International Journal of Research in Medical Sciences
Babagana U et al. Int J Res Med Sci. 2022 Mar;10(3):590-594
www.msjonline.org
pISSN2320-6071 | eISSN2320-6012
DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20220506

Original Research Article

Clinical findings and surgical outcomes of encephalocoeles

Usman Babagana[1]*, Abubakar Auwal Mohammed[2]

[1]Neurosurgery Unit, Department of Surgery, The University of Maiduguri and the University of Maiduguri Teaching Hospital, Borno State, Nigeria
[2]Paediatrics Surgery Unit, Department of Surgery, The Federal Medical Centre Yola, Adamawa State, Nigeria

Received: 05 January 2022
Accepted: 31 January 2022

***Correspondence:**
Dr. Usman Babagana,
E-mail: babaganau@yahoo.com

ABSTRACT

Background: Encephalocoeles are herniation of the brain and its meninges via a midline cranial vault defect or at the base of the skull, at the site of local mesenchymal disruption. It accounts for 10% to 20% of all the cranial and spinal dysraphisms found in 1/5000 live births.
Method: It is a 5 year retrospective study on 42 cases of encephalocoele managed in a Nigerian Tertiary Medical Centre. We extracted the clinical findings and surgical management outcomes from the patient's case notes.
Result: Ages at presentation were Neonates (24, 57.1%), ≤3 months (35, 83.3%), with mode of 2 days. Males (12, 28.6%), females (30, 71.4%), with M:F=1:2.5. Posterior (occipital) lesions (36, 85.7%), anterior (6, 14.3%). Variable size lesions (33, 78.6%), giant encephalocoeles (9, 21.4%). Hydrocephalus (16, 38.1%) included pre-excision HCP (12, 75% of 16) and post excision HCP (4, 25% of 16). Available neuroimaging was TFUSS (all, 100%), CT Scan (20, 47.6%), MRI (2, 4.8%). Surgery of excision and repair (36, 85.7%), combined craniofacial repair (6, 14.3%), additional ventriculoperitoneal shunt (16, 38.1%). Post-operative complications included CSF leak (2, 4.8%), superficial surgical site infection (3, 7.1%), meningitis (1, 2.4%), shunt infection (4, 9.5%), shunt obstruction (7, 16.7%). Outcomes were good (38, 90.5%), blind (3, 7.1%), vegetative (1, 2.4%), death (1, 2.4%).
Conclusions: Encephalocoeles are an uncommon congenital type of neural tube defect containing herniated brain tissue. Posterior (occipital) lesions are commoner (and maybe of giant size) than anterior lesions (Sincipital). Surgical outcomes are generally good.

Keywords: Encephalocoele, Occipital, Giant, Outcome

INTRODUCTION

Encephalocoeles are a type of neural tube defects (NTDs) characterised by herniation of the brain and its meninges through a defect usually in the midline of the cranial vault or at the base of the skull.[1] This CNS herniation may occur between 8 and 12 weeks of gestation at the site of local mesenchymal disruption.[2] Encephalocoeles are frequent in developing countries of Asia and Africa.[3-5] They account for 10 to 20 % of all craniospinal dysraphisms, with an incidence of approximately 1/5000 live births.[6,7]

Anatomically, 75% of the encephalocoeles are occipital, 13-15% are fronto-ethmoidal, and 10-12% are in the parietal or the sphenoidal region. Occipital encephalocoeles are common in the western hemisphere, whereas anterior (frontal and ethmoidal) are common in Southeast Asia.[8,9] Occipital encephalocoele is more common in females than males.[7] Occipital encephalocoeles may be enormous, known as a giant meningoencephalocoele (encephalocoele), when the head is smaller than the Meningoencephalocoele.[10] This is accompanied by surgical and anaesthetic challenges, especially the issues of positioning and intubation.[11]

Other anomalies often accompany Encephalocoeles in at least 60% of the patients. Notably, the neurologic problems may include Hydrocephalus, microcephaly, ataxia, developmental delay, visual issues, mental and growth retardation, and seizures.[12] Treatment of frontoethmoidal encephalocoeles should be recommended at an earlier age to avoid distortion of facial anatomy during growth.[13] Though, managing children with significant defects and herniation of a considerable brain matter into the sac can be challenging at times. In such cases, preservation of the herniated brain parenchyma can be accompanied by an expansile cranioplasty.[14] Certain factors determine the prognosis of patients diagnosed with encephalocoeles. These include the size of the sac, the contents of the neural tissue, Hydrocephalus, infections, and other pathologies that accompany the condition.[15]

This study aims at studying the presentation, surgical management, and outcomes of patients operated with encephalocoeles.

