Basal sphenoethmoidal encephalocele in association with midline cleft lip and palate: case report*

Encefalocele basal esfenoetmoidal associada a fissura labiopalatina mediana: relato de caso

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Abstract Association of basal sphenoethmoidal encephalocele with midline cleft lip and palate is extremely rare. The authors report the case of a nine-year-old girl presenting a midline facial cleft with meningocele that was noticeable through the palatine defect as a medial intranasal pulsatile mass. An analysis of clinical and radiological findings of the present case of cranial dysraphism is carried out.
Keywords: Encephalocele; Cleft palate; Congenital malformation.

Resumo Associação de encefalocele basal esfenoetmoidal com fissura labiopalatina é extremamente rara. Relatamos um caso de uma criança de nove anos de idade apresentando uma fissura facial mediana com meningocele, que era evidente através da falha do palato como uma massa mediana intranasal pulsátil. Uma análise dos aspectos clínicos e radiológicos deste caso de disrafia craniana foi realizada.
Unitermos: Encefalocele; Fissura palatina; Malformação congênita.

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INTRODUCTION

Sphenoethmoidal meningocele is a congenital defect of the anterior cranial fossa with herniation of meninges and, eventually, of parts of the brain (encephalomeningocele), determining a protrusion toward the nasal cavity through a defect in the ethmoid and sphenoid sinus. Such a condition corresponds to a subtype of basal meningoencephalocele[(1)].

Different classification have been proposed for the anterior cranial fossa encephaloceles that, in a simplified way, are divided into two major groups, according to the herniation topography. The first group includes sincipital (frontal) encephaloceles where the cranial contents herniate through the foramen cecum over the cribriform plate of the ethmoid. The second group includes basal encephaloceles with protrusion through the cribriform plate itself or through the sphenoid body[(2)].

General encephaloceles prevalence is estimated to occur in one of every 35,000 to 40,000 live neonates. The basal presentations, that are even more rarely found, represent only 2% to 10% of such encephaloceles[(3,4)]. Considering the rarity of this malformations association, whose treatment leads to high morbidity and mortality, and with few reports in the literature, the authors analyze the clinical and radiological findings of the present case of have cranial dysraphism.

CASE REPORT

A female, nine-year-old child presenting midline cleft lip and palate, hypertelorism, low height/weight development and normal intellectual development. Examination of the oral cavity, demonstrated a wide cleft palate that allowed direct visualization of a round, pulsatile cystic mass, with a brownish color, regular and smooth surface, measuring about 4 cm in diameter, located on the nasal cavity midline (Figure 1).

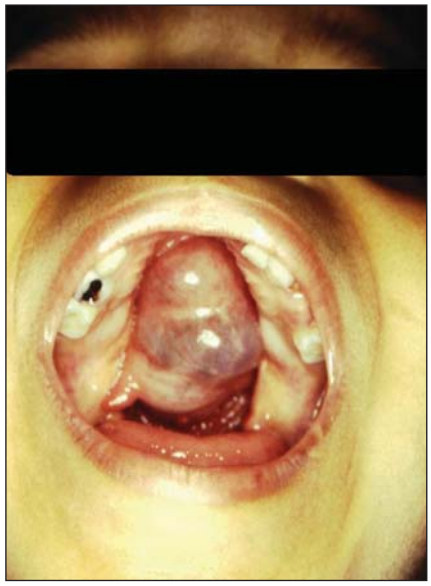

Figure 1. Basal sphenoethmoidal encephalocele. Visualization of a pulsatile cystic mass through the wide cleft palate.

Free and informed consent was obtained from the child's mother, and the Research Ethics Committee has approved the present study publication.

Computed tomography (CT) demonstrated metopic suture diastasis characterizing hypertelorism, and an empty sella turcica occupied by a cystic image. Mag-

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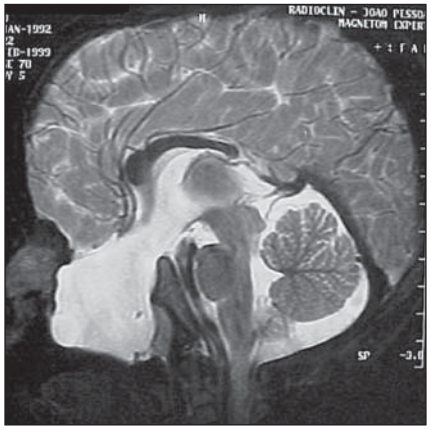

Figure 2. MRI, sagittal section demonstrating image with fluid density in the anterior region of the skull base (T2).

netic resonance imaging (MRI) demonstrated fluid density in the anterior region of the skull base, showing ethmoid and sphenoid defect on the midline with nasal extension, besides a hard palate defect (Figures 2 and 3).

