Transient focal cerebral arteriopathy of childhood following dengue fever

We report a 9-year-old right handed girl, who presented to our hospital with acute onset right faciobrachial monoplegia. She was being treated in a local hospital one week back with history of fever and myalgia. She was then evaluated to have thrombocytopenia with dengue NS1 antigen positivity. She was treated symptomatically for dengue fever, clinically became a febrile, thrombocytopenia improved and discharged home after 7 days. On the day of discharge, child developed acute onset right faciobrachial monoplegia and was referred to our hospital. On examination she had right upper motor neuron facial weakness with right upper limb grade 3/5 power. Left upper limb and lower limb powers were normal. Her blood reports showed thrombocytopenia with positive dengue IgM antibody. MRI brain showed left basal ganglia infarct [Figures 1 and 2a, b]. MR Angiography done showed left middle cerebral artery M1 segment narrowing [Figure 3]. There was no previous history of stroke and recent varicella infection in this child. Evaluation for pediatric arterial stroke was done. Investigations for primary thrombophilia like CBC, prothrombin time, activated partial thromboplastin time, factor V leiden mutation, homocysteine, and anti phospholipid lipid antibody work up, Protein C, Protein S, and Antithrombin III were negative. Lumbar puncture with CSF study done, showed normal study. Cardiology work up done, ECG, echocardiogram were normal. Imaging of brain and neck vessels ruled out dissection or Moya Moya disease. A 9-year-old child presented with the clinical picture of stroke, right faciobrachial monoplegia following dengue fever and MRI brain with MR angiogram showing left basal ganglia infarct with stenosis of left M1 segment of middle cerebral artery, a diagnosis of pediatric stroke due to transient focal cerebral arteriopathy of childhood following dengue fever was made. She was treated with pulse methyl prednisolone for 3 days along with daily aspirin to continue. Her follow-up MRI brain with MR angiography after 6 months showed resolution of left MCA M1 stenosis which confirmed transient focal cerebral arteriopathy.

Focal cerebral arteriopathy of childhood (FCA), also known as transient cerebral arteriopathy is an acute, monophasic disease causing unilateral stenosis of the intracranial cerebral arteries, mainly involving the anterior circulation. Stroke in children represents a spectrum of disorders with varying etiology, significant adverse outcomes and high risk of recurrent stroke. Owing to lack of awareness of pediatric stroke, it’s diagnosis, etiology and treatment are usually delayed or missed.

Arteriopathies are important because they are not only an important cause of childhood stroke but have a strong predilection for recurrence. An analysis of 525 cases of childhood stroke in the International Pediatric Stroke Study (IPSS) found that FCA caused 25% of arteriopathy and was the most common cause of arteriopathy. In this study, the only independent predictor of FCA was recent upper respiratory infection. The

Figure 1: MRI brain axial section FLAIR image showing left basal ganglia and internal capsule hyperintensity

Figure 2: (a and b) MRI brain diffusion weighted (DWI) high signal intensity in left basal ganglia and apparent diffusion coefficient (ADC) images showing diffusion restriction (low signal intensity)

Figure 3: MR angiogram brain showing left MCA M1 stenosis
most plausible pathogenesis underlying transient cerebral arteriopathy is a transient vasculitis induced by a preceding viral infection. Among the previously reported infections preceding transient focal cerebral arteriopathy, varicella zoster virus (VZV) accounts for the most common identifiable pathogen. Other pathogens including other herpes viruses play a role as transient focal cerebral arteriopathy continues to occur in VZV-vaccinated children.\[3\] Common etiologies described in childhood of acute ischemic stroke are vascular (dissection, trauma, fibro muscular dysplasia, Moya Moya disease, stenosis or secondary vasculitis), infections, cardiac embolism, and idiopathic.\[3\] Vessel wall imaging shows unilateral involvement (stenosis, occlusion, irregularity) of the region of bifurcation of the internal carotid artery into anterior and middle cerebral artery.\[6\] The stenosis may worsen in the first 3 months after stroke, sometimes with new neurologic symptoms, but stabilizes and can even improve by six months after initial presentation.\[3\] In addition to acute antithrombotic therapy, some children have been treated with a short course of anti-inflammatory therapy, such as high-dose corticosteroids (IV methyl prednisolone) followed by tapering oral doses for 6 to 12 weeks. In transient cerebral arteriopathy, the vasculopathy stabilizes by 6 months with no further worsening of the arterial findings.\[6\] In this case snippet we have reported a child developing a stroke due to transient focal cerebral arteriopathy of childhood following dengue fever; which to best of our knowledge has not been reported earlier. Transient focal cerebral arteriopathy should be considered in the differential diagnosis of any pediatric stroke and preceding history of recent infection should be sought in the work up of childhood stroke.

Declaration of patient consent
The authors certify that appropriate patient consent was obtained.

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Conflicts of interest
There are no conflicts of interest.

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