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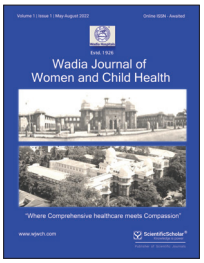

Case Series

Newer perspective in spina bifida: Limited dorsal myeloschisis

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ABSTRACT

Limited Dorsal Myeloschisis (LDM) is a relatively newly described entity in the spectrum of spinal dysraphism. Although considered rare, more cases are getting recognised with advancements in imaging technologies. We present a series of seven cases of paediatric spinal LDMs that were successfully managed surgically.

Keywords: Spina bifida, Limited dorsal myeloschisis, Saccular

INTRODUCTION

Spinal dysraphic malformations are caused by embryogenic defects of primary or secondary neurulation leading to inability to form a proper central neuraxis. First described in 1993, Limited Dorsal Myeloschisis (LDM) is a relatively new entity in the spectrum of spinal dysraphism that was initially thought of as meningocele.[1] Initially considered a rare entity, increasing number of cases are emerging nowadays due to readily available, advanced imaging technologies suggesting that it is not such a rare entity. Pang *et al*. (2010) classified LDMs into saccular and flat types.[2] True LDMs causing tethering of cord have been identified in all regions of the spinal neuraxis with maximum incidence at the lumbar level.

We present an illustrative series of 7 cases of paediatric spinal LDMs that were successfully managed by surgical approach at our centre.

CASE SERIES

Case 1

A 15-month-old girl presented with history of swelling over the neck since birth. The skin was very thin and fragile over the lesion, but was intact. Clinical examination revealed a solitary saccular swelling over the cervico-dorsal region measuring 8x9 cm. The cranial outermost portion of the swelling had a reddish, translucent, dome-like structure that was bulging out like a cap and was covered with a membrane-like structure with parched skin all around but with no defect. Base of the swelling was formed by thickened skin that was stretched with dilated veins

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suggesting chronicity of the lesion. The swelling had a broad fundus with a narrow neck palpable between the spinous processes of C6 and T2. The swelling was non-tender, soft in consistency, and uniformly transilluminating with cough impulse on crying. On examination, the patient did not have any weakness of the upper or lower limbs [Figure 1a].

MRI showed a large cystic sac-like structure measuring around 9.5 x 8.3 x 9.6 cm, protruding posteriorly through a defect at C7-D1 level. The spinal cord was tethered at that level posteriorly through a linear T2 hypointense structure, seen attaching the dorsal surface of the cord with the surface of the sac, likely a fibroneural stalk suggestive of a saccular LDM. Few ill-defined T1 hyperintense areas were seen within the sac suggestive of a fatty component. Brain screening was suggestive of small crowded posterior fossa with cerebellar tonsillar herniation with mild supratentorial hydrocephalus [Figure 1b].

She underwent surgical excision of the LDM. An elliptical incision was marked over the confluence of healthy and non-healthy skin. The meningocele sac was opened and the stalk was dissected from the inner wall of meningocele sac. A slit incision was taken over the sac releasing the cerebrospinal fluid (CSF). Release of the CSF led to decrease in the size of the sac, aiding in easy dissection [Figure 1c].

The neck of the sac could be seen passing through the laminar defect towards the dura. Normal dura was identified cranial and caudal to the neck. Dura was opened and the insertion of the fibroneural stalk inserting into the dorsal sac was identified. The sac was opened from above and the fibroneural stalk was resected from the dorsal thecal sac [Figure 1d].

Post-operative period was uneventful. No worsening or new neurological onset deficit was noted.

Case 2

A 7-month-old girl presented with a swelling over the lumbosacral region since birth. On examination, a solitary lumbosacral LDM was noted with normal bilateral lower limb movements. The skin over the swelling was very thin but intact with brilliant transillumination. Fibroneural stalk could be seen very clearly on MRI and also during the surgery. The patient underwent surgical excision of the LDM with an uneventful post-operative period [Figure 2].

