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Original Research Article

Outcome of surgery in Occipital Encephalocele in tertiary care hospital

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Abstract: The aim is to analyse the factors which determine prognosis and assess the outcome of surgery in occipital encepahlocoele. 20 Patients of occipital encephalocoele presented in our Paediatric surgery unit in tertiary care centre in metropolitan city in India from 2008 to 2016 were included in this study. Follow-up period ranged from 3month to 9 years (Mean 3 yr 5months) for different patients. Various prognostic factors such as age at presentation and surgery, size of bony defect, contents of the sac, associated anomalies, need for Ventriculoperitoneal shunt insertion were taken into consideration. Outcome was assessed in form of mortality, wound infections, ventriculitis, shunt complications, optic atrophy and delayed milestones. Out of 20 patients presented over 9 years, 2(10%) couldn't survive till surgical repair while 3 (15 %) expired postoperatively 1 of which had documented shunt infection as cause of death. Most of the repeated admissions (in 7 of 11 who required VP shunt) were designated to shunt complications while 4 of 20 had wound infection 1 of which died and 1 had associated shunt infection as well. In Occipital encephalocoele, factors such as age at presentation, size of bony defect, contents of the sac and associated anomalies such as microcephaly, hydrocephalus, wound infection and shunt complications played major role in overall outcome.
Keywords: Occipital encephalocoele, wound infection, microcephaly.

INTRODUCTION:

Occipital encephalocoele (OE) is defined as congenital defect in cranium and dura leading to protrusion of brain substance and meninges through it in the occipital region. Occipital encephalocoele is labeled when defect is between foramen magnum and lambda while occipitocervical encephalocoele is labeled when defect involves posterior arches of first 2 cervical vertebrae [1].

Though etiology is not proven yet, it is known to be due to primary abnormal mesodermal defect [2]. The contents of the encephalocoele may include cerebrospinal fluid (CSF), meningeal structures, or brain tissue [3].

The treatment of encephalocele defects requires immediate surgical closure. During surgical intervention, the neural tissue in the sac is excised and the dura mater is restored. Generally, there is no need to restore the bony defect [4].

MATERIALS AND METHODS:

All patients of occipital encephalocoele were searched from the hospital records retrospectively from 2008 to 2016 presented in our Paediatric surgery unit in tertiary care center in metropolitan city in India. The parents were contacted and called for thorough neurodevelopmental and ophthalmological evaluation of the children. The clinical, radiological as well as surgical data were collected and summarized.

At presentation, all the patients were worked up with thorough clinical examination, ultrasound of the skull, fundoscopy and anesthesia evaluation for surgery. Except for 2 deaths before the scheduled surgery, all patients had undergone surgical repair and excision of gliotic contents of the sac followed by primary watertight closure of the dura and layered wound closure thereafter.

Patients with documented hydrocephalus on ultrasound were subjected to ventriculoperitoneal (VP) shunt procedure postoperatively after 5-7 days of the first procedure. Postoperative ophthalmic evaluation was not done in all patients. For documentation purpose of the study, it was done at the time of current follow up.

RESULTS:

1. Gender

Out of total 20patients, 18 (90%) were pure occipital while 2 (10%) was occipitocervical encephalocoele. 14 (70%) were girls while the rest (6 i.e.30%) were boys.

