Post Infectious Multicystic Encephalomalacia
Encéfalomalacia Multicística Pós-Infecciosa

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ABSTRACT
The term multicystic encephalomalacia is used to describe the presence of necrosis areas that turn into cystic lesions inside the brain. It is usually seen in the first year of life. We describe a case of a newborn with a large occipital encephalocele surgically treated. He complicated with Staphylococcus aureus meningoencephalitis that was treated with Vancomycin. Control computed tomography twenty-four days after surgery showed significant multicystic encephalomalacia, which is a uncommonly rapid progression, since it usually takes months for a complete cystic degeneration. The pathophysiological mechanism that we propose is a direct aggression of the bacterial infection and a disturb of the brain perfusion. Secondary to a Staphylococcus aureus infection stablished a severe infection of CNS inducing a disrupt in the vascular autoregulation. Multicystic encephalomalacia is an uncommon disease of the childhood and has a very poor outcome, early diagnosis is very important for family counseling.

Key words: Multicystic encephalomalacia; Encephalomalacia; Encephalocele; Meningoencephalitis

RESUMO
O termo encefalomalácia multicística é usado para descrever a presença de áreas de necrose que evoluem para lesões císticas no parênquima cerebral. Acomete principalmente crianças menores de um ano de vida. Descrevemos o caso de um recém-nascido com uma grande encefalocele occipital tratada cirurgicamente. Apresentou complicação de meningoencefalite por Staphylococcus aureus tratada com vancomicina. A tomografia de crânio de controle, após vinte e quatro dias do procedimento, revelou uma grave encefalomalácia multicística. Normalmente são necessários alguns meses após o agravamento para o aparecimento da degeneração cística completa, portanto, foi uma evolução muito atípica pela rápida evolução. Acreditamos que o mecanismo fisiopatológico envolvido é a ação direta da infecção bacteriana e um distúrbio de perfusão cerebral secundário a uma falha no mecanismo de auto regulação, causado pela infecção grave do sistema nervoso central pelo Staphylococcus aureus. A encefalomalácia multicística é uma doença incomum e está associada a um mau prognóstico. O diagnóstico precoce é muito importante para o aconselhamento familiar.

Palavras-chave: Encefalomalácia multicística; Encefalomalácia; Encefalocele; Meningoencefalite

INTRODUCTION

The term multicystic encephalomalacia (MCE) is used to describe the presence of areas of necrosis that develop into cystic lesions inside the brain. This lesion is usually due to severe asphyxia and/or hypotension, but may also be secondary to meningoencephalitis, abusive head trauma¹,²,⁴,⁵,⁷,⁹, twin-to-twin transfusion and more rarely to carbamyl phosphate synthetase 1 deficiency (urea cycle enzyme) and deficiencies of sulfite oxidase (mitochondrial enzyme encoded by the SUOX gene)⁶,⁸.

Multicystic encephalomalacia is an uncommon disease of the childhood and has a very poor outcome, with early diagnosis being important for family counseling⁴.

CASE REPORT
A 2-hour-old newborn was referred to Department of Neurosurgery at Santa Casa de Belo Horizonte, Brazil, after being diagnosed with large occipital encephalocele (Figure 1). No medical care was given before birth.

On examination, the head circumference was in normal size, and both fontanels were pulsatile and not tense. The infant accepted...
breast feed, and had no neurological deficit. A large liquid-filled sac originating around the external occipital protuberance was present, with a 41-centimeter diameter. The sac was soft, fluctuant, translucent, and its walls were intact.

Computed tomography scan (CT) (Figure 2) showed a bone defect in the occipital region, with mild brain herniation. The ventricular system was within normal limits.

Forty-eight hours after birth, the sac was surgically removed and the defect repaired, the procedure was uneventful. On the seventh day after surgery, the child presented fever and mild signals of infection at the surgical site. There was no signal of cerebrospinal fluid leak. Transfontanelle ultrasound revealed no signals of hydrocephalus. Lumbar puncture was suggestive of bacterial infection and a posterior culture was positive for *Staphylococcus aureus*. Antibiotic therapy was initiated with Vancomycin. There was good response, with resolution of the fever and infections signs and maintenance of the cephalic perimeter.

Control CT twenty-four days after surgery showed severe multicystic encephalomalacia (Figure 3).

The hallmark of this case is the very fast evolution to multicystic encephalomalacia secondary to a neurosurgical complication to correct an encephalocele.

Figure 1: A newborn with a large (41 cm in the larger diameter) occipital encephalocele.

Figure 2: An CT showing the normal aspect of the ventricular system.

Figure 3: An axial CT showing severe multicystic encephalomalacia.

**Discussion**

According to Stannard et al., MCE is occasionally present at birth, and it is usually seen in the first year of life. Orejón de Luna et al. observed that 58% of the patients were diagnosed within the first month of life, 32% between 1 and 12 months-old, and 11% older than 1 year-old. Naidich et al. characterized multicystic encephalomalacia as: 1) moderate atrophic ventricular dilatation; 2) intact ventricular walls with thickened subependymal glial layer; 3) multiple cysts of irregular size and shape distributed throughout the cerebral white matter and the inner layers of the cortex bilaterally; and 4) relative sparing of the orbital surfaces of the frontal lobes, the temporal lobes below the superior temporal gyri, the basal ganglia, the cerebellar hemispheres, and the spinal cord (except
for secondary degeneration of corticospinal tracts).

Orejón de Luna et al. published a series of nineteen children diagnosed of multicystic encephalomalacia. The causes were twin pregnancy, 12 cases; perinatal hypoxia, 4 cases; perinatal infection, 1 case; cardiac arrest, 1 case; and 1 patient with multiple embolism of unknown cause. The clinical development was unfavorable. Two children died (10.5%). They studied the outcome of 15 patients. Eleven patients (73.3%) showed severe deficit and 4 (26.6%) mild or moderate deficit. All the 15 patients (100%) presented developmental delay, microcephaly (73.3%), spastic tetraplegia (73.3%), seizures (46.6%) and hemiparesis (26.6%).

Multicystic encephalomalacia secondary to bacterial infection was also described by Ries et al. A case of a premature baby with Enterobacter sakazakii meningitis and large bilateral hypo- and hyperdensities suggesting massive hemorrhagic and ischemic intracerebral infarctions leading to multicystic encephalomalacia.

Weidenheim et al. presented eight cases of multicystic encephalomalacia over a 34-year period and searched for herpes viruses as a possible etiological agent. They performed immunocytochemical methods, in situ hybridization and polymerase chain reaction (PCR) to search for herpes simplex virus types 1 and 2 (HSV1 and HSV2), cytomegalovirus (CMV), varicella zoster virus (VZV), Epstein-Barr virus (EBV) and JC variant of papovavirus (JCV). Only one case showed evidence of latent HSV infection by PCR. CMV, VZV, JCV and EBV were not detected. Arteriopathy was noted in one case. The widespread nature of the lesions and their association with perinatal ischemia suggested that severe hypoxia might be the more common etiology of MCE. Term infants seem to be especially susceptible to this type of cerebral damage.

In our case, we observed a complete cystic degeneration of the brain comparing two CTs within a 24-days interval (Figure 2 and 3). This is a uncommonly rapid progression, since it usually takes months for a complete cystic degeneration. The physiopathological mechanism that we propose is a direct aggression of the bacterial infection and a disturb of the brain perfusion, because of a disrupt in the vascular autoregulation induced by a severe infection of the CNS secondary to a Staphylococcus aureus wound infection.

REFERENCES


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