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Case Report

Imaging of Multicystic Encephalomalacia of Infancy (MICE): A Case Report

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Abstract

Multicystic encephalomalacia is a diffuse lesion of the brain in which cerebral parenchyma is replaced by cysts of varying sizes in the perinatal period. The prognosis is grave and prompt recognition of this condition with the help of neurosonography at birth helps in further management. It is stressed that multiple cystic encephalomalacia has a characteristic appearance on ultrasound and all children should be screened routinely so as to detect it at an early stage. We present a case report of an infant showing multiple cysts spaces in brain secondary to hypoxic insult.

Keywords: Hypoxia, Multicystic Encephalomalacia, Neurosonography

1. Introduction

Multicystic encephalomalacia (MICE) is a condition defined anatomically by the presence of multiple cavities in the greater part of both cerebral hemispheres, generally seen in the first year of life. It is associated with profound neuromotor delay and consequent grave prognosis. Typically, the posterior fossa is spared with usually no connection to the ventricular system. It may be clinically silent; and is discovered when the infant fails to achieve the early milestones of neuromotor development. Various causes which include asphyxia, meningitis and twin-to-twin transfusion are responsible for this disorder[1]. Neurosonography has been described as the primary investigative modality and is considered superior to CT[2].

2. Case Report

A full term infant delivered at 40 weeks gestation delivered of normal vaginal delivery with APGAR score: 2, 2, 3 and Weight 3042 g, Head Circumference 35 cm was transferred to tertiary care rural hospital at 1 day of age after severe postnatal acidosis and asphyxia with a 24-hr history of seizures. High doses of anticonvulsants were required for persistent seizures. The baby developed marked neuromotor retardation. Clinical assessment revealed a positive Mono reflex, inability to hold the head, and a very poor suck reflux.

The baby came in the Radio-Diagnosis department for the Neurosonography and was investigated. The cranial ultrasound was performed through the anterior fontanel. Coronal section of the brain revealed multiple thin-walled cysts within the brain parenchyma bilaterally (Fig.1). The ventricles were mildly dilated, each measuring 1.2 cm at the frontal horns (Fig. 2 & 3). The brain parenchyma was reduced to thick septae between the cysts. The cerebral vessels were seen traversing between the cysts. The cyst walls per se showed no vascularity. All these features were suggestive of Multiple Cystic Encephalomalacia.

The baby also underwent Computed Tomography and the findings were similar to those on Neurosonography.

Figure 1: Coronal section of the brain shows multiple thin-walled cysts are noted within the brain parenchyma bilaterally.

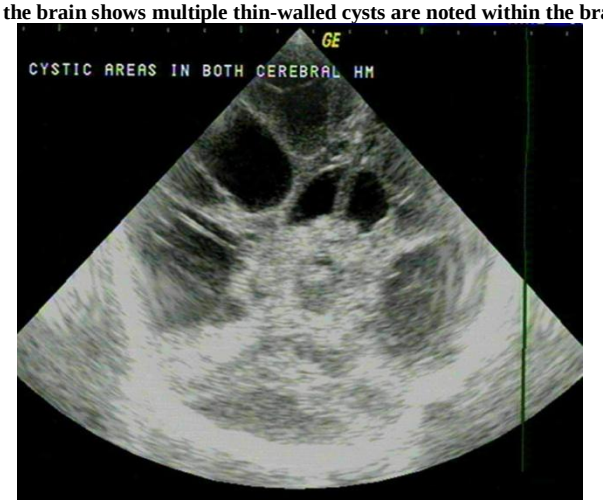


Figure 2: Coronal section through the frontal horns: The numerous cysts replacing the brain parenchyma are seen again. The ventricles are mildly dilated, each measuring 12 mm at the frontal horns.

