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Case Report

A Lipomyelomeningocele with Tethered Cord Syndrome Associated with Scoliosis and Clubfoot: A Rare Case Report

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Abstract

Background: A lipoma in limbosanal region can be associated with myelomeningocele with tethered cord. Tethered cord means abnormally low conus medullaris with abnormally thick filum terminale attached to lower sacral region. **Objective:** The case was discussed because there is rare association between lipomyelomeningocele with tethering of cord, scolosis and clubfoot. The operation was done by 'S' shaped transverse incision crossing over the middle of the lump. Excision of lipoma was done. Neural placode was identified and dethering of cord and repair of sac was done. **Results:** The muscle power after surgery was improved. There was no bladder disturbance and gait was intact. **Conclusion:** There was rare association of many congenital anomalies. Early diagnosis and surgery prevent future cord infarction and scoliosis, due to undue traction of cord.

Key words: Lipoma; Myelomeningocele; Tethered cord; Clubfoot; Dethering of cord; Excision of tumor; Repair of sac.

INTRODUCTION

Incidence of spina bifida with meningocele or lipomyelomeningocele is seen in 1–2/1000 live births (0.1%–2%). Risk can increase to 2%–3% if there is one previous birth with myelomeningocele and 6%–8% after two affected children. The risk is also increased in families where close relative (e.g., siblings) have given birth to myelomeningocele (MM) children, especially when on the mothers side of the family. Incidence may increase in times of war, famine or economic disasters, but it may be gradually declining overall.[1] Transmission follows non-Mendelian genetics and is probably multifactorial.

Hydrocephalus develops in 65%–85% of patients with myelomeningocele and 5%–10% of myelomeningocele patients have clinically overt hydrocephalus at birth. Over 80% of myelomeningocele patients develop hydrocephalus before the age of 6 months. Myelomeningocele patients will have an associated chiari type 2 malformation, chiari malformation. Closure of the defect may convert a latent hydrocephalus to active hydrocephalus by eliminating route of egress of cerebrospinal fluid (CSF).[2]

Early closure of myelomeningocele defect is not associated with improvement of neurologic function but evidence supports lower infection rate with early closure. Myelomeningocele should be closed within 24 hrs whether or not membrane is intact.[2]

