# *A CASE OF OCCIPITAL MENINGOENCEPHALOCYSTOCELE IN A FULLTERM NEONATE*

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## Abstract

**Background:** The neural tube defect is one of the congenital anomalies which is frequently found in developing countries. A large occipital meningoencephalocystocele poses difficulties in surgical intervention. This is because the sac usually contains large amounts of cerebrospinal fluid, meninges, brain tissue, which leads to a higher rate of surgical complications, as well as a difficult positioning at intubation to minimalize the complications.
**Case Presentation:** To report a case of neonate with a large occipital meningoencephalocystocele, which was managed with surgery by removing the extra cranial tissue containing the necrotic brain tissue and the meninges. The patient was discharged with normal wound healing, but ventriculoperitoneal shunt was not performed. Feeding difficulties and poor growth were observed several months after surgery with obvious neurological problems.
**Conclusions:** A through management of meningoencephalocystocele involves many aspects include treatment after surgery was performed.

**Keywords:** occipital encephalocele, meningoencephalocystocele, excision, neonate

## Background

Meningoencephalocystocele is a type of encephalocele, a rare congenital anomaly due to neural tube defect. This anomaly is marked by the protrusion of neural element, such as brain tissue, along with meninges, and cerebrospinal fluid due to the involvement of ventricular system, which extends through a defect in the skull.[1] This congenital anomaly occurs in 0.8–5.6 per 10,000 live births.[2] This defect occurs due to the failure of ectoderm layer to separate from the neuroectoderm during organogenesis. Although the exact cause of the condition is still unknown for certain, environmental factors are also thought to have a role.[3,4] This defect can be detected through ultrasonography during antenatal care, and a complete evaluation of other abnormalities, whether it is an intracranial or extra cranial anomaly needs to be performed after the diagnosis is established.

The treatment depends on the size, location and severity of the defect. Imaging studies are needed to know the severity of the defect, followed by surgery to repair it. The prognosis for patient with meningoencephalocystocele depends on the size and location of the sac, the presence of brain tissue inside the sac, and

other accompanying brain abnormalities. Other influencing factors include hydrocephaly, infections, and other comorbid pathologies. Occipital encephalocele has an extremely high morbidity and mortality in spite of well given treatment before and after surgery.[5]

**Case Presentation**

A one-day female neonate admitted to the neonatal emergency unit with a swelling at the back of the head noted at birth. The baby was born in a primary health center, through normal vaginal delivery, with a little difficulty during labor due to breech presentation. The mother did not have regular antenatal care. The baby was a first-born child; history of congenital anomaly was denied. She was born full term with a birth weight 2,6 kg. She cried immediately, there was no bladder and bowel dysfunction. There were no convulsions or signs of meningeal irritation, and she was breast-feeding well. Upon local examination, the swelling was spherical, sized 11x6x6 cm, with mild erythema at the center, with a head circumference of 28 cm. The consistency of the swelling was soft with a negative trans illumination test (Figure 1). The vital signs were within normal limits, there was no sign of respiratory distress, and there was normal bladder and bowel function.

Figure 1. The occipital meningoencephalocele in a full-term neonate

The neurological examination revealed that the patient was conscious and had a normal suckling reflex. There were no limbs weakness and good muscular tone. The primitive reflexes were normal and symmetrical. The light reflex was normal.

**The Diagnosis and Important Findings**

The results of laboratory evaluations were within normal range. A head CT-scan showed a defect on the right occipitoparietal bone accompanied by a protruded sac with mixed density (8,65 – 33,75 HU), the brain tissue and the ventricular system were visually distorted, and it was concluded to be a meningoencephalocystocele (Figure 2).

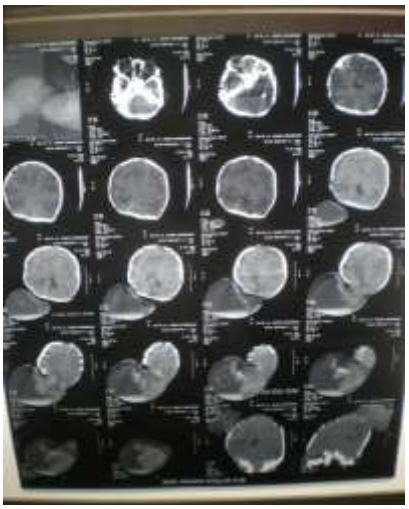

Figure 2. Head CT-scan

**Work Up**

The patient was laid in prone position on the operation table, supported on a soft doughnut roll (Figure 3). The surgery aimed to make an excision and to repair the sac. The excision was done by removing the ekstracranial tissues (Figure 4), and then closing the defect with durafascial flap. Careful observation and monitoring the baby's vital sign was performed throughout surgery.

