Giant Encephalocele: A Study of 14 Patients

A.K. Mahapatra

Department of Neurosurgery, AIIMS, New Delhi, India

Key Words

Giant encephalocele • Surgery • Outcome • Cranioplasty

Abstract

Background: Giant encephalocele is a rare condition and few published reports are available in the English literature. It is a challenge to neurosurgeons, even today. This series consists of 14 patients with giant encephaloceles treated at our institute. **Material and Observation:** Over a period of 8 years, from 2002 to 2009, 110 patients with encephaloceles were managed at our institute. Amongst them, 14 were children with giant encephaloceles. All patients had CT/MRI or both prior to surgery, and all were operated upon. Four patients were neonates, under 1 month of age, and 9/14 patients (64%) were under 3 months. The youngest child was a newborn baby aged 2 days. Except for 1 with an anterior encephalocele, the rest were patients with occipital encephaloceles. A CT scan was performed on 5 and an MRI on 1 patient. Both CT and MRI scans were performed on the other 8 patients. MRI/CT showed hydrocephalus in 10/14 patients. Of these, 7 required ventriculoperitoneal (VP) shunt, and the remaining 3 with mild to moderate hydrocephalus did not. Of the 7 patients who underwent VP shunt, 5 had a shunt during the encephalocele repair and 2 had a postoperative

shunt for increasing hydrocephalus. **Results:** Other associated anomalies recorded were acquired Chiari malformation in 3 patients, secondary craniostenosis with microcephaly in 5, and syringomyelia in 1 patient. All the patients underwent repair of encephalocele and 4 had suturectomy of coronal suture for the secondary craniostenosis. There were 2 postoperative deaths due to hypothermia. Among the 12 surviving patients, 9 had a good outcome and 3 had poor mental development. The present study shows overall good outcomes in 9/14 (66%) patients. Copyright © 2012 S. Karger AG, Basel

Introduction

Giant encephalocele is a rare clinical condition. Even today it is a formidable challenge to neurosurgeons [1, 2]. The challenge is due to the amount of brain tissue, which forms the content of the encephalocele, intraoperative blood loss and intra- and postoperative hypothermia. The condition is rarely reported in the literature. Except for a few case reports [1–5], no series has been published in the literature. Current analysis deals with 14 cases of giant encephaloceles treated at our center.

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Fig. 1. Clinical photograph of a case of occipital giant encephalocele.

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Fig. 2. Clinical photograph of a case of anterior fontanel encephalocele.

Fig. 3. CT scan showing occipital giant encephalocele with bone defect.

Fig. 4. MRI of occipital encephalocele.

Materials and Methods

This was a retrospective study at our institute, over an 8-year period, from 2002 to 2009. During this period, a total of 110 encephaloceles were treated, among them 14 patients with giant encephaloceles. All patients had CT, MRI or both, to evaluate the associated brain anomalies and to plan the surgical procedure. All patients were followed up at the outpatient department.

The patients ranged in age from 2 days to 4 years. Nine patients were under 3 months and among them 4 were neonates of less than 30 days. All the neonates weighed between 2 and 2.5 kg. Only 2 patients were over 1 year of age. Boys outnumbered girls, as there were 9 boys and 5 girls. Except for 1 patient who had anterior fontanel encephalocele, the remaining 13 had encephalocele in an

occipital or suboccipital location (fig. 1). In the 1 patient with anterior encephalocele, the encephalocele was coming through the anterior fontanel (fig. 2).

All the patients had imaging by CT or MRI (fig. 3). Five patients had CT and 1 patient had MRI only. The remaining 8 patients had both MRI and CT. CT provided additional information regarding the bony defect. Imaging revealed hydrocephali in 10 patients, of which 7 were gross or significant, and in 3 patients ventriculomegaly was considered mild to moderate. Three patients had Chiari II malformation and 1 patient had syringomyelia. Five patients had associated secondary craniostenosis, all with fused coronal suture and closed anterior fontanel. MRI revealed the content of the encephalocele (fig. 4), and in 3 patients there was no brain tissue inside the sac. CT scans were performed

to evaluate the size of the bone defect. In 3 patients the defect in the occipital bone was small and measured less than 3 cm in diameter. In these 3 patients MRI/CT did not show brain matter as the content of the sac.

