Blake's Pouch Cyst with Corpus Callosum Dysgenesis

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Abstract

Blake's pouch cyst is cystic malformation of posterior fossa characterized by infracerebellar cyst, absence of communication between the fourth ventricle and the subarachnoid space, and hydrocephalus. The child may present with clinical features depending on extent of hydrocephalus and may have atypical associations. Here we are presenting a case of three year male child having blake's pouch cyst along with corpus callosum dysgenesis, and global developmental delay.

Keywords: Blake’s Pouch; Corpus Callosum; Hydrocephalus

Introduction

Blake's pouch cyst (BPC), a cyst like structure i.e. posterior ballooning of the inferior medullary velum (thin sheet of tissue formed by ependyma and piamater of the tele choroidea) into the cisterna magna, below and posterior to the vermis, which communicates with fourth ventricle. Its etiology is probably failure of regression of Blake's pouch secondary to the non-perforation of the foramen of Magendie [1,2].

The clinical presentations of BPC include macrocephaly with or without neurological deficits to hydrocephalus as the age increases. Neuroimaging plays an important role in differentiating BPC and other posterior fossa cystic malformations [3]. Here we are presenting a three year male with blake's pouch cyst along with corpus callosum dysgenesis and global delay.

Case Report

A three year male child presented in outpatient department of pediatric medicine with complaint of inability to walk without support and inability to speak words, clearly suggestive of developmental delay. The child was born through emergency lower segment caesarean section due to failed progression of labour. There is also history of delayed cry at birth for which child got admitted in neonatal intensive care unit (NICU) for two days and got discharged successfully. The developmental history is suggestive of delayed achievement of milestones. There is no history of any trauma, consanguinity, neuropsychiatric manifestations, constipation or urinary retention.

On examination, child was active with stable vitals. Child had global developmental delay (neurodevelopmental score: motor and adaptive < 20, language: 60, personal social: 40). Anthropometry was done which suggested of weight: 10.8 kg (Expected: 14.3 kg), head circumference was appropriate for age whereas, height was between -1 standard deviation and -2 standard deviation. On physical examination, there was obvious frontal bossing but no spine deformity and child had pes planus foot deformity. The central nervous system examination revealed slightly decreased muscle tone with normal deep tendon reflexes along with bilateral extensor plantar reflex. The child has no involvement of cranial nerve or any sensory deficit; ophthalmological examination did not suggest any ocular or extra ocular movement abnormality. There was no evidence of papilledema. Cardiovascular, respiratory and abdominal examinations were normal.

Complete blood counts, renal, liver function tests and thyroid profile were normal. Magnetic Resonance Imaging of brain suggests evidence of T1/T2 CSF attenuating retrocerebellar cyst communicating to fourth ventricle along with thinning of corpus callosum suggestive of corpus callosum dysgenesis (Figure 1a and 1b).

Discussion

BPC was first described by Tortori-Donati, et al. in 1996. Blake’s pouch cyst is a cystic posterior fossa anomaly. Its differential diagnosis are Dandy Walker Syndrome, arachnoid cysts, cisterna magna [4]. These anomalies are differentiated on basis of neurological imaging (Table 1).

The mass effect due to BPC depends on enlargement of cyst. The posterior fossa is typically normal in size. Due to the enlargement of the fourth ventricle, the fourth ventricle choroid plexus is stretched inferior to the vermis along the anterosuperior aspect of the cyst. The stretched choroid plexus is best visualized as an enhancing structure on sagittal contrast-enhanced T1-weighted images [5]. The cerebellar vermis is normal in size and not rotated, allowing the differentiation between BPC and Dandy Walker malformations. MRI typically does not allow the visualization of the cyst wall that may be seen to some extent using a fast imaging employing steady-state acquisition (FIESTA) sequence. BPC remains under diagnosed as it may be asymptomatic at earlier ages.
Blake’s Pouch Cyst with Corpus Callosum Dysgenesis

<table>
<thead>
<tr>
<th>Disease</th>
<th>Vermis (size)</th>
<th>IVth ventricle (size)</th>
<th>Posterior fossa (size)</th>
<th>Hydrocephalus</th>
</tr>
</thead>
<tbody>
<tr>
<td>BPC</td>
<td>Normal</td>
<td>Enlarged</td>
<td>Normal</td>
<td>Yes (majority of the patients)</td>
</tr>
<tr>
<td>DWM</td>
<td>Hypoplastic</td>
<td>Enlarged</td>
<td>Enlarged</td>
<td>Yes (majority of the patients)</td>
</tr>
<tr>
<td>PFAC</td>
<td>Normal</td>
<td>Normal or reduced</td>
<td>Normal</td>
<td>Possible</td>
</tr>
<tr>
<td>MCM</td>
<td>Normal</td>
<td>Normal</td>
<td>Inconsistently enlarged</td>
<td>No</td>
</tr>
</tbody>
</table>

Table 1: Clinical manifestations of different types of posterior fossa cysts.

BPC: Blake’s Pouch Cyst; DWM: Dandy-Walker Malformation; MCM: Mega Cisterna Magna; PFAC: Posterior Fossa Arachnoid Cyst.

BPC could be congenital or acquired and may be differentiated by susceptibility weighted imaging. BPC is believed to result from a failure of embryonic assimilation of the area membranacea anterior within the tela choroidea associated with consequent lack of perforation of the foramen of Magendie [5,6]. Acquired BPC is caused by obstruction of initially fenestrated Blake’s pouch by blood products or fibrin sheaths after intraventricular hemorrhage or central nervous system infection and appear like malformed BPC [1,7,8]. Rarely, BPC may be caused by trauma or infection. Macrocephaly in neonatal period is the most common presenting feature of BPC [3]. BPC, however, may also present later in life without hydrocephalus. It can also present with symptoms of syringomyelia.

Conclusion

Endoscopic third ventriculostomy is a safe and effective treatment in symptomatic patients. Shunt surgeries are also advocated but with higher rate of complications [8]. Although optimal treatment and its outcome remains unknown, the child needs neuropsychiatric assessment in follow up later.

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None.

Conflict of Interest

None.

Bibliography


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