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Risk and prognostic factors in patients with congenital encephalocele

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Abstract

Background and objective Encephaloceles are considered to be a spectrum of neural tube defects with a herniation of the brain and the meninges through a bony skull defect to the exterior.

Methods This retrospective and prospective study was carried out on 30 patients with congenital encephaloceles either vault or basal. The risk factors and the prognostic factors were assessed.

Results There was a statistically significant association between the content of the sac, outcome and seizures. There was a presence of neural tissue associated with worse outcome and seizures. There was a statistically significant association between the size of the sac and outcome with seizure. There was a statistically insignificant association between site of defect and parent consanguinity, folic acid intake, drug history, seizures, outcome, gender, family history and a statistically significant association between the site of the sac and hydrocephalus. There was a statistically significant association between the outcome and presence of Hydrocephalus, microcephaly and seizures.

Conclusions There was statistically significant association between the worst outcome and presence of hydrocephalus, microcephaly and seizures, and there is correlation between site of the defect in occipitocervical region and development of hydrocephalus.

Keywords Prognostic factors, Congenital encephalocele, Hydrocephalus

Introduction

An encephalocele is a congenital herniation of intracranial contents throughout a cranial defect. These intracranial contents, which protrude through the defect, may include meninges, cerebrospinal fluid (CSF), and/or brain tissue [1, 2].

The estimated worldwide prevalence is 0.8–4 per 10,000 live births. Encephaloceles are generally classified based on the anatomical location where 75% of encephaloceles are located in the occipital region, 13–15% are

situated in the frontal ethmoidal region, and 10–12% are in the parietal or the sphenoidal area [3–5].

The etiology of congenital encephaloceles is not fully understood but many risk factors may be associated such as genetic factor, environmental factors like maternal nutrition deficiencies such as folic acid deficiencies [2].

Encephaloceles are considered to be a spectrum of neural tube defects caused due to failure of neurulation [2, 6].

Clinical presentations vary depending on each major subtype of encephalocele. Most of the patients present with swelling in the head region, depending on the location and size of the encephalocele (Fig. 1). Patients can also present with seizures, developmental delay, vision impairment, microcephaly and features of raised intracranial pressure [7, 8].

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Fig. 1 Clinical photograph shows two encephalocele (post-parietal and occipital)

For a new born with an encephalocele, the prognosis depends on many factors including the size and contents of the sac, presence of microcephaly, hydrocephalus [1, 3, 5, 10, 11].

Hydrocephalus carries a significant morbidity in these children, reported to occur in 60–90% of patients with occipital encephaloceles. Hydrocephalus is frequently present preoperatively or develops after the surgical repair of occipital encephaloceles [1, 3, 9, 12].

The diagnosis is usually prenatally by ultrasonography and magnetic resonance imaging (MRI), but after birth, MRI (magnetic resonance imaging) is unequivocally the modality of choice for imaging a case of encephalocele. MRI helps to delineate the location of the encephalocele and characterize the hernia sac (based on the size and contents) and CT (computed tomography) may also be used to display the bone anatomy [2, 13].

The management of encephalocele defects requires immediate surgical closure. During surgical intervention, the neural tissue in the sac is excised and the dura must be closed in a watertight manner (Fig. 2) and associated hydrocephalus if present can be managed with VP shunt [3, 12, 14–16]. The aim of this work was to detect the risk factors for the congenital encephalocele and to illustrate the possible prognostic factor.

