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MAGNETIC RESONANCE IMAGING OF MULTICYSTIC ENCEPHALOMALACIA IN INDIA

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ABSTRACT
Multicystic encephalomalacia is a rare subtype of encephalomalacia commonly seen in paediatric age group with hypoxic ischaemic encephalopathy. Since it has poor prognosis, it is essential to make early diagnosis. Radiological imaging plays an important role in diagnosis. We present our findings of multicystic encephalomalacia in term infant evaluated with MR imaging.

Key words: Multicystic encephalomalacia, HIE, MRI

INTRODUCTION

Multicystic encephalomalaciais the final stage brain injury characterised by multiple fluid filled cavities with septations associated with gliosis. Multiple lesions denotes diffuse brain parenchymal injury. The cause is multifactorial and commonly seen in paediatrics results in neuromotor developmental failure. A male term infants were brought with complaints of developmental delay and seizures[1-5]. History of GDM, perinatal asphyxia, multiple episodes of seizures and delayed milestones in postnatal period was given.

Materials and methods:

Twenty-one patients who presented with convulsion, mental-motor retardation and microcephaly and had evidence of multicystic encephalomalacia on MR images were included in this study. MR imaging patterns and clinical findings were reviewed. Consequently, we correlated MR imaging findings and clinical outcome.

On examination, the Patients had microcephaly, increased muscle tone in all four limbs, bilateral cortical thumb, exaggerated deep tendon reflexes, ankle clonus, bilateral extensor plantar reflexes and global developmental delay. Provisional diagnosis of cerebral palsy with spastic quadriplegia and global developmental delay was made and referred for magnetic resonance imaging of brain.

Results:

All patients had cortical thinning, white matter destruction, atrophy and gliosis. Tetraplegia was seen in 13 out of 15 patients with mixed type cerebral palsy in two patients with diffuse or symmetric involvement on MR imaging. Both of the patients with mixed type cerebral palsy had basal ganglia involvement on MR imaging. Hemiplegia was seen in two patients with asymmetric involvement on MR imaging.

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Microcephaly was seen in 13 patients with diffuse or symmetrical, and in one patient with asymmetrical, involvement. Microcephaly and tetraplegia was seen in all patients with cerebellar and basal ganglion involvement.

MRI brain study without i.v contrast was performed with following sequences SE and FSE technique, T1W, T1W, FLAIR, axial, T1Wsagittal section, DWIADC and MRA.Multiple cystic lesions of irregular shapes and different sizes noted in bilateral cerebral hemispheres which appear hypointense in T1W, hyperintense in T2W and hypointense in FLAIR sequences.

Sepatations in cystic lesions appear isointense. Bilateral lateral ventricles are dialated.Bilateral thalami,brainstem and posterior fossa contents appear normal. No evidence of calcifications. Bilateral internal carotid and right vertebral artery appear hypoplastic.With above said findings , radialogical diagnosis of multicystic encephalomacia was made.

Fig: 1a, b, c : (T1W, T2W and FLAIR axial) show classical MR features of multicystic encephalomalacia

