Lumbosacral Lipomyelomeningocele

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Lipomyelomeningocele, is a closed neural tube defect, that usually occurs in the lumbosacral area as a single lesion but can be associated with other spinal dysraphism [1] and Caudal regression syndrome.

Represent a unique population within the spectrum of spinal dysraphism.

Pathogenesis

Premature separation of the neuroectoderm from the ipsilateral surface ectoderm allowing mesenchymal tissue to invade into the central canal, or “premature dysjunction” theory, was proposed for the pathogenesis of dorsal type lumbosacral lipoma. To test this theory, the unilateral neural fold was incised using Hamburger and Hamilton stage 12 or 13 chick embryos. Among 35 embryos evaluated, 15 showed abnormal findings, and of these one showed findings which suggested lumbosacral lipoma: a back lump, blending of the neuroepithelium and mesenchyme through indistinct basement membrane and vertebral body abnormalities. The other 14 embryos showed abnormalities including blunt tails, open neural tube defects, incomplete closure of the dorsal neuroepithelium with intact skin, skin dimples, disorganized gray matter, scoliosis, ectopic neuroepithelium and an accessory spinal cord. The results revealed that the incision of the unilateral neural fold in the early chick embryo may produce a lesion suggestive of lumbosacral lipoma, a finding which supports the premature dysjunction theory. This method needs further refinement to overcome technical difficulties, high mortality, and a low yield before being adopted as an experimental model for lumbosacral lipoma [2].

Pathology

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

Natural history

The natural history of LMMC remains poorly defined. The description and prevalence of the presenting orthopaedic clinical signs and symptoms for LMMC have been infrequent and often documented only in general terms.

Clinical features

Many patients with lumbosacral lipoma are asymptomatic; however, a significant proportion will have neurological deficits present at birth and most develop neurological symptoms over time.

Implication of these deficits with respect to natural history and management are not well understood.

Diagnosis

New dynamic MRI-based parameters to establish the presence and magnitude of tethered cord syndrome (TCS) have been defined. oscillatory frequency (OF) measured the extent of loss of translational cord displacement in supine and prone positions; delta bending angle (ΔBA) defined the relative angulation of conus with lower spinal cord, and sagittal and axial root angles represented ventral nerve root stretching. The difference in OF or ΔBA was minimum in the group with thick filum terminale and progressively increased in the groups with lipomyelomeningocele and meningomyelocele [4)]

Butterfly vertebra may be associated.

Treatment

Untethering surgery.

Surgical technique

From Naidich TP, McLone DG, Mutluer S. A new understanding of dorsal dysraphism with lipoma (lipomyeloschisis): radiologic evaluation and surgical correction. AJNR. 1983; 4:103–116 [5)]

1. mobilize the subcutaneous mass, it funnels down through the deep fascia

2. open last intact vertebral arch (work from normal dura)

3. identify the fibrovascular band that crosses the most cephalic widely bifid lamina

4. sectioning the fibrovascular band frees the dural tube and releases the sharp kink in the superior surface of the meningocele

5. taking care to preserve dorsal nerve roots, the dura is incised anterior to the dura-lipoma junction

6. similar procedure is carried out with arachnoid membrane

7. dural/arachnoid incisions are continued around entire extent of tethered conus

8. cord and placode are untethered;

9. Lipoma is then trimmed as completely as possible, intentionally leaving some fat behind to avoid injury to dorsal surface of placode. Superior extension along dorsal surface of cord or into central canal is debulked as much as is safely possible

10. the placode is reformed into a closed neural tube

11. close the pial margins

12. the dura is closed (primarily if possible, or using fascia lata graft if too much tension is placed on folded placode)

An expansile dural graft should be incorporated in cases of lipomyelomeningocele in which primary dural closure does not permit free flow of CSF [6].

Continuous eCUSA-based stimulation of the cauda equina during LMMC resection is a feasible mapping technique with potential added value improving safety of untethering. Future studies evaluating extension of untethering, as well as the rates of retethering and long-term neurological and urological outcomes, are warranted [7].

Outcome

Lumbosacral lipomas (LSL) of the 'dorsal' type have been associated with more favourable outcomes compared with other conus region lipomas.

