LATERAL ENCEPHALOCELE

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Introduction

Protrusion of the meninges or cerebral matter through defects in the calvarium located in a lateral position is not common. Most of the cases of cranio-spinal dysraphism have a posterior mid line defect, the majority being lumbo-sacral. Our case is an example of lateral cranial meningocele which is rare. We came across only two case reports of lateral frontal encephalocele in literature [1].

Case Report

Baby Guddi, a two months old infant, was brought by her parents with a history of having a swelling on the left side of her head since birth. The swelling had gradually been growing with the child. It was as if the child had two heads! On examination there was a cystic swelling 12 x 6 x 4 cm in the region of the left temple extending on the frontal region anteriorly and the parietal region posteriorly [Fig 1]. The swelling was fluctuant, transilluminant and conducted an impulse on crying. Clinically a diagnosis of an encephalocele was made. An ultrasound confirmed the cystic nature of the swelling.

The infant was taken up for surgery. An elliptical incision was made along the superior margin of the cyst which was dissected free of the surrounding skin. At the base was a narrow neck protruding through a small defect between the frontal and temporal bones. There were no neural elements forming the contents of the

cyst. The neck was clamped and the cyst excised. This was followed by water tight closure of the dura. Post operatively the wound healed well (Fig 2). CSF analysis collected per-operatively revealed clear fluid with WBC 6/mm³; Proteins 510 mg%; Sugar 67 mg% and chloride 108 meq/L. This closely resembled the analysis of the fluid from the cyst which showed WBC 8/mm³; Proteins 470 mg%; Sugar 23 mg% and chloride 115 meq/L.

Histopathology revealed a lining of flattened to cuboidal epithelium, with papillations at a few places. The wall was composed of connective tissue.

Discussion

The sequence of events in human CNS ontogenesis follow a complex pattern which starts at around the 18th day of gestation with the formation of neural plate and carries on till early post-natal period when neuronal migration, axosomatic and axo-dendritic synaptic connections take place. Interwoven in this neural sequence is the development of the vertebral column and the skull from the mesoderm. Cranio-spinal dysraphic states can be due to failure of fusion of bone in the region of the vertebral column-spina bifida (occulta, cystica or aperta) or due to improper development of the skull-cranium bifidum. The incidence of the latter condition is about one-tenth that of the for-

Fig. 1 : Posterior view of giant lateral temporal encephalocele

Fig. 2 : Photograph after removal of the lateral encephalocele

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mer. Our case is one of cranium bifidum. In such cases the lesions, regardless of whether they are a meningocele or contain neural tissue, are usually classified together as 'encephalocoeles'. In western countries the lesion in cranium bifidum involves the occipital bone in the midline posteriorly in 85% of cases. In Asia, the majority of encephalocoeles are anterior and involve the frontal or nasal bones, but still in the midline. In our case the involvement was lateral, which is rare. Matson [2] in his celebrated series of 1396 cases of spinal dysraphism, found an incidence of 0.3% for lateral meningoceles; and of these, almost all were lumbo-sacral. From India, Sharma et al [1] have reported a series of 619 cases of spinal dysraphism. They have been able to trace 10 cases of lateral cervical meningocele and only 2 of lateral frontal meningocele in world literature. In 'Lehman syndrome' multiple lateral meningoceles are associated with presence of wormian bones, malar hypoplasia, downslanted palpebral fissures, high narrow palate and cryptorchidism [3]. However, in our case no other

associated anomaly was clinically detected.

Defective calvarial ossification can result in encephalocoeles presenting at atypical sites. An anterior basal encephalocoele obstructing the nares can be mistaken for a nasal polyp and removal attempted. In our case the calvarial defect was between the frontal and temporal bones. A high index of suspicion that a swelling at the ectopic sites could be an encephalocoele must be maintained and an awareness of such a lesion will avoid a sense of bewilderment and proper management will lead to a normal development of the infant.

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