Surgical Procedure and Outcome

All the patients were subjected to surgical repair. In 2 patients, hemoglobin was low; hence they were discharged and admitted later when hemoglobin was normalized. Amongst the 10 patients with hydrocephalus, 7 were considered suitable for ventricular diversion. Five patients had low-pressure ventriculoperitoneal (VP) shunt insertion during surgery for the repair of the encephaloceles. Two patients required VP shunt procedure within 2 weeks of surgical repair and before discharge from the hospital, as their postoperative noncontrast CT revealed further enlargement of the ventricle.

All the patients underwent repair of the encephalocele. In 3 patients, where there was no brain tissue, repair was simple closure after excising the sac. In 1 patient, torcula and part of the transverse sinuses were in the encephalocele sac, which was gently pushed forward and repair was done. In the remaining 9 patients with occipital encephaloceles, the herniating brain was partially excised and the rest was pushed gently into the cranial cavity (fig. 5a–c). In the 5 cases of craniostenosis, wide craniectomy was carried out around the coronal suture by a separate bicoronal skin incision (fig. 6a, b). This was to avoid raised intracranial pressure (ICP) in the postoperative period. In 4 patients, cranioplasty was carried out to cover occipital bone defect, either by removing the bone around the coronal suture (2) or using methyl methacrylate (2). Two patients who developed cardiac arrest during surgery were successfully revived and surgery was completed. Both patients had good postoperative recovery and were discharged within 7 days.

There were 2 postoperative deaths, both due to hypothermia during surgery, despite the use of a warming blanket. The remaining 12 patients had an average 7-day postoperative stay in the hospital. The duration of the stay ranged from 3 to 15 days. The 2 patients who required a postoperative VP shunt remained in hospital for 10 and 13 days, respectively. One patient with a wound infection required prolonged antibiotics and was discharged after 15 days. Follow-up ranged from 6 months to 6 years. The patient with an anterior fontanel encephalocele required two shunt revisions in the first 2 years. However, his mental development was poor when last followed up at the age of 6 years. One patient with occipital encephalocele who had VP shunt required shunt revision 4 years after surgery. Overall, mental development was satisfactory in 9 and poor in 3 patients. Thus, 33% of patients had a significantly poor mental development. A clinical profile of all patients can be seen in table 1.

Discussion

Encephaloceles are not uncommon; however, giant encephalocele, where the encephalocele reaches a large volume, is rare [1, 3, 4, 6]. These conditions can be termed massive encephalocele [6], large encephalocele [1, 7] and giant encephalocele [2–5], which of course mean the same thing with similar challenges [1, 2, 4, 7]. They pose a great

Fig. 5. **a** Intraoperative photograph of a case of occipital encephalocele showing flap dissection. **b** Intraoperative photograph showing dissection of sac containing occipital giant encephalocele. **c** Intraoperative photograph of occipital giant encephalocele after excision of sac.

