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Persistent Urinary Incontinence with Progressive Limb Weakness in A 6yr Old Girl as A Presenting Feature of Lipomyelomeningocoele with Tethered Cord Syndrome: the Role of MRI In Diagnosis And Management

Idris S.K[1], Abba A.A[2]

[1]Departments of Radiology Bayero University Kano
[2]Aminu Kano Teaching Hospital, Kano-Nigeria

ABSTRACT

The objective is to report a rare case of lipomyelomeningocele presenting as persistent urinary incontinence with progressive lower limb weakness. Lipomyelomeningocele lies within the spectrum of closed neural tube defects and represents a rare but complex neurological disorder that may present with neurological deterioration secondary to an inherent tethered spinal cord. Diagnosis can be challenging in a resource-limited settings but the availability of magnetic resonance imaging (MRI) proved to be essential in making accurate diagnosis. The case of a 6year old girl with history of 5 months persistent urinary incontinence, progressive lower limb weakness and history of recurrent fever over the last 3 months is presented. Physical examination revealed a 3x2 cm soft, nontender mass in the lower back just above the gluteal cleft. Decreased muscle power of about 4/5 was noted in both lower limbs and the tone was also increased. Lumbosacral MRI revealed a well-defined T1 and T2 hyperintense mass in the sacral aspect of the thecal sac, measuring 27.1mm x 16.6mm in size. The mass was hypointense on STIR sequences. The neural placode-lipoma interface was located outside the spinal canal. There was tethering of the cord which extended inferiorly and terminated at S3 level, it however showed normal thickness and signal intensity. Myelosequence confirmed spina bifida. MRI findings confirmed Lipomyelomeningocoele with tethered cord syndrome.

Keywords: Urinary Incontinence, Limb Weakness, Lipomyelomeningocoele, Tethered Cord, MRI

Corresponding Author:
Name: Abba AA
Address: Aminu Kano Teaching Hospital, Kano-Nigeria
Email: drabbaadamu@gmail.com
Contact: +2348065463048

INTRODUCTION

Lipomyelomeningocoele is a form of closed spinal dysraphism which is a rare neural tube closure defect characterized by the presence of a subcutaneous fatty mass (lipoma) just above the intergluteal cleft that extends through a defect in the lumbodorsal fascia, vertebral neural arch, and dura. This painless lesion can occur anywhere along the spinal canal but usually is found in the sacral or lumbar region (1,2). It is, therefore, a rare but complex neurological disorder that may present with neurological deterioration secondary to an inherent tethered cord (3).

Globally, the prevalence has been found to range between 0.3 and 0.6 per 10,000 live births (2,3). Female to male ratio of 1.5 to 1 is also reported globally (3).

Lipomyelomeningocoele is the commonest cause of congenital tethering in which the lipoma inherently tethers the cord to the adjacent dura and soft tissue (3-5). Tethered cord syndrome (TCS) is a developmental abnormality of the neuroaxis due to a stretch-induced functional disorder of the spinal cord where its caudal portion is anchored by an inelastic structure, generally situated in the lumbosacral cord, leading to neurological damage such as leg weakness, low back pain, scoliosis, sensory and motor problems as well as loss of bowel or bladder control (3-6).

CASE REPORT

K. I. is a 6-year-old female child brought to the Pediatric Emergency Unit (EPU) on account of a 5 months history of progressive lower limb weakness, urinary frequency and incontinence. There was associated history of recurrent fever over the last 3 months. There was no preceding or associated history of trauma. The patient was not a known sickle cell disease patient and there was no previous history of similar illness among her other 4 siblings. No history of maternal hypertension, diabetes and alcohol intake or drug abuse during the index case pregnancy. No maternal or paternal family history of congenital anomaly. Labour and delivery were said to be uneventful and the baby cried immediately after birth. However, a small lump was noted on her lower back just above the gluteal cleft immediately after birth but was said to be reassured by attending midwife and therefore did not seek further specialist evaluation. She attained all milestones without difficulty and was enrolled in a primary school about 4 months prior to initial presentation.

Examination revealed a healthy-looking girl, not pale, afebrile and anicteric. She exhibited a slightly unsteady gait but still could walk unsupported. Further examination revealed a 3x2 cm soft, nontender mass in the lower back just above the gluteal cleft. A slightly decreased power of about 4/5 was noted in both lower limbs and the tone was also increased. Examination of the respiratory and cardiovascular system was essentially normal. A working diagnosis of a closed neural tube defect was made and patient referred to the Neurosurgery unit. Initial lumbosacral x-ray done showed spina bifida occulta. Lumbosacral MRI revealed a well-defined T1 weighted and T2 weighted hyperintense mass in the sacral aspect of the thecal sac, measuring 27.1mm x 16.6mm in size (Figure 1). The mass appeared hypointense on STIR sequences (Figure 2a). There is associated dilatation of the thecal sac and the mass is seen communicating posteriorly with the skin at this level. The neural placode-lipoma interface was located outside the spinal canal (Figure 2b). There is tethering of the spinal cord which extended inferiorly and terminated at S3 level, but however showed normal thickness and signal intensity. Myelosequence confirmed spina bifida (Figure 3). These characteristic MRI findings, confirmed the diagnosis of lipomyelomeningocoele with tethered cord syndrome.