After the surgery, there were no evidence of infections and cerebrospinal fluid leakage. The wound healed perfectly, and there were no signs of raised intracranial pressure during hospitalization. She began breast feeding in a week after surgery. A little difficulty was noted at the time, but she was getting better with time.

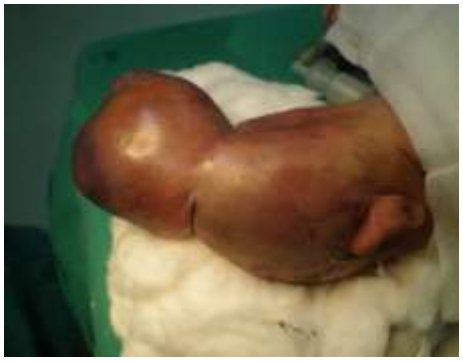

Figure 3. The preoperative view of the patients under general anesthesia

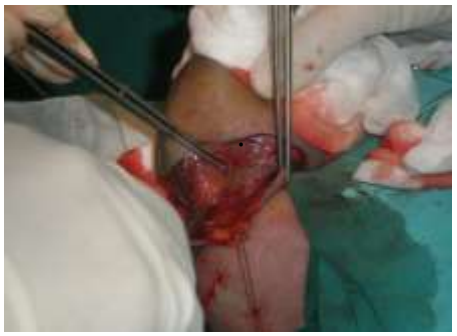

Figure 4. The excision of extra cranial tissue, and the sack containing non-functional brain tissue after removal

**Follow Up and Outcome**

Two weeks after surgery, the patient was discharged. The head circumference increased by 2 cm, but it was still considered as microcephaly. The prognosis of this anomaly and the possibility of ventriculoperitoneal shunt

have been informed to the mother, but the family refused the procedure.

5 months after surgery, the patients came back experiencing feeding difficulties in sucking and lip closure. A physical examination revealed that the patient was undernourished, with delayed development in four domains of child development, and there were signs of hydrocephalus. However, the family refused hospitalization.

**Discussion**

The neural tube defect (NTD) is the leading cause of central nervous system congenital anomaly due to the failure of its closure during the fetal period. The exact cause of NTD is unknown, although some factors such as hyperthermia, drugs, malnutrition, chemical substances, obesity, maternal diabetes, radiation, folic acid deficiency, teratogenic agents, trypan blue, arsenic, and genetic determinants are known to influence the development of the central nervous system, which can cause encephalocele.[6,7] In this case, there was a possibility of nutritional deficiencies, but no objective measurement was performed at the time.

Other malformation or chromosomal anomalies can be found in 60% of patients with encephalocele. These patients have a higher risk of having hydrocephalus due to aqueduct sylvii stenosis, Chiari malformation, or Dandy-Walker syndrome.[4,7]

The mesodermal defect which cause a defect on calvaria, duramater, and the separation of ectodermal surface with neuroectoderm are the main components of meningoencephalocystocele. This causes herniation of CNS, brain tissue and meninx. It can happen in the occipital (75%), frontoethmoidal (13-15%), parietal (10-12%) or sphenoidal region.[3,4,8]

The presence of encephalocele lowers the survival rate of newborns to 21%, and only half of them will survive. Around 75% of the survivors will suffer from mental retardation, and around 3% have recurrence after surgery.[3,5]

In this case, the diagnosis of meningoencephalocystocele was based on medical history and physical examinations and was confirmed by a head CT-scan. Generally, the neonate was active and there were no signs of neurological defect. She had positive primitive reflexes and a good sucking reflex, although it was slightly hampered by the mass. Meningoencephalocystocele is usually accompanied by other craniofacial abnormalities or brain malformations. The symptoms include neurological defects, hydrocephalus, spastic paralysis, microcephaly, ataxia, developmental disorders, visual problems, mental and growth retardation, and seizure. As the

presence of associated intracranial pathologies, including brain malformations, is predictive for the cognitive outcome.[9] In this neonate, we found microcephaly due to the protrusion of brain tissue into the encephalocele pouch. During hospitalization, the baby did not experience seizures or have any other noticeable neurological abnormalities.

Meningoencephalocystocele was managed through corrective surgery. The treatment of choice is excision of the sac through surgery, followed by dural and cranial defect reparation. The outcome of this surgery depends on the location and the size of defect. The VP-shunt placement is meant to drain excess cerebrospinal fluid. The final result depends on the amount of normal brain tissue left inside the skull after surgery. The surgery mostly to facilitates the baby's nursing. Furthermore, less brain tissue in the sac made the excision of the sac easier and safer.[2,10]

In our patient, the head circumference increased by 2 cm after surgery, but was still in the category of microcephaly. This can be caused by the accumulation of CSF due to aqueduct sylvii stenosis, which can be corrected through the implementation of a VP-shunt. Unfortunately, in this case, the VP-shunt was not performed.

