

A Brief Review of Giant Occipital Encephalocele

An encephalocele is a congenital neural tube defect (NTD) characterized by herniation/protrusion of the intracranial contents (meninges, brain, and part of the ventricles) through a defect in the cranium.^[1] Encephalocele is caused by failure of the closure of the cranial part of the developing neural tube during the first few weeks of fetal life. An encephalocele is termed as “giant encephalocele” when the size of encephalocele is larger than the size of the head.^[2] Giant encephalocele is also termed as “massive encephalocele” and “large encephalocele.”^[3] Giant encephaloceles are most frequently observed at occipital region.^[2,3] Giant occipital encephalocele is a rare entity, and mostly reported as case reports.^[4,5] The internet search revealed two series published on giant occipital encephaloceles.^[3,6] Mahapatra published a series of 14 cases of giant encephaloceles, which included thirteen cases of giant occipital encephaloceles, and one case of anterior encephalocele.^[3] Ozdemir *et al.* recently published a series of four neonates who underwent surgical repair of giant occipital encephaloceles.^[6]

NTDs are among the most common of all birth defects in human and approximately 300,000 babies are born with NTDs globally each year.^[7] During the last few decades, there is significant decline in the incidence of NTDs worldwide; however, the incidence is still much higher in the developing countries.^[7] Encephalocele has an estimated prevalence of 0.8–5.0 per 10,000 live births.^[8,9] Occipital encephalocele is the most common of all encephaloceles.^[1,2] Depending on the anatomical location of encephalocele, it can be named as occipital, anterior, parietal, temporal, and vertex encephalocele. Encephaloceles are also classified based on their anatomical location of the skull defect.

Associated anomalies are reported to occur in 36%–60% of the infants born with encephaloceles, and include cranial and extracranial anomalies.^[1,3,8] Giant occipital encephalocele associated with Chiari malformation Type III is extremely rare entity.^[4,5] Chiari malformation Type III is characterized by the herniation of the posterior fossa contents, including the cerebellum, brain stem, and fourth ventricle, and in some cases, the upper cervical spinal cord through a low occipital and/or upper cervical osseous defect.^[4] Bulut *et al.* found approximately 30 published cases of Chiari Type III malformation, to which few more cases have been added in literature.^[4,5]

Clinically, giant occipital encephaloceles are obvious and suspected due to the characteristic swelling. The

size of the encephalocele may vary from few centimeters to a giant swelling. The clinical examination consists of examination of the encephalocele/swelling (size, extent, location), size of bony defect, size of the head for clinical suspicion of hydrocephalus or microcephaly, associated cranial, and extracranial anomalies.^[3-5]

Investigations to detect NTDs including encephaloceles during antenatal period are; antenatal fetal sonography, antenatal fetal magnetic resonance imaging (MRI), and measurement of maternal serum alpha-fetoprotein. During antenatal fetal sonography, most of the cases of encephaloceles are easily diagnosed on cranial views with brain protruding through cranial defect. Fetal MRI serves as an accurate tool to delineate not only the protruded encephalocele, but also the fetal brain, and associated cranial anomalies.^[5,8]

Postnatally, computed tomography scan, and MRI of the head including the encephalocele are the investigations needed for the evaluation of occipital encephaloceles and associated intracranial anomalies (hydrocephalus and other brain abnormalities), although MRI of the head is preferred investigation.^[1-6] If there is an additional-associated anomalies suspected or detected during clinical examination, other investigations are also required. Magnetic resonance angiography and magnetic resonance venography are the additional radiological investigations required in selected cases to delineate cerebral vascular pattern.^[5,6]

In general, most of the prenatally diagnosed neurosurgical anomalies/malformations are best managed after birth by an appropriate medical and surgical therapy. Only few of the neurosurgical anomalies may require and benefit by antenatal fetal intervention. The commonest neurosurgical anomaly treated during antenatal period with a high success rate is myelomeningocele.^[9] The limited number of fetal repair of encephaloceles has also been reported in the literature.^[9]

Surgical procedures for giant occipital encephalocele are a challenging task, not only for neurosurgeons, but also for the anesthesiologists. These challenges are due to its site, large size, contents, associated intracranial anomalies, intraoperative blood loss, intraoperative hypothermia, and prolonged anesthesia.^[3-6,10] The problems faced by anesthesiologists during anesthesia management of patients with giant occipital encephaloceles are difficult tracheal intubation, instability in hemodynamic, excess blood loss, intraoperative hypothermia, and cardiorespiratory complications.^[10] Occipital encephaloceles are best

operated in the prone position. Partial aspiration of cerebrospinal fluid (CSF) is also a better option to reduce volume of giant occipital encephalocele to facilitate the endotracheal intubation.^[3]

In general, operative repair for giant occipital encephalocele consists of excision of the excess meninges, partial or complete excision of protruded brain tissue, watertight closure of the dural defect and approximation of the skin.^[3-6] Mahapatra reported that five of thirteen cases of giant occipital encephaloceles also had associated craniostenosis; and therefore, craniectomy and/or cranioplasty were also required.^[3]

Associated hydrocephalus is frequently present with giant occipital encephaloceles or developed following the surgical repair of it, and CSF diversion in the form of ventriculoperitoneal shunt is also an additional surgical procedure required.^[3,5] Complications observed following repair of giant occipital encephaloceles are CSF leak, postoperative meningitis, wound infection, wound dehiscence, and hydrocephalus.^[3,5] Mortalities have also been reported following surgical repair of giant occipital encephaloceles.^[3]

The prognosis of the children treated for giant occipital encephaloceles depends on many factors and includes the size, amount of brain tissue involved in the encephalocele, associated intracranial (microcephaly and hydrocephalus), and extracranial anomalies, and the treatment offered for the correction of the disease.^[3,6] Associated hydrocephalus and microcephaly carries a significant morbidity in these children. A significant proportion of these children lack normal developmental milestones, and also has neurological deficits.

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