

A rare case surgery repair for pediatric sphenoethmoidal encephalocele with single-step transfrontobasal approach



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ABSTRACT

Background: Basal encephalocele (BEC) is the rarest type of all encephaloceles. Several methods were used to close and repair the bony defect and prevent CSF leakage. Here we present a case of a 1-month-old pediatric patient who presented with a sphenoethmoidal basal encephalocele with a single transfrontobasal approach.

Case Presentation: A female infant one-month-old with diagnosed large sphenoethmoid encephalocele (basal encephalocele). From the 3D CT scan reconstruction, we found a defect in the planum sellae and ethmoid region with a maximum line of 2.84 cm x 2.89 cm. We performed a transfrontobasal approach by using titanium mesh/plate. After follow-up, we did not find CSF leakage.

Conclusion: Uncertainty persists on the best way to restore BEC. In this instance, we used single TBF approaches to seal and repair the basal encephalocele. There are no signs of infection or CSF leakage throughout the follow-up.

Keywords: Basal Encephalocele, Transfrontobasal Approach, Sphenoethmoid encephalocele.

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INTRODUCTION

Basal encephalocele (BEC) is reported to be the rarest type of all encephaloceles.^{1,2} With its incidence at 1 in every 35,000 – 40,000 births, it has been previously reported to be a challenging case in which half of all operated cases were reported to have failed, with most ending in death.¹⁻³ This is due to certain factors, namely their deep location and rarity. Moreover, patients with basal encephaloceles are more common and have been diagnosed with several other congenital abnormalities, including midfacial malformation and other minor anomalies. These anomalies include hypertelorism, anterior cranium bifidum, nasal cleft, cleft lip, cleft palate, and bifid uvula.^{4,5} Aside from these, central nervous involvement is not an impossibility and symptoms such as seizures, developmental delay, vision impairment, ataxia, microcephaly, spastic paraplegia, and signs of raised intracranial pressure.⁶

Although newer techniques and developments in medicine have paved the way to treat basal encephaloceles

with higher success and lower chance of failure and mortality, it is important to note that this does not negate the fact that this case still poses the risk of treatment failure for all neurosurgeons.^{3,7} This necessitates pediatric neurosurgeons to understand basal encephaloceles – the features, pathophysiology, diagnostics, and therapeutic options.

Based on those mentioned above, this case study aims to evaluate a 1-month-old pediatric patient who presented with a sphenoethmoidal basal encephalocele and multiple comorbidities.

CASE PRESENTATION

A one-month-old female infant with a chief of complained fluid discharge from the nasal cavity was clear like water. The patient also has febrile following the main complaint. The patient was the first child to be delivered by cesarean due to induction failure, with a weight of 4000 grams and a body length of 51 cm. The patient's mother had CMV IgG (+) and Rubella IgG (+). On examination, FOC head circumference was 38 cm (within normal limits) with

open and flat at major and minor. From the 3D CT scan reconstruction, we found a defect in the planum sellae and ethmoid region with a maximum line of 2.84 cm x 2.89 cm (Figure 1). The patient was diagnosed with a large sphenoethmoid encephalocele (basal encephalocele/BEC). We closed the defect with titanium mesh/plate with one screw fixation (Figure 2). We performed the closure of the dura mater with a periosteal graft and filled it with fibrin glue. The BEC was repaired with a transfrontobasal approach using titanium mesh/plate. Postoperative follow-up in 2 weeks after surgery, there is no fever and sign of CSF leakage.

DISCUSSION

Several mechanisms have been proposed to explain the occurrence of basal encephalocele. The most widely accepted theory suggests the occurrence of neuroschisis after neural tube closure. Thus, basal encephalocele is often complicated by midfacial malformation and minor anomalies, such as hypertelorism, anterior cranium bifidum, nasal cleft, cleft lip, cleft