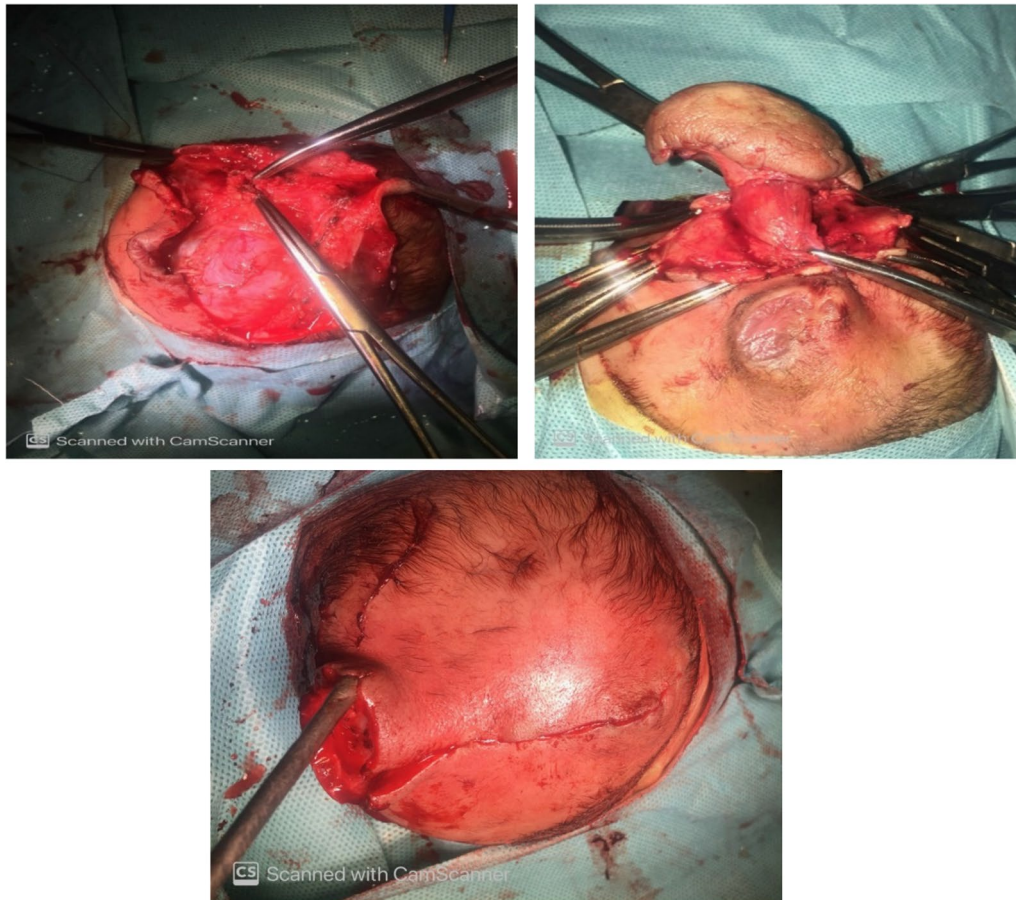


Fig. 2 Surgical repair of encephalocele

Patients and methods

This retrospective and prospective cohort observational study was carried out on 30 patients with congenital encephaloceles either vault or basal. Our study was done at neurosurgery department at Sohag University Hospital. The study was done after approval from the scientific ethics committee. An informed written consent was obtained from all patient guardians.

Exclusion criteria were encephaloceles associated with major craniofacial abnormalities.

All patients were subjected to: maternal evaluation using structured questionnaire during interview at outpatient clinic about the following risk factors; mother age, degree of consanguinity, family history of neural tube defect, folic acid intake during pregnancy, maternal illness (like TORCH infection and anemia), nutrition, maternal education level, mode of delivery. Patient evaluation which include clinical assessment (history of present illness such as age, gender and NICU admission, general examination by obtaining necessary information about the cardiovascular, respiratory, gastrointestinal and genitourinary systems and exclusion of other congenital anomalies, neurological examination for presence or absence of any neurological deficit was specifically noted after acquiring neurological examination of each patient, local examination for the following factors due to their prognostic value: site,

size of encephalocele (Fig. 3) (the size was classified into atretic which is a small encephalocele that just can be seen by eyes, large about or more than the size of the head of the patients and medium in between these two sizes), content of the sac, associated hydrocephalus, associated seizures and sac rupture with CSF leak); investigations such as MRI brain and MRV were done in all cases except in two cases that admitted with sac rupture with CSF leak (Figs. 4 and 5).

