Case Report

Supra- and infra-torcular double occipital encephalocele

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\textbf{A B S T R A C T}

An encephalocele is a protrusion of the brain and/or meninges through a defect in the skull that is closed or covered with skin. Occipital encephaloceles are the most frequent type in North America and Western Europe, where about 85% of encephaloceles take this form. To the best of our knowledge, there are only three other reported cases of double occipital encephaloceles in the literature. The current study reports a double and both supra- and infra-torcular occipital encephalocele in a neonate and discusses the importance of preoperative neuroimaging studies to optimize the outcome. The patient was a 1-day-old male child who was identified by prenatal ultrasound to have two occipital encephaloceles. The patient underwent a closure of the occipital encephalocele on the second postnatal day. The infant tolerated the procedure well and was extubated on the first postoperative day. The child continues to do well during follow-up.

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\textbf{E n c e f a l o c e l e o c c i p i t a l d o b l e i n f r a - y s u p r a - t o r c u l a r}

\textbf{R E S U M E N}

Un encefalocele es una protrusión del cerebro o de las meninges a través de un defecto craneal que está cerrado o cubierto con la piel. El encefalocele occipital es el tipo más frecuente en EE. UU. y Europa occidental, donde alrededor del 85% de los casos de encefalocele adoptan esta forma. Según nuestro conocimiento, solo existen publicados otros 3 casos de encefalocele occipital doble en la literatura. El presente estudio describe un encefalocele occipital doble en un neonato que además se presentaba tanto supra- como infratorcular. También se analiza la importancia de los estudios de neuroimagen preoperatorios para
Introduction

An encephalocele occurs as a post-neurulation phenomenon\(^1\) and it is a protrusion of the brain and/or meninges through a defect in the skull that is closed or covered with skin. The pathogenesis is believed to be the combination of diminished skull base growth and normal growth of posterior fossa structures.\(^2\) Abnormal venous drainage patterns then develop secondarily from the pre-existing encephalocele.\(^3\) It is a relatively rare neural tube defect. The presence of multiple (two or more) neural tube defects along the neural axis is extremely rare and mostly encountered in separate locations.\(^4-6\) The current study presents a neonate, who had two encephaloceles that arose from two different bone defects. The superior encephalocele was supra-torcular and the other was infra-torcular. To the best of our knowledge, this is only the fourth case of double occipital encephalocele published in the English literature.\(^5,7,8\)

Case report

History

The patient was a 1-day-old male child who was identified by prenatal ultrasound to have two occipital encephaloceles. The mother underwent a Cesarean delivery at 36 1/7 weeks of gestation. The patient had Apgar scores of 6 and 8 at 1 and 5 min, respectively. He weighed 2980 g and had a head circumference of 31.2 cm. Both encephaloceles were in the occipital region, and the superior and inferior sacs measured 9 cm × 8 cm and 10 cm × 9 cm, respectively (Fig. 1).

Imaging

A CT scan was performed and two different defects in the occipital bone were identified. Superior and inferior defects had diameters of 15 mm × 17 mm and 21 mm × 22 mm, respectively (Fig. 2). Hydrocephalus was not present. The MR imaging revealed that the superior defect contained portions of the occipital lobe and the inferior defect contained cerebellar tissues (Fig. 3). In addition, MR venography was performed preoperatively (Fig. 4). There were no dural sinuses displaced through the sacs.

Operation

The patient underwent a closure of the occipital encephalocele on the second postnatal day. The patient was placed in the prone position. Two horizontal incisions were made on each of the encephaloceles. Subcutaneous dissection was completed and the dural sacs were revealed. Dural incisions were made and the superior and inferior dural sacs were cut. Cerebrospinal fluid (CSF) was drained and then apertures were enlarged. The gliotic, dysplastic, neural tissues, which...
Fig. 3 – In the sagittal MRI, the superior defect contained portions of the occipital lobe and inferior defect of the cerebellar tissue.

are extensions of the occipital lobe through the superior sac and of the cerebellum through the inferior sac, were resected flush to the bone and care was taken to preserve large draining veins that were noted at the entrance to the skull (Fig. 5).

Postoperative course

The infant tolerated the procedure well and was extubated on postoperative day one. He stayed in the newborn intensive care unit for two days, after which he was transferred to neurosurgery service. No hydrocephalus was observed in the postoperative MR imaging on day 7 and the head circumference was 32 cm on day 12.

