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Lipomyelomeningocele: A Case Report

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Abstract: ***Introduction: Lipomyelomeningocele (LMMC) is a neural tube defect with neural elements incorporated into the spinal lipoma with the prevalence ranging from 0.3-0.6 per 10000 live births. Lipomyelomeningocele is a form of closed spinal dysraphism presenting as a subcutaneous fatty mass over the inter-gluteal cleft. It is a lipoma fused to a dorsally split spinal cord. It can present with cutaneous manifestations, musculoskeletal features, urological features and neurological features. Case: Our patient is a 2 year old girl with no history of complications at birth who was referred to the neurosurgery outpatient department with complaints of swelling in the lower back and a cutaneous dimple since birth. An initial ultrasound at birth had showed features of spinal dysraphism. The patient was asymptomatic with normal strength of the lower extremities. The preoperative study included MRI of the lumbosacral spine with T1 weighted image (plain and contrast) and screening of the brain and was correlated with CT scan at the level of L2 to L5. It showed closed spinal dysraphism with lipomyelomeningocele from L2 to L5 level with tethered spinal cord, herniated tissue measures 11.5 x 4.4 x 7 cm, sacral agenesis with an open defect of 3cm with ossified bony component in the subcutaneous plane at L2-L5 level measuring 5 x 3 cm on the left side. The patient underwent repair of the lipomyelomeningocele under general anesthesia. Conclusion: The management of lipomyelomeningocele is challenging with surgical management being done in patients with progressive symptoms. The clinical management of the lesion depends on the severity. Asymptomatic patients can be treated conservatively with regular follow up. Surgical intervention is planned in symptomatic patients such as sensorimotor deficits, progressive worsening of musculoskeletal abnormalities. Early surgery in asymptomatic patients can prevent the development of neurological deficits.***

Keywords: Lipomyelomeningocele, Meningocele, Myelomeningocele, Lipomeningomyelocele, Spina bifida, Spinal lipoma, Spinal dysraphism, Neural tube defects

1. Introduction

Lipomyelomeningocele (LMMC) is a neural tube defect with neural elements incorporated into the spinal lipoma. The presentation of LMMC varies ranging from mild cutaneous lesions, musculoskeletal abnormalities to sensorimotor deficits and urological dysfunction[1]. The prevalence ranges from 0.3 - 0.6 per 10000 live births[2].

2. Case Report

A 2 year old girl with no history of complications at birth was referred to the neurosurgery outpatient department with complaints of swelling in the lower back and a cutaneous dimple since birth. An initial ultrasound at birth showed features of spinal dysraphism. The patient was asymptomatic with normal strength of the lower extremities.

3. Specific Investigations

The preoperative study included MRI of the lumbosacral spine with T1 weighted image (plain and contrast) and screening of the brain and was correlated with CT scan at the level of L2 to L5. It showed –

- Posterior spinal defect measuring 3cm (cc) noted from L2 to L5 level containing lipomatous tissue with neural elements.
- Herniated tissue measures 11.5 cm (CC) x 4.4 cm (AP) x 7 cm (IR)
- Spinal cord seen to be ending at L3/L4 level
- Enlargement of subarachnoid space and posterior extension of cauda equina attached to lipomatous tissue through spinal defect at L2 to L5 level. This area is predominantly hyperintense on T1 and T2; and hypointense on IR images. No evidence of post contrast enhancement.
- Rest of the vertebral bodies show normal alignment and signal intensity
- Sacral agenesis
- Corresponding CT cuts reveal open defect of 3cm with ossified bony component in the subcutaneous plane at L2 - L5 level measuring 5 x 3 cm on the left side
- Closed spinal dysraphism with lipomeningomyelocele from L2 to L5 level with tethered spinal cord
- Screening of brain and cervix - dorsal spine is within normal limits.