Case 3

A 5-month-old boy presented with a swelling over the lumbar region since birth. On examination, a solitary lumbar LDM was noted over the back along with bilateral club foot. The skin over the swelling was intact but scarred. Attachment of the fibroneural stalk to skin could be clearly elicited during

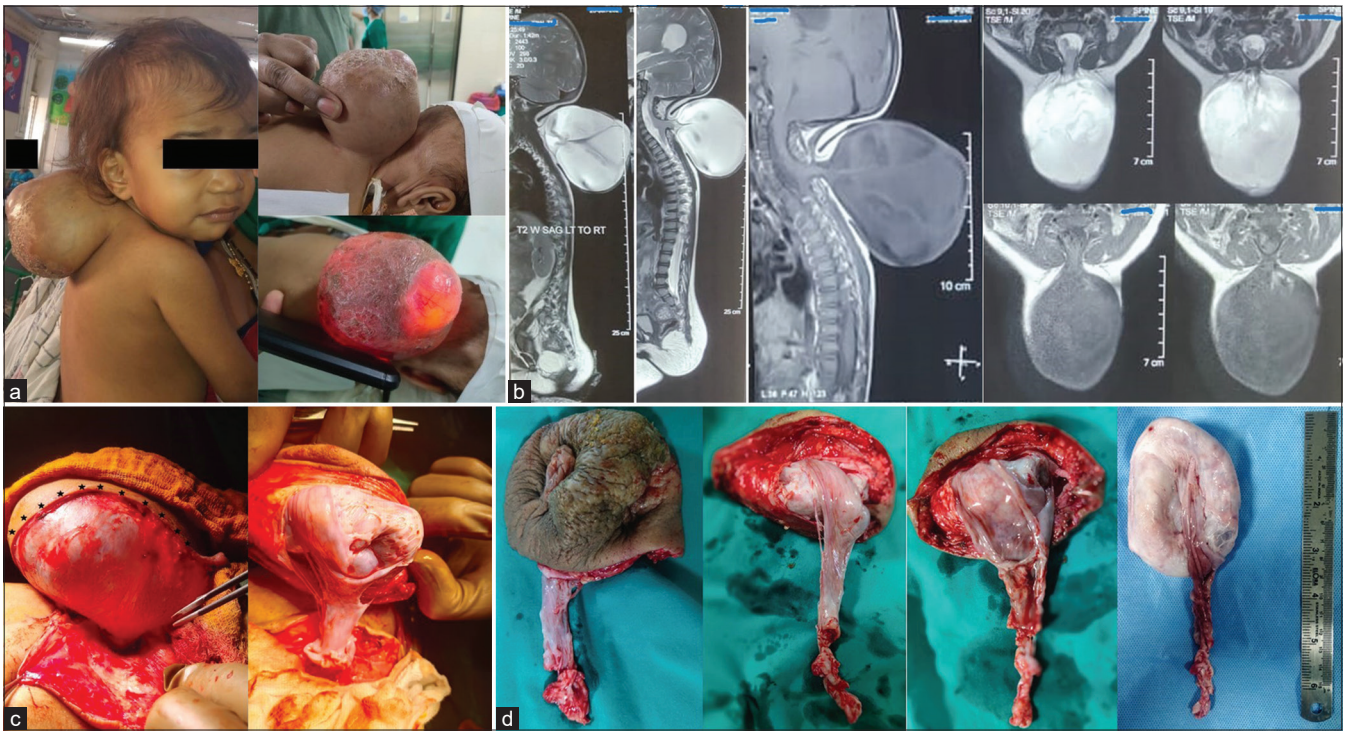

Figure 1: (a) A solitary saccular swelling over the cervico-dorsal region, (b) MRI was suggestive of a large cystic sac-like structure protruding posteriorly through the defect at C7-D1 level, (c) an elliptical incision was marked over the confluence of healthy and non-healthy skin, and (d) insertion of the fibroneural stalk inserting into the dorsal sac.

transillumination. CSF - filled sac was carefully dissected from the surrounding tissues during surgery. The patient had an uneventful post-operative period [Figure 3].

Case 4

A 7-month-old boy presented with a solitary lumbar LDM with normal bilateral lower limb movements. The skin over the swelling was thin but intact. Large CSF - filled membranous dome was carefully dissected from the surrounding tissues during surgery. Surgical excision of the LDM was done using intraoperative neuromonitoring. Post-operatively, the boy recovered without any complications. [Figure 4].

Case 5

A 5-month-old boy presented with an unusual cauliflower-shaped lumbar LDM with normal bilateral lower limb movements. The patient underwent surgical excision of the LDM under intraoperative neuromonitoring with uneventful post-operative recovery [Figure 5].

Case 6

A 13-year-old girl came to us with a solitary lumbar LDM with weakness of both (left > right) lower limbs. The skin over the swelling was thickened. MRI showed a solitary flat L2 level LDM associated with split cord malformation (SCM) Type 2 and dorsolumbar scoliosis. The long fibroneural stalk usually seen in saccular type is absent in these children who have a flat LDM. The patient underwent surgical excision of the LDM under intraoperative neuromonitoring with an uneventful post-operative period [Figure 6].