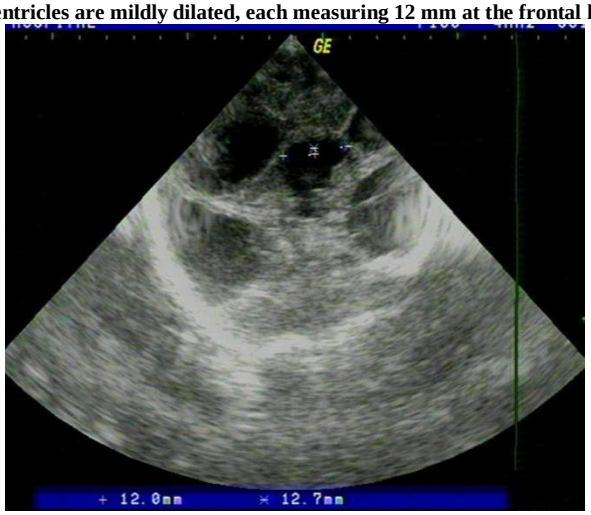
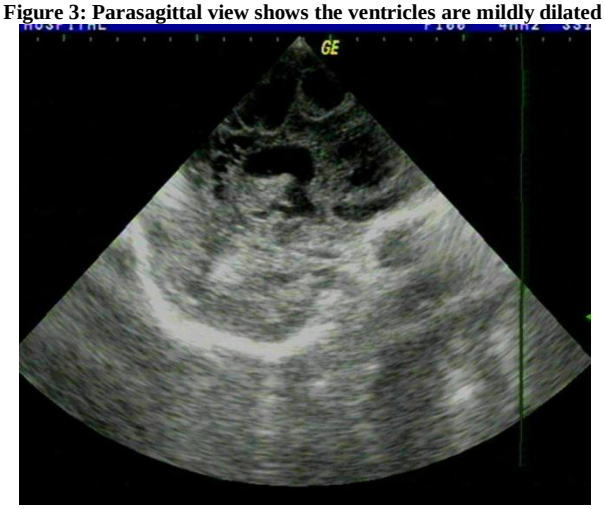


Figure 3: Parasagittal view shows the ventricles are mildly dilated



3. Discussion

Encephalomalacia is an area of focal brain damage which when diffuse may present as large areas of multisystem degeneration. The tendency of human brain to undergo dissolution and cavitation is a function of the timing of insult. It had been described under a variety of synonyms in paediatric and pathological literature[3]. Most cases show the lesions distributed in the region of anterior and middle cerebral arteries sparing cerebellum and medulla. Brain hypoxia is the common etiological factor in the development of multiple cysts[4]. Stannard and Jimenez found no communication between cyst and ventricular system in their four cases of MICE [1].Pathologically the damaged area has astrocytic proliferation and glial septations. The presence of pus in multiple cysts suggests that either bacterial cerebritis or septic embolism resulted in the formation of multiple cysts in both cerebral hemispheres.

The cystic deformity has been recognized in life by sonography, ventriculography, pneumoencephalography, and computed tomography (CT). Amongst all imaging modalities, cranial sonognaphy is preferred in MICE, apart from radiation protection due to:
1. Diagnosis of the condition.
2. Reliably assess the size of the cysts and the need for cyst shunts.
3. Monitor response to treatment.
4. No radiation exposure.

Neurosonography reveals presence of multiple, thin walled cysts replacing the brain parenchyma. There may or may not be associated ventriculomegaly or interventricular haemorrhage. If the encephalomalacia is secondary to intracranial infections, calcifications may be seen. The brain parenchyma is reduced to thick septa between the cysts. NECT demonstrates the numerous bilateral cysts, compatible with pattern of extensive bilateral infarction and liquefaction of the brain parenchyma.

Cranial US provide a convenient, noninvasive, relatively low-cost screening examination of the hemodynamically unstable neonate at the bed-side. The examination also imparts no radiation. Sonography is sensitive for the detection of haemorrhage, periventricular leukomalacia (PVL), and hydrocephalus. Doppler interrogation and the assessment of resistive index (RI) provide additional information on cerebral perfusion. Normally, the RI decreases with increasing gestational age, and thus correlation with gestational age is necessary for accurate interpretation of RI results. Decreased RI is noted to be an abnormal finding and is postulated to be caused by impairment in cerebral autoregulation and subsequent decreased cerebrovascular resistance and increase in end-diastolic flow. However, sustained asphyxia with subsequent development of intracranial haemorrhage or diffuse cerebral oedema and loss of forward diastolic flow result in increased RI and is indicative of a poor outcome[5,6,7].

Sonography is operator dependent, however, and less sensitive to structural abnormalities in the cerebral convexity and in the brainstem[8]. Parenchymal abnormalities, such as PVL and cerebral oedema, identified at US are also often nonspecific[9]. Gupta *et al* also emphasised that sonography is diagnostic for MICE and does not want further investigation.

4. Conclusion

Cranial sonognaphy should be considered in the assessment of all neonates and infants who suffer from asphyxia, intracerebral haemorrhage, and intracranial infection. As the prognosis of multiple cystic encephalomalacia is grave, prompt recognition will facilitate timely family counselling. Enlargement of the cysts can be detected and the effectiveness of cyst taps or cystopenitoneal shunts recorded. To prevent inordinate increase in head size, more than one cyst shunt may be required.

Hence, sonography is diagnostic for multiple cystic encephalomalacia (MICE) and does not warrant any further investigations.

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