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Posterior Reversible Encephalopathy Syndrome in a child after Severe Traumatic Brain Injury

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ABSTRACT:

Pediatric brain is anatomically and physiologically different from adult brain. Child's brain is more vulnerable to the effects of a brain injury and takes longer to recover. Because the child's brain is still developing, injury may alter the course of development of the brain and its functions. The child's brain is less "set" than the adult's, its plasticity offers hope to damaged tissue and areas. It is very rare to have posterior reversible encephalopathy syndrome as a complication of traumatic brain injury in children.

Key Words: Posterior reversible encephalopathy syndrome, Traumatic brain injury, Neuroimaging, Seizure.

Posterior reversible encephalopathy syndrome (PRES) is a clinic-radiological syndrome characterized by various symptoms including headache, seizure, altered consciousness and visual disturbances. It was first described in 1996 by Hinchey et al. in 1996. [1] This syndrome is also known as reversible posterior cerebral edema syndrome or posterior leukoencephalopathy syndrome.[2] High blood pressure, eclampsia, infection, kidney disease, and certain autoimmune diseases are the main causes of posterior reversible encephalopathy syndrome.[3,4] The common clinical manifestation is encephalopathy of varying grades has been reported in 28–94% of patients.[3] These range from mild confusion, cognitive deficits, somnolence, stupor, and coma. The main diagnostic tool is neuroimaging. Brain imaging is the cornerstone in confirming a diagnosis of PRES. Although vasogenic edema can be visualized on non-contrast computed tomography (CT) in some patients, however brain MRI, especially T2-weighted diffusion weighted imaging (DWI) and fluid attenuated inversion recovery (FLAIR) sequences are much more sensitive.[5] The treatment for PRES is supportive: removal of the cause or causes and treatment of any of the complications, such as anticonvulsants for seizures. Complications include focal neurologic deficits from ischemic injury, epilepsy, and life-threatening conditions, such as tonsillar herniation, as reported in children.[6]

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Case Description:

A 3-year-old girl with an alleged history of falling injury from first stored building while playing was brought by her father to emergency around an hour after the accident. The child was in a state of unconscious with a history of clenching of teeth, sustained injury over right frontal region, right orbital swelling and bruising, and injury over right leg with swelling of right thigh. Pediatric size Philadelphia hard cervical collar placed. Spine was stabilized. The child's ABCs were immediately assessed. She was barely opening eyes to trapezius squeeze and was making incomprehensible sounds. She was extending all four limbs on painful stimulation. The monitor showed a pulse rate of 88 beats per minute and BP of 100/80 mm Hg and SPO2. The child was intubated using the rapid sequence intubation protocol with a 4.5 mm size un-cuffed endotracheal tube and fixed at 12.5 cm and connected to mechanical ventilator ACVC mode. Post intubation vitals were stable. Capillary blood sugar was 225 mg/dL. She was not febrile and the right pupil was 2 mm sluggish reactive and left pupil was 2 mm in size with normal reaction. A secondary survey was done as ABCs were stabilized. There was bulging of right frontal region and periorbital swelling and bruise. We got an arterial blood gas, trauma protocol CT scan was done, and along with a non-contrast CT scan of the brain. The child was loaded with 500 mg of Levetiracetam. The CT scan of head showed acute thin subdural hematoma along bi-parietal and bilateral occipital convexities with diffuse cerebral edema (Figure 1A). Diastatic fracture of the sagittal suture was seen with fracture line extending to mid frontal bone where it bifurcates and extends further to the right orbital roof anteriorly. Posteriorly, the fracture line extends to mid to right off midline aspect of the occipital bone (Figure 1B). Soft tissue swelling and subgaleal hematoma at frontal and right parieto-occipital scalp regions were noted with right orbital swelling. (Figure 1A)

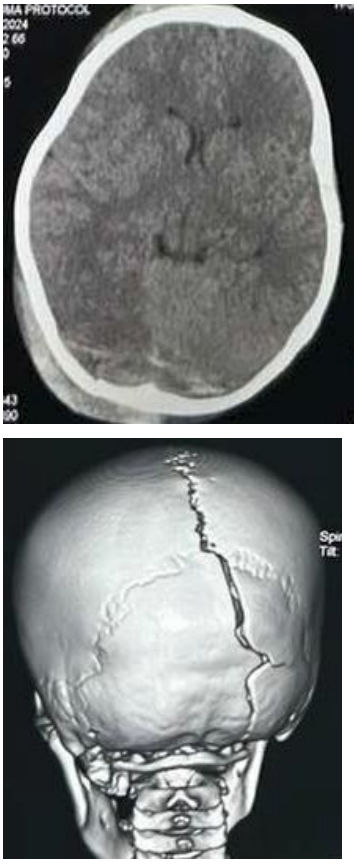

Figure 1A: NCCT head axial view showing acute thin subdural hematoma along bilateral parietal and occipital convexities with diffuse cerebral edema