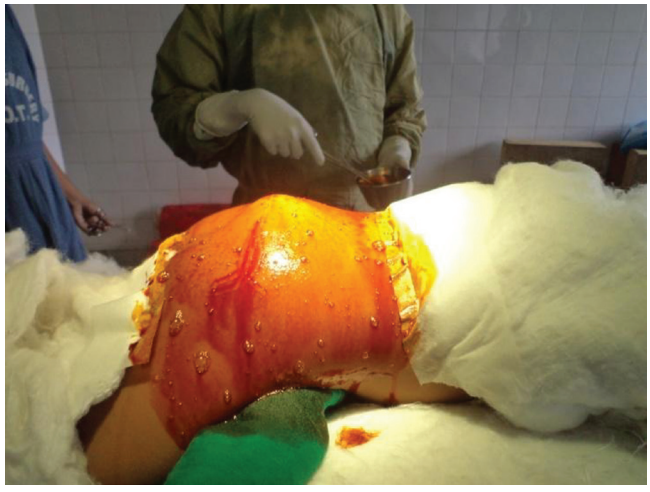

Figure 1: Preoperative lipomyelomeningocele

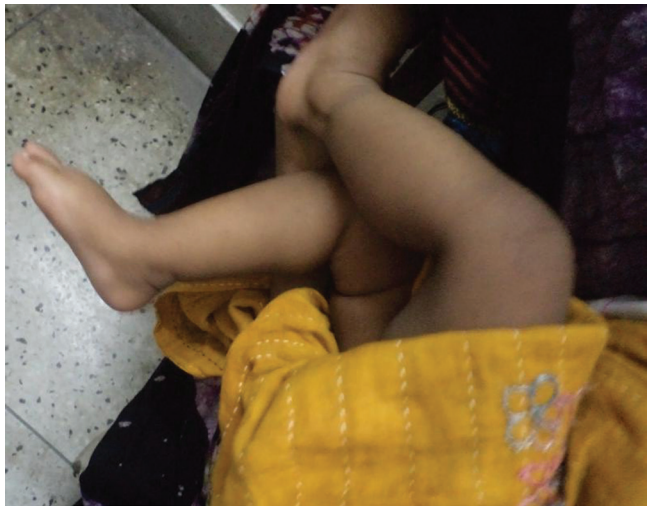

Figure 2: Clubfoot at right side

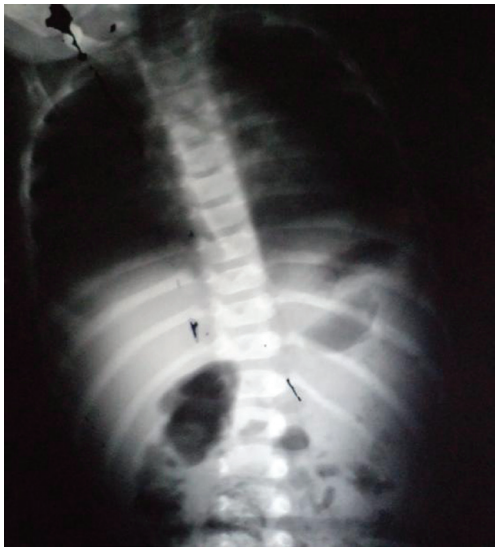

Figure 3: Scoliosis of thoracolumbar spine

Case report

An 11-month old girl had been admitted in the Department of Neurosurgery, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, with the complaints of progressive swelling at right side of buttock, since birth. Swelling was progressive in nature. Solid and cystic consistency, spherical in shape, 12 cm × 10 cm in diameter and 35 cm in circumferences was observed (Figure 1).

The child developed progressive weakness of both lower limbs. Right side was more prominent than left side. Since birth wasting of muscle of both lower limb and more on right leg, calf, thigh muscle, deep tendon reflex were equivocal. Limping gait and a clubfoot was seen on the right side. (Figure 2).

Plain X-ray of lambosacral and dorsal spine showed scoliosis of thoracolumbar spine (Figure 3).

MRI of limbosacral spine showed a lipomyelomeningocele with tethered cord (Figure 4).

There was no hydrocephalus or other anomalies. Her mother's previous child died after birth. Neither a history of consanguineous marriage of her parents nor a history of obstructed labor was recorded. Her mother did not give the history of antenatal checkup. We operated the patient under general anesthesia. With all aseptic precaution 'S' shaped transverse incision was given at lumbosacral region. Subperiosteal dissection was done. Extradural lipoma was excised (Figure 5).

Sac was opened and then repaired. Dethering of cord was done, proper hemostasis was done and a drain was kept in situ. Wound was closed in layers (Figure 6).

There was wound dehiscence at postoperative period. Daily dressing was done and wound was closed by secondary suture. Patient's muscle power remained same as before. For clubfoot, patient was sent to orthopedics specialist. She was advised physiotherapy for limb weakness. After 1 month follow-up, the patient's muscle power improved. She could walk normally without limping.

DISCUSSION

A subcutaneous lipoma that passes through a midline defect in the lumbodorsal fascia, vertebral neural arch and dura merges with an abnormally low tethered cord.[3] These may be terminal, dorsal or transitional.

The intradural fatty tumor may also be known as lipoma of the cauda equine. In addition to being abnormally low, the conus medullaris is split in the midline dorsally usually at the same level as the bifid spine and this dorsal myeloschisis may extend superiorly under intact spinal arches.[4] There is a