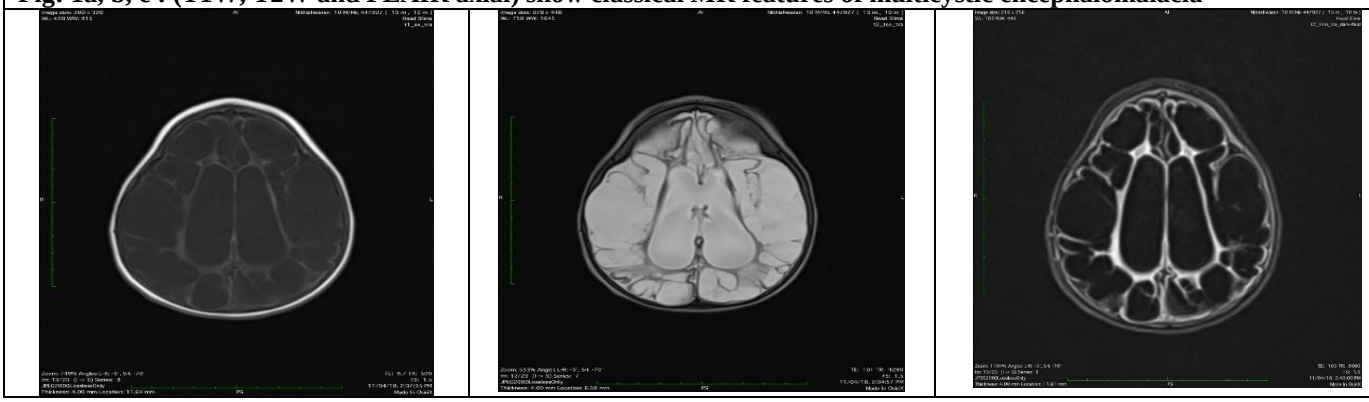


Fig: 2 a, b : (T1W sagittal and T2W coronal) show normal cerebellum, brainstem and thalami.

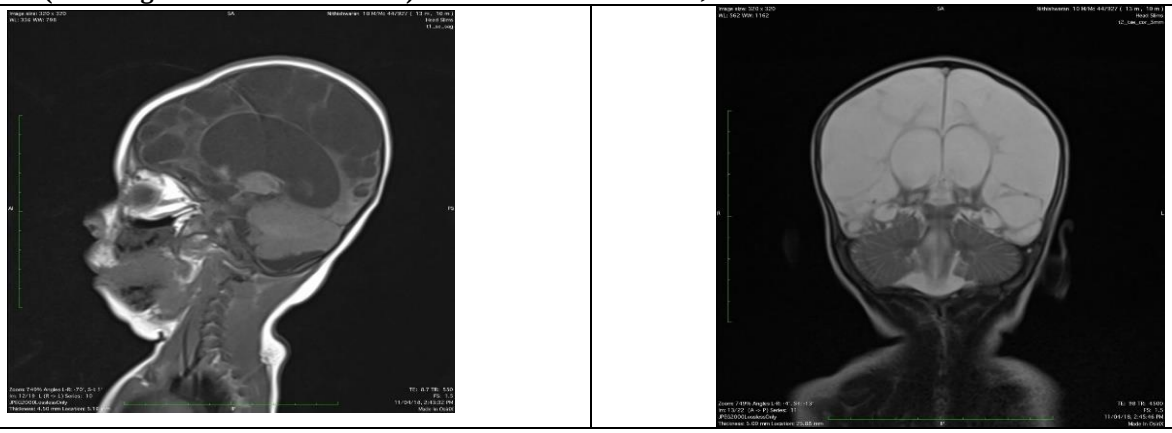
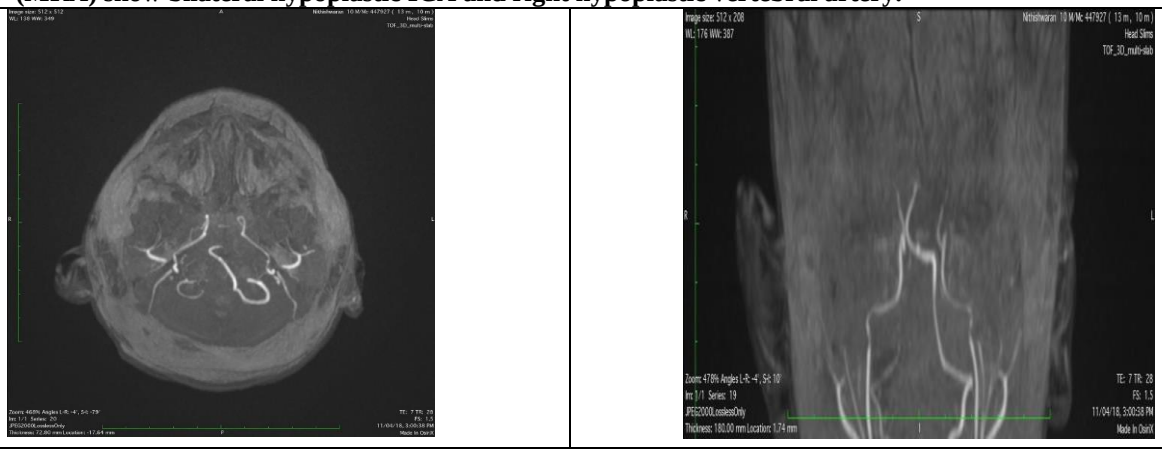


