MR FEATURES IN A CLASSICAL CASE OF MULTICYSTIC ENCEPHALOMALACIA AND ITS DIFFERENTIAL DIAGNOSIS

Rajul Rastogi

Yash Diagnostic Centre, Yash Hospital & Research Centre,Civil Lines, Kanth Road, Moradabad, U.P.-244001, India

Abstract : *Multicystic encephalomalacia is a pediatric entity and a rare disease in childhood where the brain tissue is substituted by cavities of variable sizes. The cause is the insult to the brain tissue. This disease has a very poor outcome, for this reason early diagnosis is very important. Therefore, we are presenting classical radiological features of multicystic encephalomalacia, its differential diagnosis is also discussed.*

INTRODUCTION

Encephalomalacia represents the end stage injury to the brain tissue in the late fetal or early neonatal life. It is multifactorial in etiology. It is characterized by fluid filled cavities associated with gliosis. The cavities show internal septations. The multicystic variety is characterized by several such cavities and represents the diffuse insult to the brain tissue. This entity needs to be differentiated from the closely resembling porencephaly and hydranencephaly.

CASE REPORT

A 6 month old infant presented to our hospital with the complaint that the child is not doing well for few months. Clinical examination of the child revealed delayed milestones, past history, antenatal and perinatal history were unremarkable. Laboratory tests did not reveal any significant abnormality.

The child was referred to the radiology department for evaluation of the brain. Cranial sonography revealed multiple fluid filled cavities with internal septations in both the cerebral hemispheres. Thalami, brainstem and cerebellum appeared normal. Lateral ventricles were dilated.

MR imaging of the brain also revealed multiple fluid filled cavities with internal septations in both the cerebral hemispheres involving primarily the frontal and parietal lobes. The cavities were hypointense on T1W1 and hyperintense on T2W1. Septations appeared isointense on T1W1 and hyointense on T2w1. The cavities did not reveal any significant internal heterogeneity. Thalami, brainstem and posterior fossa were normal. Lateral and third ventricles were dilated with bending of the lateral walls of the lateral ventricles (Figure 1a to 1c)

Based on the above findings, the radiological diagnosis of classical multicystic encephalomalacia was made.

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[1,2]. There is formation of multiple cystic cavities of variable sizes with multiple glial septatitons in the area of necrosis[2]. It is pathologically characterized by astrocytic proliferation and glial septations in the damaged areas of the brain.

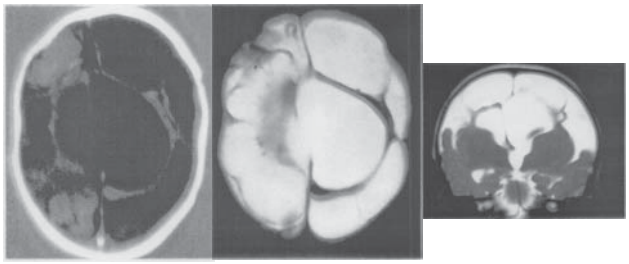

Figure 1a-1c: Axial T1w1 and axial & coronal T2W MR images show classical features of multicystic encephalomalacia.

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. However, if the insult is due to mild to moderate hypotension, the watershed zones intervascular boundary of the cortex and white matter are affected In contrast, when the insult is due to severe hypotension, the entire cortex is affected with sparing of the deep periventricular white matter only. In cases of infection, only the involved region of the brain is necrosed.

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[3]. 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[4] 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.

On MR, the affected areas (areas of reactive astrocytosis and the areas of tissue injury) appear hypointense on T1 and hyperintense on T2 paralleling fluid[3]. All patients have cortical thinning, white matter destruction, atrophy and gliosis. Basal

Correspondence: Dr. Rajul Rastogi,
e-mail: drsharastogi@gmail.com

ganglia or cerebellar involvement may be seen MR imaging. Microencephaly and spastic tetraplegia develops mostly in patients with diffuse involvement, whereas hemiplegia in patients with asymmetric involvement. The clinical outcome is worse in patients with cerebellar and brainstem involvement. Therefore, the symmetry of lesions and cerebellar or brainstem involvement might be used as a prognostic indicators[5].

The important differential diagnosis of multicystic encephalomalacia is porencephaly and hydranencephaly.

Porencephaly refers to focal cavities with smooth but shaggy walls and minimal surrounding glial reaction[1,2,7]. To differentiate it from schiencephaly (also known as agenetic porencephaly), it is often referred to as encephaloclastic porencephaly. The latter is the result of insult to the area of brain in late gestation, perinatal or postnantal period[1,8,9].

On imaging, encephaloclastic porencephaly appears as smooth walled cavities that the isointense to CSF on all sequences. The cavities are devoid of internal features as septae and the surrounding brain is of normal signal intensity (figure 2).

Figure 2: Axial T2W MR images shows a large porencephalic cyst in the left cerebral hemisphere.

Hydranencephaly is a condition in which most of the brain mantle has been damaged liquefied and resorbed[10] and can be considered as porencephaly of the nearly the entire brain. Thin walled sacs containing CSF lined by leptomeninges replace the cerebral hemispheres[1,2]. Multiple causes have been advocated including vascular and infectious (toxoplasmosis and CMV).

Clinically, the head may be normal small or large in size. The child is always mentally retarded.

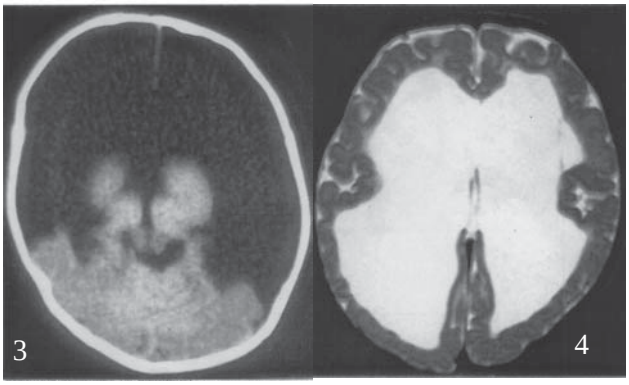

Figure 3: Axial CT image shows a classical case of hydranencephaly with relative sparing of thalami and cerebellum.

Figure 4: axial T2W MR image shows dilated lateral ventricles surrounding by cortical mantle in a case of hydrocephalus.

On imaging, the cerebral hemispheres are nearly completely replaced by CSF[1,6]. The thalami are usually preserved. The inferior and medal aspects of the frontal and temporal lobes may also be preserved. The brainstem is usually atrophic. The cerebellum is almost always normal (figure 3). 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 (figure 4).

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