

## Late Presenting Meningocele: Review of Ten Cases



### Medical Science

**KEYWORDS :** lumbosacral meningocele, Spina bifida, Neural tube defect

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### ABSTRACT

*Introduction: Spina bifida is a spinal cord malformation that occurs as a defect of the neural tube (ie, the embryonic structure that develops into the spinal cord and brain). Usually presents since birth and ideally operated within 48 hrs of birth. The present study is a compilation of cases who presented quiet late and were operated successfully.*

*Material and Method: The present study is a retrospective analysis of ten cases of lumbosacral meningocele operated at more than 3 months age at lifeline hospital and R.K. Mission hospital Varanasi during 2010-2015. A detail record was made of the presenting features, neurological signs, investigations, surgery and outcome.*

*Results: All patients presented to us after 3 months age with no or very little neurological deficit with normal overlying skin. MRI was done in all the cases. All the patients were operated and recovered very well.*

*Conclusion: lumbar meningocele is a congenital condition, although should be operated within 48hrs of life, some ignored cases with intact and healthy skin cover may grow normally and can be operated at any age.*

### Introduction

Spina bifida is a spinal cord malformation that occurs in varying degrees of severity. Classified as a defect of the neural tube (ie, the embryonic structure that develops into the spinal cord and brain), it was recognized as long as 4000 years ago.

Neural tube defects have a range of presentations, from still-birth to incidental radiographic findings of spina bifida occulta. Myelomeningocele, a form of spina bifida, is visible at birth. Patients with myelomeningocele present with a spectrum of impairments, but the primary functional deficits are lower limb paralysis and sensory loss, bladder and bowel dysfunction, and cognitive dysfunction.(1)

### It is classified as

#### *Spina bifida cystica*

Spina bifida cystica can occur anywhere along the spinal axis but most commonly is found in the lumbar region. In this condition, the spine is bifid and a cyst forms. A meningocele, a cystic swelling of the dura and arachnoid, protrudes through the spina bifida defect in the vertebral arch. A person with a meningocele may have no neurologic sequelae. Spina bifida cystica causes a problem when cord tissue extends into the meningocele, in which case the cyst is called a myelomeningocele. According to Menelaus, the myelomeningocele form of spina bifida cystica is the most significant and common type of spina bifida, accounting for 94% of cases. Another form of spina bifida cystica, the most severe type in fact, is the myelocele, or myeloschisis, variety, in which the open neural plate is covered secondarily by epithelium and the neural plate has spread out onto the surface.

#### *Spina bifida occulta*

The term spina bifida does not actually refer to spina bifida occulta, which may exist in a very large number of healthy adults.

#### *Syringomeningocele*

The term describes a hollow center, with the spinal fluid connecting with the central canal of the cord enclosed by a membrane with very little cord substance.

#### *Syringomyelocele and syringomyelia*

Syringomyelocele is a type of spina bifida in which protrusion of the membranes and spinal cord lead to increased fluid in the central canal, attenuating the cord tissue against a thin-walled sac. Syringomyelia, or hydrosyringomyelia, is the presence of

cavities in the spinal cord, which may result from the breakdown of gliomatous new formations.

Magnetic resonance imaging (MRI) of the spine and brain is helpful in neurologic assessment and provides a baseline for comparison in future investigations, especially in the context of progressive neurologic deterioration. MRI provides considerable detailed information regarding the spinal cord and its malformations, including low-lying or tethered cords.

Antibiotics, sac closure, and ventriculoperitoneal shunt placement are the standard of care for spina bifida and are implemented in the perinatal period in 93-95% of patients. Supportive care alone may be recommended in cases associated with an irreparable sac, active gross CNS infection or bleeding, and/or other gross congenital organ anomalies causing life-threatening problems.

Closure of the myelomeningocele is performed immediately after birth if external cerebrospinal fluid (CSF) leakage is present. In the absence of CSF leakage, closure typically occurs within the first 24-48 hours. The surgery can be delayed for several days without additional morbidity or mortality, giving families more time to deal with the emotional impact of their child's condition. This delay also gives parents more time to learn about myelomeningocele and to therefore better participate in the decision-making process regarding their child's treatment.

### Material and methods

We retrospectively analysed ten cases of lumbar meningocele who presented to us after 3 months of age at R.K. Mission Hospital and Lifeline Hospital Varanasi during 2010-2015. A detail record was made of the presentation, investigations, surgery and outcome.

