# 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 meningomeyelocoele , complicated meningomyelocole , 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 .

## II. CASE DISCUSSIONS

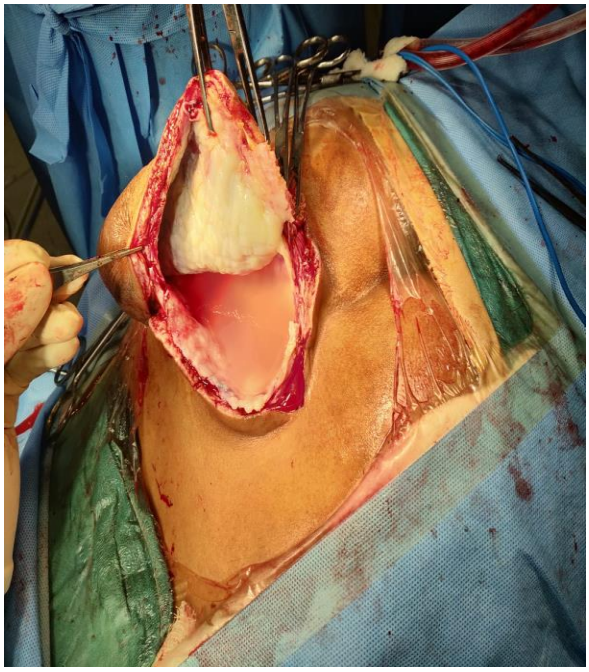
CASE 1) 46 year old female patient , P2 L2 ,clinical symptoms of swelling over the back present since birth , progressively increased to attain the size of a foot ball , with watery discharge from past 10 days .

Neurological examination shows no gross motor or sensory deficits with retained bladder and bowel function .

MRI lumbo sacral spine : meningocole with tethered cord

Operative details : meningocole sac opened , with large gush of csf, detethering of the sac done with primary

closure.



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 necessiaty being cosmetic purpose .

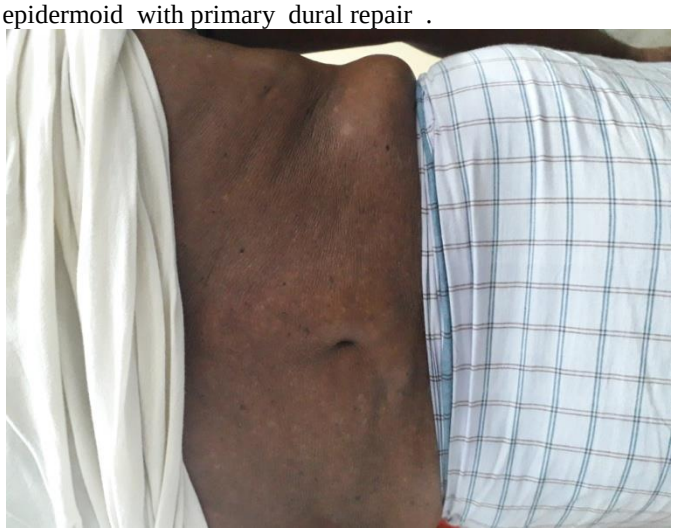
MRI lumbosacral spine : meningocole

Operative details: meningocole 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 ( spatic 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 .

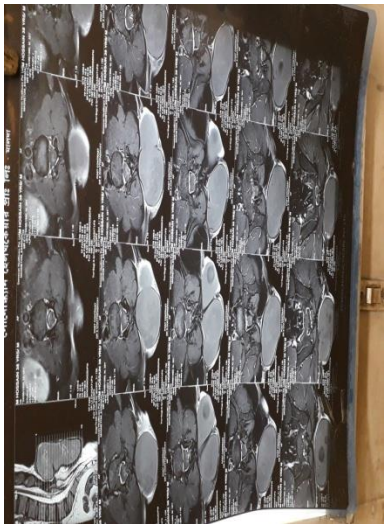
Operative details : Elliptical incision with dissection of the sinus tract, laminectomy with removal of intra medullary epidermoid with primary dural repair .