CT brain was done in all cases to assess the intracranial contents for gross brain structure, ventricular size and bone defect, surgical treatment: We dissect the sac from the surrounding normal skin, then excision and repair of herniated part of the brain which was nonviable, then we close the dura mater in a watertight manner. In the final step, we close the subcutaneous tissue and the skin (Fig. 6), follow-up daily for all patients during hospital stay (average 5 day) and monthly in outpatient clinic for at least 6 months (Fig. 7) assessing patient's surgical wound healing, head circumference, an increase in the ventricular size on imaging studies and neurological deficit and assessing motor and verbal developmental milestones. Children who reached all milestones appropriately were considered to have normal neurological status, whereas children who did not meet all milestones were considered to have developmental delay.



Fig. 3 Clinical photograph of partial encephalocele

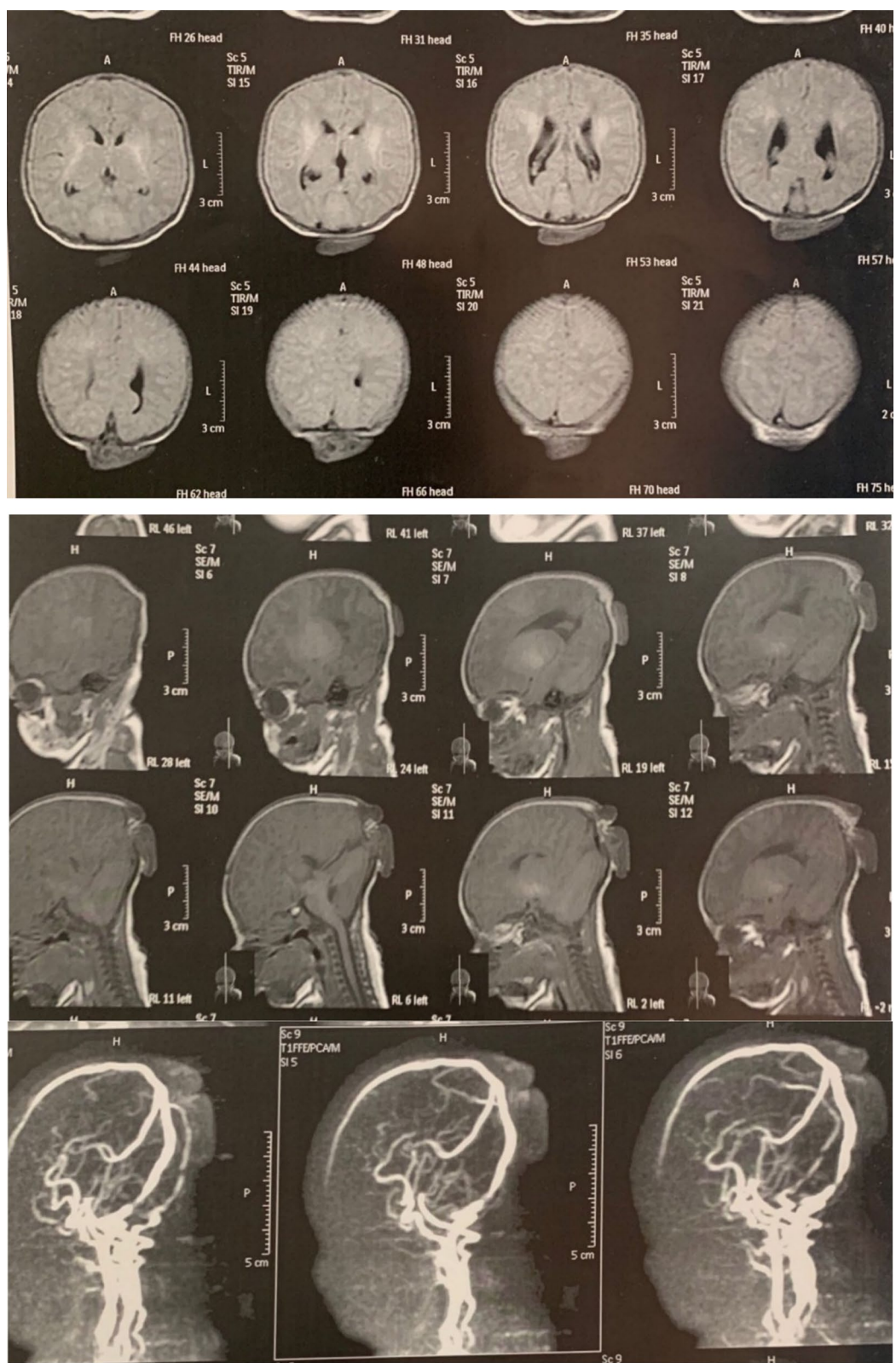


Fig. 4 CT brain, MRI brain and MRV of our case

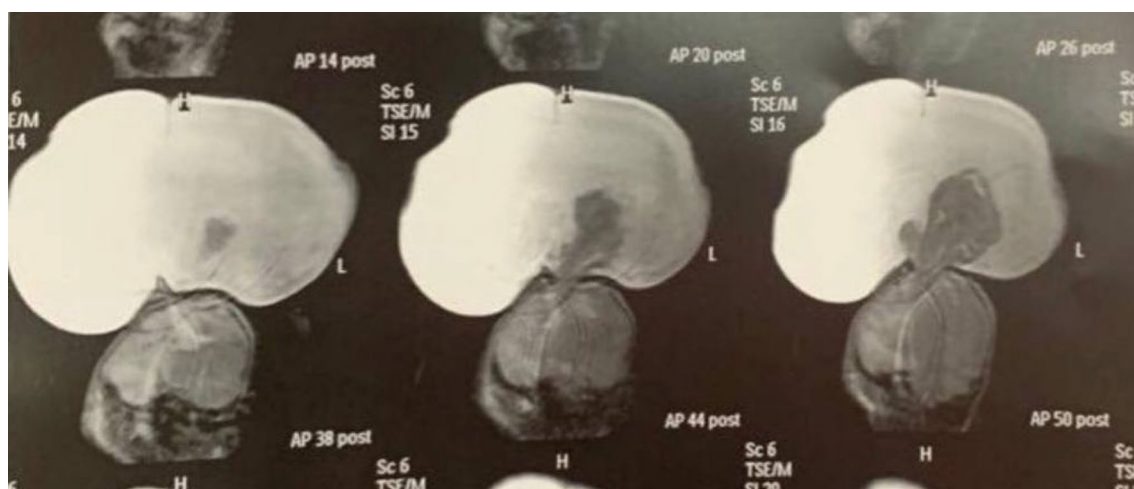


Fig. 5 MRI T2 (coronal view) of parietal encephalocele our case study



Fig. 6 Intraoperative picture

Statistical analysis

Data were collected and entered to the Statistical Package for Social Science (IBM SPSS) version 26. Pearson Chi-square was used to find out the significance of association between independent and dependent variable. A two tailed P value < 0.05 was considered significant.

Results

Table 1 shows maternal risk factors.

Table 2 shows clinical characteristics of encephalocele, cranial anomalies, complications of encephalocele, outcome of the studied patients.

There was a statistically significant association between the content of the sac, outcome and seizures. There was a presence of neural tissue associated with worse outcome and seizures. There were statistically significant associations between the size of the sac and outcome with seizure (Table 3).

There was a statistically insignificant association between site of defect and parent consanguinity, folic acid intake, drug history, seizures, outcome, gender, family history and a statistically significant association between the site of the sac and hydrocephalus (Table 4).



Fig. 7 The patient at the age of 2-year

There was a statistically significant association between the outcome and presence of Hydrocephalus, microcephaly and seizures (Table 5).

Discussion

Encephaloceles most probably occur as a result of mesodermal abnormality that causes a defect in the calvarium and dura through which protrudes the brain tissue. This usually occurs at 8–12 weeks of gestation [3, 10, 17, 18].

Younger mothers have been reported to be associated with encephaloceles [4, 19]. As confirmed in this study, 19 mothers (63.33%) were less than 25 years.