The prognosis of encephalocele depends on the location, size, and the content of the sac. The absence of brain tissue in the sac is a good prognostic factor. Poor prognosis includes giant posterior encephalocele, the presence of hydrocephalus and other systemic abnormalities. Patients with anterior encephalocele have a higher rate of survival rate, compared to those with a posterior encephalocele.[1,3,10]

The most common complications of the operation are infection, meningitis, brain damage, and hydrocephalus. Recurrence can happen after surgery. Most neonates with occipital encephalocele will suffer from hydrocephalus. Visual dysfunction, motoric abnormalities and intelligence problems are common in these cases. The mortality rate of this case approximately 30% despite the application of appropriate treatments. In this case, the prognosis is poor due to herniation of brain tissue, which will most likely cause some serious neurological defects. The involvement of the ventricular system in this case can lead to hydrocephalus, which will worsen the prognosis.[1,2,9,11]

**Summary**

A case of a female neonate with meningoencephalocystocele was reported. The diagnosis was based on the result of a head CT-scan. The surgical intervention performed consisted of

meningoencephalectomy (excision) without a VP-shunt was performed. The prognosis of this patient in terms of quo ad vitam was dubia, quo ad sanationem and ad functionem were poor.

**Informed Consent**

Authors declare no conflict of interest. The family was agreed for the publication of this case and signed a written informed consent.

**References**


1. Pahuja, HD, Deshmukh, SR, Lande, SA, Palsodkar, SR, Bhure, AR. Naesthetic Management of Neonate with Giant Occipital Meningoencephalocele: Case Report. Egyptian Journal of Anaesthesia. 2015. Available at www.sciencedirect.com.
2. Ganapathy, A., Sadeesh, T., Swer, MH., Rao, S. Occipital meningo-encephalocele with Cleft Lip, Cleft Palate and Limb Abnormalities- A Case Report. Journal of Clinical and Diagnostic Research. 2014; Vol. 8(12). Available at www.jcdr.net.
3. Raja, RA., Qureshi, AA., Memon, AR., Ali, H., Dev, V. Pattern of Encephaloceles: A Case Series. Journal of Ayub Med Coll Abbooabad. 2008: 20(1). Available at www.ayubmed.edu.pk/JAMC/PAST/20-1/Raja.pdf.
4. Khan AN., Turnbull, I., Macdonald, S., Sabih, D. Encephalocele Imaging. 2019. Available from : https://emedicine.medscape.com/article/403308-overview#a1.
5. Kiymaz, N., Yilmaz, N., Demir, I., Keskin, S. Prognostic factors in patients with occipital Encephalocele. Pediatric Neurosurgery. 2010; 46: 6–11.
6. Jeong, DH., Kim, CH., Kim, MO., Chung H., Kim, TH., Jung, HY. Arnold-Chiari Malformation Type III with Meningoencephalocele: A Case Report. Annals of Rehabilitation Medicine. 2014. Available at http://dx.doi.org/10.5535/arm.2014.38.3.401.
7. Kinsman SL & Johnston MV. *Congenital Anomalies of the Central Nervous System* in *Nelson Textbook of Pediatric*, Kliegman RM e al. 18th Edition. Saunders Elsevier. Philadelphia. 2007. Hal: 2447.
8. Jain, K., Sethi, SK., Jain, N., Patodi, V. Anaesthetic management of a huge occipital meningoencephalocele in a 14 days old neonate. Ain-Shams Journal of Anesthesiology. 2018. Available at https://doi.org/10.1186/s42077-018-0005-7.
9. Kasprian, GJ., Paldino, MJ., Mehollin-Ray, AR., Shetty, A., Williams, JL.,

Lee, W., Cassady, CI. Prenatal Imaging of Occipital Encephaloceles. Fetal Diagnosis and Therapy. 2015; 37:241-248.

10. Ahlawat, G., Chhabra, S., Ahlawat, MS. 2014. Occipital Meningoencephalocele - Anaesthetic Challenges: A Case Report. World Journal of Pharmaceutical Research. Volume 3, Issue 3. Available at www.wjpr.net.
11. Alwahab, A., Kharsa, A., Nugud, A., Nugud S. Occipital Meningoencephalocele case report and review of current literature. Chinese Neurosurgical Journal. 2017. Available at www.biomedcentral.com.