Table 1. Clinical profile of the patients

No.	Name of patient, age, year of admission	Site of encephalocele	CT; MRI	Associated finding	Surgery	Immediate result	Long-term follow-up
1	B.Z., 2 days, 2004	anterior fontanel	CT; MRI	hydrocephalus	VP shunt and repair	discharged 7 days; good	6-year follow-up: fits; mental retardation; 2 shunt revisions 2004–2006
2	Ans, 5 months, 2005	occipital anterior fontanel closed	CT	hydrocephalus; craniostenosis	excision repair; craniectomy; cranioplasty by autologous bone graft; VP shunt	discharged on 5th day; good	2½ years: mental retardation
3	Aman, 26 days, 2002	occipital	CT; small occipital bone defect	no brain tissue inside the sac; hydrocephalus	excision; 2nd surgery; VP shunt	postoperative ventilated; shunt on 7th day; discharged on postoperative day 14	good outcome at 4 years
4	Asha, 3 months, 2006	occipital	CT; MRI	no hydrocephalus; craniostenosis	craniectomy; excision; cranioplasty by methyl methacrylate	discharged on 7th day; good	good outcome at 3 years
5	K.S., 10 days, 2003	occipital	MRI	no hydrocephalus	excision and repair	expired due to hypothermia	patient expired 1st postoperative day
6	S.S., 9 months, 2003	occipital	CT; MRI	polygyria; microgyria craniostenosis; no hydrocephalus	excision and repair; craniectomy; cranioplasty by autologous bone	postoperative stay 10 days	5-year follow-up: poor mental state
7	Neetin, 2½ months, 2004	occipital	CT; small occipital defect	hydrocephalus; no brain tissue in sac	VP shunt; excision and repair	discharged on 5th day; good	good outcome at 4-year follow-up
8	S.M., 4 months, 2005	occipital	CT	no hydrocephalus; craniostenosis	excision and repair; craniectomy	discharged on 5th postoperative day; good	3-year follow-up: good condition
9	S.G., 3 months, 2002	occipital	CT; MRI	hydrocephalus	VP shunt; repair of encephalocele	discharged on 7th postoperative day; good	patient was well; shunt block 2006; VP shunt revised; good mental development
10	K.S., 1 month, 2007	occipital	CT; MRI	hydrocephalus	VP shunt; excision and repair	discharged on 3rd day	good at 2-year follow-up
11	B.S., 1 year, 2008	occipital	CT; MRI	hydrocephalus	excision and repair; postoperative shunt 4th day	discharged on 12th day	1-year follow-up: good
12	S.B., 4 years 2008	occipital	CT; MRI	no hydrocephalus	repair of encephalocele	discharged on 8th day	good condition at 1 year
13	S.F., 4 months, 2009	occipital	CT; MRI	microcephaly; craniostenosis; small bone defect; no hydrocephalus	excision and repair; craniectomy	discharged on 3rd day	6-month follow-up: good normal development so far
14	R.K., 20 days, 2009	occipital	CT; MRI	microcephaly; no hydrocephalus	excision repair, patient had intraoperative hypothermia	ventilated; died on 1st postoperative day	expired 24 h following surgery

Fig. 6. a Postoperative clinical photograph showing occipital wound following excision and repair of giant occipital encephalocele. **b** Bicoronal skin closure after craniectomy in a patient with giant occipital encephalocele with craniostenosis.

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challenge to the neurosurgeon, neuroradiologist and neuroanesthetist during surgery. This series highlights the clinical features and management problems of these patients.

The exact incidence of giant encephalocele is not known. The literature only provides case reports [1–3, 7]. This is the only series of giant encephaloceles analyzed in the world literature so far apart from one previous series published by our institute in abstract form [5]. The current study includes 14 patients with giant encephaloceles, amongst a series of 110 patients with encephaloceles, treated during the period 2002–2009. The vast majority of patients (11/14) were under 6 months, and 4 were neonates. Only 2 patients were over 1 year of age. By and large these patients typically present as neonates and infants [3, 4, 6] as the external swelling produces nursing and feeding problems. One of our patients was a 2-day-old baby admitted with a leaking anterior fontanel encephalocele [3]. Generally, the patient's relatives seek advice at birth; however, as the experience is limited, proper advice cannot be offered; hence many patients do not present either at birth or during the neonatal period. However, by the time the baby is between 3 and 6 months of age, they attend our institute having taken advice from several doctors and even neurosurgeons. The relatives seek dedicated pediatric neurosurgical and neuroanesthetic care, which is offered at our institute.