The families of most of the children lived in rural areas [1]. That match with our study. In our study 24 (80%) of patient were living in rural and 6 (20.0%) were living in urban.

Encephaloceles have occurred more in families with a lower education level [20, 21]. In our study, according to maternal education only 4 (3.33%) of patients’ mothers continued post-high school education. In contrast to other study, mother’s education level was not related to NTDs [22].

According to mode of delivery, 10 (33.33%) of patients were delivered vaginally, but 20 (66.67%) were delivered by cesarean section. In other study [1], twenty-one (66%) were delivered via vaginal delivery, and 11 (34%) via cesarean section.

In our study, according to mother’s disease, 6 mothers had maternal diseases, 1 (10.0%) had anemia, 1 (3.33%) had antiphospholipid syndrome, 1 (3.33%) had epilepsy and 1 (3.33%) had urinary tract infection.

Table 1 Maternal risk factors

Mother age (years)	25.23 ± 6.07
Gestational age	
Preterm	5 (16.67%)
Full term	25 (83.33%)
Maternal education	
> high school	3 (10.0%)
< high school	17 (56.66%)
Illiterate	10 (33.33%)
Residence	
Rural	24 (80.0%)
Urban	6 (20.0%)
Family history	4 (13.33%)
Folic acid intake during pregnancy	21 (70.0%)
Consanguinity	18 (60.0%)
History of maternal illness	
Anemia	3 (10.0%)
Antiphospholipid syndrome	1 (3.33%)
Epilepsy	1 (3.33%)
Urinary tract infection	1 (3.33%)
Total	6 (20%)
History of maternal drug use during pregnancy	
Antibiotics	1 (3.33%)
Antiepileptic	1 (3.33%)
Clexane	1 (3.33%)

Data are presented as mean ± SD or frequency (%)

Regard as mother drug use 1 (3.33%) received antibiotics, 1 (3.33%) received antiepileptic and 1 (3.33%) had clexane.

Table 2 Clinical characteristics of encephalocele, cranial anomalies, complications of encephalocele, outcome of the studied patients

Sex	
Male	14 (46.67%)
Female	16 (53.33%)
Location	
Occipital	20 (66.7%)
Occipitocervical	6 (20%)
Parietal	2 (6.7%)
Basal (transehmoidal)	1 (3.3%)
Frontoethmoidal	1 (3.3%)
Size	(9.10 × 9.38) ± (8.18 × 8.04)
Content	
Neural tissue	19 (63.33%)
Head circumference	
< 35 cm	7 (23.33%)
≥ 35 cm	23 (76.66%)
Sac rupture with CSF leak	2 (6.67%)
Cranial anomalies	
Corpus callosum dysgenesis	6 (20.0%)
Dandy–Walker malformation	2 (6.67%)
Preoperative hydrocephalous	2 (6.67%)
Microcephaly	7 (23.33%)
Total	17 (56.66%)
Extracranial anomalies	
Cleft lip	2 (6.67%)
Klippel feil syndrome	2 (6.67%)
Microtia	1 (3.33%)
Polycystic kidney	1 (3.33%)
Cardiac anomalies	2(6.67)
Complication	
Wound infection	2 (6.67%)
CSF leaking from repaired wound	3 (10.0%)
Postoperative hydrocephalus	5 (16.67%)
Seizures	6 (20%)
Outcome	
Good	13 (43.3%)
Delayed development	14 (46.7%)
Died	3 (10.0%)

Data are presented as mean ± SD or frequency (%). CSF: Cerebrospinal fluid

In other study, 13 (41%) mothers had upper respiratory tract infections, 6 (19%) had lower respiratory tract infections, 11 (34%) had experienced genitourinary infections, 19 (59%) of the mothers were exposed to 1 or more TORCH infection and 12 (38%) mothers used analgesics and antibiotics in the first trimester [1].

Evidence suggests that folate supplementation in the first 6 weeks of gestation may prevent NTDs and reduce the risk of another affected pregnancy [20, 23].

