Complications of Unattended Spina Bifida Presenting in Adults

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Abstract- Spinal bifida is a congenital neural tube decade, has been described in literature since 19th century. There has been gross development of management of spina bifida in the form of prenatal repair, MOMs trial. Since the introduction of folic acid in primary health care, there is a significant reduction in incidence of spina bifida. In the era where pediatrics neurosurgeons are faced to manage the neonatal and pre natal repair of MMC, we are challenged to manage patients with adult spina bifida with complications. In this advanced neurosurgical era this is rare challenge to face. We described 4 patients with neglected complications of spina bifida, their clinical presentations and management.

Index Terms- meningocoele, spina bifida, epidermoid cyst, tethered cord.

I. INTRODUCTION

Spina bifida varies in its presentation from occulta, aperta to overt manifestations. Incidence of spina bifida had dramatically reduced over past 2 decades after the implementation of perinatal consumption of folic acid.

In the era where the world is witnessing the role of FOETAL surgery in the management of spinal bifida, we are challenged with adult presentations of meningomyelocele, complicated meningomyelocele, dermal sinus with intra medullary epidermoid cyst.

Materials and methods

This is a retrospective study done at BIN, IPGMER.

We had retrospectively collected the data over 3 years duration.

We had traced the pre operative, operative and post operative track records, follow up after 1 year, with telephonic conversations for all the patients.
CASE 2) : 23 year old male, unmarried, with complaints of swelling over the back since birth, with out any CSF leakage without any neurological deficits, with only necessity being cosmetic purpose.

MRI lumbosacral spine : meningocoele

Operative details: meningocoele sac opened, with reduction of the contents and primary closure of the sac.

CASE 3) : 52 year old male patient with complaints of recurrent pus discharge (2 weeks) from a small swelling over the mid back which has been present since birth, with progressive weakness of lower limbs (spastic paraparesis) with intact bladder and bowel control.

MRI DORSOLUMABR SPINE : dermal sinus D 10 level with intra medullary hyperintense signals, Epidermoid cyst, with syringomyelia extending to D 2 level.

Operative details: Elliptical incision with dissection of the sinus tract, laminectomy with removal of intra medullary epidermoid with primary dural repair.

CASE 4) : 55 year old male, with 2 married children, with progressive increase in swelling which was present since birth with occasional whitish discharge from the swelling, clinical neurological examination did not reveal any deficits.

MRI LUMBOSACRAL SPINE : meningocoele with tethered cord with probable multiple heterogenous signal mixed mass that occupied the sac.

Operative details: meningocoele sac is opened and we had found a mature teratoma (with fatty tissue, hair).
III. DISCUSSIONS

Spina bifida is a complex congenital condition with an estimated prevalence of between 3.06 and 3.13 cases per 10,000 live births, not including cases of spina bifida occulta (1). The introduction of mandatory folic acid supplementation and early prenatal diagnosis accompanied by the subsequent termination of affected fetuses has lead to a decrease in the incidence of spina bifida. (7, 6)

The term spina bifida simply refers to splitting of the vertebral arches. This splitting can be isolated (spina bifida occulta), or it may include the meningeal sac (meningocele) or the meningeal sac plus portions of the spinal cord and/or spinal nerves (myelomeningocele) (5). When excess lipomatous tissue is involved, the condition is referred to as lipomeningocele or lipomyelomeningocele, depending on the involvement of the nervous tissue. Associated conditions that do not necessarily involve splitting of the vertebral arches, but often do, include diastematomyelia (split cord), diplomyelia (duplicated cord), myeloschisis (flatten malformed cord), and fatty filum (lipomatous tissue surrounding the filum terminale). (4)

Spinal dysraphism manifests as an incomplete fusion of the neural arch, varying from the occult to more severe open neural tube defects (NTD). Meningocele is the simplest form of open NTD characterized by cystic dilatation of meninges containing cerebrospinal fluid without any neural tissue. (9) The majority of meningoceles are identified and treated perinatal. We describe the delayed presentation of a meningocele and its complications and the reasons behind their late presentations in adulthood with relevant review of the literature. (8)

In developed countries MOMS trail has focused the attention of training neuro surgeons in foetal surgery (3)

In developing countries like INDIA we still face the challenge of managing the neglected spina bifida and its complications in adults.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Symptoms</th>
<th>Reason for presenting late</th>
<th>Reason for presenting now</th>
<th>Complication</th>
<th>Intra and post op period</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>46</td>
<td>CSF leak</td>
<td>Uneducation</td>
<td>CSF leak</td>
<td>None</td>
<td>MMC repair with detetherig</td>
</tr>
<tr>
<td>2</td>
<td>23</td>
<td>Cosmesis</td>
<td>Poverty, lack of knowledge</td>
<td>Cosmesis</td>
<td>None</td>
<td>MMC repair</td>
</tr>
<tr>
<td>3</td>
<td>52</td>
<td>Paraparesis</td>
<td>Poverty, lack of knowledge</td>
<td>Paraparesis</td>
<td>Transient paraplegia for 48 hours</td>
<td>Sinus tract excision with complete removal of epidermoid cyst</td>
</tr>
<tr>
<td>4</td>
<td>55</td>
<td>Whitish discharge</td>
<td>Undedication</td>
<td>Whitish discharge</td>
<td>Mature teratoma</td>
<td>mmc</td>
</tr>
</tbody>
</table>

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IV. CONCLUSION

Most of the spina bifida are corrected in childhood, but rarely we can see those neglected patients in adults. Here we had come across 4 patients over 3 years period at Eastern India(10). Most of them hailed from poor socio–economic status with lack of medical knowledge.

One patient had come across for the purpose of cosmesis.

Remaining three patients had presented in their 40/50s after the development of complications.

It is very rare thing to notice that 46 year old lady has completed her pregnancy and delivery, gone unnoticed by medical examinations and presented with csf leak after rupture of sac.

It has not been reported on the mature teratoma withing the sac, which is quite unexpected.

Social stigma in adolescence is one of the major factor too for their late presentations.

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