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Original Research Paper

Neurosurgery

INCIDENCE, PRESENTATION AND MANAGEMENT OF ENCEPHALOCELE AT A TERTIARY LEVEL CENTRE.

Dr Dinesh Kumar Rajput — Associate Professor, Department of Neurosurgery MLB MC Jhansi.

Dr Sucheta Rajput* — Assistant professor, Department of Gynaecology and obstetrics MLB MC Jhansi. *Corresponding Author

Dr Vijendra Maurya — Associate Professor, Department of Neurosurgery, Kalyan Singh Superspeciality Cancer Institute , Lucknow.

ABSTRACT Encephaloceles are herniation of cranial contents through defect in calvarium. It may be from occipital region to frontal region and also to base of skull. Aim of surgery in these cases is reduction of brain contents and watertight closure of duramater and repair of bone defect. **Aims And Objectives-** To know epidemiology of encephaloceol, its presentations, imaging features, surgery and surgical nuisance. **Material And Methods-** Retrospective study was carried out in MLB medical college from June 2017 to June 2022. All patients suffering with encephaloceol were recorded for clinical features, imaging and surgical procedures with their outcome and complications. **Results** – In our study 50 cases of encephaloceol were operated during this time duration. Out of which 30 cases (60%) were boys and 20 cases (40%) were girls with age ranging from 2 days to 5 years. Most common presentation was occipital (n=35, 70 %) followed by, Parietal (n=5, 10%) Nasofrontal (n=5, 10%) and Nasofrontoethmoid (n=5, 10 %). Most common associated congenital anomalies were chiari malformations, meningomyelocoels, syrinx and hydrocephalus. 45 patients (90%) were discharged well after surgery. Two patients developed cerebrospinal fluid leaks followed by meningitis. Three patients developed hydrocephalus after surgery, in two of them VP shunt was done but in one patient presented in late stage with extensor response so could not saved. One patient developed severe sepsis and ARDS, did not survive after surgery, after prolonged intensive stay. **Conclusion-** In our study, most common type of encephalocoel is occipital and they have better surgical outcome with early surgery. Poor outcome is because of other comorbid associated anomalies.

KEYWORDS : Encephalocele, meningomyelocele, surgical management.

INTRODUCTION-

Encephaloceol is congenital malformation associated with neural tube defect disorder and characterized by herniation of brain tissue through defect of calvarium. Depending on position of encephaloceol they are divided into occipital, parietal, frontonasal and frontonasoethmoid.These are common congenital problem and seen in worldwide in neurosurgery practice. The overall incidence of encephaloceol is about 0.8-3.0/10000 live births. Most of encephalocoels are occipital; approximately 75 percent followed by frontoethmoid (13-15%), parietal (10-12%) and followed by sphenoidal.Frontoethmoidal encephaloceol are relatively high incidences (1:5000 live births) in Southeast Asia countries like Malaysia, Myanmar and Thailand. Basal encephalocoels are very rare.MRI brain is ideal imaging modality to diagnose which part of brain is herniated and also to know other associated anomalies. MR angiography and MR venogram are very helpful in Diagnosing arterial and venous anatomy, venous sinuses and torcula. By the help of venogram, proper surgery is planned as sometimes torcula herniated through occipital encephaloceol. CT scan head with three-dimensional reconstruction are helpful in delineating bony defects so planning for bone reconstruction during surgery.

Management of these encephalocoels require multidisciplinary team which includes neurosurgeon, plastic surgeon, maxillofacial surgeon and neuroanaesthestist and paediatric intensivist. Encephalocoels must be operated as soon as possible in early age of life. Aim of surgery is resection of herniated brain tissue with meticulous care and water tight dural repair followed by reconstruction of bone defect. Bone defects can be reconstructed with the help of autologous bone graft or by titanium mesh.

Aim of present study is evaluation of clinical features of encephaloceol cases and management of these cases. Results of our study, and knowledge may be applied in further studies and further clinical trials and surgical approaches.