However, in our study 21 (70.0%) of patient received folic acid during pregnancy so we did not find any correlation between folic acid intake and encephalocele occurrence similar to the observations made by Rowland [24] and Mahajan [25].

In our study, family history was positive in 4 (13.33%) patients. In other study, family history was positive in (6.7%) of cases [11]. A second encephalocele case was observed in one family in our study.

Consanguinity degree was positive in 18 (60.0%) patients. Compared to other study, consanguinity was found in 67.08% of cases [26].

In our study, neural tissue was present in 19 patient (63.33%). In other study, neural tissue was present in 16 (53.3%) of 30 patients [10], and in other study it was present in (67%) [9]. In our study, there is a statistically significant association between the content of the sac and worse outcome.

In our study, size of encephalocele ranged from atretic to large encephalocele and there is statistically significant association between the size of the sac and outcome. All atretic encephaloceles in our study have good outcome. However, in other reports [11, 13] the size of the sac did not correlate with the prognosis.

Our findings are compatible with the Matson and Ingraham classification [27]; all of our patients presented with midline encephalocele and occipital region were predominant (66.7%) occipital, (20%) occipitocervical (6.7%) parietal, (3.33%) frontoethmoidal and basal (3.33%).

In other study of 30 encephalocele sacs, 27 were in the occipital region (90%), 1 was in the parietal region (3.33%), and 4 were in the frontal region (13.33%), [1] and in other study, 40% occipital, 26.7% occipitocervical, 20% vertex, 10% frontal, and 3.3% nasal [11].

In our study, there is correlation between site of the defect in occipitocervical region and development of hydrocephalus.

In our study, according to cranial anomalies 6 (20.0%) of patients had corpus callosum, 2 (6.67%) of patients had Dandy–Walker malformation and 2 (6.67%) of patients had preoperative hydrocephalous, and according to extracranial anomalies 2 (6.67%) of patients had cleft lip, 2 (6.67%) of patients had Klippel Feil syndrome, 1 (3.33%) had microtia, 1 (3.33%) of patients had polycystic kidney and 2 (6.67) of patients had cardiac anomalies.

Table 3 The statistical relationships between variables with the content and size of the defect

Variables	Content of the sac		Total	P value
	Neural tissue present	Neural tissue absent		
Sex				
Male	9	5	14	0.919
Female	10	6	16	
Consanguinity	12	6	18	0.643
Family history	2	2	4	0.552
Folic acid intake	15	6	21	0.160
Drug history	1	2	3	0.256
Hydrocephalus (preoperative and postoperative)	5	2	7	0.612
Seizures	6	0	6	0.037*
Outcome				
Good	4	7	11	0.047*
Delayed development	12	4	16	
Died	3	0	3	

Variables	Size of the sac			Total	P value
	Atretic	Medium	Large		
Sex					
Male	3	8	3	14	0.433
Female	3	12	1	16	
Consanguinity	4	11	3	18	0.707
Family history	1	3	0	4	0.697
Folic acid intake	3	16	2	21	0.240
Drug history	1	2	0	3	0.690
Hydrocephalus (preoperative and postoperative)	2	4	1	7	0.792
Seizures	0	3	3	6	0.009*
Outcome					
Good	4	7	0	11	0.028*
Delayed devolvment	0	12	4	16	
Died	0	3	0	3	

Data are presented as frequency, * significant as P value ≤ 0.05

Compared to other study, cranial anomalies were 13.3% of patients with corpus callosal agenesis, 6.7% with Dandy–Walker, 3.3% with right cerebellar agenesis, 3.3% with arachnoid cyst, and 3.3% with hydrocephalus [11].

In our study, 56.66% of patient had cranial anomalies compared to other study 61% of patients had cranial anomalies [9] and in other study, congenital brain anomalies were detected in (51.9%) of patients [20].

In our series of patients, hydrocephalus was observed in 2 patients (6.67%) who were treated by placing VP shunt before sac repair while 5 patients (16.67%) developed hydrocephalus after surgery that was again successfully managed by VP shunt as second surgery.