Lipomyelomeningocele is a complex congenital anomaly of the neural tube often associated with significant complications, both before and after surgical treatment. These complications can affect the nervous system, musculoskeletal system, and other bodily functions. Below is an overview of the most common and significant complications:

1. Neurological Complications Tethered Cord Syndrome: Progressive stretching of the spinal cord as the patient grows, leading to neurological deficits such as: Pain (localized or radiating). Paresthesias or anesthesia. Weakness or motor deficits in the lower limbs. Bladder and bowel dysfunction (incontinence or retention). Syringomyelia: Formation of fluid-filled cavities within the spinal cord,

leading to sensory and motor impairments. Neuropathic Pain: Persistent or chronic pain due to nerve root irritation or compression. 2. Orthopedic Complications Scoliosis: Abnormal curvature of the spine due to underlying spinal dysraphism or neurological imbalance. Foot Deformities: Clubfoot or cavovarus deformities. Joint Contractures: Limited range of motion in lower limb joints due to muscle imbalance or spasticity. 3. Urological Complications Neurogenic Bladder: Bladder dysfunction leading to: Urinary incontinence. Urinary retention. Increased risk of urinary tract infections (UTIs). Vesicoureteral reflux and kidney damage if unmanaged. Urinary Stones: Secondary to stasis or infection in a neurogenic bladder. 4. Skin and Wound Complications Ulceration or Pressure Sores: Result of impaired sensation in the lower limbs, leading to chronic non-healing wounds. Postoperative Wound Issues: Increased risk of infection or delayed healing after surgical correction. 5. Muscular Weakness and Fatigue Lower Limb Weakness or Atrophy: Due to nerve root involvement or chronic tethered cord syndrome. Gait Abnormalities: Difficulty walking, leading to fatigue or reliance on assistive devices. 6. Recurrent Lipoma Growth Residual or Recurrent Lipoma: Incomplete resection of the lipoma can lead to retethering and recurrence of symptoms. 7. Psychological and Social Impacts Chronic Pain and Disability: May lead to depression, anxiety, or reduced quality of life. Educational and Occupational Challenges: Long-term physical and neurological impairments may limit social and occupational opportunities. Management of Complications Neurological Monitoring: Regular MRI and clinical evaluations to detect tethering or syringomyelia. Physical and Occupational Therapy: Address muscle imbalances, prevent joint contractures, and improve mobility. Urological Care: Proactive bladder management (e.g., clean intermittent catheterization, urodynamic studies). Orthopedic Interventions: Bracing, surgery, or physiotherapy to correct deformities. Pain Management: Use of gabapentinoids, opioids, or spinal cord stimulators in severe cases. Surgical Revision: For recurrent tethered cord or unresolved complications. Early recognition and multidisciplinary management of these complications are critical to optimizing outcomes and improving the quality of life for patients with lipomyelomeningocele.

Meta-analysis

The aim of a study is to compare the outcomes of surgical and conservative treatments of pediatric asymptomatic lumbosacral lipomas, and to address whether the patients can benefit from prophylactic surgeries. The literature reports of surgical and conservative treatments of child asymptomatic lumbosacral lipomas were reviewed and collected, and a meta-analysis of the reports regarding the incidence of sphincter and lower limb dysfunctions was performed. A total of five literatures were collected, containing a total of 403 patients, among which 124 patients received conservative treatments with 32 (25.81%) cases developing neurological dysfunctions during follow-up, and 279 received prophylactic surgical treatments with 30 (10.75%) patients developing neurological dysfunctions in follow-up, the difference being statistically significant ($P \leq 0.05$). For pediatric asymptomatic lumbosacral lipomas of the three major subtypes, the limited source of literature so far suggests that prophylactic surgery is superior to conservative strategy in preventing the patients from neurological deterioration. Larger patient cohorts, randomized studies, and longer length of follow-ups are needed for further corroboration [8].

Case series

Lumbosacral lipomyelomeningocele case series.

Case reports

An 8-day-old infant girl presented to the clinic with a sacral dimple. Imaging demonstrated a tethered spinal cord with low-lying conus medullaris and an LMM. A firm mass was noted in the subcutaneous lipoma. Detethering surgery and removal of the lipoma and mass were performed at the age of 6 months. Pathological examination identified the mass as cartilage, fat, and immature nephroblastic tissue consistent with NR tissue or, less likely, a teratoma with renal differentiation.

This presentation of an LMM associated with an immature teratoma or NR poses a risk of malignant transformation in patients. As a result, careful surgical dissection, resection, and close clinical follow-up are recommended for these patients [9].

Lumbosacral Lipomyelomeningocele with Lateral Attachment to Neural Placode [10].

