

## Case Reports

# FRONTO ETHMOIDAL MENINGO ENCEPHALOCLE; STUDY OF TWO CASES AT THE DEPARTMENT OF NEUROSURGERY, BANGABANDHU SHEIKH MUJIB MEDICAL UNIVERSITY

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### Abstract

**Background:** In meningoencephalocele, meninges, cerebrospinal fluid (CSF) and brain parenchyma come out through the defect of cranial bone.

**Objective :** These case were discussed because these are rare case and are difficult to approach. The operations were done by bifrontal incision and bifrontal craniotomy. Dura was opened, CSF was sucked out. Degenerated brain were removed with proper haemostasis and the dural adhesion was dissected. Water tight close were done dura were plicated. Part of temporal bone were taken under temporal muscle were replaced bony gap which were fixation.

**Results:** Post operative period were uneventful. There were no CSF leakage or other complications.

**Conclusion:** These are rare congenital anomalies, early diagnosis and surgery can save patient life.

**Key words:** Meningoencephalocele, bicoronal, craniotomy, dural plication, bone fixation.

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### Introduction:

Cranium bifidum is a defect in the fusion of the cranial bone, it occurs in the midline and is most common in the occipital region. If meninges and CSF herniate through the defect, it is called a meningocele. If meninges and cerebral tissue protrude, it is called an encephalocele.

Encephalocele also known as cephalocele is an extension of intracranial structures outside of the

normal confines of the skull. One case was seen for every five cases of spinal myelomeningocele.<sup>1</sup> A nasal polypoid mass in a newborn should be considered an encephalocele until proven otherwise.

Fronto-ethmoidal AKA sincipital, 15% of encephaloceles; external opening into face in one of the following 3 regions (i) nasofrontal external defect in the nasion (ii) naso-ethmoidal defect between nasal bone and nasal cartilage (iii) naso-orbital defect in the antero-inferior portion of medial orbital wall.

A transnasal approach to a basal encephalocele may be fraught with intracranial hemorrhage, meningitis or persistent CSF leak. Usually a combined intracranial approach and transnasal approach is used.

Frontoethmoidal meningoencephaloceles are common in many Southeast Asian countries, occurring in 1 in 5000 live births.<sup>2,3,4,5</sup> These lesions often affect poor, rural children in developing countries but their etiology is still poorly understood.<sup>6,7,8</sup> In addition to the obvious facial deformity of the fronto ethmoidal meningocele (fMEC) itself, affected children may also have

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telecanthus,<sup>9,10</sup> amblyopic eyes, and epiphora.<sup>11</sup> Furthermore, some of the children have neurological complications or associated brain anomalies,<sup>12,13</sup> although most are mentally normal.<sup>14,15,16</sup>

Several papers have been published on fMEC treatment in countries with modern medical infrastructures,<sup>17,18</sup> such as Thailand.<sup>20,21</sup> However, fMECs are common in many poorer and less-well-developed countries where such facilities are not available. This puts a different emphasis on the development of effective fMEC treatment programs. For example, in Cambodia the main focus of fMEC management should be on the development of low-cost but effective procedures by adapting treatment programs to the country's social, financial, and medical resources.<sup>22</sup>

They have published previously on this topic,<sup>23,24</sup> and we report here on a series of 200 patients who were surgically treated for fMECs between 2004 and 2009 using

limited surgical materials and equipment. To our knowledge, this is the largest series of fMEC cases published to date. This study also represents the appraisal of a humanitarian and teaching program. We discuss several issues surrounding fMECs, focusing on the development of a sustainable program of treatment and on surgical procedures that can be adapted to local conditions.

It may be that a combination of environmental factors with a genetic predisposition could explain the pathogenesis and geographical distribution of these malformations.<sup>25,26</sup> Familial cases are very rare among previously published series<sup>27</sup> and in our own experience. From our own series of cases, we have demonstrated that the date of conception, with predominance for the wet season, may play a role.<sup>24</sup> This was also shown in a case series in Burma 25 years ago.<sup>28</sup> Among environmental factors, fungal and teratogenic agents such as aflatoxin or ochratoxin, found in moldy rice during the wet season, could be involved.<sup>15</sup> But it must be acknowledged that currently the etiology of fMEC is largely unknown.

### Case report- I

A two years old male child from remote area of Dhaka had been admitted at the department of Neurosurgery at Bangabandhu Sheikh Mujib Medical University with the complaints of slowly progressive swelling at mid fronto-nasal region since birth, swelling extended to

mid forehead to nasal region. The swelling was progressive in nature. His father did not give history of consanguines marriage. No history of antenatal checkup of his mother. Mother did not take folic acid. He had one brother and sister and were healthy. His birth history was normal vaginal delivery and no obstructed labour. He had no any other congenital anomalies. Size of swelling is about 5 cmx2.5 cm and circular in shape. Swelling were pulsatile, cough impulse was present. Swelling is cystic in nature and trans illumination test is negative. The patient had no neurological deficit. Computerized tomography scan (CT scan) of brain showed fronto-ethmoidal meningoencephalocele. All general parameters are normal.

