Giant Occipital Meningoencephaloceles: Challenges in Management and Our Experience

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Abstract

Introduction: Giant encephalocele are encephalocele more than the size of head from which it arises. They pose a management challenge in view of risk of associated anomaly, challenges in anaesthesia and surgical management. There is no literature available from peripheral tertiary care centres from India apart from few case reports.

Methods: This case series comprises of seven cases operated at our institute from 2014 to 2021. After routine clinical examination, patients subjected to recommended radiological investigations. Patients were anaesthetised with standard protocol with intubation in lateral or supine position with head hanging at edge of table and supported at table. Surgery carried out in lateral position with complete excision of sac with CSF diversion if pre existing hydrocephalus was present.

Results: Out of seven cases, 3 were males and four female with age range from 12 days to 14 Mo. All patients were anaesthetised with standard protocol and had excision of sac with CSF diversion if required with satisfactory outcome in post operative phase and follow up. No death was recorded.

Conclusion: With careful assessment and all recommended investigation with standard practice of anaesthesia and surgery, acceptable outcomes can be expected. Emphasis should be laid for good antenatal folate supplementation and possible antenatal diagnosis of meningoencephaloceles.

Key words: CSF diversion, difficult intubation, giant encephalocele, Surgery

Introduction

Meningoencephalocele is the herniation of brain tissue and meningeal membranes along with collection of cerebrospinal fluid (CSF) through a bone defect in the skull. Embryologically it is thought to be a mesodermal defect.[1,2] An encephalocele larger in size than the size of head from which it arises is termed as a giant encephalocele.[3] Giant encephaloceles are most frequently observed at occipital region.[4] Giant occipital encephalocele is a rare entity and mostly reported as case reports. In Southeast Asia, the incidence of meningoencephalocele is one in 5000 live births (Creightonetal. 1974).[5] The causative mechanisms of such birth defects may be genetic and environmental factors or may be their interactions. There has been a protective effect of folate supplementation to mother during pregnancy.[6,7] With advancement in clinical experience, antenatal diagnosis of meningoencephalocele is feasible.[8] About 40-60 % of giant occipital encephalocele are associated with other congenital anomalies.[9,10] The optimal management of encephaloceles requires a multidisciplinary team consisting of neurosurgeon, neuroanesthesiologist, paediatric, maxillofacial, and plastic surgeon working in synchronisation. The challenges in anaesthetic management includes securing of airway because of inability of good extension, maintenance of perioperative fluid management, electrolyte disturbances, hypothermia, blood loss, associated birth defects and risks associated with aspiration pneumonitis.[11] Surgery aims at excision of sac, tight dural closure, reconstruction of bone defect along with management of hydrocephalus if any.[4] At times other surgical specialities and rehabilitation expertise is required on individual case to case basis. There

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are a lot of challenges in successful management of giant occipital encephalocele which we have tried to address as per our experience in managing this rare clinical entity.

