Case report

Clinico-anatomical correlations in asphyxiated babies who developed spastic quadriparesis at one year of age

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The neurologic correlates of Hypoxic Ischemic Encephalopathy in the newborn period and on follow of the high risk babies depend on the topography of the neuropathologic lesions. Selective Neuronal Necrosis, Parasagittal Cerebral Injury, Periventricular Leucomalacia and Focal Ischemic Necrosis are the four basic lesions occurring in Hypoxic Ischemic Encephalopathy[1]. Considerable overlap in the occurrence of these lesions often act as persistent confounders in predicting the neurologic sequelae.

Here we discuss the outcome of five babies who were delivered vaginally following uncomplicated pregnancies. Prolonged labour was the cause of intrapartum hypoxic insults and Apgars at 1 and 5 minutes were 0 to 3. Neonatal Neurologic Syndrome associated with clinically significant Encephalopathy persisted for 7 to 10 days in all the babies. They were treated at Level III NICU for Sarnat Stage II encephalopathy. Their mean duration of NICU stay was 15 days. Seizures occurred within first 24 hours of life and were controlled by Phenobarbitone and subsequently Phenytoin. However the other problems associated with severe asphyxia like meconium aspiration, hypoglycaemia, hypocalcemia, Persistent pulmonary hypertention, renal failure, need of ventilation for secondary apnoea were not encountered in anyof the cases. All the babies had spastic quadriparesis, microcephaly, growth retardation and developmental delay at one year follow up. Motor and Mental Developmental Quotient (MoDQ and MeDQ respectively), were calculated using Developmental Scale For Indian Infants (DASII)[2] which is based on Bayley Scales of Infant Development II norms.

Case I- The child had massive periventricular infarctswith consequent dilatation of the ventricles. The child had severe spasticity with contractures of tendoachilles and adductors of the hip. The ischemic insults were mainly periventricular and hence expected to involve the descending motor tracts subserving the lower limbs- but lateral extension into centrum semiovale and corona radiata resulted in involvement of upper extremities and cognitive development as well[3]. There were associated recurrent chest infections, feeding problems due to pseudobulbar palsy and refractory seizures. The child also developed choreoathetoid movements with dystonia possibly due to deep nuclear variety of neuronal necrosis involving the Thalamoputaminalregion[4]. Thus he exhibited Mixed spastic dyskinetic features. Background activity in EEG was depressed and undifferentiated with randomly superimposed abnormal waves. Visual Evoked Potential (VEP) showed prolonged P100 latency. Brainstem Auditory Evoked Response (BAER) showed persistent severe bilateral sensorineural hearing loss (SNHL). MoDQ and MeDQ scores were 30% and 25% respectively, thereby signifying severe developmental delay.

Case II- The child had occipito parietal encephalomalacia with loss of periventricular white matter. Fundoscopy was normal but the child had cortical blindness and delayed response in VEP.

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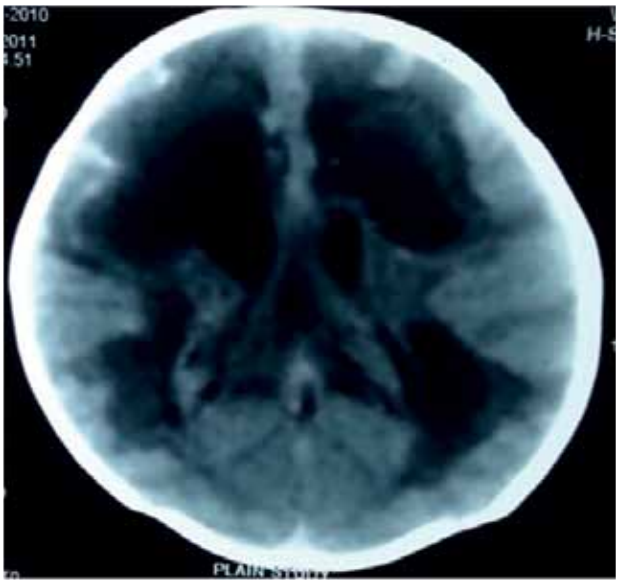

Figure 1- Massive periventricular ischemic injuries with subsequent loss of cerebal white matter resulting in ventriculomegaly. Ischemic injuries also involve the deep gray matter resulting in thalamic tissue loss and dilatation of third ventricle.