Fetal lipomyelomeningocele was suspected during the second-trimester ultrasound and confirmed by magnetic resonance imaging. The pregnancy took its course and a term neonate was delivered. At 2 years of age lipomyelomeningocele surgical removal was performed. The patient is now 4 years old and, despite neurogenic bladder, is a healthy boy with normal psychomotor development for his age. This case illustrates the favorable prognosis of this entity and the importance of prompt diagnosis and multidisciplinary counseling [11].

A patient with lipomyelomeningocele (known in utero) presented for MRI characterization prior to surgical procedure at three months of age. Cross-sectional imaging revealed a spinal dysraphism of the lower lumbar spine, with a posterior spinal defect spanning L4 to S2 subcutaneous fat intrusion, and distal spinal cord extrusion. An osseous excrescence was also appreciated, articulating with the left iliac bone. This case demonstrates the youngest known lipomyelomeningocele with accessory limb and the abnormal growth of multiple tissue types at the site of spinal dysraphism-a potential consequence of dedifferentiated cell proliferation originating from a secondary neural tube defect or rachipagus parasitic twinning [12]

Case report from the HGUA

Q12378

A 29-year-old male with a history of lipomyelomeningocele surgically corrected during infancy, presenting with progressive neurological symptoms in adulthood. Imaging revealed significant spinal anomalies, including a low-lying conus medullaris, lipomas, and syringomyelia. The patient underwent neurosurgical intervention for tethered cord syndrome, with slow but favorable postoperative recovery. This case emphasizes the challenges of long-term management in patients with congenital neural tube defects and highlights the importance of a multidisciplinary approach.

Introduction

Lipomyelomeningocele is a rare neural tube defect characterized by the presence of a lipoma associated with spinal dysraphism. Although early surgical intervention aims to prevent severe neurological deficits, complications such as tethered cord syndrome may develop later in life. This case report outlines the presentation, surgical management, and postoperative outcomes of an adult with these sequelae.

Case Presentation

Patient Details:

A 29-year-old male with a history of lipomyelomeningocele surgically repaired at 18 months of age presented with progressive neurological symptoms.

Symptoms at Presentation:

Discomfort and paresthesia in the left lower limb. Numbness extending from the knee to the foot, with anesthesia in the left foot. Fatigue and reduced walking tolerance. Chronic ulceration of the second toe on the left foot, non-healing for two months.

Physical Examination:

Strength: Preserved in both lower limbs. Reflexes: Normal deep tendon reflexes. Sensory findings: Dysesthesias in the left L3 dermatome. Imaging Studies:

MRI Lumbar Spine:

Low-lying conus medullaris at L3. Lipoma intramedullary at L2-L3 and epidural at L4-L5. Syringomyelia at L1-L2. Mild central disc protrusion at L5-S1 with foraminal narrowing. Small foraminal hernia at L4-L5, without significant stenosis.

Management and Surgery The patient underwent surgical release of the tethered cord to alleviate neurological symptoms and prevent further deterioration.

Postoperative Course:

Early recovery:

Good wound healing without infection or cerebrospinal fluid leakage. Successful removal of urinary catheter with spontaneous voiding. Gradual initiation of ambulation, though residual dysesthesias persisted.

Pain Management:

Controlled with oral analgesia.

Follow-up findings:

Improved ambulation with no significant motor deficits. Persistent paresthesia and chronic ulceration in the left foot.

Discharge Plan: The patient was discharged home once pain was controlled, and ambulation was initiated. He was advised to continue physiotherapy, wound care for the ulcer, and outpatient follow-up with neurosurgery and neurology.

Discussion

Lipomyelomeningocele, a complex form of spinal dysraphism, poses significant long-term risks, including tethered cord syndrome. Symptoms such as paresthesia, fatigue, and chronic ulceration in this patient correspond to tethering of the spinal cord and nerve root compression.

Surgical release of the tethered cord is the primary treatment to prevent neurological progression, but long-standing deficits, such as neuropathic pain and trophic changes, may persist. This case highlights the importance of multidisciplinary care, including physical therapy, dermatological support, and pain management, to address residual deficits and improve the patient's quality of life.

Conclusion

This case underscores the challenges of managing long-term sequelae in patients with lipomyelomeningocele. While surgical intervention can halt the progression of neurological symptoms, comprehensive follow-up is essential to address residual deficits and optimize functional outcomes.

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