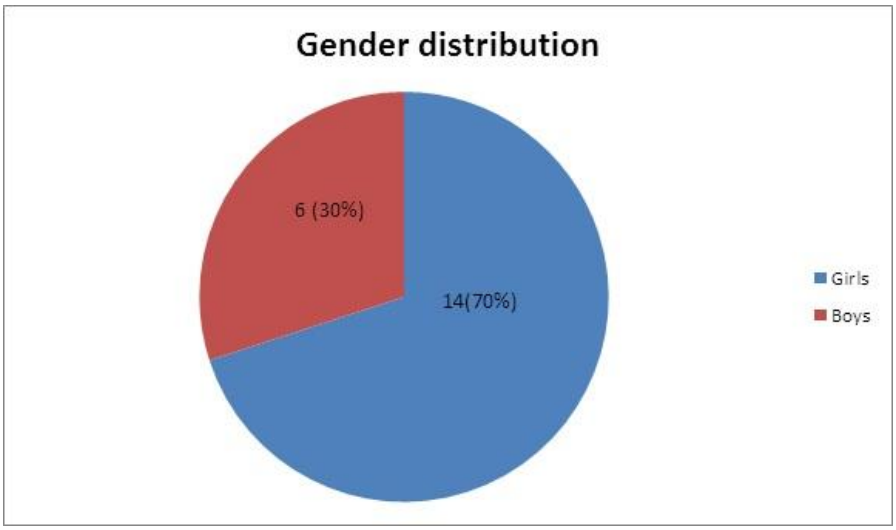

Fig –1. Gender distribution

2. Associated anomalies

Associated malformations of CNS or other systems didn't show major effect on survival. Anomalies such as microcephaly, cerebellar/occipital lobe hypoplasia per say didn't affect survival except for a death in microcephaly patient due to shunt complication. One patient had cognitive impairment in associated bilateral hypodense cerebral hemispheres and cerebellar hypoplasia. A patient with associated cardiac malformation (ASD and PDA) was operated for PDA (Clipping done) and has survived till date i.e. at 3 years age. Intracystic hemorrhage patient didn't need VP shunt in postoperative period and survived till date i.e. 8 years age. Day 5 presenter with ruptured encephalocoele sac and open meningomyelocoele didn't survive beyond 2days further and couldn't be operated.

One patient with associated Dandy Walker syndrome who had late presentation at 9 months and had shunt infection within a month of shunt procedure had died at 1 year of age due to failure to thrive and pneumonia.

3. Age at presentation and mortality

13 (65 %) presented within first 5 days of life and all were operated within 1 to 7 days of admission. 2 (15.3%) of these 13 early presenters died preoperatively while 2 (15.3%) operated patients died later in life. Rest 7 (35%) presented beyond 5 days ranging from 12 days to 2years. 1 (14.2% of late presenters) patient who had presented at 9 months died at 1 year due to failure to thrive and pneumonia.

Age at presentation	Number of patients	Pre/Intra operative deaths	Postoperative deaths
Early (0-5 days)	13	2	2
Late (12 days – 2years)	7	0	1

Fig –2. Relation of age at presentation to mortality

4. Infection

4 (22.2%)patients had evidence of wound infection. 2 of these infected patients had presented on DOL 1 and 2 and operated on DOL 5 each including 1 who required debridement of the wound on POD 6 and had evidence of ventriculitis.

The other 2 had presented on DOL12 and 25 and operated on DOL 13 and 30 respectively. The second of these late presenters had evidence of ruptured sac and showed internal echoes in USG suggestive of ventriculitis preoperatively.

Out of 11(61.1%) patients who required VP shunt procedure for Pre surgery or post-surgery hydrocephalus, 2 (18.2%) developed shunt infection. This included 1 patient who had wound infection requiring local debridement.

5. Shunt complication

Out of 11 patients who required VP shunt procedure, 4(36.4%) had shunt complications apart from pure infection. These complications were- Shunt disconnection, shunt block, abdominal end migration per anus, sub-acute intestinal obstruction, etc. 2 of these died later 1 death being assigned to shunt infection.

6. Optic atrophy

3 (15%) patients had obvious optic atrophy preoperatively, 2 of these died before follow up. 1 had mildly pale optic disc (could be due to early optic atrophy) during preoperative examination. Head frank atrophy in follow up evaluation. Postoperatively no patient developed new finding of optic atrophy.

7. Development

Motor delay was found in 5 (27.8%) of 18 patients whose data could be collected at the current follow up or retrieved from records for the ones who expired. Verbal delay was found in 7(41.1%) of 17 patients with available data. All of these also had cognitive impairment along with 3 more (totally 10 i.e. 58.8%) who didn't have motor/ verbal milestone delay.

DISCUSSION:

Encephalocele is herniation of cerebral tissue, meninges, and cerebrospinal fluid (CSF) outside the confines of the skull. Although encephalocele may represent defective closure of the neural tube in gross pathology, it does not appear to occur through defective neurulation. The most widely accepted theory is derived from that of Etienne Geoffroy Sain Hilaire [5] (1827), the French naturalist, which states that encephalocele is caused by an error in mesodermal differentiation. The paraxial mesoderm, from which the meninges and skull will form, migrates in between the ectodermal layers. In cases of encephalocele formation, a neuroschisis (fissure) develops after primary neurulation, which leads to scarring and subsequent adhesion between the cutaneous and the neuroectoderm and prevents interposition of the mesoderm.

Occipital encephaloceles vary in their appearance, size, and contents, from small, atretic lesions with little or no brain elements to large lesions that may have portions of functional cerebellum, cortex, and brainstem contained within them. Nearly 75% of encephaloceles are located in the occipital region [2]. Associated central nervous system findings in patients with occipital encephaloceles include hydrocephalus, kinking of the brainstem, and an absent, rudimentary, or inverted cerebellum with the brainstem herniated posteriorly and the cerebellar vermis ventrally. A small posterior fossa or posterior fossa cysts reminiscent of a Dandy-Walker variant, along with abnormal displacement of the torcular, transverse sinuses, and tentorium, may be seen. Cortical dysplasia and callosal agenesis are frequently present.