MATERIALS AND METHODS-

This Retrospective study was conducted in MLB Medical college Jhansi we also. All cases who were admitted and were operated from June 2017 to June 2022 were included in this study. Case records of all patients were evaluated regarding age, sex and size of encephalocoels and also associated congenital cranial and systemic anomalies. All patients undergo three-dimensional reconstruction CT brain and face, MRI brain, MR angiogram, MR Venogram. All patients undergo surgical correction. Some patients need ventriculoperitoneal shunts because of hydrocephalus and some patients required shunts in second stage as they developed ventriculomegaly after encephalocele surgery.in occipital encephalocele, cases were operated in prone position while frontonasal and frontonasoethmoidal cases were operated in supine cases. In frontonasal cases, bicoronal incision given with bifrontal craniotomy. Frontal lobe retracted and atrophic brain tissue were resected and defect was repaired with pericranium and fibrin glue.

RESULTS-

Sex Distribution (n=50)

Sex	No.of patients
Male	30
Female	20

Age Wise Distribution

Age	No. Of patients
< 6 months	25
7 -12 months	10
1—3 years	10
3-6 years	05

Type Of Encephalocele

Type of encephalocele	No. Of patients
Occipital	35
Parietal	5

Nasofrontal	5
Nasofrontoethmoid	5

Associated Anomalies

Associated anomalies	No. of cases
Meningomyelocele	5
Chiari malformation	10
Syrinx	5
Dermal sinus tract	3

Post Surgery Complications-

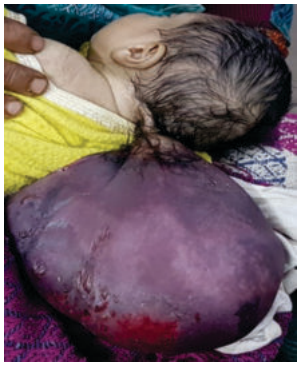
Complications	No. of patients
CSF Leak	5
Meningitis	3
Wound infection	3

Type Of CSF Diversion

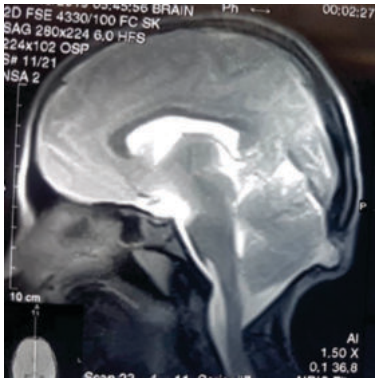
Type of CSF Diversion	No of patients
LP shunt	10
VP shunt	10

Surgical Outcomes

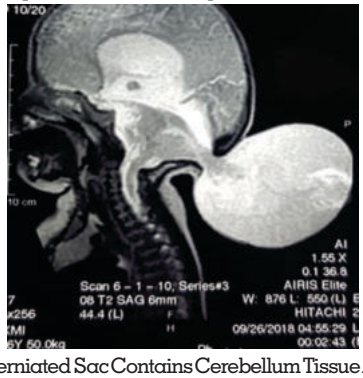
Outcome	No. of cases
Excellent	30
Good	10
Satisfactory	5
Unchanged	3
Poor	2



