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Management of the giant occipital encephaloceles in the neonates

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1. Introduction

Encephalocele is a congenital anomaly characterized by the herniation of the cranial contents through a bony defect in cranium. The incidence of encephalocele is 1–4 cases per 10.000 live births [1–3]. Approximately 75% of the encephaloceles are located in the occipital region [1,4]. Occipital encephalocele are described as giant when they are larger than the head from which they arise [2,5–7]. Giant occipital encephalocele is a rare clinical condition and the exact incidence of this pathology is not known. Herein we present four female neonates to highlight the difficulties in the management of giant occipital encephalocele. Preoperative management, surgical repair, postoperative management and follow-up for this disease are also discussed.

2. Materials and methods

This was a prospective and observational study in the level III neonatal intensive care unit (NICU) at Tepecik Research and Training Hospital, over an 3-years period, from November 2012 to January 2016. Our NICU unit is internationatal destination for the treatment rare and complex medical conditions in neonates. Our recognized team of physicians, fellows, surgeons, and nurses offers state-of-the art care and an outstanding record of success in nearly 100 clinical specialties. Each year, >10,000 births occurs and nearly 1500 neonates admitted to our NICU. During this study period, a total of four giant occipital encephaloceles were admitted to our hospital. The clinical symptoms, radiological features, operative approaches, preoperative care, intraoperative findings, postoperative management and prognosis were noted prospectively. These patients were diagnosed with giant occipital encephalocele based on their clinical findings and cranial magnetic resonance imaging (MRI) study. All cases were surgically excised and diagnoses were confirmed by surgery. Children were assessed by the Denver Development Screening Test-II (DDST-II) for psychomotor delay [8]. The DDST-II evaluates four areas: personal-social, fine motor, gross motor, and language. The developmental outcome scores are set for these four skills and total development. Scores between age-normative values and 20% of those values were considered near-normal. Scores between 20% and 30% of age-normative were considered as borderline. Scores higher than age-normative values (>30%) indicate the presence of a significant delay [8]. Head circumference, cranial nerve function, muscle strength, muscle tone, coordination, posture, and reflexes were evaluated during the neurological examination. Motor development was compared to age-normative values. Neurological functions were classified as normal, mild disability (the presence of an abnormality on examination, which does not lead to a significant impairment in function), moderate disability (abnormality, which leads to functional impairment), and severe disability (severe functional impairment requiring special assistance at all times) [9].

3. Results

There were total of four patients who underwent resection of their giant occipital encephalocele during this period. The clinical, demographical, radiological and surgical data obtained and all cases are presented in Table 1. Also, the details are summarised in below.

The procedure was performed on four females. The mean gestational age of the neonates was 37.2 weeks (3 terms, 1 preterm). The mean birth weight was 3235 g (from 2760 to 3680 g). The mean size in four giant occipital encephaloceles was 16.5 × 20.5 cm. All the patients had imaging by MRI. Imaging revealed the content of the giant occipital encephalocele (Case 1, Case 2, Case 3), and in one patient there was no brain tissue inside the sac (Case 4). Hydrocephaly was present in one patient (Case 4) at the time of diagnosis. MR angiography (MRA) and MR venography (MRV) were performed to evaluate the cerebral arterial or venous anomalies for our Cases 1 and 4. In our Case 1, MRA showed a posterior cerebellar artery and basilar artery into the defect.

All neonates underwent repair of the giant occipital encephalocele. In one neonate (Case 4), where there was no cerebral and cerebellar tissue, repair was closure after excising the sac. In one neonate (Case 1), posterior cerebral artery and basilar artery were into the encephalocele sac, which was preserved and gently pushed forward. In this neonate, the herniating brain was partially excised and the rest was reposited

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Table 1
Reported four cases with giant occipital encephaloceles.

No	Name of patient, gestational age, sex	Birth weight (gram)	Size of sac (cm)	Radiology	Associated findings	Time and technics of surgery	Operative time (minutes)	Length of hospital (number of days)	Long term follow-up
1	BBU, 38 weeks, female	3500	20 × 24	MRI, MRA, MRV	Herniation of the occipital lobe, occipital horn of the lateral ventricle, cerebellar vermis, cerebellar tissue, posterior cerebellar artery and basilar artery into the sac, corpus callosum agenesis, no hydrocephaly	7 days, intubation via lateral position, excision repair, duraplasty with galeal graft, no cranioplasty	165	13	40 months, motor and mental retardation, visual disability, good cosmetic result
2	FBİ, 38 weeks, female	3680	16 × 20	MRI	Herniation of the occipital lobe, no hydrocephaly	5 days, intubation via lateral position, preserved occipital lobe, duraplasty with galeal graft, no cranioplasty	105	7	34 months, normal motor and mental development, good cosmetic result
3	EBÖ, 38 weeks, female	3000	14 × 18	MRI	Herniation of the occipital and cerebellar lobe, dilatation of the posterior horns of lateral ventricles, below position of the tentorial incisura, thick tectum, no hydrocephaly	3 days, intubation via lateral position, preserved occipital and cerebellar lobe, duraplasty with galeal graft, no cranioplasty	140	16	30 months, normal mental and motor development, good cosmetic result
4	LBT, 35 weeks, female	2760	16 × 20	MRI, MRV	No brain tissue inside the sac, a patent right-deviating superior sagittal sinus, mesencephalic retraction, deformation of the fourth ventricle, dilated lateral ventricles	7 days, intubation via lateral position, duraplasty with galeal graft, no cranioplasty, tracheostomy (30 days), third ventriculostomy (50 days)	105	117	18 months, poor mental and motor development with tracheostomy, good cosmetic result