METHODS

This retrospective study lasted five (5) years between January 2017 and December 2021. All cases of encephalocoele who had surgical intervention (excision and repair) were included in the study. Excluded were those who had excision and repair in another centre but referred to us for other surgery and those lost to follow up. The study was conducted in Federal medical centre (a tertiary medical facility), located in Northeastern Nigeria, in Yola, Adamawa state. Relevant data such as age at presentation, sex, clinical types based on the location of encephalocoele, the size of the lesion, presence of seizure and hydrocephalus at presentation, available radiologic evaluation, operative method, postoperative complications, and follow up conditions were extracted from the patient's case notes. The data were entered into a pre-formed proforma and analysed using IBM SPSS version 25.0. Simple frequency tables were generated, percentages, mean, and standard deviations were calculated. Associations were determined using the Student t test for continuous variables and Chi square test for categorical variables. A p value of <0.05 was considered statistically significant.

RESULTS

We managed a total number of 42 patients with various encephalocoeles during the study period. Ages at presentation ranged froma few hours to 6 months with a Mode of 2-day olds. Out of the 24 patients that presented within the neonatal period (≤1 month), 15(62.5%) presented within the first three (3) days of delivery. There were 12 males and 30 females, with a male:female ratio of 1:2.5. Nine (21.4%) patients had giant occipital (posterior) encephalocoele, while the others were of variable sizes and sited either anteriorly or posteriorly. Hydrocephalus was found in 16 patients, with 12 observed before the surgery and four (4) after the surgery. In addition to trans fontanelle ultrasound scan (100%), computed tomography scan (CT scan) was available in about half of the patients, while only two (2) patients did magnetic resonance imaging (MRI). Various clinical findings among the 42 patients are shown in (Table 1).

Table 1: Clinical findings in patients with encephalocoele.

Parameters	N	%
Age at presentation (months)		
≤1	24	57.1
2-3	11	26.2
4-5	05	11.9
≥6	02	4.8
Gender		
Males	12	28.6
Females	30	71.4
Clinical types		
Anterior	6	14.3
Posterior	36	85.7
Size of lesions		
Variable size	33	78.6
Giant size	9	21.4
Presence of seizures		
Yes	02	4.8
No	40	95.2
Presence of hydrocephalus		
Pre excision	12	28.6
Post excision	04	9.5
Available investigations		
Trans fontanelle ultrasound scan	42	100
Head computed tomography	20	47.6
Magnetic resonance imaging	02	4.8

Table 2: Postoperative complications and outcomes.

Complications	N	%
Excision and repair		
Cerebrospinal fluid leak	02	4.8
Superficial surgical site infection	03	7.1
Meningitis	01	2.4
Post ventriculoperitoneal shunt		
Shunt infection	04	9.5
Shunt obstruction		
Proximal	06	14.3
Distal	01	2.4
Postoperative outcome		
Good	38	90.5
Clinically blind	2	4.8
Vegetative state	01	2.4
Death	01	2.4

The patients with posterior (occipital) lesions had excision and repair with or without choroid plexus

excision for those whose excised parenchyma included parts of the lateral ventricle. The ones with anterior lesions had combined craniofacial excision. The most common complication was proximal shunt obstruction followed by shunt infection, superficial surgical site infection, CSF leak, meningitis, and distal shunt obstruction, as shown in (Table 2).

Figure 1: A) Occipital giant encephalocoele, B) frontal (sincipital) ulcerated encephalocoele in a new-born, C) brilliantly trans illuminating occipital encephalocoele coexisting with a cervical meningocoele.

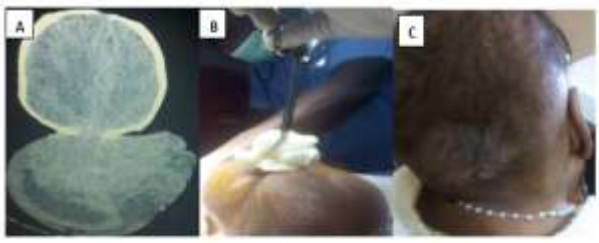

Figure 2: A) CT Scan finding of largely parenchymal mass and some cystic component in a giant occipital encephalocoele, B) intraoprative picture of our technique of holding up(supporting) a giant occipital encephalocoele with the patient in a prone position, C) post excision scare in occipital region.

