizing angiopathy. Ivano et al reported brain infarction following trauma in an 8 month infarct with preexisting SLV.[10] They postulated that underlying lenticulostriate vasculopathy makes infant predispose to vascular obstruction following head trauma. Lingappa et al postulated that stress across mineralized lenticulostriate arteries during minor trauma predispose individual to thrombosis. There is an age specific predisposition at particular stage of mineralization.[4] These mineralized vessels are prone to blockage after minor trauma and remain asymptomatic at later ages beyond 2-3 yrs, though mineralization persists.

2007 National institute for Health and Clinical Excellence guidelines for assessment and management of head injury recommends CT as primary imaging modality in children with focal deficit following head injury.[11] Though MRI is preferred imaging modality for stroke, it fails to indentify mineralization on gradient recalled echo sequence and vascular abnormalities on MR angiography. Conventional CT done for head injury with 5 mm thin axial sections fails to appreciate true nature of vascular mineralization. MDCT with thin sections help in identifying nature of vascular mineralization by multiplanar reconstruction.

Children are vulnerable to minor head injury which results in translating, stretching and distorting forces. Impact on skull leads to opposing movement of brain parenchyma with resultant stretching and shearing effect on vessel due to high movement of inertia. This causes traumatic endothelial intimal lesion, followed by fibrin accumulation, leukocyte reaction and formation of wide thrombus occluding the lumen vessel. Obstruction due to thrombus results in ischemia of cerebral parenchyma with clinical symptoms after a symptomless latency period.[12]

Predisposition of stroke in pediatric age following minor trauma is due to particular anatomical characteristic of arteries. Lenticulostriate arteries, terminal branches of anterior communicating arteries responsible for flow to caudate nucleus, globuspallidus, putamen and internal capsule create an acute angle with MCA which is more pronounced in child-hood than adulthood.[13] Thalamo- perforating arteries and choroidal arteries may also be involved. Lateral perforators make a more acute angle than medial perforators, which hence can readily undergo stretching during head trauma. Lateral perforators are short in children hence susceptible to stretching. Intraparenchymal segment of lenticulostriate arteries (connected to brain parenchyma) and extra parenchymal segment of lenticulostriate arteries (connected to MCA) are relatively fixed. Between these two segments, there is a small mobile segment which undergoes stretching by trauma with resultant vasos-pasm and/or thrombosis causing ischemia in the involved vascular territory. In children, sphenoid bone is not fully developed and hence brain has greater mobility than skull base in case of violent deceleration. These factors facilitate stretching of lenticulostriate arteries by traumatic force causing vasospasm and thrombosis.[14] Ischemia of basal ganglia following minor head trauma in children under 18 months of age is caused by vasospasm of lenticulostriate arteries which manifest as nausea, vomiting, drowsiness and hemiparesis. The clinical picture is known as Juvenile head trauma syndrome (JHTS).[13]

Conclusion

This case report described mineralizing angiopathy causing basal ganglia stroke in infants after minor trauma as a distinct clinico-radiological entity. MDCT with sagittal and coronal reconstruction is mandatory to diagnose this condition as it detects vascular calcification along course of lenticulostriate arteries. Prognosis in unilateral stroke is good,while infarcts with bilateral (recurrent) stroke have significant long term disability.

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