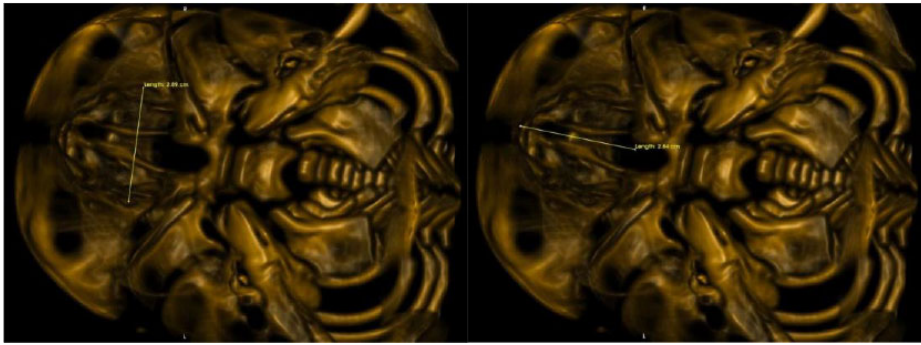


Figure 1. Defect in the planum sellae and ethmoid region with a maximum line of 2.84 cm x 2.89 cm.

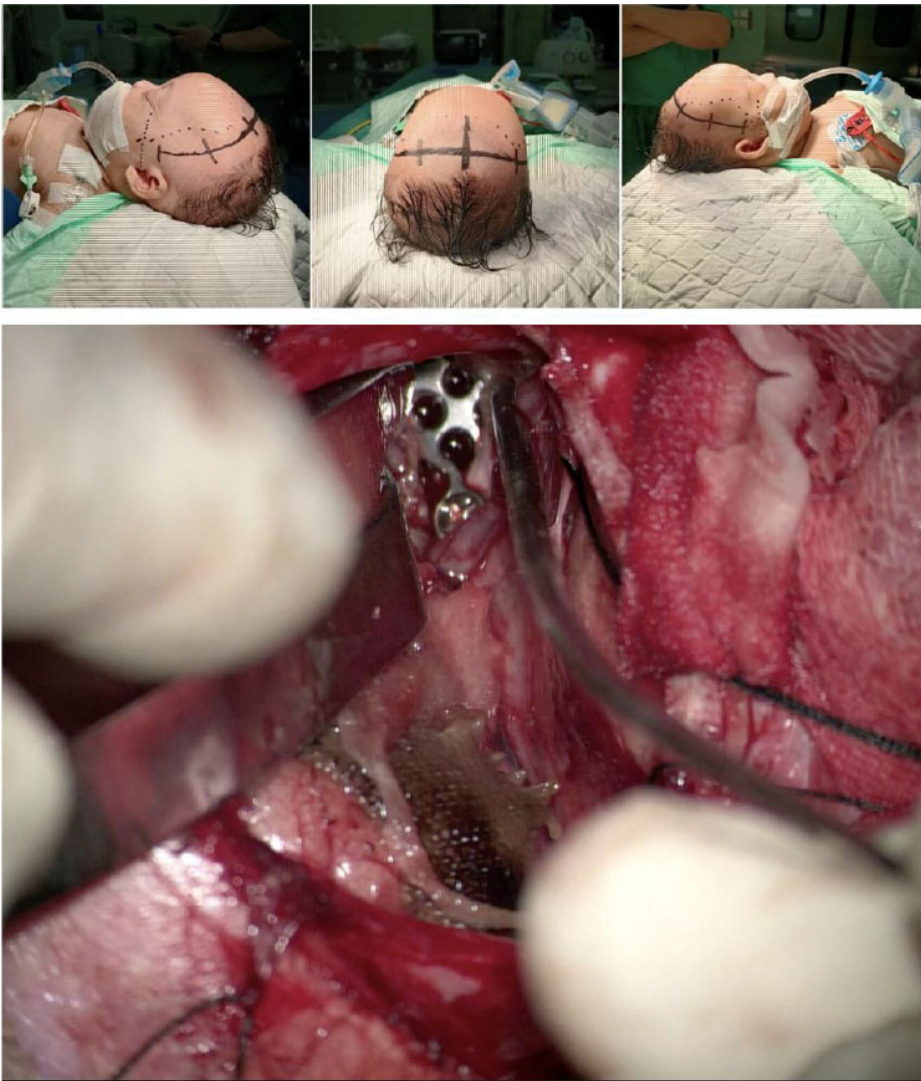


Figure 2. Coronal skin incision and elevation of the pedicled periosteum flap (Upper) and defect closure with titanium mesh/plate with one screw fixation (lower).

palate, and bifid uvula, and most cases are diagnosed at birth.⁸

BEC were classified into 3 subtypes—sphenoidal, sphenothmoidal, or ethmoidal—based on the location of the midline bone defect.^{3,9,10} The size of the

BEC was classified as small, medium, or large by the size of the skull base bone defect as measured by a midsagittal reconstructed 3D CT image. Bone defects less than 1 cm, between 1 and 2 cm, and more than 2 cm were defined as small,

medium, and large, respectively.³ In this patient, we found a skull-based defect in planum sellae and ethmoid region with a maximum line of 2.84 cm x 2.89 cm, so we classified this patient as a large sphenothmoidal BEC.