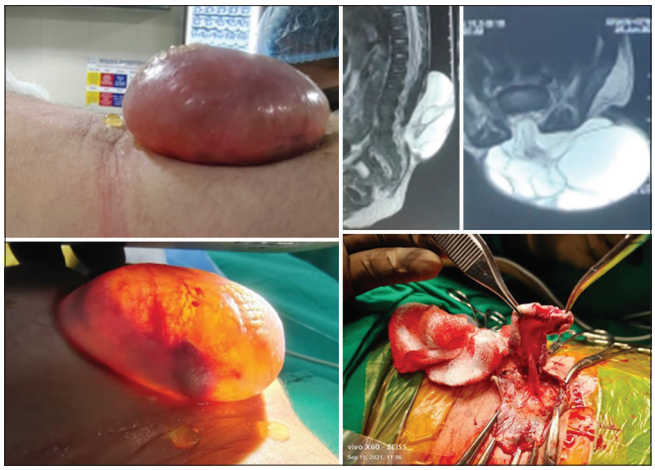

Figure 2: A solitary lumbosacral LDM showing brilliant transillumination with fibroneural stalk.

Case 7

A 7-month-old boy presented with a swelling over the nape of neck since birth. On examination, a solitary cervical LDM was noted with normal power in all four limbs. MRI showed a solitary flat C3-C4 level LDM. During surgery, the LDM was excised in toto. The absence of any CSF - filled sac/tract is notable [Figure 7].

DISCUSSION

LDM is a relatively newly recognized form of spina bifida that is characterized by a fibro-neural stalk between the thecal sac and the inner part of the skin.

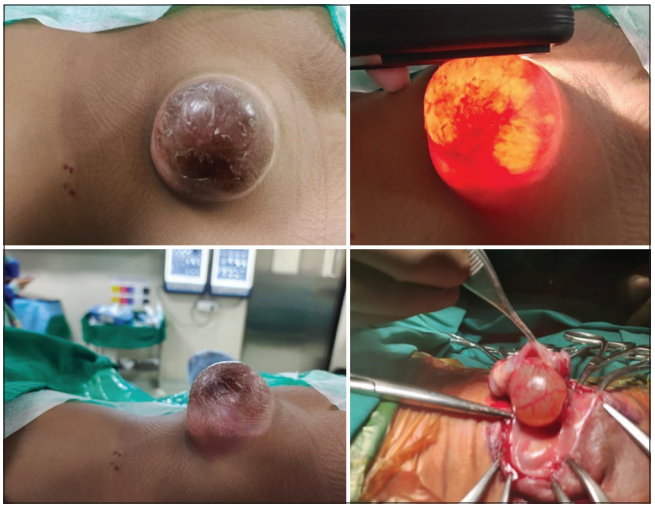

Figure 3: A solitary lumbar LDM showing brilliant transillumination with CSF-filled sac.

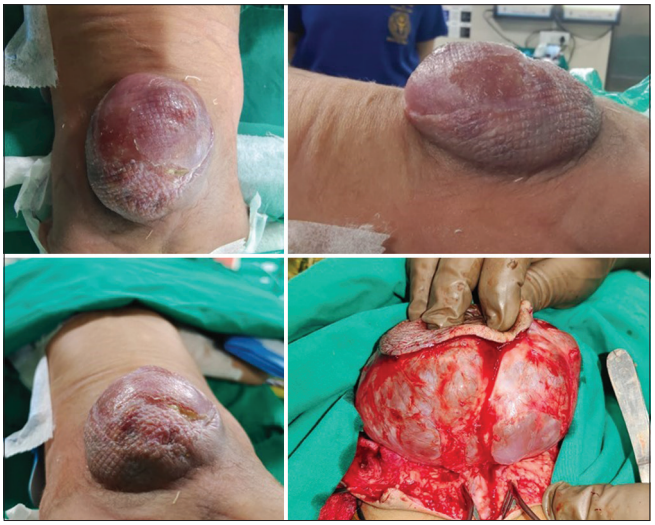

Figure 4: A large solitary lumbosacral LDM showing a large CSF filled membranous dome.