The patient's chronological age was nine years, while her osseous age was five years. Basal human growth hormone (HGH) levels after clonidine treatment was 0.2 ng/ml (reference: 0 to 10 ng/ml).

DISCUSSION

Data reporting such defects association are scarce in both the Brazilian and foreign literatures.

Geographic and racial factors influence both the frequency of this condition and the encephalic area to be involved, showing endemic levels in the Southern Asia. At two health centers in Bangkok, Thailand, in the period from 1992 to 1996, 120 cases of frontoethmoidal encephalomeningocele (sincipital). However, the basal presentation is rarely observed in all of the racial groups[5].

Clinically, basal encephaloceles may manifest as a mass on the nasal cavity midline causing, or not, obstruction, feeding difficulties, cerebrospinal fluid leakage and meningitis. Such a condition may be associated with cleft lip and palate, microphthalmos, bifid nose and optic nerve coloboma. Considering the presence of a nasal mass, glioma, dermoid cyst and polyp should be taken into consideration in the differential diagnosis[1].

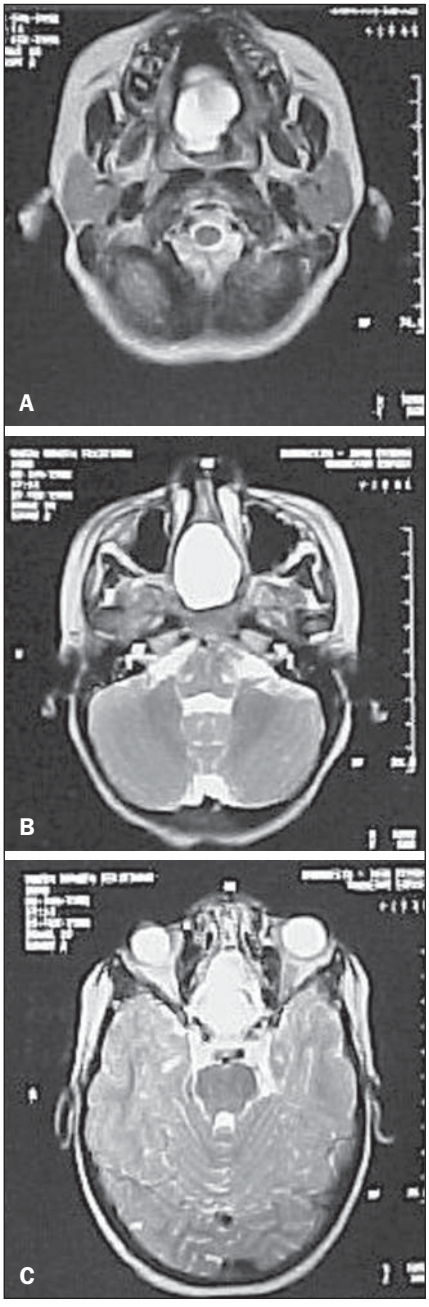

Figure 3. MRI, axial section demonstrating showing ethmoid and sphenoid defect on the midline with nasal extension, besides a hard palate defect (T2).

Advanced imaging studies are extremely necessary both to confirm de diagnosis and to analyze the hernial sac contents. Non-contrast-enhanced skull CT with 3D reconstruction is important to assess the skull base defect, and RMI plays a critical role in the analysis of the hernial sac elements[4].

In the present case, preoperative MRI played a key role, providing valuable information such as the location of vital structures within the hernial sac. Only cerebrospinal fluid was observed within the hernial sac, without the presence of cerebral structures, rigorously characterizing a basal meningoceles. The empty sella turcica explains the hypothalamic-hypophyseal failure that may be present in 60% of cases[6].

Additionally, MRI has been largely utilized in the intrauterine diagnosis of central nervous system anomalies for its higher sensitivity as compared with ultrasonography[7,8].

The prognosis, in such cases, is reserved. Only a little more than half of treated cases present a favorable evolution. In a study including 114 patients, 59% have presented normal development, 18% have presented some degree of physical or mental deficiency, and 23% have a severe deficiency[2].

Additionally to the rarity of the present case, it is important to highlight the high índex of morbidity, mortality and risk for extremely severe complications in cases where the condition is equivocally diagnosed as a dermoid cyst or nasal polyp and the surgeon inadvertently attempts to perform its resection.

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