Acute encephalopathy: A novel presentation of mineralizing microangiopathy of childhood

Sir,

Mineralizing angiopathy (MA) of childhood is increasingly being recognized as an important cause for arterial ischemic stroke following minor trauma/fall in healthy children.[1,2] It typically presents with acute onset focal neuro deficit with or without seizure following minor

trauma. Neuroimaging usually reveals unilateral infarcts in the basal ganglia with mineralization (calcification) in the corresponding lenticulostriate arteries. This is seen as punctate calcification in the axial cuts of computed tomography (CT) scan of the brain and is clearly seen on coronal and sagittal reconstructions. Magnetic resonance imaging (MRI) shows acute infarct and the magnetic resonance angiography (MRA) in these cases is normal. Very rarely mild encephalopathy has been reported when the infarct is large.[1] However, severe encephalopathy with coma has not been reported in MA. We recently saw two toddlers who presented with acute encephalopathy following minor trauma with neuroimaging suggestive of bilateral basal ganglia stroke and three-dimensional (3-D) coronal reconstructions demonstrating calcified lenticulostriate arteries, thus confirming MA as a cause for stroke.

Case 1 was a 48-month-old developmentally normal boy born to a nonconsanguineously married couple, presented with loss of consciousness after a fall from the bed while playing. He showed abnormal flexion to painful stimuli. Over a period of next 2–3 days, the child's Glasgow coma scale score improved with eye opening to pain, nonspecific sounds, and withdrawal to pain. After 3 days, the child showed paucity of movements in the right upper and lower limb and deviation of angle of the mouth toward the right. The CT scan of the head showed bilateral basal ganglia hypodensities along with punctate calcification in the putamina [Figure 1d]. MRI of the brain showed bilateral asymmetric basal ganglia stroke but the MRA was normal [Figure 1a-c]. Bilateral putaminal signal changes were asymmetrical pointing to a vascular cause and prompted a 3-D coronal reconstruction that demonstrated calcified lenticulostriate arteries, thereby, confirming MA as the underlying cause for the stroke [Figure 1e and f]. Echocardiography, serum homocysteine levels, cerebrospinal fluid (CSF) findings including lactate, tandem mass spectrometry, serum ammonia were normal. Human immunodeficiency virus (HIV) serology, sickling test, and CSF, cytomegalovirus (CMV), polymerase chain reaction (PCR) were negative. The child was put on antiplatelet therapy in the form of oral aspirin with iron supplementation and physiotherapy. The child improved over a period of 4–6 weeks, became ambulatory, and had mild hemidystonia.

Case 2 was an 18-month-old boy who presented with altered sensorium for 5 days after falling from the walker while playing. CT scan showed bilateral basal ganglia infarcts along with putaminal calcification. A 3-D sagittal reconstruction showed calcified lenticulostriate arteries confirming MA. MRA was normal.

MA as an entity, causing acute basal ganglia stroke, has been described in the last 3 years. The calcified lenticulostriate arteries which are brittle predispose these children for basal ganglia stroke following minor head trauma. MRA is usually normal, and thus helps differentiating this condition from the common arteriopathies.[3] Presentation

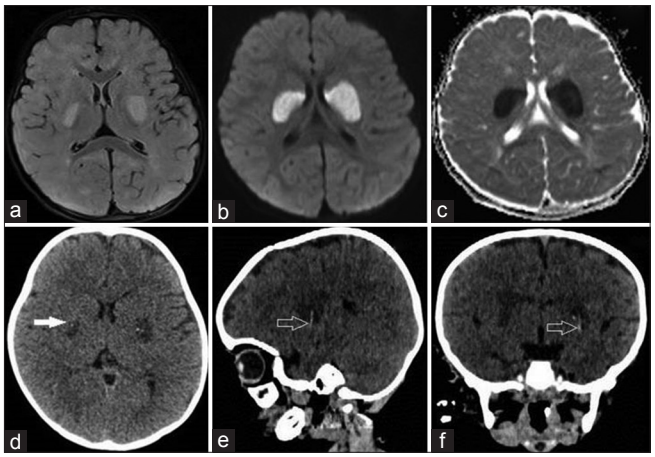

Figure 1: (a) Axial FLAIR MRI image of case 1 at the level of basal ganglia showing asymmetric hyperintensities in bilateral putamina (b and c) Diffusion weighted image showing diffusion restriction and reversal on ADC mapping suggesting acute infarct (d) Axial CT of the head showing asymmetric hypodensities in bilateral putamina with calcifications in the hypodensities (white arrow) (e and f) 3-D sagittal reconstruction and 3-D coronal reconstruction of the brain of case 1 showing calcified lenticulostriate arteries (hollow arrows)

as acute encephalopathy with coma due to bilateral basal ganglia strokes has not been reported earlier in literature; however, it is possible, as we have demonstrated in our two cases. Bilateral basal ganglia involvement can suggest various underlying disorders such as inborn errors of metabolism, degenerative diseases, sequelae of acute insults, dysmyelinating diseases, and neurofibromatosis type 1.[4] MRI changes in these diseases shows symmetrical involvement of bilateral basal ganglia unlike in MA, where it is asymmetrical.

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Conflicts of interest
There are no conflicts of interest.

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References

1. Lingappa L, Varma RD, Siddaiahgari S, Konanki R. Mineralizing

angiopathy with infantile basal ganglia stroke after minor trauma. Dev Med Child Neurol 2014;56:78-84.
2. Yang FH, Wang H, Zhang JM, Liang HY. Clinical features and risk factors of cerebral infarction after mild head trauma under 18 months of age. Paediatr Neurol 2013;48:220-6.
3. Kieslich M, Fiedler A, Heller C, Kreuz W, Jacobi C. Minor head injury as cause and co-factor in the aetiology of stroke in childhood: A report of eight cases. J Neurol Neurosurg Psychiatry 2002;73:13-6.
4. Ho VB, Fitz CR, Chuang SH, Geyer CA. Bilateral basal ganglia lesions: Pediatric differential considerations. Radiographics 1993;13:269-92.

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