DISCUSSION

From the 42 patients we managed, the majority (35, 83.3%) presented within three (3) months of delivery, with neonates (≤1 month old) constituting a large number (24, 57.1%). This large number presenting within three (3) months is similar to the findings of Ramdurg, Babagana and Raja, while Amadi found older (up to sixmonths) children.[16-19] Our patients were probably younger because of early referral. Most of them were delivered in a primary health care facility with no expertise in the early care of such patients. This finding contrasts with the findings of Charu that found it in 20.4% of the infants (younger than one month).[20] We found dominance of female (71.4%) patients over males. This dominance agrees with other studies: Rehman, Kabre, Gyan and Shokunbi, whom all found female dominance over males. Contrary to ours, Charu found slight male dominance (53.4%).[20-24]

Posterior (occipital encephalocoele) was the most common lesion in our study, followed by the anterior (Sincipital). This is similar to the Nigerian findings of 92.5%, 82.4%, and 70%, respectively, by Gyan, Babagana, and Shokunbi. Reports from other climes by Raja (80%), Charu (67%) and Warf (48%) also indicate that the common lesions are occipital in locations.[17,18,20,23,25] The reverse is the case in Southeast Asia (Mahapatra and Hoving).[8,9] Frontal (Sincipital) Encephalocoeles were very few. It conforms tothe reports by Softa (17 in 6 years), Raja (3 in 3 years), and Esperence (2 in 10 years).[18,26,27] However, Mahapatra and Hoving reported that frontal lesions were commoner.[8,9] We found 9 (21.4%) cases of giant Encephalocoele, nearly similar to the finding by Gyan (28%) in Sokoto, Nigeria. All were occipital, while Gyan found 1 in the parietal region.[23] In our finding, presentations with seizures were rare (2, 4.8%). We probably had non that presented with CSF leak or high-grade fever, a recognised cause of the seizure. Parmar found fewer seizures (2, 2.35%), but Kabre and Rehman found a higher number presenting with seizures (14%and 15%) respectively, while Bui found more (17%) in patients with occipital encephalocoeles.[21,28,29]

Hydrocephalus was found in 16 patients (38.1%), with the majority diagnosed before the excisions. Preceding the excision was found in 12 patients (28.6% %) of the 42 patients, were found on the patients with occipital lesions. The association of hydrocephalus with the occipital lesion is 60% to 90%, as seen by Gamache, unlike ours (100%).[30,31] We found Hydrocephalus in only 3(18.75%) out of the nine (9) giant occipital encephalocoelesin our series, while Gyan reported only 1 (7.1%) out of 14 in his series on giant encephalocoele in Sokoto, Nigeria.[23] Our finding of pre excision hydrocephalus (12, 28.6%) outnumbered the ones from a report by Parmar (8.57%).[28] We found 4 (9.5%) post excision Hydrocephalus far more than what Parmar got (1, 2.85%), but nearly equal to the finding of Rehman (4, 8%), and just short of half the result by Ehab (21%).[21,28,32] All had trans fontanelle ultrasound scan and additionally computed tomography (CT Scan) in about 47.6% and magnetic resonance imaging (MRI) in only 2 (4.8%). The fewer CT scan and MRIs maybe because it is either not available (CT Scan and MRI are about 15 Km and 250 Km away from our hospital) or expensive as the cost of CT Scan and MRI is about 50% and 100% the cost of operating on these patients.

The patients with preoperative Hydrocephalus had Ventriculoperitoneal Shunt with the excision in the same sitting (we called this "2 in 1"). Those with occipital (including the giant occipital lesions) had standard excision and repair, while those with anterior lesions had craniofacial excision and repair. One (1) patient with giant encephalocoele (containing significant brain tissue) had a modified cranial compartment enlargement by making radial incisions about 4 cm long on the occipital bone to allow the fanning out of the bone in other to accommodate the minimally excised brain parenchyma. None had the need for autologous bone cranioplasty as

