DOI: 10.7860/JCDR/2021/49792.15119

Case Series

Paediatrics Section

Mineralising Angiopathy of Lenticulostriate Vessels in Childhood: Series of Four Cases

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ABSTRACT

Mineralising lenticulostriate vessels' angiopathy is becoming an important recognisable cause of ischaemic stroke in healthy children post trauma or fall. The usual presentation is a focal neuro-deficit of acute onset following trivial trauma in infancy with or without seizures/dystonia. The authors conducted a retrospective chart review of children diagnosed with stroke in a tertiary care paediatric hospital. Based on clinical and radiological features, children diagnosed with mineralising angiopathy with basal ganglia stroke in the last two years were identified, data was retrieved from the hospital's electronic database and included in the study. Also, the patients were contacted telephonically in case of missing data. Out of the four cases, three were less than 18-month-old, and one was 52-month-old. All were normal before illness and had a history of trivial trauma {except one who had Road Traffic Accident (RTA)} followed by hemiparesis. Investigations for other aetiologies of stroke were non contributory. With antiplatelet therapy and iron supplements, most did well with an excellent functional outcome at follow-up. None of the children had recurrence during follow-up. In a typical case, an extensive work-up is not required.

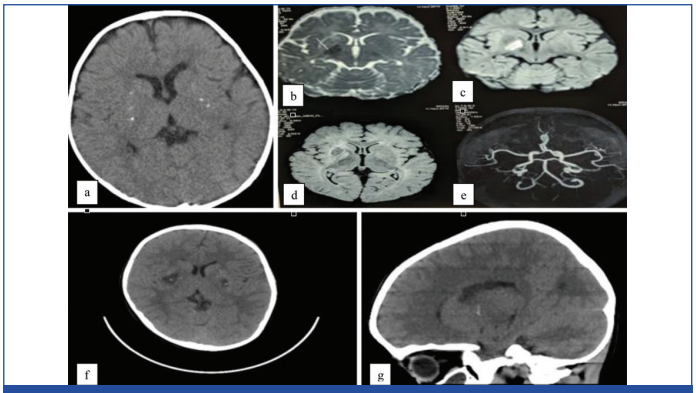
Keywords: Basal ganglia infarct, Stroke, Trivial trauma

CASE SERIES

Four cases with classical features were included in the present study cohort. The presentation was acute in all of them. Three were less than 18 months of age and had a history of trivial trauma (fall from bed, fall while walking), presented with hemiparesis. The fourth case is a 52-month-old, previously normal child who had a history of RTA, following which the child developed weakness, seizures, dystonia and neuro-imaging suggested hypodensity in bilateral basal ganglia with linear and punctate calcifications. All except one had microcytic hypochromic anaemia. Magnetic Resonance Imaging (MRI) brain with Magnetic Resonance (MR) angiogram was done in two of them and were suggestive of gangliocapsular infarct in MRI brain and MR angiogram was normal. Three had undergone non contrast CT brain with oblique and coronal reconstruction. Computed Tomography (CT) brain showed punctate calcification in basal ganglia in axial cuts and reconstructed images in saggital and coronal section demonstrated linear calcifications along the vessels. Secondary aetiological work-up in three of them was negative, including prothrombotic work-up in two. The 2-Dimentional Echocardiography (2D ECHO) was normal in all. They were treated with aspirin (3 mg/kg/day), iron supplements, and physiotherapy. Three of them had near complete recovery in follow-up. The clinical picture with imaging findings [Table/Fig-1] and follow-up data are summarised in [Table/Fig-2].

DISCUSSION

Stroke is an alarming condition in children. It is amongst one of the top 10 common causes of childhood deaths [1]. Minor bumps on the head are very common in childhood, but cerebral infarction post trauma is an extremely rare sequela. Pertaining to childhood age, a head injury can be an important cause for acute ischaemic stroke. In children who are of below 18 months of age, mild head trauma is quite common. The most common region affected following trivial trauma is striatocapsular and the basal ganglia region [2]. Mineralising angiopathy can be explained as a condition characterised by cerebral parenchymal calcifications in lenticulostriate vessels. Affected children present with hemiparesis and facial paresis soon after trauma, and there is a typical finding of infarcts and calcification in basal ganglia on neuroimaging.



[Table/Fig-1]: (a) (Patient1)- CT brain showing punctate calcification in basal ganglia bilaterally. (b-e) (Patient 2) MRI diffusion weighted images (c) showing diffusion restriction over right gangliocapsular region and (b) Apparent Diffusion Coefficient (ADC) images showing corresponding changes suggesting acute infarct. T2 flair is showing hypointense region with hyperintense rim in right basal ganglia (e) normal MR angiogram of brain (f) (Patient 4) CT Brain showing bilateral hypodensity in basal ganglia region with punctate hyperdensities resembling soft calcification (HU 50-90) (g) Same patient with sagittal CT image reconstruction showing linear calcification in basal ganglia.