The child continues to do well during follow-up. He was seen every six months until he was 3 years old. He was showing signs of developmental delay, but was walking without assistance and had equal use of all extremities. His head circumference was 48.2 cm (between the 10th and 15th percentiles). The patient was lost to follow-up after three years.

Discussion

Encephaloceles are less common than other neural tube defects. Their prevalence has been estimated at 0.8–5 per 10,000 live births.6 Occipital encephaloceles are the most
frequent type in North America and Western Europe, where about 85% of encephaloceles take this form. Their incidence ranges between 1 in 3000 and 1 in 10,000 live births. On the other hand, in Southeast Asia and parts of Russia and Central Africa, anterior encephaloceles are more frequent 1 in 3500 to 1 in 5000 compared to the occipital type. Males and females are equally affected. 9

Neural tube defects arise due to failures in neural folds opposition and fusion during primary neurulation process. Classical theory explains that the neural tube closure starts from the mid-cervical region, and continuously reaches to the rostral and caudal ends in a bidirectional, zipper-like fashion. 5,6,8,10,11 However, this theory is unable to explain the presence of double neural tube defects or defects in the cervical region. 8 Van Allen et al. and Nakatsu et al. proposed multiple sites of closure of the neural tube. 12,13 Recently Sivanes et al. and O’Rahilly et al. have also suggested multiple closure sites. 14,15 These theories may explain the presence of a double neural tube defect in separate locations; however, the occurrence of such lesions in the same region remains unexplained. Goyal et al. suggested a Y-shaped closure of the zipper at the rhombencephalic-mesencephalic junction (site B of the model proposed by Nakatsu et al.). 7 Their proposal based may explain the presence of the double encephalocele in the current case.

The classification of encephaloceles is based on the cause and anatomical location of the skull defect. Primary encephaloceles are congenital and present at birth. Primary encephaloceles are divided into three major types including sincipital (fronto-ethmoidal), basal (trans-sphenoidal, sphenoid-ethmoidal, and sphenoid-orbital), and occipital (supratorcular and infra-torcular). Secondary encephaloceles are acquired and commonly due to trauma or a postsurgical defect. 16 Occipital encephaloceles are sub-classified as supratorcular or infra-torcular. 17

Neuroimaging should be performed to evaluate the intracranial components of the malformation and identify any associated brain vascular anomalies. Computerized tomography (CT) scans are effective to detect the extent of neural herniation. Neuroimaging will also detect hydrocephalus, if present. It is important to preoperatively assess the venous anatomy because the sacrifice of venous structures may lead to cortical infarction or to the development of hydrocephalus. 16,19 In the current patient, MR venography showed no dural sinuses within the encephaloceles. Although larger venous structures were seen in the superior sac, no large draining veins were observed in the inferior encephalocele.

During the repair of encephalocele, there are two important decisions. The first is the decision to sacrifice the venous structures within the encephalocele. Sacrificing large veins may potentially increase the risk for hydrocephalus and venous infarct. The researchers of the current study took care to preserve large draining veins at the entrance to the skull. Intraoperative observance may not always prove to be correct. Venous imaging prior to surgical closure is useful to determine the variations of venous anatomy and individualize the surgical strategy based upon venous anatomy and anomalies to optimize the outcome.

The other important point is to make a judgment to excise the brain partially or to attempt to return all the contents into the intracranial cavity. Sometimes the head is small and the volume of the herniated brain is large so that it is not possible to close the sac without excising brain tissue. The researchers decided to excise the dysplastic neural tissues within both sacs after combining intraoperative observations with preoperative MR imaging. In the patient presented herein, the sacrifice of the dysplastic neural tissue did not result in any adverse events.

Conclusion

Encephaloceles are not merely skull defects with herniating neural tissue. Abnormalities of brain development such as variations in venous anatomy accompany encephaloceles. The management of large encephaloceles constitutes a challenge that involves unique solutions for each case, such as to excise herniating neural tissue or not. Combining intraoperative observance with preoperative MRI and MR venography will help surgeons in determining the appropriate course. The patient presented herein is interesting because he had two occipital encephaloceles that arose from two different bone defects—one with an infra-torcular location and one with a supra-torcular location. Preoperative MRI and MR venography provided the optimal opportunity for determining the surgical strategy.

Disclosure

We confirm that this work has not been published or considered to be published, in whole or in part, in any other journal and permit the reproduction of copyrighted material.

REFERENCES