Case Report

Double Encephalocele: An Unusual Presentation

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The occurrence of multiple neural tube defects in a single patient is rare; the cases reported in the past are mostly of thoracic and lumbosacral region. Double occipital encephaloceles are rare; only four cases have been reported till date. Here we report a case of double encephalocele—one in the vertex and the other in the occiput, and a literature study on similar reports.

Keywords: Double encephalocele, hydrocephalus, neural tube defects

INTRODUCTION

Encephalocele is the protrusion of the cranial contents beyond the normal confines of the skull through a defect in the calvarium. The presence of multiple neural tube defects (NTDs) along the neural axis is extremely rare and mostly encountered in separate locations.[1] Here we present a case of double cranial encephalocele in a 2-year-old boy with congenital hydrocephalus.

CASE HISTORY

A 2-year-old boy who was antenatally supervised but not diagnosed with any anomaly came with complaints of two swellings at the back of the head since birth. He also complained of progressive increase in size of the head with delayed milestones. At presentation, he had no head holding, had no speech, and was unable to walk even with support. On examination, he had bulging anterior and posterior fontanels with two swellings in the cranium—one in the vertex and the other in the occipital region. The vertex swelling was 4 × 4 cm, and the caudal one 2 × 2 cm. Both were skin covered. Non contrast computed tomography of the head showed a porencephalic cyst with ventricular dilatation, which was more on the left than the right [Figure 1A and B]. Patient underwent a left ventriculoperitoneal (VP) shunt [Figure 2A]. The size of the swellings mildly decreased in the follow-up at 3 months [Figure 2B]. He subsequently underwent repair of encephaloceles and is presently doing well at 1-year follow-up with mild improvement in his milestones.

DISCUSSION

The occurrence of multiple NTDs in a single patient is extremely rare. The exact worldwide incidence is not known. Many of these cases have been reported at thoracic and lumbosacral levels.[2] NTDs arise due to failures in neural folds opposition and fusion during primary neurulation process. Classical theory explains that the neural tube closure starts from the mid-cervical region, and continuously reaches to the rostral and caudal ends in a bidirectional, zipper-like fashion. However, this theory is unable to explain the presence of double NTDs or defects in the cervical region.[3] Van Allen et al.[4] and Nakatsu et al.[5] proposed multiple sites of closure of the neural tube. The multisite neural tube closure model suggests the existence of multiple closure points or “zippers”—under the control of one or more genes—in normal neural tubes in humans.[4,5]

The reported cases in literature have been enumerated in the tabulated form [Table 1]. Goyal et al.’s[6] case can be explained if there is an existence of neural tube closure in Y-shaped fashion (zipper) at rostral end. Singh et al.[7] did an exhaustive search of the existing theories and concluded that the explanation given by Goyal et al.[6] was an assumption and had no scientific basis, and that it certainly challenges our existing knowledge of neural tube closure. Canaz et al.[8] discussed the importance of preoperative neuroimaging studies to optimize the outcome.

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CONCLUSION
Multiple NTDs are rare. Early surgery as for other encephaloceles and meningomyeloceles are associated with better appearance, easier nursing care, lower rates of trauma and ulceration, and prevention of neurological impairments.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will

Table 1: Details of patients reported with double encephaloceles

| Sl. No | Author            | Comments                                                                 | Outcome                                               |
|-------|-------------------|--------------------------------------------------------------------------|-------------------------------------------------------|
| 1     | Goyal et al. [6]  | Double MMC located at suboccipital region, adjacent to each other like twin MMC | Good recovery with no hydrocephalus at 2 years follow-up |
| 2     | Singh et al. [7]  | Double meningoencephalocele in the occipital region                       | No neurological sequelae                              |
| 3     | Ramdurg et al. [3] | Claimed to having reported first case in world literature of double occipital encephalocele with a split pons | Uneventful recovery                                  |
| 4     | Canaz et al. [8]  | Double occipital encephalocele, each arising from a bony defect separately—one on top of the other | Some signs of developmental delay in 3-year follow-up |
| 5     | Index case        | Double occipital encephalocele with hydrocephalus with porencephalic cyst | Delayed milestones at 2 years of age                  |

MMC = Meningomyelocele
not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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