Materials and Methods

We analysed seven consecutive cases of giant occipital encephaloceles operated at Department of Neurosurgery, Government Medical College, Kota from 2014 to 2021. The size of sac in all the patients was larger than the size of head of the patient. All patients were assessed with a detailed clinical history including presenting complaints, any symptoms of hydrocephalus, motor weakness or bladder bowel involvement, respiratory complaints or any other symptoms referable to other associated congenital anomaly. Examination included looking for size of head, size of sac, status of anterior fontanelle, examination of sac for possible contents, pupillary light reflex and response to follow light, respiratory efforts and rate, movement of limbs, any facial anomaly, syndactyly, club foot, cutaneous marker over back. The patients underwent radiological investigations including MRI Brain with whole spine screening, MR Venogram brain, USG whole abdomen, 2 D Echo, visual evoked response. After thorough assessment of all investigations and examination findings, plan of management was decided to go for excision alone or with CSF diversion in same sitting. Prognosis of possible outcomes explained well to parents and informed consent was obtained. Intraoperative data including technique and position for intubation, any complication or hemodynamic alteration, blood loss was recorded and post operative data including outcome of surgery, any complication including CSF leak, meningitis, hydrocephalus etc were recorded. We had a standard anaesthesia protocol for all children. All patients were attached with pulse oximeter, paediatric BP cuffs, EtCO2 monitor, ECG leads, two IV cannulas and body warmer were kept. All patients were given glycopyrrolate 2microgm/ kg iv, Fentanyl 1.5 microgm/ kg iv, Propofol 2 mg/kg iv and muscle relaxation was maintained with Succinyl choline 2mg/kg iv. Intubation was done either in lateral position or in supine position with head hanging at the edge of table and supported by an assistant and intubation done with adequate size tube and fixed. For maintenance anaesthesia oxygen, sevoflurane, atracurium intermittent bolus were given. If hydrocephalus present CSF diversion with a low pressure ventriculoperitoneal Chhabra type shunt was done in supine position followed by excision of sac in lateral position. After painting and draping, an elliptical incision was taken at the base of sac, after careful dissection base of sac was exposed and excision of sac was done at the base. After excision contents were reduced back to cranial cavity and dural closure was done in a water tight manner and bone defect was covered with a gelfoam. Proper sterile dressings applied after skin closure and patient was turned supine for extubation which was facilitated after reversal of residual paralysis with neostigmine. Patients were shifted to post operative ICU and watched for all vital parameters including signs of hydrocephalus in post operated period for non shunted patients. If hydrocephalus developed in post operated period confirmed on NCCT brain, CSF diversion was planned. A follow up assessment was done till 5 years.

Results

We had total seven cases of giant encephalocele from 2014 to 2021, three male and four female. The age of presentation ranged from 12 days to 14 Mo with average age of presentation being 8.2 Mo. The presentation of all patients was gradually increasing occipital swelling since birth. The size of encephalocele was larger than the head of the patient (Figure 1). There was associated club foot (Figure 2) in one case and acyanotic congenital heart disease on 2 D echo in two cases. Out of seven patient four patients were having hydrocephalus associated upon MRI brain (Figure 3 a and 3 b). Two of the patients were diagnosed antenatally on USG as having some occipital swelling. There was no folate intake in antenatal period in two cases. Five out of seven cases were intubated well in lateral position, for two cases due to high anterior location of larynx and inability to extend well due to large size of sac; larynx was not well visualised on lateral position hence supine position with head hung on the edge of table and sac well supported by an attendant was used (Figure 4a and 4b). Two patients had intraoperative rhythm disturbances, no patient had significant hemodynamic changes resulting in any shock, asystole etc. Two patients needed intraoperative blood transfusion. No event of significant hypothermia noted. All patient had complete excision of sac at the base and dura reduced back to cranial cavity after a water tight closure and covered with layer of pericranium over it, and defect covered with gelfoam. The contents of sac in five patients were meninges with CSF while two patients had associated small amount of redundant brain tissue. Four of the patients who had hydrocephalus pre existing had CSF diversion in form of low pressure ventriculoperitoneal shunt during same sitting while two patients who developed hydrocephalus in post operated day 4 and post operated day 6 had CSF diversion in similar manner on Day 5 and day 6 respectively. One patient did not require any CSF diversion. Two patients had a complication of wound infection with no meningitis in post operated period which was managed with regular dressing and culture specific antibiotics, two patients had CSF leak on day day 4 and day 6 of surgery and on CT had hydrocephalus. Leak was managed with CSF shunting. Rest of the patients had an uneventful post operated course.

Patients are followed up till 5 years post surgery except one patient who had a follow up till 18 Mo post surgery then lost in follow up. All patients are having satisfactory outcome in follow up visits with actively moving all four limbs. One patient had seizure episodes at 3 Mo follow up and is continued with anticonvulsants. No mortality reported in our series.