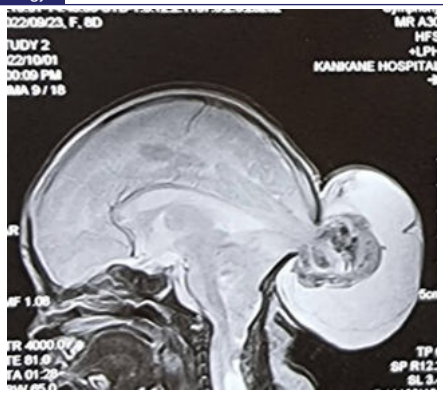
Legand 1- Large Sac In Encephalocele



Legand 2- Very Small Sac In Encephalocele



Legand 3- Herniated Sac Contains Cerebellum Tissue And Torcula



Legand 4 – Sac Containing Cerebral And Cerebellar Parenchyma With Torcula

DISCUSSION-

Encephalocoels are common congenital disorder, in which a portion of brain is herniated through defect. Regarding etiopathogenesis of encephaloceles, different theories were proposed, out of which most accepted one is theory proposed by e Klerk and DeVilliers. In his theory they suggested that adhesions between the neuroectoderm and surface ectoderm prevent the normal growth of mesoderm, so not able to form skull. These are also supposed because of deficiency of poor intake of folic acid during pregnancies. That is why, rare in west and still very common in developing and backward countries. Encephaloceol are classified as per site of herniation of brain tissue, as given in table form.

Classification Of Encephaloceles

Type	Site of herniation	Location of mass
I. Occipital		
II. Frontal		
Sincipital	Fonticulus nasofrontalis	
Nasofrontal	Foramen cecum	
Nasoethmoidal	Medial orbital wall	Forehead: Nasal bridge
Naso-orbital		Nasal bridge
Basal	Cribriform plate	Orbit
Transethmoidal	Between ethmoid and sphenoid	
Sphenoethmoidal	Craniopharyngeal canal	Intranasal
Trans sphenoidal	Superior and inferior orbital fissure	Nasopharynx
Sphenomaxillary	Fonticulus nasofrontalis	Nasopharynx

Frontal encephalocoel are divided into sincipital or frontoethmoidal (60%) and basal (40%). The site of the sincipital encephalocoel is at the cranial end of defect and defect is at the area of foramen Caecum at the junction of frontal and ethmoid bone. Encephalocoel sac may be sessile or pedunculated and skin overlying sac may be normal, may be shiny or thick or wrinkled.

Sincipital encephalocoel are further divided into nasofrontal, Nasoethmoidal nasoorbital.

Each of patient must be evaluated carefully with clinical examination and thorough investigations to plan for surgery and to achieve highest quality results and reducing morbidity or disfigurement. The aim of surgical correction is closure of open skin defects to reduce infection and desiccation of viable brain tissue, removal of non-viable extracranial part of brain with meticulous dissection and water tight closure of dura with reconstruction of skull defect with autologous bone or mesh.

Frontoethmoidal encephalocoel must be treated early to prevent facial deformities

Occipital encephalocoel is very common. In our study also, occipital encephalocoels was present in 60 % cases. the size of encephalocoel sac may be vary from very small to quite large and contains CSF and atrophied brain, which can be excised safely without causing much harm to patients. The size of bony defect may also variable.

In our series, male are affected more (60%) as compared to other studies. Age of presentation vary may be few days to six years. In our study, patients are referred from large geographical area as MLB Medical college had been selected as referral centre under RSBK program. That is why this does reflect true incidence and prevalence in our population.

In large sac encephalocoels surgery is planned earlier as there are chance to develop sore followed by infection. In some cases (n==) hydrocephalus was present and VP shunt was done in same sitting. In other few cases, hydrocephalus developed in later cases (n=) which is similar to other studies also.

In our study (90%) cases did well in postoperative period similar to other studies.

Four of our cases developed wound dehiscence and CSF leak which were managed conservatively. One of our patients died because of severe meningitis followed by sepsis in postoperative period.

CONCLUSION –

This study elaborated that encephalocele are very comlex congenital anomalies and management of such cases is very challenging. Multidisciplinary management is required while dealing such cases to achieve best outcome in these cases. A team of expiranced neurosurgeon, maxillosurgeon, plastic surgeon, neuroanaesthetic and pediatric intesivist is required. In cases of encephaloceol, surgical outcome depends upon type of encephalocoel, nature of associated brain anomalies and other comorbid systemic anomalies.MRI brain, MR angiogram MR venogram along with CT Face with 3D reconstruction are best imaging. Surgery is mainstay of treatment which must be done early as soon as possible.

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