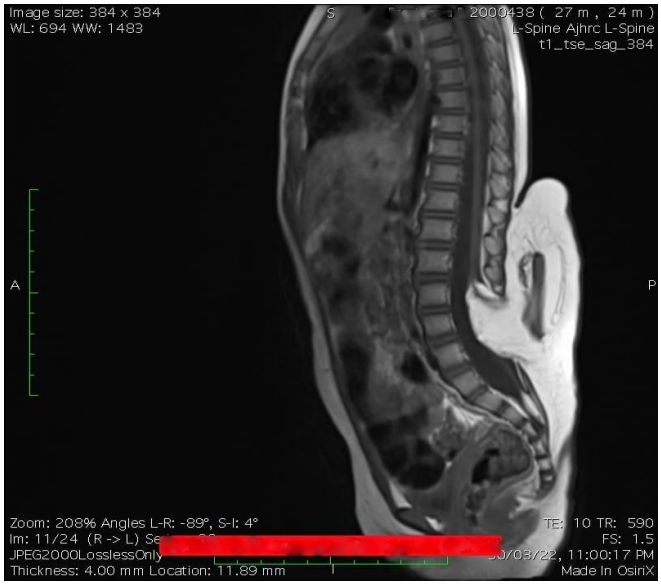

Figure 1: MRI image of the spinal cord showing closed spinal dysraphism with lipomyelomeningocele from L2 to L5 level with tethered spinal cord

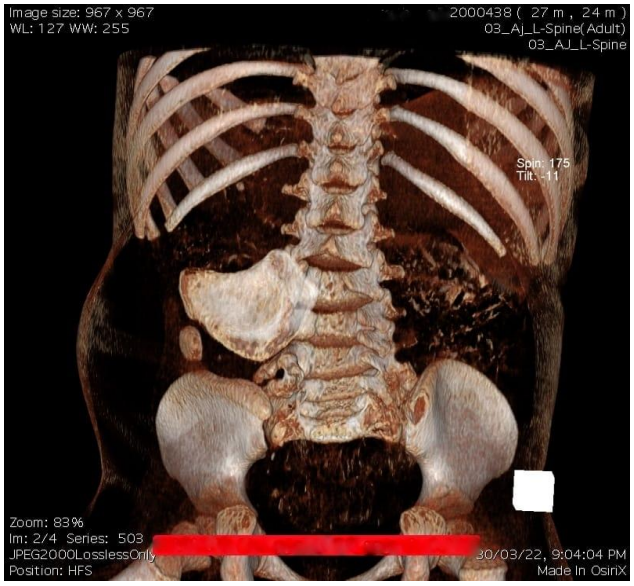

Figure 2: CT scan at the level of L2 - L5 shows open defect with ossified bony component in the subcutaneous plane at L2 - L5 level

Treatment:

The patient underwent repair of the lipomyelomeningocele under general anesthesia with an S shaped incision placed over the midline. Dissection was done upto the dorsolumbar fascia to reach the isolated bone fragment.

Intraoperative finding:

- The bone fragment had cartilaginous content at the centre.
- Poor development of the muscles in the midline
- Absent vertebra in the midline
- Lipomatous content noticed to be entering the cord from the medial side

A small part of the lipoma adherent to the cord filament was left behind. Duraplasty performed using dorso - lumbar fascia and reinforced with fragments of bone nibbled off the isolated bone fragment. Closure was done in layers. Post - operative period was uneventful and the patient was discharged on post - operative day 12 with no complications.

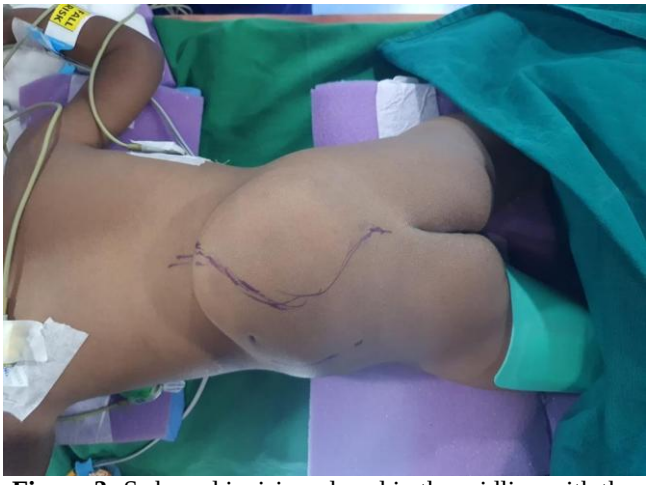

Figure 3: S shaped incision placed in the midline with the patient in prone position

