

## LEARNING BY IMAGES

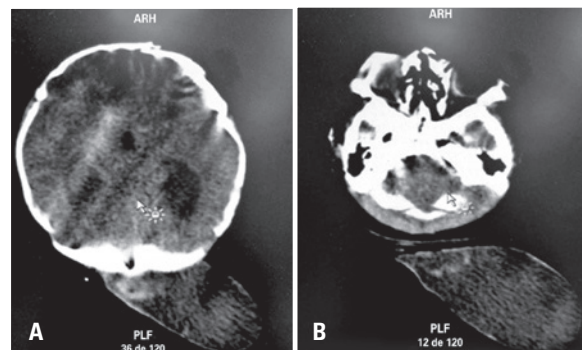
# Congenital occipital myelocele

## Mielocele occipital congênita

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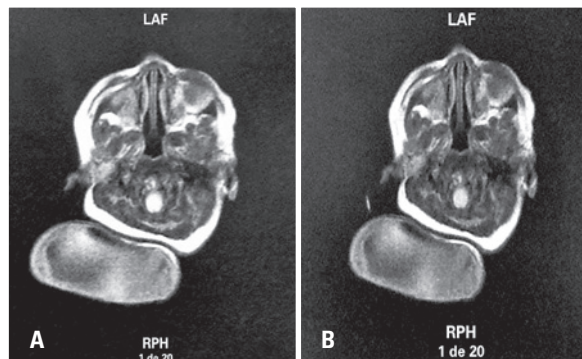
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**Figure 1.** Cranial computed tomography with occipital subcutaneous cystic formation to the left with licorice attenuation of content associated with occipital bone micropuncture in the midline and cortex isodense mass, therefore, suggesting encephalomeningocele



**Figure 2.** Newborn with hernia sac in occipital region



**Figure 3.** Cranial magnetic resonance imaging showing median suboccipital bone defect with extrusion of superior aspect and cerebellar vermis, cerebrospinal fluid and meninges (encephalomeningocele)

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## INTRODUCTION

Congenital encephalocele is a neural tube closure defect that presents herniation of cranial contents because of a cranial congenital malformation, the occipital encephalocele is the most common form of this defect.<sup>(1)</sup> Encephalocele often occur because of occurrence of occipital bone defect within the fourth week of embryogenesis and it can be extended for foramen magnum and affect the posterior arch of the atlas.<sup>(2)</sup> Hernia sac content varies and worst prognosis is large brain content inside the sac.<sup>(3,4)</sup> Etiology of this malformation include genetic and environmental factors, such as folate deficiency that is prevented with the use of folic acid supplements during pre-conception period until 12 weeks of gestation, and also for poor prenatal care.<sup>(5,6)</sup> Although this malformation varies in several demographic regions, neural tube closure defects incidence is approximately 1 in 1,000 live births.<sup>(7,8)</sup>

Both adequate prenatal care and imaging tests are importance to identify variations of this affection. For this reason, clinical case reports and their on encephalocele images are important.

## DESCRIPTION OF CLINICAL CASE

A 19-years-old healthy primigravida mother with no remarkable clinical history, who reported no use of folic acid or iron supplementation during the preconception and prenatal period.

During prenatal period, ultrasonography tests according to second and third trimester of gestation showed a single and live fetus with cystic formation in posterior cervical region that could indicate a cystic hygroma and occipital encephalocele, in addition to severe bilateral ventriculomegaly.

Because of the features of the case, we decided to undertake a cesarean section in week 39 of gestation. This was a female newborn weighting 3,100g, measuring 45cm, with head circumference of 33cm and Apgar score of 9.9. The patient evolved clinically stable without interurrences. However, an encephalocele was observed (Figure 1).

A computed tomography (Figure 2) and a cranial magnetic resonance imaging (Figure 3) were requested after the child birth. Tests results showed occipital encephalomeningocele with median suboccipital bone defect with extrusion of content similar to cerebrospinal fluid and meninges.

## DISCUSSION

Encephalomeningocele is a congenital malformation caused by neural tube closure defect. This disease has a poor prognosis because it can cause infection in central nervous system, in addition to advanced stages motor and sensory deficiencies.<sup>(9,10)</sup>

Cranial computed tomography and magnetic resonance imaging are tests used to diagnose this type of disease, define hernia sac content, and to evaluate the best surgical approach.<sup>(9)</sup>

In our case, the mother did not receive folic acid supplementation during preconceptional period up to 12 weeks of gestation, which is a risk factor for evolving of this disease<sup>(5)</sup> in newborns.

Of note, to perform imaging tests adequate to gestational age is paramount to diagnose as early as possible affections such as the encephalomeningocele.

During prenatal tests a clinical feature compatible with encephalocele was identified, but only after the birth more specific tests were done such as cranial computed tomography and magnetic resonance imaging. Results showed a clinical picture different from encephalocele, the encephalomeningocele, being this latter more severe and with worse prognosis because it evolves a larger parcel of cerebral content, such as brain and meninges.<sup>(9)</sup>

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