2 patients had a history of CSF leak and 4 had microcephaly as the coronal sutures were fused. One patient also had a cleft lip, which was repaired before the patient was referred to the neurosurgery unit.

MRI is the most important modality of imaging; however, most of the patients were already investigated prior to coming to our center. In this study, both MRI and CT were performed on 8 patients, CT scan only was carried out on 5 and MRI only was done on 1 patient. It is important to understand that CT not only gives radiation risk to the neonate, but it has less value in delineating soft tissue anomalies like micro- and macrogyria and heterotopia. CT has the only distinct advantage of showing bone defects, which is the limitation in MRI and tentorial anomaly. The other reason why CT was performed before the patients came to us was that CT scans are less expensive, less time-consuming and easily accessible in small towns, where an MRI facility may not be available.

Large numbers of brain anomalies are associated with encephaloceles, especially with giant encephaloceles [3, 4, 6–9]. In cases of occipital giant encephaloceles, the evaluation of the transverse sinus and torcula are also important. In one of our giant occipital encephalocele, the venous sinuses and torcula were inside the encephalocele sac. This information helps the surgeon to plan the surgery well and handle the structures during surgery without damaging them. Chiari II malformation was present in 3 patients and 10 patients had associated hydrocephalus. In 3 patients MRI did not reveal any brain tissue inside the encephalocele sac.

Surgical repair is a great challenge in patients with low birth weight and with prolonged anesthesia [1, 2, 4, 7]. Intubating the patient is difficult with giant occipital encephaloceles. In 50% of these children partial aspiration of CSF was carried out to reduce the volume, which facilitated the intubation process during anesthesia. Seven

of our patients required VP shunt, 5 at the time of repair of encephalocele. In 2 patients postoperative shunt insertion was undertaken as the ventricle further dilated. Two of our patients had transient cardiac arrest during anesthesia. Both patients were successfully revived, surgery was completed and there were no postoperative complications.

During the repair of encephalocele it is important to make a judgement to excise the brain partially or to try to return all the contents into the intracranial cavity. Sometimes the head is small and the volume of the herniated brain is large so that it is not possible to close the sac without excising the brain tissue. In 7 (50%) patients a significant amount of herniated brain (50–70%) was excised to achieve a lax brain, at the time of closure. Some authors have described closure without excising the brain by providing expansible cranioplasty [1]. In the present study, craniectomy performed around the coronal suture and an anterior fontanel was created in 5 patients to avoid postoperative raised ICP, even though VP shunt was carried out. The management of postoperative raised ICP in a newborn is unpredictable and the baby can develop sudden respiratory arrest. In 4 patients cranioplasty was carried out at the site of the occipital bone defect. In 2 of these cases an autologous bone graft was taken from the region of the anterior fontanel, and in the remaining 2 a methyl methacrylate mould was made to cover the bone defect, to protect the brain and to prevent reherniation. This

method helped the patients and none of them had postoperative or long-term problems.

Intraoperative blood loss, hypothermia, bradycardia and cardiac arrest are well known [2, 4, 7]. Two of our patients had cardiac arrest; however, both did well and were discharged within 7 days. Two patients with severe hypothermia approaching 32°C did not recover and expired within 24 h. One of our patients had a wound infection and remained in hospital for 2 weeks postoperatively. At long-term follow-up, 66% of the patients had good mental function.

Conclusions

Giant encephaloceles are rare and pose a unique challenge. The present study is the largest series in the English literature. With dedicated effort good results are likely in 65–70% of cases. We recommend early surgery to avoid rupture or skin excoriation. MRI is the investigation of choice, which can delineate all intracranial anomalies. Large encephaloceles can be aspirated under anesthesia to facilitate intubation and the positioning of the patient in the operating room. A liberal craniectomy is ideal in patients with secondary craniostenosis to avoid postoperatively raised ICP. Judicious management of intraoperative blood loss and hypothermia are the keys to success.

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