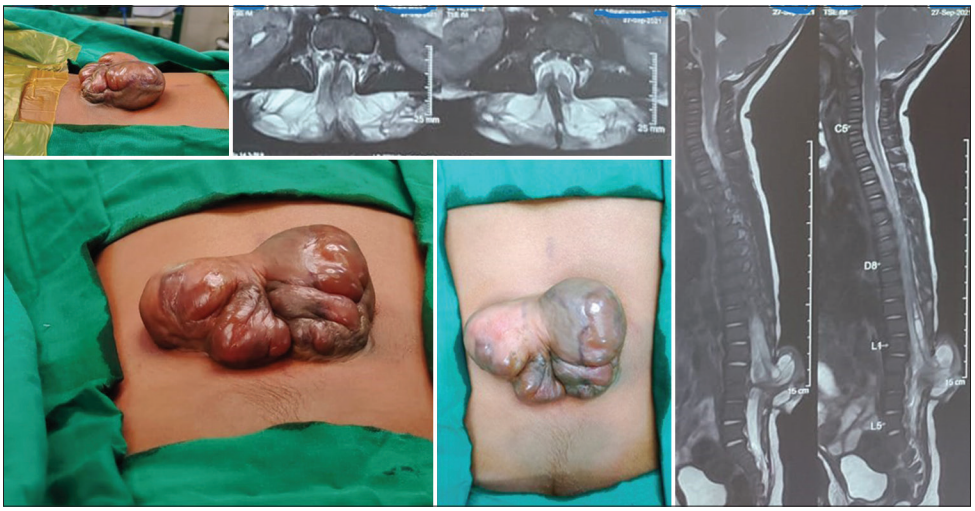

Figure 5: An unusual cauliflower-shaped lumbar LDM.

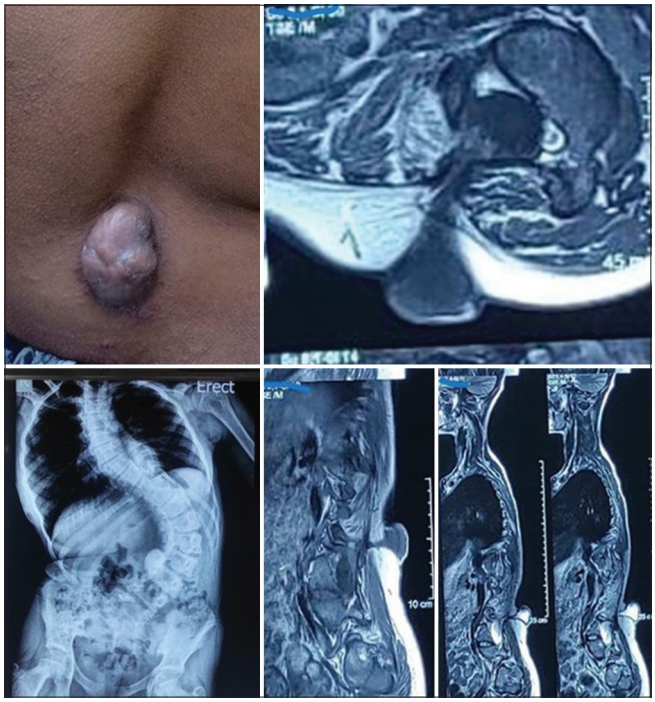

Figure 6: A solitary flat L2 level LDM associated with SCM Type 2 and dorsolumbar scoliosis (notice the absence of long fibroneural stalk that is usually seen in saccular type).

An LDM basically consists of a focal, closed, midline defect and a fibro-neural stalk.[2,3] It was initially described in 1993 by Pang et al, who later ascribed a better definition and a classification system in 2010.[2]

LDMs are considered less severe, as compared to myelomeningoceles, due to the relatively later occurrence of embryologic defect in the process of primary neurulation.[4] The absence of neural placode and the presence of closed skin differentiates it from a myelomeningocele. Most of the steps of primary neurulation including elevation, infolding, and recognition of the dorsal neural folds have happened in cases of LDM except for the final phase of fusion of the opposing neural folds.[5] This incomplete fusion of neural folds leads to the formation of a fibro-neural stalk between the skin lesion and the spinal cord.

In the past, LDMs have been confused with myelomeningoceles, meningocele and myelocystoceles. N. Muthukumar *et al.* (2007) presented a series of 14 terminal and 9 non-terminal myelocystoceles that were managed surgically.[6] A proper radiological evaluation helps in differentiating these overlapping entities. LDMs can vary from being free of any neurological deficit to causing complete quadriplegia depending on their location and tethering of the underlying cord. Preoperative absence of neurological deficits can be considered as a good prognostic marker in these cases.