OE is more common in girls. Our study showed Female: Male ratio of 2.3:1 in coherence with previously published data which says 70% of occipital encephaloceles occur in females [8].

Age at presentation in unruptured sac didn't show any difference in mortality in our study. According to the literature, repair of encephaloceles without ongoing CSF leak is an elective procedure. It should however be done as early as possible for all defects, depending on the anesthesia expertise available. A death in patient presented at DOL5 from tribal area in our state with ruptured large occipital encephalocoele and lumbosacral meningomyelocoele which was not noticed by treating general practitioner in that remote area, probably was preventable if had been referred earlier to a tertiary care centre.

61 % of our OE patients required VP shunt procedure for hydrocephalus. As per literature, hydrocephalus develops in 60% [7], 36% [9], 16%[10], 50% [11], 65%[12] in different studies of the patients with all types of encephaloceles [7].

Shunt complications such as infection, disconnection, migration, block, need for bilateral shunt were found in 5(45%) of shunt cases amounting to death in a patient. A large study conducted in California including patients over 11years showed cumulative complication rate of 32% at 5 year follow up [13].

Developmental milestones were evaluated in our study under 3 categories- motor, verbal and cognitive. Very few studies have evaluated neurodevelopment in operated OE patients. In a study by French *et al*. published in the year 2007, 17% of patients with encephaloceles had normal development, while severe mental retardation and physical delay was reported in 83% of the patients [6]. In contrast to this observation, to our surprise, our study showed only 27.8% motor delay, 41.1% verbal delay and 58.8 % cognitive impairment in which scholastic performance was taken into account in older children.

Thus, in a developing country, surgical management of a dreaded congenital malformation like occipital encephalocoele is studied in detail in this study. The results are promising and motivating to continue operating in a well formed setup with good anaesthesia backup.

CONCLUSION:

In Occipital encephalocoele, factors such as age at presentation, size of bony defect, contents of the sac and associated anomalies such as microcephaly, hydrocephalus, wound infection and shunt complications played major role in overall outcome. The overall outcome of this relatively large data was good.

REFERENCES

1. Naidich TP, Altman NR, Braffman BH, McLone DG, Zimmerman RA. Cephaloceles and related malformations. American journal of neuroradiology. 1992 Mar 1; 13(2):655-90.
2. Raja RA, Qureshi AA, Memon AR, Ali H, Dev VPattern of encephaloceles: a case series.J Ayub Med Coll Abbottabad 2008; 20: 125–128.
3. Bozinov O, Tirakotai W, Sure U, Bertalanffy H Surgical closure and reconstruction of large occipital encephalocoele without parenchymal excision Childs nerv syst 2005;21: 144-147
4. Nejmi Kıymaz, Nebi Yılmaz, İsmail Demir, Sıddık Keskin Prognostic Factors in Patients with Occipital Encephalocele Pediatr Neurosurg 2010;46:6–11
5. Saadi Ghatan Youman's textbook of Neurosurgery 6th Ed. Ch 176 p.1898
6. French BN Midline fusion defects and defects of formation; Youmans JR (ed): Neurological Surgery. Philadelphia, Saunders, 1990, pp 1164–1169.
7. Yasir Andrabi, Farideh Nijat,Mostafa El-Khashab Progressive skin necrosis of a huge occipital encephalocele Indian Journal of Plastic Surgery 41(1):82-4 · April 2008
8. Jeffrey,V., Rosenfeld and D.A.K. Watters. Neurosurgery in the tropics: A practical approach to common problems Macmillan Education Ltd, 2000
9. N. Munyi, D. Poenaru, R. Bransford and L. Albright Encephalocele – A single institution African experience East African Medical Journal Vol. 86 No. 2 February 2009
10. Shokunbi, T., Adeloye, A. and Olumid, A. Occipital encephaloceles in 57 Nigerian children: a retrospective analysis. Child's Nervous System 1990; 6:99-102.
11. Chapman, P.H., Swearingen, B. and Caviness, V.S. Subtorcular occipital encephaloceles. J. Neurosurg. 1989; 71: 375-381.
12. Lorber, J. and Schonfield, J.K. The prognosis of occipital encephalocele. Z Kinderchir. 1979; 28: 347- 351.
13. Wu Yvonne, Green Nella, Wrensch Margaret R., Zhao Shoji, Gupta Nalin Ventriculoperitoneal Shunt Complications in California: 1990 TO 2000 Neurosurgery, 2007; 61(3):557–563.