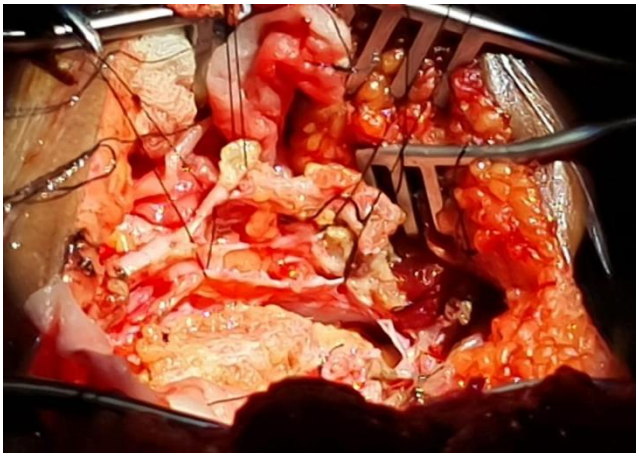

Figure 4: Intraoperative image showing the closure of the dural layer

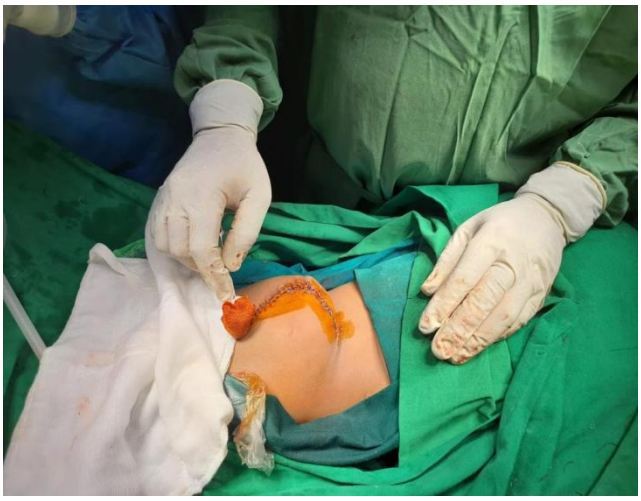

Figure 5: Post - operative image with closure of the wound in layers

4. Discussion

Lipomeningomyelocele is a form of closed spinal dysraphism presenting as a subcutaneous fatty mass over the intergluteal cleft. The predisposing factors of lipomeningomyelocele are unclear. It is a lipoma fused to a dorsally split spinal cord. The lipoma on tethering to the superficial dermal elements can manifest as tethered cord syndrome[3].

The cutaneous manifestations of spinal dysraphism can include a tuft of hair, subcutaneous lipoma, swelling, dermal sinus or a dimple[4]. They can rarely include hypertrichosis or dermal appendages. Musculoskeletal findings are scoliosis, unilateral or bilateral foot deformities, abnormal rotation or asymmetry of the foot or leg. Urological findings include incontinence, frequency, urgency or urinary tract infections. Neurological symptoms are due to tethered cord syndrome and they include back or leg pain at rest worsening with activity, weakness, sensory disturbances, gait abnormalities[1].

The clinical management of the lesion depends on the severity. Asymptomatic patients can be treated conservatively with regular follow up. Surgical intervention is planned in symptomatic patients such as sensorimotor deficits, progressive worsening of musculoskeletal

abnormalities. Those associated with multisystem abnormalities complicate the management[1]. Early surgery in asymptomatic patients can prevent the development of neurological deficits[5].

5. Conclusion

The management of lipomyelomeningocele is challenging with surgical management being done in patients with progressive symptoms. However, prophylactic surgery can prevent the development of neurological symptoms or onset of its deterioration.

References

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