Usually, LDMs are easily diagnosable at birth, however they do not require immediate surgery like in cases of myelomeningoceles. Surgery is typically advised after the child gains weight (more than 8 kg) or is at least 4 months old to be able to tolerate anaesthesia and the surgical procedure with ease. Immediate surgery is advised in cases of ruptured lesions or in cases of identifiable and worsening neurological deficit.

LDMs can be saccular or flat and can be associated with a plethora of cutaneous and neural manifestations like craters, pits, lipomas, hydrocephaus, syringomyelia, dermal sinus, split cords etc.[2] Saccular LDMs usually have a membranous dome surrounded by parched skin followed by thickened skin with dilated veins. The prolonged pressure from the CSF might be responsible for the formation of this membranous dome and parched skin. If left unattended, this can lead to ulceration and necrosis.

Almost 50% of cervical LDMs were associated with hydrocephalus in a study by Pang et al. MRI of the whole neuroaxis is ideal in these cases to rule out any other anomalies.[7]

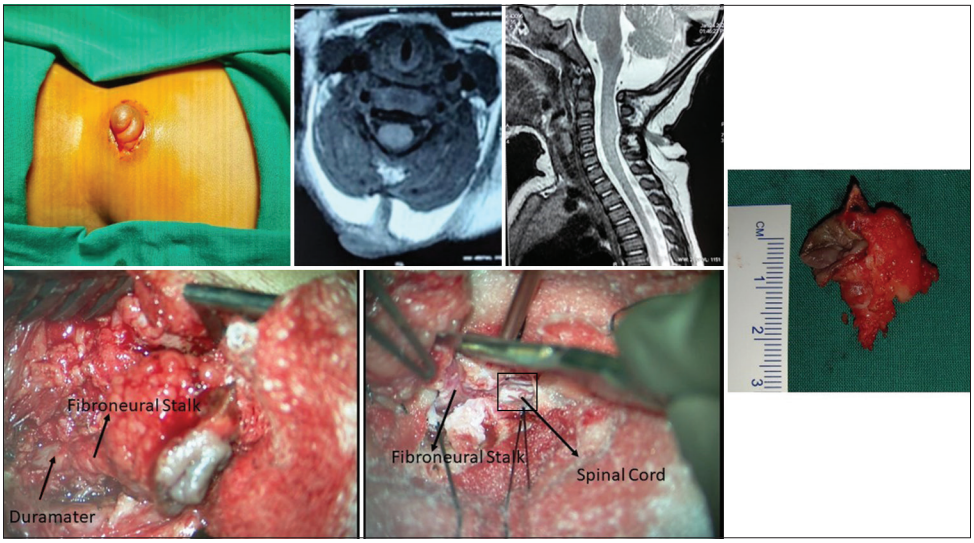

Figure 7: A solitary C3-4 level flat LDM excised in toto. (Notice the absence of any CSF filled sac/tract).

MRI helps to track down the extradural and intrathecal path of fibro-neural stalk thus guiding in the planning of surgery. The tethering of the cord by the fibro-neural stalk can be one of the reasons for tonsillar herniation.

The aim of surgical intervention in these cases is to release the tethering component and achieve good cosmetic results. Saccular LDMs are more disfiguring as compared to flat lesions and hence are brought in for medical consult early. Detethering is the principal aim of surgery to avoid any future neurological deficits. Intraoperative neuromonitoring plays an important role in these surgeries because of almost near normal limb movements in most cases. In absence of detethering, there are higher chances of recurrence.

In our series, we used intraoperative neuromonitoring and the fibro-neural stalk could be successfully detethered without any postoperative neurological deficit in any of the patients. One of the patients developed pseudomeningocele at operative site and it was managed conservatively with compressive dressing. There are high chances of wound dehiscence or infection in these cases because of thinned out skin and dermis. In our series, 2 patients had superficial wound infection, that was managed conservatively with appropriate antibiotics and wound care. The literature on LDMs is still very limited and there are a lot of critical issues yet to be understood about this entity. This condition has been confused with other dysraphic malformations in the past and hence statistical data are scarce and an actual estimate of its recurrence rate is limited.

CONCLUSION

LDM is a relatively new entity as compared to other dysraphic malformations. Accurate identification and early surgical intervention results in good outcome. However, a strict eye should be placed on the follow-up of these cases to identify recurrences.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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