### Results

Ten cases of lumbar meningocele who presented to us after 3 months of age at R.K. Mission Hospital and Lifeline Hospital Varanasi during 2010-2015 (Table 1). The mean age of presentation was 1.7 years. The male to female ratio was 3:7. All of them had swelling in the lumbar region since birth (Figure 1). Three had associated hydrocephalus which was noticed as increasing size of head and tense anterior fontanel for one month. All patients had intact skin overlying the meningocele with translucency. The swelling was fluctuant and crying impulse was present. Three children had neurological weakness in lower limbs

and parents complained of inability to stand even after one year age. Children who had hydrocephalus were lethargic with poor cry, sunken eyes and malnourished and all had papilloedema. MRI of lumbar spine and brain was done in all cases (Figure 2). In all the cases excision and repair of the sac was done (Figure 3,4,5). Patients with hydrocephalus underwent ventriculoperitoneal shunting as well. None of our patient had any new post-operative neurological deficit, recurrence, infection, csf leak or wound infection. None of the patient developed hydrocephalus after repair of meningocele who previously did not have hydrocephalus

### Discussion

Myelomeningocele is diagnosed at birth or in utero. At birth, a midline defect in the posterior elements of the vertebrae is noted with protrusion of the meninges and neural elements through an external dural sac.

Neural tube defects have a range of presentations, from stillbirth to incidental radiographic findings of spina bifida occulta. Myelomeningocele, a form of spina bifida, is visible at birth. Patients with myelomeningocele present with a spectrum of impairments, but the primary functional deficits are lower limb paralysis and sensory loss, bladder and bowel dysfunction, and cognitive dysfunction(1).

Although spina bifida occulta is common and almost always without consequence, some developmental abnormalities may occur such as a spinal cord lipoma or a fibrous cord that can cause subtle or rare neurologic signs.

In our series, all the ten cases were neglected cases with poor socioeconomic background mostly females. Although the abnormality was very well noticed at birth but due to very little or practically no neurological deficit the problem was overlooked by the family and the local care givers. As in all the cases the overlying skin was quite normal children could sustain the abnormality without infection and ulceration. Patients with hydrocephalus presented to us somewhat earlier because those children became lethargic, irritable, feeding poorly and had developmental delay.

The most obvious finding on physical examination is some degree of motor and sensory loss(1). Most patients with myelomeningocele have a flaccid paraparesis below the spinal cord lesion.

In our study three patients who could not stand even after one year age they were also shown to us relatively earlier as the parents could correlate the problem with the meningomyelocele. However six patients presented to us either because of social or the cosmetic cause when they became older started mixing in society or school.

Hydrocephalus can be tracked with serial cranial ultrasonograms (in infants) or computed tomography (CT) scans. A CT scan of the head is appropriate to evaluate for possible recurrent hydrocephalus or a change in the size or function of the ventricles, which may be affected even with normal pressure hydrocephalus.

Magnetic resonance imaging (MRI) of the spine and brain is helpful in neurologic assessment and provides a baseline for comparison in future investigations, especially in the context of progressive neurologic deterioration. MRI provides considerable detailed information regarding the spinal cord and its malformations, including low-lying or tethered cords. In all our cases we did MRI of the lumbar spine and brain and could get to know about content of sac, position of conus and associated hydrocephalus. However we could not detect any other neurological abnormality.

By definition, all children with a myelomeningocele have a tethered cord on MRI, but only about 20% of children require an operation to untether the spinal cord during their first decade of life, during their rapid growth spurts. In our series also all children had low lying and tethered cord. Thus, the MRI scan must be placed in context of a history and examination consistent with mechanical tethering and a resultant neurologic deterioration.

The ventricular system has a characteristic shape, with small frontal and large occipital horns, which are typical in patients with spina bifida.

In the United States, antibiotics, sac closure, and ventriculoperitoneal shunt placement are the standard of care for spina bifida and are implemented in the perinatal period in 93-95% of patients. Supportive care alone may be recommended in cases associated with an irreparable sac, active gross CNS infection or bleeding, and/or other gross congenital organ anomalies causing life-threatening problems.

Patients with spina bifida require extensive, active, interdisciplinary treatment by a trained and coordinated team. Neonatal neurosurgery is followed by monitoring of head size and condition for potential hydrocephalus, evaluation of sphincters, and progression toward an appropriate bowel and bladder regimen.<sup>[31, 32]</sup>

Closure of the myelomeningocele is performed immediately after birth if external cerebrospinal fluid (CSF) leakage is present. In the absence of CSF leakage, closure typically occurs within the first 24-48 hours. The surgery can be delayed for several days without additional morbidity or mortality, giving families more time to deal with the emotional impact of their child's condition. This delay also gives parents more time to learn about myelomeningocele and to therefore better participate in the decision-making process regarding their child's treatment.