Yang FH et al., in their study amongst 16 of the infants less than 18 months of age, found that almost 62% of infants developed significant neurological signs and symptoms within 72 hours of minor trauma [3]. The first symptoms encountered were hemiparesis, facial paresis, and convulsions, further these children had basal ganglia calcification. Study done by Lingappa L et al., suggested that this is a distinct clinico-radiological syndrome [4]. They reported 22 previously normal infants who presented with stroke after minor trauma and had basal ganglia calcifications. Eighteen had a history of falls, and 16 of them presented within eight hours of fall. Twenty had hemiparesis as presenting complaint, and 16 developed dystonia within two to four days of fall.

Gowda VK et al., published a series of 38 children with similar presentation and radiological profiles [2]. They also reported two cases in the same family with second-degree consanguinity, indicating the possibility of genetic factors. The present study cohort has similar results in comparison with above studies. However, the present cohort had a 52-month-old child who presented similarly at a later age and CT showed characteristic findings similar to

Case No.	Sex/Age	History suggestive of fall	Onset of weakness post trauma	Neurological deficit	Associated relevant findings	Neuroimaging (Basal ganglia calcification)	Treatment given	Recovery	Neurodevelopmental outcome	Maximum duration of folow-up	Recurrence
1	1.5 years/ Female	Yes	6 hours	Power 1/5 left U/L and left L/L	Left side hemiparesis, Microcytic hypochromic anaemia	Punctuate linear calcification in BG bilateral	Aspirin Iron	Fully recovered at 6 month follow-up	Normal	18 months	Nil
2	9 months/ Male	Yes	Immediate	Power 2/5 left U/L and left L/L	Left Hemiparesis, Microcytic hypochromic anaemia	Right BG	Aspirin Syp B12, Iron	9 months	Residual weakness+, Left L/L power 4/5, circumduction gait	11 months	Nil
3	16 months/ Female	Yes	6 hours	Power 2/5 right U/L and right L/L	Right hemiparesis	Bilateral BG calcification - punctate	Aspirin Physiotherapy	Lost to follow-up	Fair (Residual weakness+telephonic follow-up two months back)	Lost to follow-up Last contact by telephone 2 months back	Nil
4	4.4 years/ Male	Yes	Immediate	Power 2/5 all four limbs	Dystonia + (onset after 24 hrs), Weakness+bilateral, pallor+and seizures	Bilateral BG and high parietal region	Aspirin Pacitane Iron Anticonvulsant	Dystonia improved, No seizures	Residual weakness +, power of 4+/5 in both L/L.	7 months	Nil

[Table/Fig-2]: Clinical presentation, imaging and follow up profile of the present cohort.
U/L: Upper limb; L/L: Lower limb; BG: Basal ganglia

mineralising angiopathy in infancy. Therefore, the authors suggest that some children may remain asymptomatic during infancy and may present later with more severe trauma (RTA in this case).

The pathogenesis of ischaemic stroke after minor head injury in children is being explored. A proposed hypothesis during infancy is that mechanical forces cause rapid brain shift during trivial trauma, leading to excessive stretching of arteries [2], leading to disruption between the fixed intracerebral portion of the vessel and the extracerebral mobile portion. This leads to the formation of thrombus, which is mainly attributed to shearing stress due to vasospasm, leading to intimal lesion in the vessel [5]. Mineralising microangiopathies are also known to be found secondary to some infectious causes such as echovirus, cytomegalovirus, Epstein Barr virus, and mycoplasma [6]. One other possibility would be that trivial injury may cause temporary middle cerebral artery spasm, and if the child is excessively crying, that leads to a decrease in pCO2, which may further aggravate the arterial spasm [7].

According to guidelines of the National Institute of Health [8], CT is a primary investigation in children having focal deficits after head injury. CT scan may show punctate calcifications in basal ganglia. Thin slice multiplanar reconstruction CT should be done to delineate linear calcification along lenticulostriate vessels. Conventional MRI sequences are not found to be of great help than CT scans, but in T2 sequences, particularly in Susceptibility Weighted Imaging (SWI), these will show as artifacts in affected areas [9]. However, whenever a gangliocapsular infarct is detected in MRI in infancy, gradient sequences and multiplanar CT should be done [10].

Most of the children affected by this disease have a complete recovery and don't have recurrences. The role of aspirin and the duration of therapy is unclear. Some had a recurrence and developed persistent residual weakness and dysarthria [3,4]. None of our patients had a recurrence during the short duration of follow-up. To understand the natural history, larger cohorts and longer follow-up would be required. Establishing a disease-specific registry would be of importance for this. It would also allow us to have a reasonable understanding of the duration of antiplatelet therapy.

CONCLUSION(S)

Mineralising lenticulostriate vasculopathy presents as ischaemic basal ganglia stroke following trivial trauma and is a distinct clinico radiological entity with good outcome. Thin section spiral CT with multi planar reconstruction is the investigation of choice to pick linear calcifications. Paediatricians should be aware of this rare entity and extensive work-up is not required.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Apr 07, 2021
- Manual Googling: Apr 16, 2021
- iThenticate Software: May 21, 2021 (3%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Apr 06, 2021**
Date of Peer Review: **Apr 17, 2021**
Date of Acceptance: **May 21, 2021**
Date of Publishing: **Jul 01, 2021**