advocated by Bozinov.[33] We found seventeen (17) complications among nine (9) patients, with four (4) patients who had two (2) complications each and two (2) who had three (3) complications each. Post excision CSF leakages were observed in two (2) patients who subsequently developed hydrocephalus, proximal shunt obstruction and superficial surgical site infection. The post excision CSF leak (4.76%) was twice the findings of Rehman and Parmar respectively.[21,24] Our superficial Surgical Site infection finding was three times less than that reported by Parmar (22.85%), probably because we had only one (1) preoperative wound infection, a risk factor for postoperative infection.[28] One of our patients developed meningitis (2.4%). Parmar observed five (5) times (11.42%) more meningitis and local abscess cases than ours.[28] Seven (7, 16.7%) patients had shunt obstruction, mainly of the proximal type, in the ones with (3) giant hydrocephalies that had VP Shunt and in 4 others. Our finding regarding shunt infection was (9.52%). Parmer found lesser cases of shunt obstructions and infections (2.35%), respectively.[28] Babagana, in the same Hospital but from different studies and periods found fewer shunt obstruction and infections of 12.5% and 8.3%, respectively.[17] We found good outcomes in most of our patients (90.5%). Two had blindness probably from the excised visual cortex of the giant encephalocoele or Knobloch syndrome.[33-34] One became vegetative after developing a venous brain infarction, While the other one died from anaesthesia related complications.

Limitations

Limitation of current study is that it is a retrospective one with a small number of participants characterised by missing case notes. Improvement is likely with a prospective study involving more participants.

CONCLUSION

Most of our patients presented early, lesions were common among females, and the commonest encephalocoeles are occipital ones. Despite the Scarcity of computed tomography and magnetic resonant imaging, patients had various surgical interventions with good outcomes, except for a few that developed shunt-related complications, one death, one blindness. It confirms that occipital encephalocoeles are commoner than others in our environment and that most can be managed safely with few complications even when neuroimaging is not readily available.

ACKNOWLEDGEMENTS

Authors are thankful to all the doctors in paediatrics department and paediatric surgery unit of Federal medical centre Yola, Adamawa State, Nigeria.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

REFERENCES

1. David DJ. Cephaloceles: classification, pathology, and management a review. J Craniofacial Surg. 1993;4:192-202.
2. McComb JG. Encephaloceles, In: Youmans JR, eds. Neurological surgery: a comprehensive reference guide to the diagnosis and management of neurosurgical problems. 4th ed. Philadelphia: WB Saunders; 1996:829-42.
3. Oucheng N, Lauwers F, Gollogly J, Draper L, Joly B, Roux FE. Frontoethmoidal meningoencephalocele: appraisal of operatedcases. J Neurosurg Pediatr. 2010; 6(6):541-9.
4. Sanoussi S, Chaibou MS, Bawa M, Kelani A, Rabiou MS. Encéphalocèleoccipitale: aspects épidémiologiques, cliniques et thérapeutiques : à propos de 161 casopérésen 9 ans à l'Hôpital national de Niamey. Afr J Neurol Sci. 2009;28(1):26-31.
5. Warf BC, Stagno V, Mugamba J. Encephalocele in Uganda: ethnic distinctions in lesion location, endoscopic management of Hydrocephalus, and survival in 110 consecutive children. J Neurosurg Pediatr. 2011;7(1):88-93.
6. Chapman PH, Swearingen B, Caviness VS. Subtorcular occipital encephaloceles: Anatomical considerations relevant to operative management. J Neurosurg. 1989;71:375-81
7. Singh AK, Sharma MS, Agrawal VK, Behari S. Surgical repair of a giant naso ethmoidal encephalocele. J Pediatr Neurosci. 2006;1:293-4.
8. Mahapatra AK, Agrawal D. Anterior encephaloceles: a series of 103 cases over 32 years. J Clin Neurosci. 2006;13:536-9.
9. Hoving EW. Nasal Encephaloceles. Child's Nerv Syst. 2000;16:702-6
10. Kumar V, Kulwant SB, Saurabh S, Richa SC. Giant occipital meningoencephalocele in a neonate: A therapeutic challenge. J Pediatr Neurosci. 2017;12:46-8.
11. Mahapatra AK. Management of encephalocele. In: Ramamurthi R, eds. text book of operative neurosurgery. New Delhi: BI Publications Pvt. Ltd.; 2005:1;279-90.
12. Stoll C, Alembik Y, Dott B. Associated malformations in cases with neural tube defects. Genet Couns. 2007;18:209-15.
13. Macfarlane R, Rutka JT, Armstrong D, Philips J, Posnick J, Forte V, Humphreys RP, Drake J, Hoffman HJ. Encephaloceles of the anterior cranial fossa. Pediatr Neurosurg. 1995;23:148-58.
14. Mohanty A, Biswas A, Reddy M, Kolluri S. Expansile cranioplasty for massive occipital encephalocele. Childs Nerv Syst. 2006;22:1170-6.
15. Kıymaz N, Yılmaz N, Demir I, Keskin S. Prognostic factors in patients with occipital encephalocele. Pediatr Neurosurg. 2010;46:6-11.