Table 1 summarises experience of our results in present series.

Patient 7	14 Mo/M	Taken	No	Nil	No	Supine	No	Day 6 Post surgery	CSf, meninges,	CSF leak	Continued uneventful
Patient 6	4Mo/M	Not taken	Yes	Nil	No	Lateral	No	Day 5 post surgery	CSf, meninges,	CSF leak	Continued uneventful
Patient 5	6Mo/F	Taken	Yes	Club foot	Present	Lateral	No	Low pressure VP shunt in same sitting	CSf, meninges, redundant brain tissue	Uneventful	Seizures at 3 Mo follow up, continued on anti convulsants
Patient 4	10 Mo/M	Taken	No	Acyanotic congenital heart disease	Present	Lateral	Yes	Low pressure VP shunt in same sitting	CSf, meninges,	Wound infection	Lost to follow up at 18 Mo
Patient 3	11 Mo/F	Taken	No	Nil	Present	Lateral	Yes	Low pressure VP shunt in same sitting	CSf, meninges, redundant brain tissue	Uneventful	Continued uneventful
Patient 2	12 Mo/F	Not taken	No	Acyanotic congenital heart disease	Present	Supine	No	Low pressure VP shunt in same sitting	CSf, meninges,	Wound infection	5 year follow up complete, uneventful
Patient 1	12 days/F	Taken	No	Nil	No	Lateral	No	Not required	CSf, meninges,	Uneventful	5 year follow up complete, uneventful
Age/sex	Antenatal folate	Antenatal diagnosis	Associated anomaly	Pre existing hydrocephalus	Intubation	Intraoperative blood transfusion	CSF diversion		Sac contents	Post surgery complications	Follow up

Table 1 : Summary of results of giant occipital encephalocele in current series

	Mahapatra et al	Mahajan etal	Current Series
Number of cases	14	29	7
Age range	2 days to 4 years	2 days to 18 Months	12 days to 14 Mo
Sex Distribution	9 Male; 5 Females	16 Male; 13 Females	3 Males; 4 Female
Antenatal history	Not significant	Not significant	No folate intake in two cases Antenatal USG : Swelling over occipital region in 2 cases
Associated anomaly	Craniostenosis	Acyanotic heart disease, Bilateral inguinal hernia, corpus callosum agenesis, ectopic kidney, craniostenosis	Acyanotic congenital heart disease, Club foot
Intubation	No mention	26: Right lateral 03: Supine with head hanging at table and supported by assistant	05: Right lateral 02: Supine with head hanging at table and supported by assistant
CSF diversion	5: same sitting with sac excision 2: Post-operative period	6 cases	4: same sitting with sac excision 2: Post-operative period
Intraoperative Complication	Cardiac arrest, Severe hypothermia	Hypotension, hypothermia, Rhythm disturbances, hypoxia during intubation attempts	Rhythm disturbances, Need for blood transfusion
Postoperative Complications	Hydrocephalus, Respiratory depression and ventilator support	Wound infection, hydrocephalus, Seizures	Wound infection, CSF leak with hydrocephalus
Mortality	2 deaths	3 deaths	No death
Follow up	6 Mo to 6 Years follow up Shunt revised in two patients	No long term follow up data	Up to 5 years, One lost in follow up at 18 Mo follow up, one pt. had seizures at 3 Mo follow up.