In comparison with other study, hydrocephalus was observed preoperatively in four patients (36.4%) who were treated by placing VP shunt before the repair of the sac and seven children (63.6%) of 17 patients developed hydrocephalus after surgery; six of them were managed by VP shunt [3].

Microcephalus was considered as important prognostic factor for worst outcome [3, 11, 15, 28, 29]. Our study compatible with these finding six of seven patients with microcephaly has delayed development and the last was died.

According to complication of the studied patients, 2 (6.67%) patients had wound infection, 3 (10.0%) patients had CSF leak from repaired wound, 2(6.67%) patients

Table 4 The statistical relationships between variables with the site of the defect

Variables	Site of the defect					Total	P value
	Occipital	Occipitocervical	Partial	Fronto ethmoidal	Basal		
Sex							
Male	7	4	2	1	0	14	0.174
Female	13	2	0	0	1	16	
Consanguinity	10	4	2	1	1	18	0.461
Family history	3	0	0	1	0	4	0.094
Folic acid intake	15	4	1	0	1	21	0.491
Drug history	3	0	0	0	0	3	0.797
Hydrocephalus (preoperative and postoperative)	1	6	0	0	0	7	0.001*
Seizures	3	2	1	0	0	6	0.626
Outcome							
Good	10	0	0	1	0	11	0.163
Delayed development	9	5	1	0	1	16	
Died	1	1	1	0	1	3	

Data are presented as frequency, * significant as P value ≤ 0.05

Table 5 Association of some clinical features of encephalocele patients with their outcome

Variables	Outcome			Total	P value
	Good	Delayed development	Died		
Hydrocephalus	0	6	1	7	0.070
Seizures	0	4	2	6	0.019*
Microcephaly	0	6	1	7	0.070

Data are presented as frequency, * significant as P value ≤ 0.05

had sac rupture with CSF leak, 5 (16.67%) of patients had postoperative hydrocephalus and 6 (20%) had seizures.

In other study, 9 (18%) patients admitted with sac rupture with (CSF) leakage, 2 (4%) patients having rupture of sac after the admission and 1 (2%) patient admitted with the complaint of hemorrhage from the thin and shiny covering skin of the sac. Postoperatively, only 1 (2%) patient had CSF leakage from the repaired wound. Four (8%) patients developed hydrocephalus after the repair of protrude sac [28].

In our series of patients, the seizure was noted in 6 (20%) and all seizures were occurred in sac with neural tissue and there is statistically significant association between worse outcome and seizures. In other study, the seizure was noted in 8 (15%) [28] and in other study (13%) developed a seizure disorder [9].

Surgical intervention was done to all cases and involve resection of the encephalocele sac and suturing the dura mater in a watertight manner. Non-functional neural tissues commonly are excised [3, 10, 14–16, 28, 30].

Our study showed a 10% death, 53.3% delay in milestones, and 36.6% good outcome. Compared to other reports, mortality rate was 11.8% [18], 2% [28], 5.9% [3], and 10% [11].

In our study, there was statistically significant association between worse the outcome and presence of hydrocephalus, microcephaly and seizures. Compared to other study, hydrocephalus and microcephalus were considered as the two important risk factors for worst outcome [29].

Conclusions

Our studies showed that the size of the sac, the neural tissue content, hydrocephaly, microcephaly, and other pathologies that may accompany an encephalocele are factors that may determine the prognosis of patients with congenital encephaloceles and there was statistically significant association between worse the outcome and presence of hydrocephalus, microcephaly and seizures and there is correlation between site of the defect in occipitocervical region and development of hydrocephalus.

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Author contributions

AMM was involved in writing—original draft preparation; MAA, MMA, ASE were involved in formal analysis and review and editing; MMA was involved in methodology and supervision. All authors read and approved the final manuscript.

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Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Declarations**Ethics approval and consent to participate**

This study is approved by the Medical Research Ethics Committee of Sohag University Hospital. An Informed written consent is achieved from their parents.

Consent for publication

Not applicable.

Competing interests

The authors declare that they have no competing interests.

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