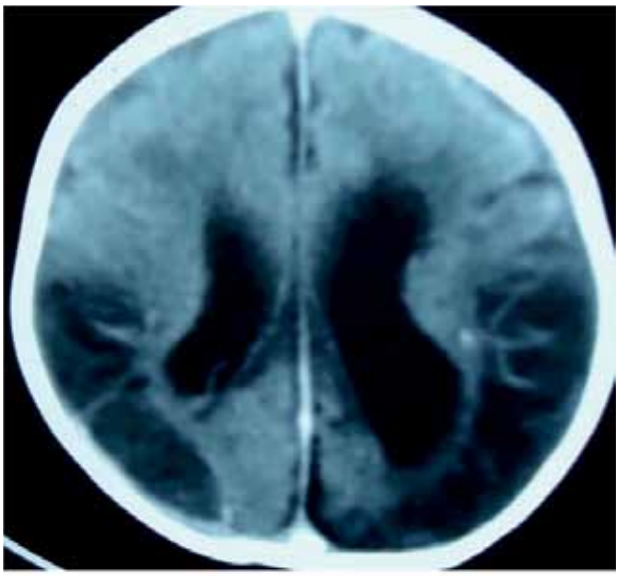

Figure 2- Decreased attenuation in Posterior parieto- occipital region along with loss of periventricular white matter with subsequent ventricular dilatation

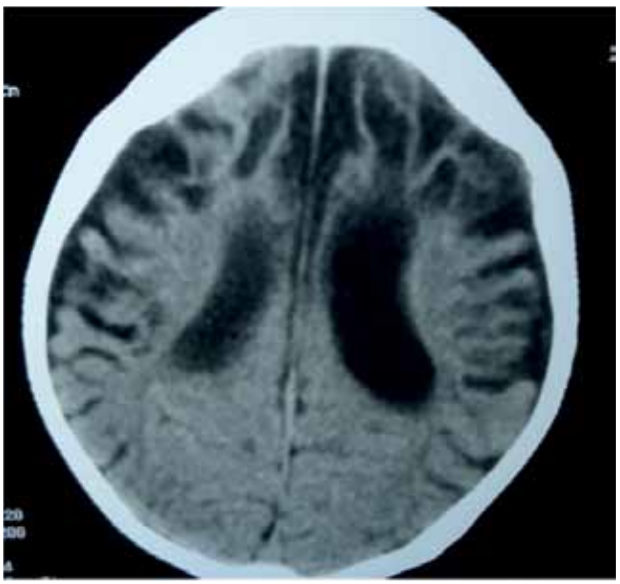

Figure 3- Strking degree of cortical atrophy manifested by prominent subarachnoid spaces and hydrocephalus ex vacuo. Prominent frontal lobe atrophy.

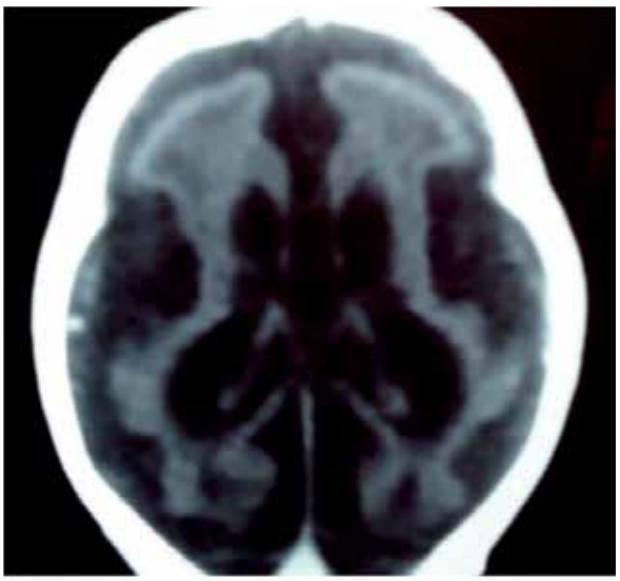

Figure 4- Extensive cortical, deep gray matter and periventricular atrophy and subsequent hydrocephalus ex vacuo and prominent subarachnoid spaces.