Figure 1B: 3D CT RECON image showing diastatic fracture of the sagittal suture was seen with fracture line extending to mid frontal bone and Posteriorly, the fracture line extends to mid to right off midline aspect of the occipital bone

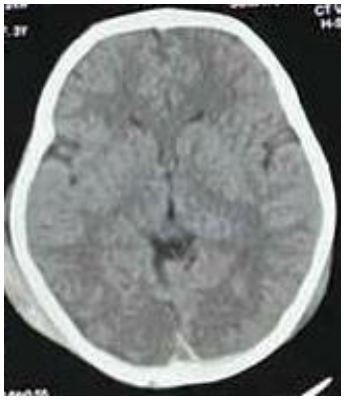

Figure 2: Repeat NCCT head axial view taken on next day showed resolving subdural hematoma and decreasing cerebral edema

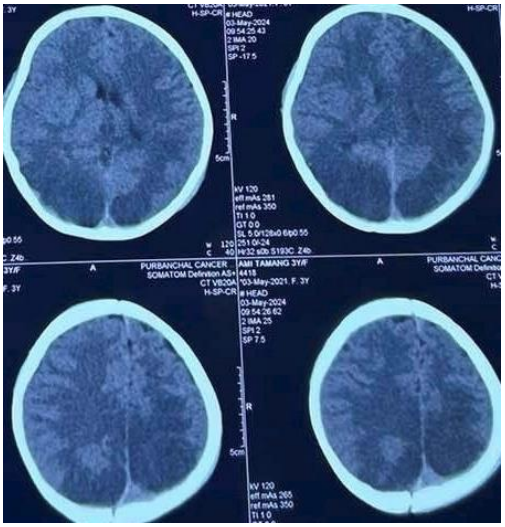

Figure 3: 2nd repeat NCCT axial views taken on 4th post admission day showed bilateral parietal-occipital hypodensity suggestive of ischemic changes

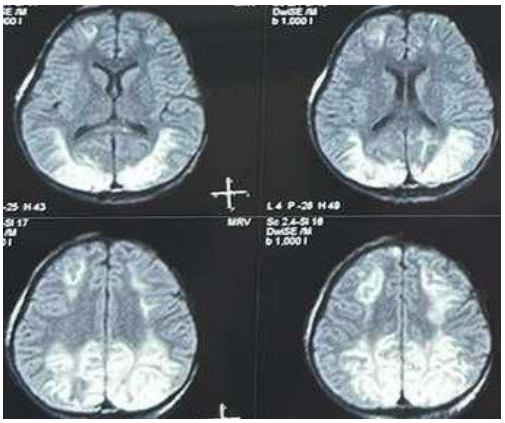

Figure 4: MRI DWI axial cuts taken on 4th post admission day showing laminar pattern of diffusion restriction in bilateral parietal and occipital region suggestive of acute laminar necrosis

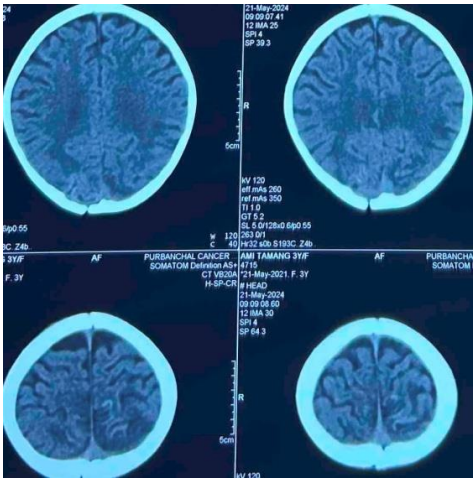

Figure 5: Repeat CT head axial views taken on 10th post admission day showing resolving cortical laminar necrosis in bilateral parieto-occipital lobes