Perioperative complications include wound infection, CNS infection, delayed wound healing, CSF leakage, additional neurologic damage to the cauda equina, and acute hydrocephalus. Long-term complications include cord tethering and progressive hydrocephalus.

In our series all the patients presented so late but all have been operated well and had no postoperative sequelae

Neurosurgical follow-up is required to recognize the complications of hydrocephalus or a possible tethered cord and to monitor any potential causes of seizure activity. In our series none of the patients developed postop hydrocephalus or progressive neurological deterioration due to tethered cord so far.

Although in a few cases hydrocephalus arrests spontaneously, 80-90% of children with myelomeningocele ultimately require shunting. Ventriculoperitoneal shunting is the preferred modality. Alternatives include ventriculoatrial and ventriculopleural shunting.

In our series three patients had hydrocephalus for which Ventriculoperitoneal shunting was done

Perioperative complications include intracerebral and/or intraventricular hemorrhage, bowel perforation, and infection. Long-term complications include infection, overdrainage or underdrainage, and obstruction of the shunt system. We have not encountered any complication so far.

### Conclusion:

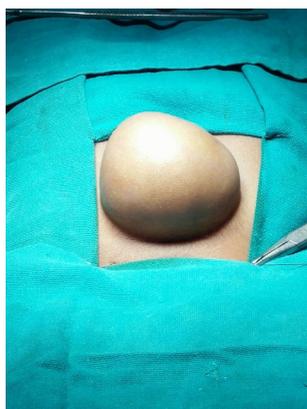
lumbar meningomyelocele is a congenital condition, although should be operated within 48hrs of life, some ignored cases with intact and healthy skin cover may grow normally and can be operated at any age of presentation with decreased risk of anes-

thetic and surgical complications.

**Table 1: Details of patients and management**

Sl. No	Age/sex	Chief complaints	Duration	CT/MRI scan	Treatment
1	3yr/F	Swelling in lumbosacral region	Since birth	Lumbosacral meningocele	Excision and repair
2	4yr/M	Swelling in lumbosacral region	Since birth	Lumbosacral meningocele	Excision and repair
3	6m/F	Swelling in lumbosacral region with increasing size of head	Lumbar swelling since birth and large head for 1m	Lumbosacral meningocele with hydrocephalus	Vp shunt placement with excision and repair of meningocele
4	18m/F	Swelling in lumbar region with difficulty in standing	Since birth	Lumbar meningocele	Excision and repair
5	3yr/F	Swelling in lumbosacral region	Since birth	Lumbosacral meningocele	Excision and repair
6	2yr/F	Swelling in lumbosacral region with difficulty in standing	Since birth	Lumbosacral meningocele	Excision and repair
7	6m/M	Swelling in lumbosacral region with increasing size of head	Lumbar swelling since birth and large head for 1m	Lumbosacral meningocele with hydrocephalus	Vp shunt placement with excision and repair of meningocele
8	1yr/F	Swelling in lumbosacral region with difficulty in standing	Since birth	Lumbosacral meningocele	Excision and repair
9	15m/F	Swelling in lumbosacral region	Since birth	Lumbosacral meningocele	Excision and repair
10	3m/M	Swelling in lumbosacral region with increasing size of head	Lumbar swelling since birth and large head for 1m	Lumbosacral meningocele with hydrocephalus	Vp shunt placement with excision and repair of meningocele

**Figure-1: lumbar meningocele with healthy overlying skin**



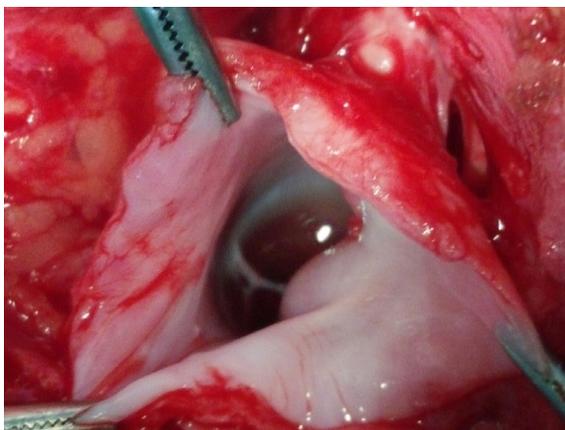
**Figure-2: MRI image of a 3yr old girl with lumbar meningocele**



**Figure-3: Sac dissected from skin, excess sac excised**



**Figure-4: Sac containing neural structures**



**Figure-5: Closed wound after sac closure****REFERENCE**

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