The swelling was operated with bicoronal scalp incision with bifrontal craniotomy. The dura were adhere to adjacent bone. Bony gap was found at fronto-ethmoidal area. Dura was opened. Spinal fluid (CSF) was sucked out. Some damaged brain mater was sucked out. Dura was closed water tight fashion. Dura and brain was dissected from adjacent bone. Dural plication was done. A piece of that bone was taken from right temporal region which was placed at bony defect and fixed with drill and silk. Proper haemostases was done. Wound was closed in layers. There were no CSF leakage and post operative period was uneventful.

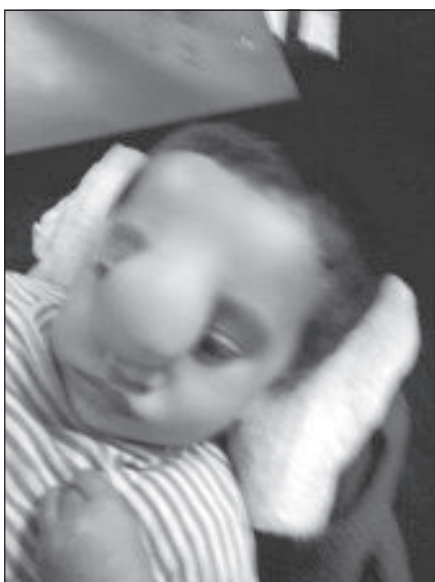


**Fig.-1:** Postoperative photograph of fronto ethmoidal encephalocele

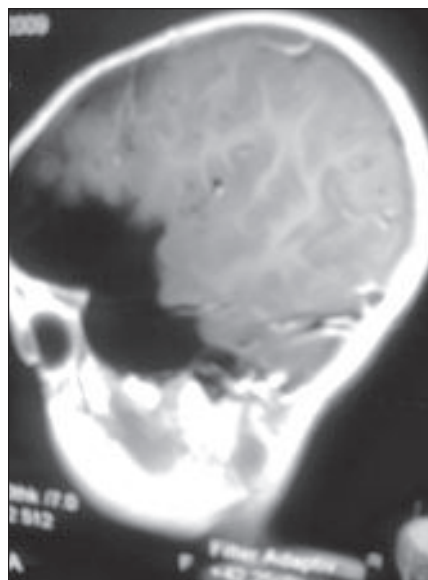
### Case report -II

A one years 5 months child had been admitted in the Bangabandhu Sheikh Mujib Medical University (BSMMU) with the complaints of progressive swelling at mid frontonasal region since birth. Swelling was enlarging in size, extended from mid frontal region to the ala of nose. Swelling was progressive in nature. She is the first issue of her parents. No history of maternal antenatal checkup. No history of consanguinous marriage of her parents. She had no other congenital anomalies. Swelling is spherical size were 5 cm x 3 cm in diameter. Swelling was pulsatile. Non tender, flactutation test is positive. Cough impulse is present. Increase the size of swelling during banding forwards and downward. Swelling is non tender and patient had no neurological deficit. CT scan and MRI of brain showed fronto ethmoidal encephalocele with arachnoid cyst at right frontotemporal region.

All general parameter are normal. Patient were operated by bicoronal scalp incision and bifrontal craniotomy and right temporal extension of craniotomy. Dura was open and CSF was sucked out. Marcipolization of arachnoid cyst was done. Dura was dissected from bony gap of fronto naso etmoidal region. Dura was sutured by water tight fassion, dura plication was done. Bony gap was replaced by bone piece from temporal region and fixed by drill and silk. Wound was closed in layers. Post operative period was uneventful.



**Fig.-2:** Preoperative photograph of fronto ethmoidal encephalocele



**Fig.-3:** MRI of brain with sagittal section shows frontotemporal archonoid cyst with encephalocele



**Fig.- 4:** MRI of brain with axial section shows frontotemporal archonoid cyst with encephalocele

### Discussion:

The condition is not classified in the category of neural tube defects<sup>12</sup> and is not associated with other neural tube defects<sup>29</sup> as seen in this series. Studies using immunohistochemical morphological analysis with neuron-specific enolase in autopsy material or surgical material showed that there was no sign of dysraphism in the underlying brain.<sup>30</sup> One possible mechanism

involves a local deficiency of the mesoderm (which forms the skull of the embryo) in combination with abnormal adhesion of the neuroectoderm to the surface ectoderm (for example, through deficient apoptosis).<sup>31,32,33</sup> The natural history of this malformation is variable among patients.<sup>22</sup> Neurological complications and congenital brain anomalies are uncommon in fMEC,<sup>28</sup> and a majority of affected children are mentally normal.<sup>15</sup> Some of our parents reported that their child's malformation did not grow at all with time, whereas others (as reported by other authors) noted that their child's malformation became progressively larger.<sup>9</sup>

Some authors therefore recommend early surgical correction to minimize the pressure effect of the mass on facial growth.<sup>3,5</sup> It is important, however, to take into consideration local resources and experience with respect to anesthesia, as smaller children and babies will be those at highest risk of anesthetic complications. The prognosis of untreated fMEC has also been debated. Historically, some authors claimed that only a minority of children who do not undergo surgery could survive to adulthood.<sup>34</sup> However, there is no recent scientific evidence to argue for or against a significantly reduced life expectancy in patients with fMEC.

### Conclusion:

Frontoethmoidal meningo encephalocele is difficult to approach. It is very difficult to avoid its postoperative complication like cerebrospinal fluid (CSF) leakage, meningitis. Proper diagnosis and timely surgery can save many lives.

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