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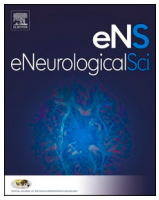

Case report

Double encephalocele with an excellent outcome postoperatively: A case report from Iraq

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ABSTRACT

Background: An encephalocele is a congenital neural tube defect that is estimated to have an incidence of 1–2 cases per 10,000 live births. There have been a few cases of double encephaloceles reported in the medical literature. We report an extremely rare case of double encephalocele with an atrial septal defect in Iraq.
Case presentation: A 2-month-old female infant presented with two swellings at the back of her head since birth. Her mother received poor prenatal care. The examination revealed a microcephaly head and two sacs in the occipital region, which were not connected and were covered completely by skin. The surgery includes a transverse incision, excision of both sacs with necrotic tissue, a duroplasty, and a water-tight dural closure. The operation proceeded without any neurological sequelae or cerebrospinal fluid leakage.
Conclusion: Double encephalocele is a congenital neural tube defect that is rarely discussed or reported in the medical literature. The management of this condition might be difficult because it requires a special approach for each patient. This case report from Iraq is used to raise awareness about this particular disorder and to motivate clinicians about the importance of early and appropriate management for such cases.

1. Introduction

"An encephalocele is a congenital neural tube defect characterized by herniation of cranial contents through a defect in the cranium and is caused by failure of the closure of the cranial part of the developing neural tube" [1]. Based on the site of the defect, it is divided into a variety of types, such as occipital, sub-occipital, parietal and basal [2]. If the condition is present at birth, it is considered a primary encephalocele; if it is acquired later in life due to traumatic or surgical causes, it is considered a secondary encephalocele [3]. It has been estimated that there are 1–2 cases of encephalocele for every 10,000 live births, with the occipital region being affected in the majority of cases [4,5]. There have been a few cases of double encephaloceles reported in the medical literature; the majority of them involved the occipital or sub-occipital region [2]. We report an extremely rare of double encephalocele with an atrial septal defect. To the best of our knowledge, this is the first reported case of double encephalocele in Iraq.

2. Case presentation

A 2-month-old female infant with two swellings at the back of her head since birth was delivered via C-section at the Baghdad teaching hospital (tertiary center) as a full-term newborn due to a failure to progress at 39 weeks of gestation. Her mother received poor prenatal care as the condition was not diagnosed prenatally and she hadn't taken folic acid supplements. The neonatologist referred the infant for neurosurgical consultation. The examination revealed a microcephaly head, as the occipitofrontal circumference was 28 cm, and two sacs in the occipital region, the left one measuring 5 × 5 cm and the right one measuring 4.5 × 5 cm, which were not connected and were covered completely by skin (as shown in Fig. 1A and B). The swellings were soft to firm in consistency, non-fluctuant, and non-transilluminant, and the anterior fontanelle was open. Computed tomography of the brain confirmed the diagnosis of double encephalocele which showed a 30 mm defect in the occipital bone with two herniated brain tissue each covered in a sac (as shown in Fig. 2A and B). The neurosurgeon decided to perform surgery; however, the infant's general well-being was unfit for general anesthesia due to the presence of an atrial septal defect and

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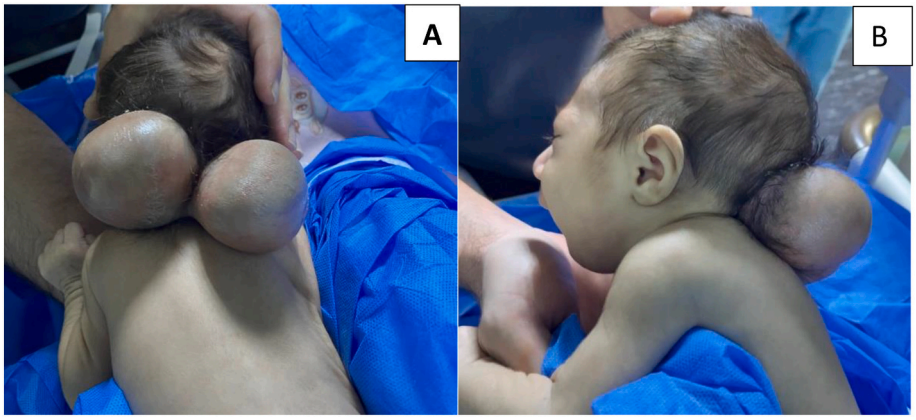

Fig. 1. Preoperatively (A) posterior view of the left and right sacs in the occipital region of the skull (B) lateral view of the left sac in the occipital region of the skull.