Child was admitted in PICU with daily pediatric consultation. She was started on intravenous antibiotics, anti-edema, and anti-epileptic drugs. A non-operative approach to decrease cerebral edema and early weaning from ventilator was adopted. The first set of results obtained two hours later was normal. On the next day morning rounds, the child was opening eyes to verbal commands, and was localizing pain. Repeat CT head

showed resolving subdural hematoma and decreasing cerebral edema (Figure 2). Close GCS, pupil and vitals monitoring was continued. The blood gas analysis was suggestive of hypocapnia with PaCO2 of 27.8 mm Hg with deranged liver function test SGOT 579.13 U/L and SGPT 372.75 U/L. On CBC picture show HB 9.2, PCV 30% and platelets count 2, 59000 and APTT 55.00 seconds. Ophthalmology, orthopedic and surgery consultations were done, and their advices were followed. During her hospital stay she was re-intubated on 4th post admission day due to sudden drop in spo2, decrease in heart rate up to 20 bpm and pinpoint pupil and 1 cycle CPR was received due to cardiac arrest followed by de-saturation and re-intubated with cuffed tube 4.5 mm and fixed at 14 cm and kept in mechanical ventilator setting ACVC mode. She had multiple episodes of tonic clonic seizure with secondary generalization, and anti-epileptic drugs were readjusted consulting pediatric team. CT head was immediately done after stabilization and showed bilateral parietal-occipital hypodensity suggestive of ischemic changes (Figure 3). Child's parents were counseled. MRI brain screening (DWI/FLAIR) was done which revealed laminar pattern of diffusion restriction in bilateral parietal and occipital region suggestive of acute laminar necrosis (Figure 4). Diagnosis of post-traumatic posterior reversible encephalopathy syndrome was made and child was managed aggressively.

Discussion:

Similar clinical presentations can also be seen with posterior circulation ischemia-infarction, vasculitis, status epilepticus; hypoglycemia, cerebral venous thrombosis, and reversible cerebral vasoconstriction syndrome.[7] Besides pounding type of headache, altered sensorium, recurrent seizure and visual disturbances are common in PRES. Cerebral auto-regulation is the ability to maintain a constant cerebral blood flow even with changes in cerebral perfusion pressure. Cerebral auto-regulation preserves cerebral blood flow leading to vasodilatation during systemic hypotension and vasoconstriction during systemic hypertension. Sudden rise in blood pressure can exceed the capacity of cerebral blood flow auto-regulation leading to cerebral hyper-perfusion.[8] This auto-regulatory failure leads to blood brain barrier interruption and leakage of plasma and macromolecules into the interstitial space through tight junctions. In our case, PRES resulting from a major hypertensive response to laryngoscopy was our strongest guess since she underwent multiple intubations during early treatment course. It is a well-known fact that laryngoscopy and intubation, especially under emergency conditions with the least support of pharmacological agents, result in a remarkable sympathetic stimulation. Although 15–20% of patients with PRES are normotensive or hypotensive [8], however the record of blood pressure in our patient was fluctuating. Another possible explanation could be the cytokine storm or brain on fire seen in first 24 hours of pediatric brain injury. Excessive circulating cytokines contribute to the injury of the microvascular endothelium which in turn increases vascular permeability. Hydrostatic leakage and extravasation or transudation of fluid and macromolecules through damaged arteriolar walls into the adjacent brain parenchyma can result in vasogenic edema. [7] A sudden increase in blood pressure leading to vasospasm and failure of auto-regulatory mechanism can predispose to PRES.[9] There are three radiological presentations of this condition: [10] (1) a holohemispheric watershed pattern with a linear involvement of the frontal, parietal, and occipital lobes predominantly, along a watershed distribution, (2) a superior frontal sulcus pattern with predominant involvement of the frontal lobes, and (3) a dominant parietal–occipital pattern in which the typical predominance of the posterior lobes

was maintained. Brainstem, cerebellum, basal ganglia, thalami, internal capsule, and splenium of corpus callosum are the atypical sites of involvement. DWI and apparent diffusion coefficient (ADC) have been found to be helpful in differentiating atypical presentations of PRES from conditions such as central pontine/extrapontine myelinolysis, non-hemorrhagic infarcts, and hypoglycemic or hypoxic encephalopathy. [10] Due to vasogenic edema in PRES, ADC shows increased values with slightly increased signal intensity on DWI, whereas the other conditions show reduced ADC values due to cytotoxic edema. Our patient demonstrated increased intensity on DWI with increased ADC values in the parietal and occipital lobes.

Management has to be done very aggressively for high blood pressure, increase ICP and recurrent seizures for better outcome. Prognosis is usually favorable. The symptoms and lesions of PRES may resolve completely if the diagnosis and treatment are prompt, as was seen in our patient, repeat ct head showed resolving edema and cortical laminar necrosis in bilateral parieto-occipital lobes (Figure 5). However, failure to diagnose may lead to irreversible infarction and death, most often as a result of acute hemorrhage or massive posterior fossa edema causing obstructive hydrocephalus or brainstem compression.

Conclusion:

We suggest that PRES should be kept in mind in children presenting with blood pressure fluctuation as a possibility with encephalopathy and seizures in the setting of severe pediatric brain injury. In complex situations, MRI (DWI/FLAIR) is important to establish such uncommon diagnosis and aggressively manage the case for favorable outcome.

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