Fig: 3a, b: (MRA) show bilateral hypoplastic ICA and right hypoplastic vertebral artery.



Discussion:

Multicystic encephalomalacia is an irregular cystic area in the brain parenchyma which is the final result of the diffuse brain insult in late gestation, during or after birth [6,7]. There is formation of multiple cystic cavities of variable sizes with multiple glial septatitons in the area of necrosis [7]. It is pathologically characterized by astrocytic proliferation and glial septations in the damaged areas of the brain. The condition may be caused by infarction, infection or trauma. They may be focal or diffuse and their distribution will depend on the cause and severity of the injury and the post conceptual age of the patient. In cases of embolic or thrombotic insult, the lesions are distributed in the territory of the major cerebral artery. Ultrasonography, within one week of the insult shows increased echogenicity in the affected areas with cystic degeneration appearing after 1-4 weeks in term infants [8]. Cranial ultrasonography is the most sensitive modality for detection of the glial septa but it lags behind MR in the overall brain evaluation. In their study, however, orejon de Luna G et al [9] concluded that cerebral ultrasonography is the imaging modality of choice in the evaluation of the multicystic encephalomalacia. CT initially shows diffuse hypodensity in the affected area which eventually becomes cystic and CSF attenuating. Septations are common and calcification may be seen 1. CT cannot reliably differentiate between porencephaly and encephalomalacia.

Multicystic encephalomalacia is a rare disorder with poor outcome, commonly seen in neonates with HIE characterised by focal brain damage mostly involving the area of anterior and middle cerebral arteries with relative sparing of brainstem, thalami and cerebellum.Possible etiological factors include brain insults, cerebral ischemia/ infection / haemorrhage/ traumatic brain injury. Pathologically it is due to liquefactive necrosis of brain parenchyma. Microcephaly and spastic tetraplegia are mostly associated with diffuse involvement of brain. USG is more sensitive and non- invasive method to diagnose but it is operator dependent and inferior to MRI in overall evaluation of brain. CT will demonstrate hypodensity followed by cystic or CSF attenuation, but it cannot differentiate porencephal from multi cysticencephalomalacia [9, 10]. MRI will show multiple cystic lesions in cerebral hemisphere iso-intense to CSF in all sequences with volume loss evidenced by ventricular dilatation. Area of distribution differ based on cause and severity of insults. Differential diagnosis include porencephaly and hydranencephaly [11,12].

Conclusion:

Microcephaly and spastic tetraplegia were developed mostly in patients with diffuse involvement, whereas hemiplegia was seen in patients with asymmetric involvement. The clinical outcome was worse in patients with cerebellar and brainstem involvement. Therefore, we supposed that the symmetry of lesions and cerebellar or brainstem involvement might be used as a prognostic indicator. Hydranencephaly, sometimes, has to be differentiated from severe hydrocephalus. There is a thin rim of cerebral tissue around the dilated ventricle than can usually be identified only on MR imaging.

As the prognosis and clinical outcome of multi cysticencehamalacia is worst, radiological imaging like MRI plays an important role. Imaging should be done in neonates and infants who suffer from asphyxia, intracerebral infection, haemorrhage and trauma to make early diagnosis and timely family counselling

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