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 : meningocole 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 meningocoeles are identified and treated perinatal. We describe the delayed presentation of a meningocoele 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 .

| ***case*** | *Age* | Symptoms | Reason for presenting late | Reason for presenting now | Complication | Intra and post op period. |
|---|---|---|---|---|---|---|
| 1 | 46 | Csf leak | Uneducation | Csf leak | None | Mmc repair with detether ig |
| 2 | 23 | Cosmesis | Poverty,lack of knowledge | Cosmesis | None | Mmc repair |
| 3 | 52 | paraparesis | Poverty,lack of knowledge | paraparesis | Transiet paraplegia for 48 hours | Sinus tract excision with complete removal of epidermoid cyst |
| 4 | 55 | Whitish discharge | uneducation | Whitish discharge | Mature teratoma | mmc |

| Author/ year | Age in years/sex | Symptoms/signs | Location of lesion/radiology | Intervention/outcome |
|---|---|---|---|---|
| Chambers and Revilla[5] | 53/male | Urinary incontinence, lower limb weakness | Lumbosacral meingocoele, bony defect at L5 on X-ray | Surgery<br>Weakness improved but not the incontinence |
| Rao and Dinakar[6] | 25/male | Pain, nocturnal enuresis<br>Loss of perianal sensation and ankle jerks | Lumbosacral meingocoele, spina bifida at L4 and L5 on X-ray | Surgery<br>Pain improved, the other deficits remained same |
| Gok *et al.* (2005)[1] | 48/male | Pain, urinary incontinence, sexual dysfunction<br>Loss of perianal sensation and ankle jerks | Myelocele, sac from L5 spina bifida. Conus reaching L5 level on MRI | Refused surgery |
| Sarda *et al.*[7] | 20/male | Asymptomatic | Meingocoele, sac from L4 to L5 spina bifida on CT scan | Operated |
| Düz *et al.* (2008)[8] | 21/male | Sensory loss, leg length discrepancy | Lumbosacral meningocoele, corpus callosal agenesis, conus at L5 | Refused surgery |
| Düz *et al.* (2008)[8] | 21/male | Dermal sinus, tethered cord syndrome | Lumbosacral meningocoele, split cord at L1, conus at L3 | Refused surgery |
| Gillis *et al.*[4] | 28/female | Known sacral agenesis<br>Swelling at age 20, headache in upright position<br>Loss of perianal sensation and ankle jerks, plantar flexion 2/5 | Sacral meningocoele at S1, conus reaching L5 level, syrinx at L2–3 on MRI | Operated<br>Reccurent CSF leaks for which lumbopritoneal shunt done<br>Neurologically same |
| Present case | 53/male | Pain radiating to lower limbs, urinary incontinence, perianal hypoesthesia | Lumbar meningocoele, conus at L3 on MRI | Operated<br>Improved in all the symptoms |

CSF = Cerebrospinal fluid; CT = Computed tomography; MRI = Magnetic resonance imaging

## 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 –economical 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 .

## REFERENCES

[1] Asher M, Olson J, Weigel J, Morantz R, Harris J, Leiberman B, Whitney W (1979) The myelomeningocele patient. A multidisciplinary approach to care. J Kans Med Soc. 80:403–408, 413 [PubMed]

[2] https://www.sciencedirect.com/science/article/pii/S1047279717302624.

[3] https://fluidsbarrierscns.biomedcentral.com/articles/10.1186/1743-8454-1-4

[4] https://link.springer.com/article/10.1007/s40141-014-0046-1

[5] https://www.ncbi.nlm.nih.gov/pubmed/26926705.

[6] https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2441590/

[7] Congenital meningocoele presenting in an adult
Raghvendra V. Ramdasi, Trimurti D. Nadkarni, and Atul H. Goel

[8] https://www.ncbi.nlm.nih.gov/pubmed/16500172

[9] Spinal dorsal dermal sinus tract: An experience of 21 cases
Ishwar Singh,* Seema Rohilla,1 Prashant Kumar,2 and Saurabh Sharma

[10] http://medind.nic.in/icb/t08/i10/icbt08i10p1086.pdf

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