Surgery as the optimal method to repair BEC is still unclear. The selection of surgical approaches for BEC treatment varies. The transnasal approach is indicated when the bone defect is small and there is no cleft palate, the transnasal approach is indicated.³ The transoral trans palatal (TOTP) approach is optimal if a cleft palate is present. Suppose the proximal edge of the BEC is difficult to expose via the TOTP approach. In that case, the TFB approach may be added in a 1- or 2-stage operation following the operation via the TOTP approach. Based on our center protocol and literature review for large sphenothmoidal BEC usually used combined approach (TOTP and transfrontobasal approach), the TOTP approach recommends itself as the standard surgical procedure for the treatment of BEC with a cleft palate, regardless of the size of the BEC.³ Endoscopic observation facilitates circumferential dissection at the proximal edge of the BEC. Transnasal endoscopic repair is indicated for small to medium BECs without a cleft palate. The TFB approach should be reserved for only certain cases. It should be combined with the TOTP approach, which best fits large BECs with intracranial or orbitofacial anomalies or huge BECs where the proximal border involves the frontonasal junction.³

In this case, we only made the transfrontobasal approach to close the defect because we thought that by using only the transfrontobasal approach, we could see the bone defect and close the defect using a titanium mesh with one screw.

A modified surgical procedure is required for ethmoidal BECs with brain parenchymal herniation. Vital structures may be involved in the BEC, and total repositioning or resection can lead to fatal complications. Palliative partial repair to restore the airway should be considered in patients according to the previous studies.^{3,11-13}

Transfrontobasal approach (TFB)

repair is required when the BEC is large and exposure of the proximal edge via the TOTP approach is difficult. The procedure can be carried out separately in 2 stages after TOTP repair or in 1 stage during the same operation following TOTP repair. Because the distal edge of the BEC is not visible during TFB repair, prior dissection of the distal edge via the TOTP approach is necessary.³

A small right frontal craniotomy is performed following a coronal skin incision and elevation of the pedicled periosteum flap. The medial one-third of the orbital rim is cut to reach the frontal base directly. The dura covering the frontal base is gently retracted and dissected. We performed the dural incision in the medial frontobasal. We identified the chiasma and olfactory nerve and performed gentle retraction carefully. Once we found and identified the defect, we carefully removed the necrotic part of the brain that was trapped in the defect. After identifying the defect, we found that the defect is 1,5cm x 1,5 cm. We closed the defect with titanium mesh/plate with one screw fixation. In addition, we also performed the closure of the dura mater with a periosteal graft and filled it with fibrin glue according to the previous study.³

There are three methods to repair basal encephalocele, TOTP, TBF and combined 2-step TOTP and TBF approaches. The optimal method to improve BEC is still unclear until now. We closed and repaired the basal encephalocele, in this case, using single TBF approaches. During the follow-up, there was no CSF leakage or sign of infection.

This reference is limited to only one successful case without complications during the follow-up period. Studies on BEC are still quite scarce due to its rarity. The best treatment requires special attention from a neurosurgeon.

CONCLUSION

There are three methods to repair basal encephalocele, TOTP, TBF and combined 2-step TOTP and TBF approaches. We closed and improved the basal encephalocele, in this case, using single TBF approaches. During the follow-up, there was no CSF leakage or sign of infection. The optimal method to repair BEC is still unclear until it is necessary to provide more references to explain the best treatment for BEC.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

ETHICS CONSIDERATION

Not applicable as no patient identity was disclosed.

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AUTHOR CONTRIBUTIONS

Dicky Lukman Rangkuti contributed to the conceptualization, data collection, writing, and editing of the study. Muhammad Arifin Parenrengi and Wihasto Suryaningtyas contributed to reviewing, editing, and finalizing the manuscript of the study.

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