16. Ramdurg SR, Sukanya M, Maitra J. Pediatric encephaloceles: A series of 20 cases over a period of 3 years. J Pediatr Neurosci. 2015;10:317-20.
17. Babagana U. Demography and management outcome of neural tube defects in a Nigerian tertiary health institution. Int J Res Med Sci. 2021;9:3263-8.
18. Raja RA, Qureshi AA, Memon AR, Ali H, Dev V. Pattern of encephaloceles: a case series. J Ayub Med Coll Abbottabad. 2008;20(1):125-8.
19. Amadi CE, Eghwrudjakpor PO. The pattern of distribution of encephalocele in University of Port Harcourt Teaching Hospital-a three year experience. Nigerian Journal of Medicine. J Nat Assoc Resid Doctors Nigeria. 2013;22(1):19-23.
20. Mahajan C, Rath GP, Dash HH, Bithal PK. Perioperative management of children with encephalocele: an institutional experience. J Neurosurg Anesthesiol. 2011;23(4):352-6.
21. Rehman L, Farooq G, Bukhari I. Neurosurgical interventions for occipital encephalocele. Asian J Neurosurg. 2018;13:233-7.
22. Kabré A. The cephaloceles: A clinical, epidemiological and therapeutic study of 50 cases. Neuro-Chirurgie. 2015; 61(4):250-4 .
23. Bot GM. Giant Encephalocele in Sokoto, Nigeria: A 5-Year Review of Operated Cases. World Neurosurg. 2020;139:51-56.
24. Shokunbi T, Adeloye A, Olumide A. Occipital encephalocoeles in 57 Nigerian children: a retrospective analysis. Childs Nerv Syst. 1990;6:99-102.
25. Warf BC, Stagno V, Mugamba J. Encephalocele in Uganda: ethnic distinctions in lesion location, endoscopic management of Hydrocephalus, and survival in 110 consecutive children. Neurosurg Pediatr. 2011;7:88-93.
26. Softa H, Dastoli P, Roger G, Couloignier V, Di Rocco F, Zérah M: Chirurgie par voieendonasale des encéphalocèlesnasales de l'enfant. Neurochirurgie. 2007;53:433.
27. Espérance BMY, N'dri Oka D, Kakou M, Bourgi H, Ba Zeze V. Les méningoencéphalocèlesen milieu ivoirien. Neurochirurgie. 2011;57:45-9.
28. Parmar HV, Adhvaryu NS, Shah JK, Trivedi B, Parmar M. A study on clinical analysis and surgical outcomes of occipital encephalocele in children. Int Surg J. 2021;8:2946-50.
29. Kabré A, Zabsonre DS, Sanou A, Bako Y. The cephaloceles: A clinical, epidemiological and therapeutic study of 50 cases. Neurochirurgie. 2015; 61(4):250-4.
30. Bui CJ, Tubbs RS, Shannon CN, Acakpo-Satchivi L, Wellons JC, Blount JP, et al. Institutional experience with cranial vault encephalocele. J Neurosurg Pediatr. 2008;1:22-5.
31. Gamache FW. Treatment of Hydrocephalus in patients with meningomyelocele or encephalocele: a recent series. Childs Nerv Syst. 1995;11:487-8.
32. Referee EA, Refaat MI, Reda M. Incidence of secondary hydrocephalus after excision of huge encephaloceles in neonates: case study. J Neurol Surg A. 2018;79:15-8.
33. Bozinov O, Tirakotai W, Sure U, Bertalanffy H. Surgical closure and reconstruction of a large occipital encephalocele without parenchymal excision. Childs Nerv Syst. 2005;21:144-7.
34. Dorlands. Dorlands illustrated medical dictionary. 32nd ed. Newzeland: Elsevier Saunders; 2011:627.

Cite this article as: Babagana U, Abubakar AM. Clinical findings and surgical outcomes of encephalocoeles. Int J Res Med Sci 2022;10:590-4.