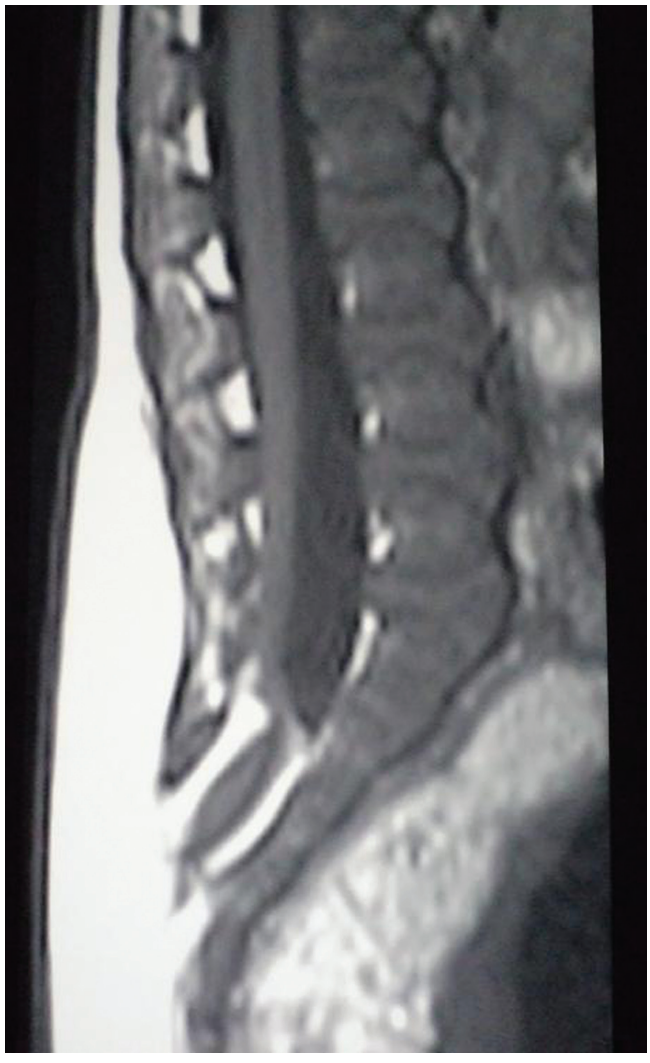

Figure 4: MRI of lumbosacral spine showed tethered cord

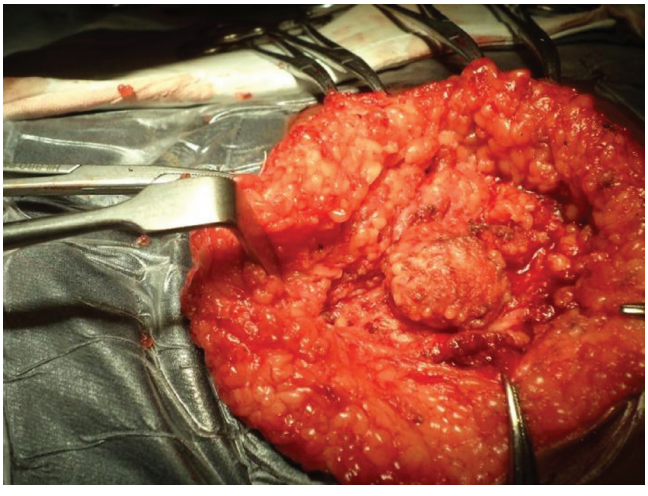

Figure 5: Peroperative photography of lipoma

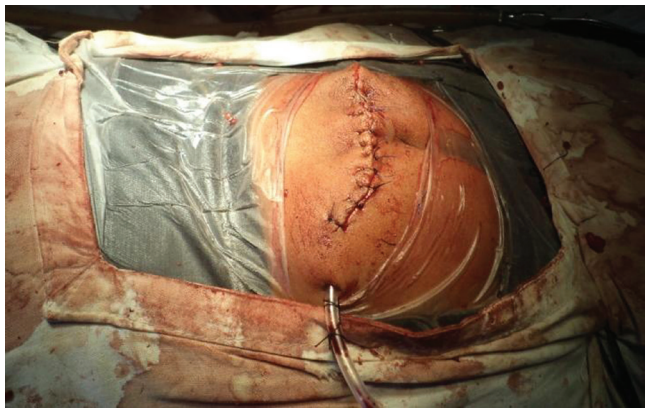

Figure 6: Peroperative photograph after closure of wound

thick fibrovascular band that joins the lamina of the most cephalic vertebrae with the bifid lamina. This band constricts the meningocele sac and neural tissue, causing a kink in the superior surface of the meningocele.

The dura is dehiscent at the level of the dorsal myeloschisis and reflects onto the placode. The lipoma passes through this dehiscence to become attached to the dorsal surface of the placode and may continue cephalad under intact arches with the possibility of extension into the central canal superiorly without dorsal myeloschisis. The lipoma is distinct from the normal epidural fat which is looser and more areolar. The subarachnoid space typically bulges to the side contralateral to the lipoma. These lipomas account for 20% of covered lumbosacral masses.

In a pediatric series, 56% presented with a back mass, 30% with bladder problems and 10% because of foot deformities, paralysis or leg pain.[5]

Almost all patients have cutaneous stigmata of the associated spina bifida: fatty subcutaneous pads (located over the middle and usually extends asymmetrically to one side) with or without dimples, port wine stains, abnormal hair, dermal sinus opening or skin appendages. Clubbing of the feet (talipes equinovarus) may occur. The neurologic examination may be normal in up to 50% of patients most presenting with skin lesion only. The most common neurologic abnormality was sensory loss in the sacral dermatomes.

Plain LS spine X-ray will show spina bifida in most cases (present in almost all by definition, but some may have segmentation anomalies instead such as butterfly vertebrae). Abnormalities of fusion and sacral defects may also be seen. The abnormally low conus can be demonstrated on myelogram/CT or on MRI. MRI also demonstrates the lipomatous mass (high signal on T1W1, low signal T2W1).

All patients should have pre-op urological evaluation to document any deficit.

Since symptoms are due to (1) tethering of the spinal cord, especially during growth spures and (2) compression due to progressive deposition of fat, especially during periods of rapid weight gain; the goals of surgery are to release the tethering and reduce the bulk of fatty tumor. Simple cosmetic treatment of the subcutaneous fat pad does not prevent neurologic deficit and may make later definitive repair more difficult or impossible.

Surgical treatment is indicated when the patient reaches 2 months of age, or at the time of diagnosis if the patient presents later in life. Adjuncts to surgical treatment include evoked potential monitoring and laser. Overall with surgery, 19% will improve, 75% will be unchanged and 6% will worsen. Foot deformities often progress regardless of surgical outcome.[6] Our case had unique presentation with multiple anomalies. The child's parents had no history of consanguinous marriage. Her mother's previous child died after live birth. The child had no hydrocephalous. She improved after surgery. Proper and early diagnosis and treatment of tethered cord with lipomyelomeningocele prevent further neurological deterioration.

CONCLUSION

A tethered cord syndrome with lipomyelomeningocele, with scolosis with clubfoot is rare association. These congenital anomalies can occur individually or together. Proper diagnosis by history taking, clinical examination, with imaging support can be done.

Early surgery can prevent cord infarction, paralysis of lower limb, bladder, bowel and sex dysfunction. Early detethering can prevent future progression of scoliosis.

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