Table 2: Results of other case series from India and current series

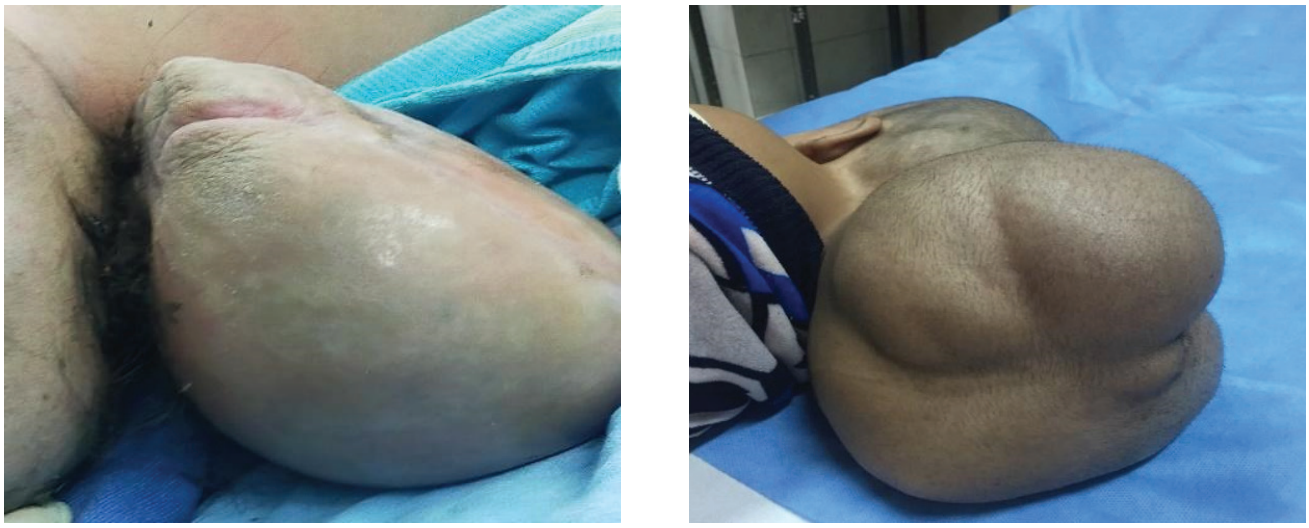

Figure 1 : Clinical picture of meningoencephalocele sac

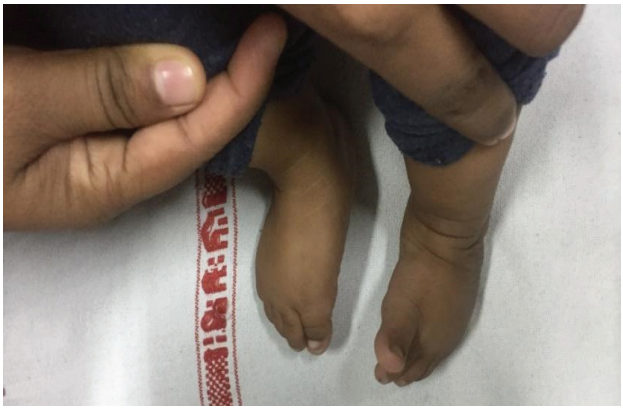

Figure 2 : Clinical picture of associated syndactyly

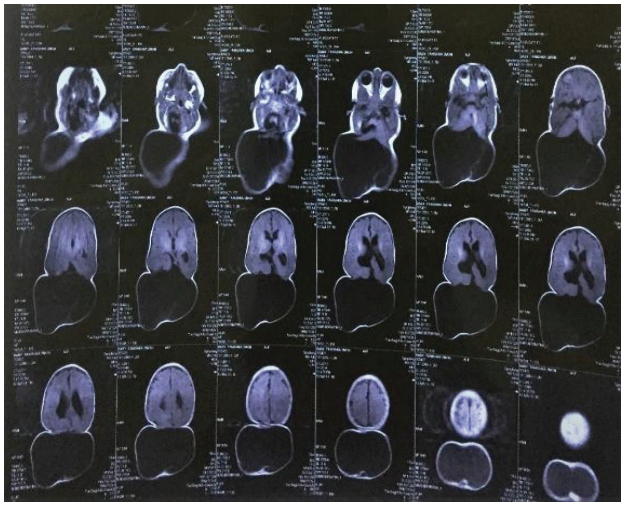

Figure 3a: MRI brain with no hydrocephalus associated

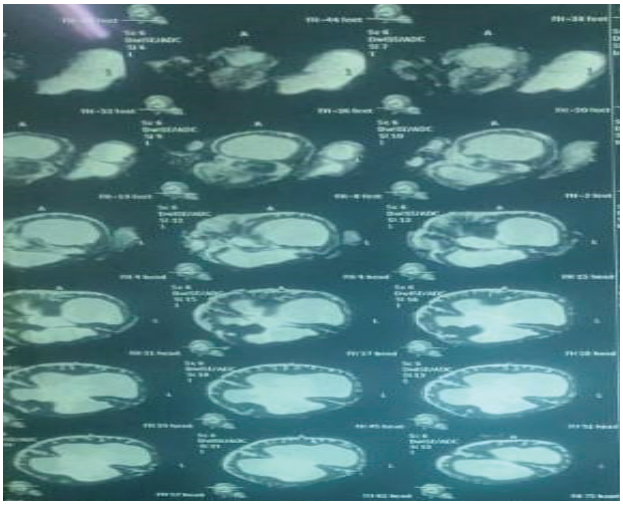

Figure 3 b : MRI brain with associated hydrocephalus

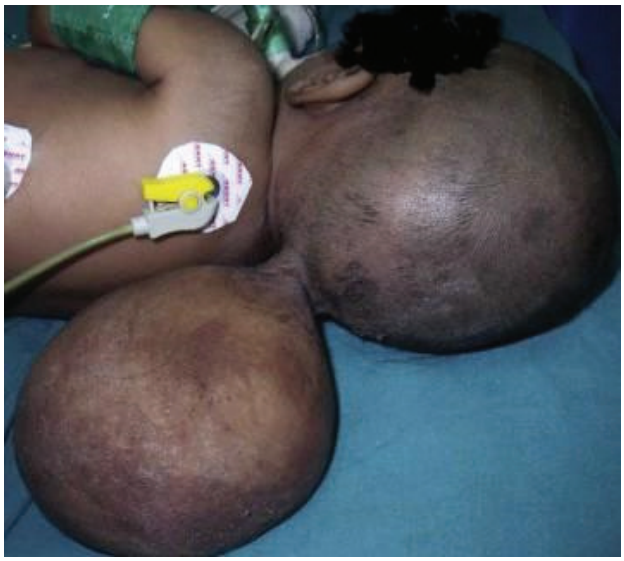

Figure 4a: Intubation in lateral position

Figure 4b: Intubation in supine position with head hanging down the table and sac supported by assistant

Discussion

Though Occipital encephaloceles are not infrequently encountered; giant occipital encephalocele having size of sac more than size of head are rare. They may also be called as massive or large encephalocele.[10,12,13] Management of giant occipital encephaloceles has been a challenge for Neurosurgeons, Neuroanaesthesiologist, and parents. There have been two case series from India Mahapatra et al[10] and Mahajan et al[11] for management challenges exclusively of giant occipital encephaloceles, we have shared our experience of managing giant occipital encephalocele at peripheral tertiary care centre of India which is to the best of our knowledge is first of its kind at peripheral tertiary care centre in India. The patient presented to us at age range of 12 days to 14 Mo which is a age comparable to Mahapatra et al and Mahajan et al. Majority of patients in our series were females which differs with sex distribution in other series. There was absence of antenatal folate intake in two cases, other series have not thrown light on antenatal folate in their patients. Two of the patients were antenatally diagnosed with occipital swelling on USG, other series does not talk about antenatal diagnosis of giant occipital encephalocele. There was a patient with club foot and two patients with acyanotic congenital heart disease in current series. The comparative results with respect to other available case series in Indian scenario has been tabulated as in table 2.

