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Case Report

Surgery Section

Congenital Interparietal Encephalocele: A Case Report

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ABSTRACT

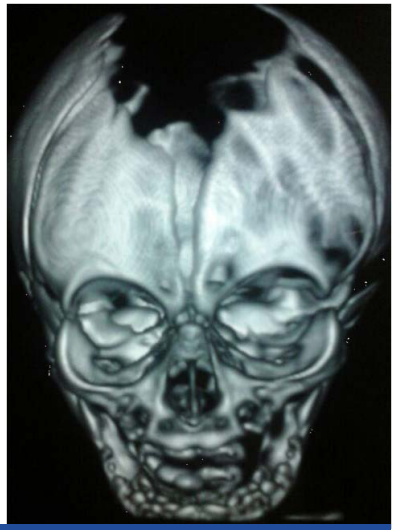
Encephalocele is a mesodermal defect in the skull bones and duramater. Parietal encephalocele is a rare congenital anomaly of newborn with variable prognostic value. The authors report a case of a very large inter parietal encephalocele with no associated other system malformations. A midline inter parietal encephalocele is much rare, earlier reported cases were posterior parietal in location. Such cases can be successfully operated upon with a very good outcome. A unique case of a 18 day neonate, with swelling over scalp was evaluated by the neurosurgical team and the patient underwent neurosurgery. In planning the strategy for management of encephalocele, one needs to take into consideration the site, size, contents, patency of CSF pathway, neurological status and other associated anomalies. Inspite of such a big encephalocele in an atypical location, excision and repair gave excellent results.

Keywords: Congenital anomaly, Parietal encephalocele, Vertex encephalocele

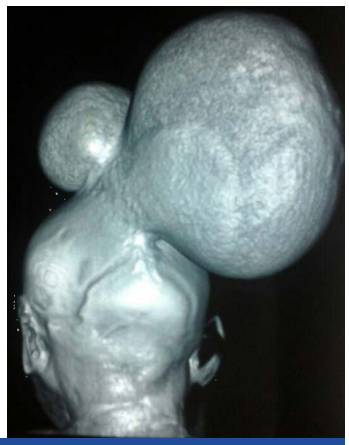
CASE REPORT

A neonate,18-day-old female baby presented with congenital scalp swelling and was evaluated by the neurosurgical team. Her mother belonged to a remote area where no prenatal ultrasonography was available; baby was delivered at a district hospital through LSCS. Scalp swelling progressively increased since birth. Complete physical examination of the baby revealed a congenital scalp swelling with no other associated congenital anomalies such as spine defect, limb defect; vertebral anal anomalies, trachea oesophageal anomalies, cardiac anomalies and renal anomalies. There was no hypertelorism or proptosis and fundus examination had no early signs of hydrocephalous. The swelling partially became lax when the patient was held erect and got tense on lying down the baby [Table/Fig-1].

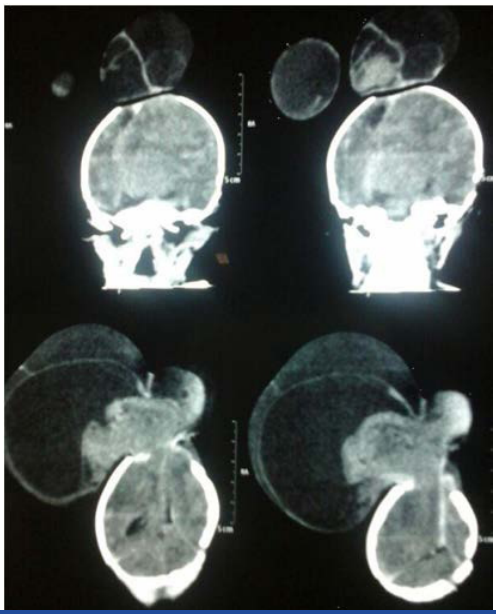
As shown in picture [Table/Fig-1] a cystic swelling of the size 9x7 inches with a daughter cyst of size 4x3 inches was located in midline, midway between frontal and occipital region. Size of the head as per age was normal with no hydrocephalous and other congenital anomaly. Detailed neurological examination and other systemic examination revealed no significant associated anomalies. Swelling was transilluminent and partly filled by aberrant brain tissue. 3D CT SCAN head showed a focal defect involving high parietal bone with herniation of CSF filled sac and neural tissue through this defect suggestive of encephalocele. There was forced sutural separation



[Table/Fig-1]: Neonate with cystic scalp swelling and a daughter cyst
[Table/Fig-2]: Antero-posterior view of skull showing a mid-line bony gap



[Table/Fig-3]: 3D CT scan of head antero-lateral view showing the location of defect
[Table/Fig-4]: 3D CT scan of head postero-lateral view showing the location of defect



[Table/Fig-5]: CT scan coronal sections showing the herniated contents of the cyst

in the midline and anterior fontanel was enlarged enormously due to the pedicle of the cyst [Table/Fig-2-5].

Excision and repair of the sac was done under general anaesthesia with prone position. Elliptical incision encircling the pedicel of the cyst was made and duramater was defined all around the sac. Sac was opened and about 800 cc dark brown fluid came out with pressure. Cyst wall was excised all around and aberrant atrophied brain tissue partially filling the cyst was excised. Superior sagittal sinus was aberrant, midline superior sagittal sinus could not be located during the surgical procedure, and probably it was obliterated or atrophied due to mass effect of tumor. Dural defect was meticulously repaired, to get a watertight closure. Child stood surgery well and post operatively child could move all four limbs without any gross neurological deficit. No ventilator support was needed post operatively. Post-operative recovery was uneventful till 4th postoperative day.

DISCUSSION

Encephalocele usually occur at occipital and anterior region while mid interparietal location is very rare [1]. Among vertex or midline parietal encephalocels, the atritic form is more commonly seen than large size encephalocele [2].Large case series on anterior encephalocele and occipital encephaloceles are present in literature [3,4]. However, large encephaloceles located in midline interparietal region are limited [5-8].

Management of encephaloceles depends on type, size and hydrocephalous associated with encephalocele. Surgical treatment is excision of sac and repair of dural defect after replacing the viable healthy brain tissue into cranial cavity.

The end result of encephalocele surgery is usually not determined by the neurosurgical procedure per se, but by the underlying brain involvement and presence or absence of other congenital defects [9].Postoperative complication that may be noted is hydrocephalous, which may not be apparent preoperatively can become apparent after the repair of an encephalocele. Giant occipital encephaloceles are associated with more underlying malformations and have poor prognosis [10-12].Parietal encephaloceles associated with hind brain malformation have poor prognosis than those not associated with brain malformations [2,6].

CONCLUSION

Among encephaloceles, inter parietal encephaloceles represents rare cases. CT image morphology is required to make a diagnosis of the underlying brain malformations.

In planning the strategy for the management of encephalocele, one need to take into consideration the site, size, contents, patency of CSF pathway, neurological status and other associated anomalies. Timely surgical intervention, as in our case has a good outcome. In spite of such a big size encephalocele excision and repair gave very good results, child was moving all four limbs and reflexes were intact. Postoperative follow up should be done at regular interval to look for hydrocephalous or other neurological deficit. Postoperatively this case doesn't revealed significant neurofocal deficit however, left sided lateral rectus paresis was noted on 6 month follow up.

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