gently into the cranial cavity. Watertight dural closure was done with the galea. In the remaining 2 patients with giant occipital encephaloceles, the herniating occipital (Case 2) and cerebellar (Case 3) tissue were not resected but the herniating brain was pushed gently into the cranial cavity. In our cases, watertight dural closure was done with a galeal graft. Cranioplasty were not performed after excision of the encephalocele sac because of the small occipital bone defect.

The mean age of the patients at the time of surgery was 5 days, ranging from 3 days to 7 days. The mean length of the combined anesthesia and surgery procedure was 2 h and 11 min (from 1 h 45 min to 2 h 45 min). One neonate (Case 3) was required blood transfusion during surgery (20 ml). The mean hospital stay, determined by the neonatology and neurosurgery team, was 38.7 days (from 7 to 119 days), which also included the tracheostomy and third ventriculostomy for Case 4.

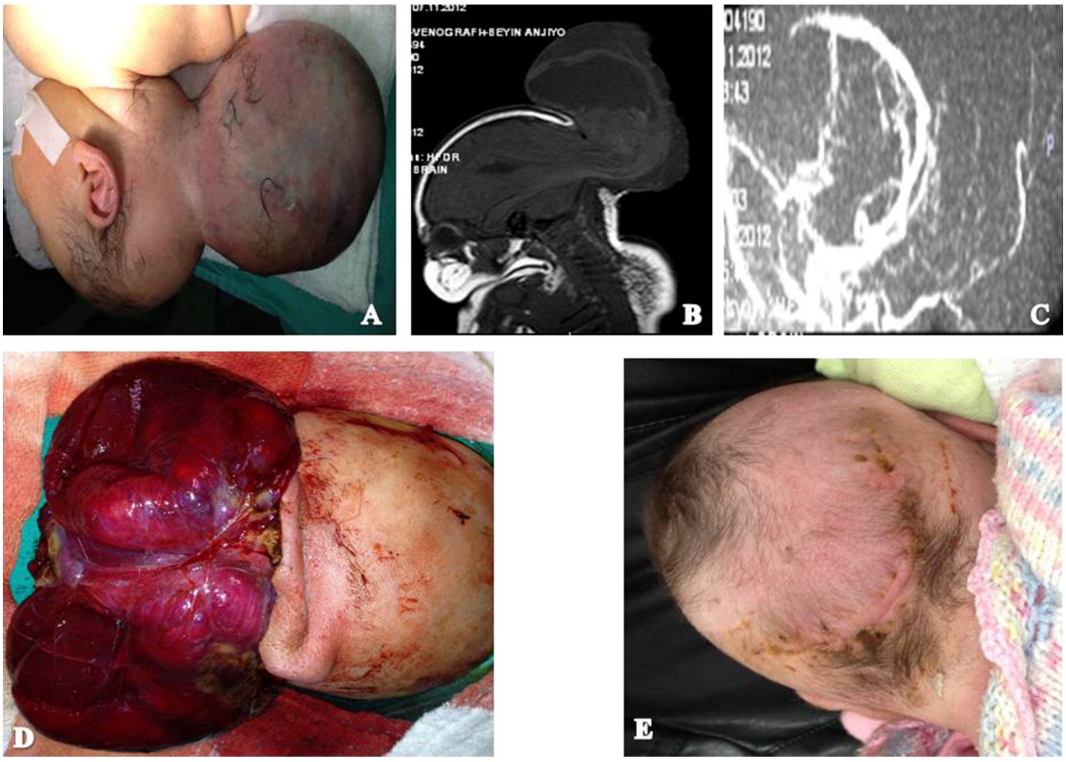

Fig. 1. A) Preoperative photograph of the Case 1 of giant occipital encephalocele, B) T_1-weighted sagittal MRI of the brain showing a herniation of the occipital lobe, occipital horn of the lateral ventricle, cerebellar vermis, dysmorphic cerebellar tissue into the sac, C) MRA revealing a posterior cerebellar artery and basilar artery into the defect, D) intraoperative photograph showing dissection of sac containing occipital giant encephalocele, E) clinical photograph of the child demonstrating a satisfactory cosmetic result.