BAER showed persistent severe bilateral SNHL. These findings were attributed to damage to the posterior parietal- occipital region where resides mainly associative functions, especially relating to auditory and visual input and output[3]. EEG showed multiple foci of abnormal high voltage sharp waves. MoDQ and MeDQ scores were 58% and 50% respectively (moderate developmental delay).

Case III- The child had cortical atrophy manifested by prominent subarachnoid spaces and accompanying cerebral white matter injury, evidenced by ventricular dilatation. Significant damage to the frontal lobes resulted in prominent Frontal Release Syndrome.The child exhibited obligatory palmar and plantar grasp reflexes although they should have disappeared by 3 and 9 months of age. Rooting ,

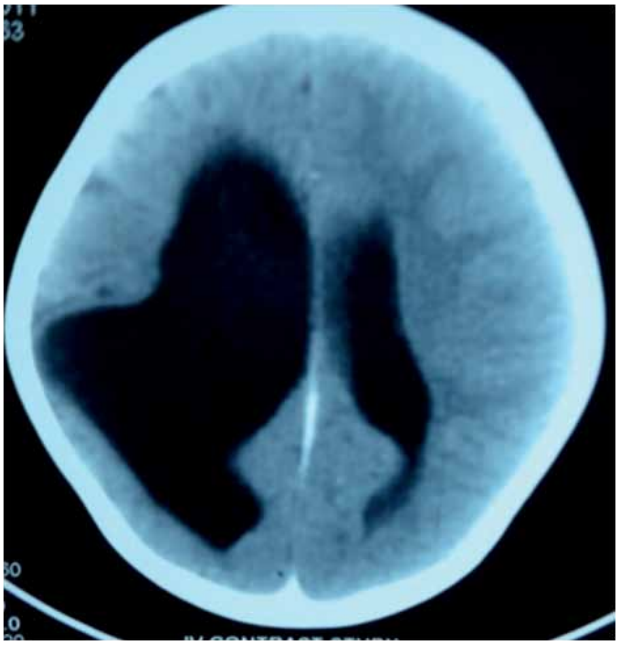

Figure 5- Diffuse loss of virtually all cerebral tissuenin thedistribution of right Middle Cerebral Artery territory and virtual absence of peritrigonal white matter on left side such that cortical gray matter virtually abuts the ventricular wall in that region.

sucking, swallowing reflexes were also exaggerated to the extent of wolving reflex[5]. Mac Carthy'sreflex persistently produced blinking response even on tapping over the vertex. He suffered from seizure disorder despite having normal EEG. BAER initially showed moderate degree of SNHL, but normalised by one year of age. Fundoscopy and VEP were normal. MoDQ and MeDQ scores were 60% and 58% respectively (moderate developmental delay).

Case IV- The child had diffuse cerebral atrophy and hydrocephalus- ex- vacuo with significant loss of both gray and white matter. Severe spastic quadriparesis coupled with contractures, cortical blindness (no response in VEP), profound SNHL in BAER and depressed background activity accompanied by lack of differentiation of normal frequencies in EEG marked the grim prognosis of the child. Fundoscopy revealed optic atrophy. MoDQ and MeDQ scores were 20% and 16% respectively (severe developmental delay).

Case V – The child had asymmetric spastic quadriparesis (left side more affected than right side) possibly due to greater loss of periventricular neurons on right side. EEG revealed cortical dysrhythmia, fundoscopy and VEP were normal. BAER at 3 months had revealed bilateral moderate SNHL- which had normalised by one year. MoDQ and MeDQ scores were 72% and 70% respectively- thereby suggesting that rarely infants with apparently severe bilateral cerebral destruction can have remarkable preservation of neurologic function[6].

In all these cases, spasticity relates to cortical damage (the pyramidal cells beingparticularly vulnerable to selective neuronal necrosis in term infants) and to damage of the pyramidal tracts descending through the periventricular white matter resulting in ventriculomegaly.Hearing deficit is attributed to involvement of inferior colliculus[7] which stands out in terms of vulnerability to hypoxic ischemic insults and also possible involvementof cochlear nucleus and superior olivary complex[8]. VEP abnormalities reflect the affection of geniculocalcarine radiations with consequent dilatation of occipital horns of lateral ventricles[9]and necrosis of the better differentiated neuronsofcalcarine cortex[3].

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