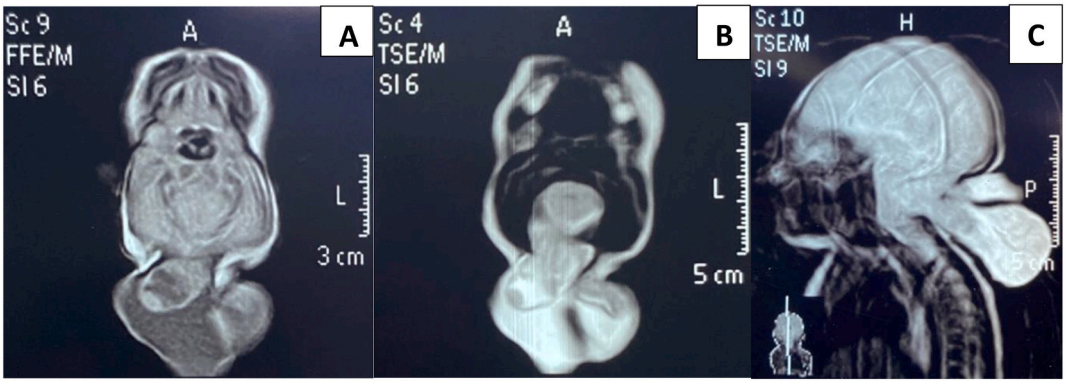

Fig. 2. Computed tomography scan of the brain showing (A, B) on axial view a 30 mm defect in the occipital bone with two herniated brain tissue sacs (C) on sagittal view a defect in the occipital bone with two herniated brain tissue sacs.

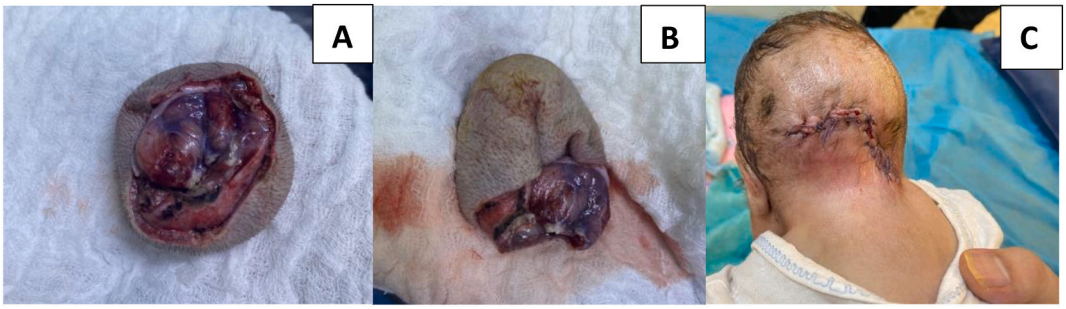

Fig. 3. Postoperatively (A, B) The excised sacs containing neuronal tissue (C) The occipital region of the head showing lazy S-type suturing of the repaired defect.

physiological jaundice, so the surgery was postponed. Two months later, after the infant's condition had stabilized, the surgery was done under general anesthesia in a prone position. The surgery includes a lazy S incision, excision of both sacs with necrotic tissue, a duroplasty, and a water-tight dural closure (as shown in Fig. 3A and B). Using vicryl suture 4/0 (as shown in Fig. 3C), The operation proceeded without any neurological sequelae or cerebrospinal fluid leakage. The procedure was well tolerated by the infant; she did not require admission to a neonatal intensive care unit and was discharged home three days later, afebrile and well-nourished on breast milk. There was no similar family history. And the parents had the first degree of consanguinity.

3. Discussion

A double encephalocele is a rare and serious condition, and for now, only four cases have been reported; ours is the fifth. This is the first time that a case of double encephalocele has been reported in Iraq, and the doctors involved in the case are calling for increased awareness and education about the condition to prevent further cases.

