



Congenital Occipital Encephalocoele- technically challenging

SUKIRTI CHAUHAN

Department of Neuro Surgery, MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract : Introduction Neural tube defects in the brain present as encephalocoeles. It is basically a developmental defect where the developing brain and meninges covering the brain and spinal cord are supposed to close ideally but do not do so leading to their outpouching .. Occipital encephalocoeles herniate through a skull defect between the lambda and the foramen magnum.

Case report- A 1yr old boy baby with no history suggestive of any perinatal insult and having a soft tissue swelling at the back of the head since birth was brought to us in the neurosurgical outpatient department. It was a 5cm by 3 cm occipital encephalocoele. It had skin discoloration. It was not pulsating, not warm, not tender, no cough impulse. CT brain showed the contents of the mass as having both a soft tissue component and hyperdensity similar to CSF. It was in continuity to the brain and there was a well defined bony defect in the occiput bone around 2x2 cm. Magnetic resonance venogram was done and it was noted that the swelling was arising near the torcula but lack of continuity could not be ruled out. Treatment was by surgery. A vertical elliptical incision and 1cm craniotomy was done to enlarge the bony defect. Durotomy done at the base of the mass and clear CSF noted. Mass excised in toto and sent for histopathology examination and the dural defect closed. Post-operatively, baby had no neurological deficit and wound healed well with a pseudomeningocele for which a right ventriculo-peritoneal shunt was done. We present this case in view of the technically challenging surgery since the occipital encephalocoele was very close to the torcula as noted in the MRV. As neurosurgeons we were well prepared with adequate blood preoperatively.

Keyword : Occipital Encephalocoele, torcula, MRV

INTRODUCTION

Neural tube defects in the brain present as encephalocoeles. It is basically a developmental defect where the developing brain and meninges covering the brain and spinal cord are supposed to close ideally but do not do so leading to their outpouching. This outpouching in the cranial area is called the encephalocoele. Approximately 1 per 5,000 live births is the incidence of encephalocoeles (1)

in which occipital encephalocoeles account for almost 80% of the encephalocoeles. They are more commonly seen in the female child. Occipital encephalocoeles herniate through a skull defect between the lambda and the foramen magnum. There is a high propensity for associated anomalies of the nervous system apart from the occipital encephalocoele.

CASE REPORT

A 1yr old boy baby, first child of non-consanguineous parents born through normal vaginal delivery with no history suggestive of any perinatal insult and having a soft tissue swelling at the back of the head since birth was brought to us in the neurosurgical outpatient department. It



It was a 5cm long fleshy ellipsoid shape soft tissue mass with a diameter of around 3cm at the midline above the foramen magnum and just below the lambda. It has skin discoloration (reddish patches on mass and around the mass). It was not pulsating nor had any scars, sinuses or dilated veins on it. It was not warm, not tender, no cough impulse. Soft to firm in consistency.

Imaging showed herniation of the meninges with CT brain showing the contents of the mass as having both a soft tissue component and hyperdensity similar to CSF. Rest of the anatomical development of the brain appeared appropriate for the age of the patient. It was in continuity to the brain and there was a well defined bony defect in the occiput bone around 2x2 cm. Magnetic resonance venogram was done and it was noted that the swelling was arising near the torcula but lack of continuity could not be ruled out. The content of the occipital encephalocoele did not show any brain tissue but the minimal outpouching of the torcular herophili could not be ruled out. The rest of the brain appeared normal with no definitive anatomical deformity. There was no evidence of hydrocephalous.

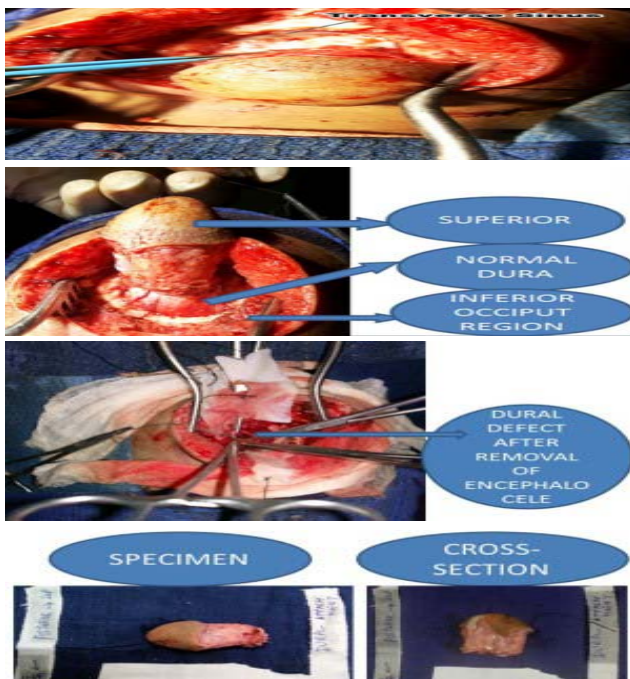


MRV



MRI

The condition of the patient was explained in detail to the parents. Treatment was by surgery. Patient was put in prone position under paediatric neuroanaesthesia care with proper fluid management and normothermia. A vertical elliptical incision was made and along with the adherent skin, the mass was delineated. 1cm craniotomy was done to enlarge the bony defect and normal dura noted all around it. Durotomy done at the base of the mass and clear CSF noted. Mass excised in toto and sent for histopathology examination and the dural defect closed.