About the development of occipital meningoencephalocele, various theories have been put forward; the most acceptable one is by De Klerk and De Villiers. This theory suggests that there is hindrance to normal ingrowth of mesoderm to grow skull normally due to adhesions between neuroectoderm and surface ectoderm.[14] There may be associated congenital anomalies in these patients including club foot, facial anomalies, urogenital anomalies, rectal anomalies, congenital cardiac diseases and therefore a thorough clinical examination to rule out all these conditions along with supportive investigations including 2 D echo and USG should be done.[15,16] MRI brain with whole spine screening is the investigation of choice for occipital encephalocele patients where we can look for the sac, its possible contents, margins of defect, hydrocephalus if any, thickness of cortical matter, spinal dysraphism anomaly at other levels, other intracranial associated anomalies like corpus callosum agenesis, aqueductal stenosis, migration disorders can be ruled out. As a routine all patients should undergo MR venogram brain to rule out venous sinuses or torcula as a content of sac and any compression over them. Visual evoked response is important to rule out visual cortex as a content of sac and prognostication to patient.[10,5,4] Meningoencephaloceles are one of the congenital anomalies which can be definitively diagnosed in antenatal period and foetal interventions are feasible for its antenatal management. Maternal serum alpha fetoproteins, Antenatal USG and Foetal MRI are useful for definitive diagnosis in antenatal period.[9] Anaesthetic management of these cases poses a challenge in terms of paediatric age group, associated anomalies, inability to maintain adequate extension for endotracheal intubation, hypothermia, intraoperative fluid management, narrow margin of blood loss. For intubation there may be preference for lateral position, supine with head hanged on edge of table and supported by an assistant. Alternatively, fibreoptic bronchoscopy, video laryngoscopy, optical stylets or simply laryngeal mask airways with a bougie are a good back up to secure the challenging airway in such patients.[5,11] The complete surgical excision with careful dissection of sac and repositioning of contents back into cranial cavity again poses a challenge to surgeons. The usual position for surgery can be lateral or prone. An elliptical incision at the base of sac followed by careful dissection of membranes to expose the sac and make it free. Laying open the sac and repositioning the contents back, water tight closure of dura and reinforcing it with a cover of pericranium prevents reherniation in long term.[5,10,11] There has been a practice of cranioplasty using autologous split bone graft or methyl methacrylate grafts but in current series we had a practice of placing a gelfoam over a defect and no case of reherniation reported so far to us. At times due to large sac contents its not feasible to reduce them completely and excision of brain tissue may be warranted or some authors prefer to go for expansile cranioplasty[12]. There may or may not be a requirement of CSF diversion with ventriculoperitoneal shunt in same sitting with excision of sac or in post operative period.[5,6,11,17] We have four ventriculoperitoneal shunts during same sitting and two cases on post surgery day 5 and 6 respectively for CSF leak and hydrocephalus on NCCT brain. The complications seen in post operative phase include fever, wound infection, CSF leak, meningitis, respiratory complications, hydrocephalus, seizures.[10,11] The prognosis of outcome depends on contents of sac, weight of child, need for CSF diversion, associated anomalies, avoidance of infection.[3, 5, 9,11] As a developing nation, we have still many cases of neural tube defects or occipital meningoencephaloceles as compared to western world. We should aim to intensify the preventive measures including antenatal folate supplementation to all expected mothers along with prenatal screening with maternal serum alpha fetoproteins and antenatal USGs[9,17] and offered prenatal management with appropriate referrals to affected mothers.

Conclusions

Giant occipital encephaloceles are a management challenge. Careful assessment of possible challenge in

preoperative phase with all relevant investigations and involvement of multidisciplinary specialities can give acceptable results. We recommend an earlier intervention with preoperative optimisation and a liberal approach for CSF diversion for good surgical outcome. We also recommend to intensify antenatal folate coverage in all expectant mothers and to have a recommended antenatal screening measures to have the best management options to the affected children.

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Consent: Proper consent has been taken from legal guardians of patients to use their data for academic publication with assurance of non disclosure of identity to the best of the extent.

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