The case report describes a newborn female infant with a double encephalocele involving the occipital bone of the skull. Additionally, the infant had an atrial septal defect, which is a hole in the wall separating the right and left atria of the heart. These defects can cause blood to flow improperly, leading to heart problems and other complications.

In a double encephalocele, there are two separate areas of the brain that have protruded through the skull. This can cause a range of symptoms, including hydrocephalus, neurological deficits, and cognitive impairments. It is often accompanied by other birth defects, such as facial abnormalities and defects in the spine or skull [6].

There is no clear explanation for this rare case, but as it exists with a

cardiac septal defect, it suggests that a teratogenic exposure during the childbirth period could be the cause [7].

Treatment for a double encephalocele typically involves a combination of surgical intervention, medication, and rehabilitation. The first step is to repair the opening in the skull and return the protruding brain tissue to its proper position. This may be done through a combination of open surgery and minimally invasive techniques [8].

Medications may be prescribed to manage symptoms such as hydrocephalus and reduce the risk of infection. Rehabilitation may be necessary to address any neurological deficits and help the individual achieve the best possible quality of life.

The prognosis for a double encephalocele can vary depending on the specific case and the extent of the damage to the brain. In some cases, the condition may be life-threatening or result in severe developmental delays. In others, the individual may be able to lead a relatively normal life with appropriate treatment and support [8].

4. Conclusion

Double encephalocele is a congenital neural tube defect that is rarely discussed or reported in the medical literature. The management of this condition might be difficult because it requires a special approach for each patient. This case report from Iraq is used to raise awareness about this particular disorder and to motivate clinicians about the importance of early and appropriate management for such cases.

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CRediT authorship contribution statement

Ali Tarik Abdulwahid: Conceptualization, Methodology, Software, Supervision. **Ahmed Dheyaa Al-Obaidi:** Data curation, Writing – original draft. **Mustafa Najah Al-Obaidi:** Visualization, Investigation. **Hashim Talib Hashim:** Conceptualization, Methodology, Software.

Declaration of Competing Interest

We declare that we have no conflict of interest.

References

[1] S. Zahid, A. Khizar, Giant occipital encephalocele: a case report, surgical and anesthetic challenge and review of literature, Egy. J. Neurosurg. 36 (2021) 38, https://doi.org/10.1186/s41984-021-00136-8.

[2] C.K. Shah, R.Y. Lee, S. Jeph, In-utero diagnosis of double Encephalocele - imaging features and review of literature, J. Radiol. Case Rep. 15 (12) (2021) 1–9. Published 2021 Dec 1, https://doi.org/10.3941/jrcr.v15i12.4230.

[3] A. Horcajadas, A. Palma, B.M. Khalon, Frontoethmoidal encephalocele. Report of a case, Neurocirugia (Astur: Engl Ed) 30 (2) (2019) 94–99, https://doi.org/10.1016/j.neucir.2018.02.006.

[4] E. Tan, S. Makaranka, N. Mohamed, N. Cavale, Occipital encephalocele in a neonate: a case successfully managed by excision and formation of a reverse visor scalp flap, BMJ Case Rep. 13 (1) (2020) 1–4. Published 2020 Jan 21, https://doi.org/10.1136/bcr-2019-232127.

[5] M. Mikayilli, T. Hasanov, G. Demirci Otluoğlu, D.E. Nacitarhan, Z.O. Toktaş, A. Çolak, Congenital lateral encephalocele-case report, Childs Nerv. Syst. 36 (12) (2020) 3119–3122, https://doi.org/10.1007/s00381-019-04436-1.

[6] Carolina M. Valdez, et al., Encephalocele—radial, cardiac, gastrointestinal, anal/renal anomalies: novel evidence for a new condition? Am. J. Med. Genet. A 164 (5) (2014) 1085–1091.

[7] U. Froster-Iskenius, P. Meinecke, Encephalocele, radial defects, cardiac, gastrointestinal, anal, and renal anomalies: a new multiple congenital anomaly (MCA) syndrome? Clin. Dysmorphol. 1 (1) (1992) 37–41.

[8] Cruz Alejandro J. Matos, Orlando De Jesus, Encephalocele, in: StatPearls [Internet], StatPearls Publishing, 2021.