Post-operatively, baby had no neurological deficit and wound healed well with a pseudomeningocele for which a right ventriculo-peritoneal shunt was done. Histopathological examination was unremarkable. Patient on one year follow up is normal with normal development of milestones.

DISCUSSION

Nearly about 50% of the encephalocoeles have associated congenital anomalies (2) ranging from common anomalies like corpus callosum agenesis, Dandy-Walker cyst, Chiari malformation, Klippel1. Feil anomaly (3) and iniencephaly to even tumours (4) in the occipital encephalocoele. We need to precisely assess the location of the occipital encephalocoele as a part of planning of the surgical procedure.

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities

Since the site of occipital encephalocoele can range from the lambdoid suture to the foramen magnum, the proximity to important vascular structure needs to be studied especially the torcula Herophili which is the site of confluence of sinuses. Magnetic Resonance Venogram helps in depicting the relationship of the torcula with the occipital encephalocoele and also the contents of the protruding mass. Another investigation, the Visual Evoked Response (VER) is useful to determine the functional visual cortex(5) in the occipital lobes if the content of the occipital encephalocoele is a part or whole of the occipital lobe. Hydrocephalous is more common in the posterior encephalocoele with almost 60% patients developing it after definitive surgical management. The contents of the occipital encephalocoele can range from occipital lobe, brainstem to rarer cases of cerebellum or torcula(6). Torcula as a content is more challenging in view of the fact that injury might lead to cerebral deep venous system thrombosis with devastating sequelae. This might require the operating team to be standby for a venous reconstruction(7) by vascular graft if the need arises. The reason for post op hydrocephalous is explained by the changes in cerebrospinal fluid dynamics after the excision of the occipital encephalocoele.

We present this case in view of the technically challenging surgery since the occipital encephalocoele was very close to the torcula as noted in the MRV. As neurosurgeons we were well prepared. The patient attenders need to be made well aware of the risks of operating near the torcula. As per literature there is scarcity of data telling good prognosis about surgeries involving the torcula and ending up with a mortality. We hence need to be prepared with adequate blood pre-operatively and appropriate corrective measures instrumentation in the event of a Torcula tear or damage needing a graft(8) or an attempted primary repair none of which have promising results. As advocated by Mahapatra et al. preservation of the neural and vascular elements is important. Walia et al.(9)suggested that although the gliosed and ischemic herniated neural tissues can be excised, greater caution is to be exercised to preserve the venous sinuses that course through it.

CONCLUSION

We present a common case of occipital encephalocoele giving primary importance to appropriate investigations of ruling out adhesion to vascular structures which is many at times not included in the basic investigation battery of tests of the resident neurosurgeon.

BIBLIOGRAPHY

- Mealey J. Jr, Dzenitis AJ, Hockey AA. The prognosis of encephalocoeles. J Neurosurg. 1970;32:209-18
- Brown MS, Sheridan-Pereira M: Outlook for the child with a cephalocoele. Pediatrics 1992; 90:914- 919
- Cohen MM Jr, Lemire RJ: Syndromes with cephalocoeles. Teratology 1982
- Rao SA, Rao VRK, Mandalam KR, Gupta AK, Kumar S, Joseph S: Corpus callosum lipoma with frontal encephalocoele. Neuroradiology 1990;32:50-52.
- Mahapatra AK, Agrawal D. Anterior encephalocoeles: A series of 103 cases over 32 years. J Clin Neurosci. 2006;13:536-9.
- Mahapatra AK. Skull base anatomy, radiology and management. In: Samii M, editor. Skull Base Anatomy, Radiology and Management. Basel: S Karger; 1994. Pp. 220-3.
- Sindou M. Meningiomas invading the sagittal or transverse sinuses, resection with venous reconstruction. J Clin Neurosci. 2001 May;8 Suppl 1:8-11.

Nagashima M, Kobayashi S, Takemae T, Tanaka Y. Total resection of torcular herophili hemangiopericytoma with radial artery graft:case report. *Neurosurgery*. 1995 May;36(5):1024-7.

Walia B, Bhargava BP, Shandu K. Giant occipital encephalocele. *MJAFI*. 2005;61:293-4