Surgical repair of the lipomyelomeningocoele and untethering of the spinal cord was done, which involved the removal of the lipomatous tissue, repair of myelomeningocele, untethering of the spinal cord and thecal sac reconstruction. The immediate post-operative condition was satisfactory.

The patient was discharged 2 weeks later, on follow-up and

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physiotherapy. Up to the time of reporting the case, there was no any post-surgical complications documented.

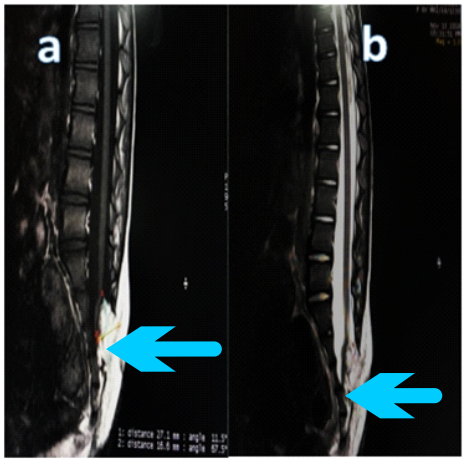


Figure 1: Sagittal lumbosacral MRI showing TIW (a) and T2W (b) hyperintense mass in the sacral segment of dilated thecal sac, communicating posteriorly with the skin and tethering the cord at S3 level (Blue Arrows).

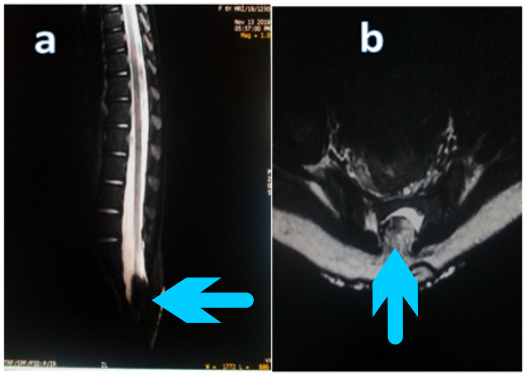


Figure 2: (a) Sagittal T2 STIR lumbosacral MRI showing lesion appearing hypointense due to fat suppression (Blue Arrow). (b) Axial T2W lumbosacral MRI at the level of S3 vertebra showing neural placode-lipoma interface located outside the spinal canal (Blue Arrow).

DISCUSSION

Lipomyelomeningocoele is a type of closed spinal dysraphism which typically presents at birth or during infancy as a lipomatous mass over lower back contiguous with a neural defect and, therefore, lies within the spectrum of closed neural tube defects which represents a rare but complex neurological disorder that may present with neurological deterioration secondary to an inherent tethered cord (1,3,4,6). Several types of closed spinal dysraphisms result from embryological abnormalities during primary neurulation. Those that arise from premature disjunction result in fusion of the spinal cord with fatty elements, the most common of which is a lipomyelomeningocele (2,3,7) In premature disjunction, mesoderm can migrate into the neural tube before it is fully closed, disrupting the neurulation process. This mesoderm then differentiates into fat and forms a border between the neural placode and the now entrapped lipoma (2,3). The neural placode-lipoma interface, which is the connection between the spinal cord



Figure 3: Myelosequence confirming spina bifida at the level of the Lipomyelomeningocoele (Blue Arrow).

and the lipoma, can lie outside, within, or at the edge of the spinal canal. Lipomyelomeningocoele is characterized by a placode-lipoma interface located outside the spinal canal (1-4).

Plain radiograph is an initial screening tool because it is widely available and affordable, however, it is non-specific and inconclusive as it may only show spina bifida (1,2). However, a detailed MRI is the definitive imaging modality of choice for the evaluation of lipomyelomeningocele and can give an accurate diagnosis (2,4,6,7). The anatomical detail of the placode-lipoma junction is clearly shown in relation to the normal spinal cord which further reinforced the diagnosis of lipomyelomeningocoele of the presented case.

Lipomyelomeningocoele is the most common cause of tethered cord (3-5), which is the presenting feature of the index case. Because the natural progression of lipomyelomeningocoele is causing neurologic deficits, and/or urological disturbances, prophylactic surgery is recommended for these lesions by some authors (3,5.6).

Surgical objectives in a lipomyelomeningocoele repair include removal of the adipose mass, identification of the defect in the lumbosacral fascia for release of the tether (untethering), possible release of the filum terminale, preservation of neural elements, and prevention of retethering of the spinal cord (2,3.6). The lipomatous component is intimately associated with the neural placode, which will preclude complete resection of the lipomatous component without neurological injury (2,3,7). Lipoma resection may be achieved utilizing cautery, laser, or ultrasonic aspiration. Additionally, if primary repair of the dura cannot be achieved, duraplasty may be warranted (3).

The clinical conditions of the presented case had improved within 2 months after surgical removal of the lipomatous tissue and untethering of cord. No post-surgical complications were reported.

CONCLUSION:

Lipomyelomeningocele is a rare congenital anomaly with insidious onset but can have progressive serious consequences if it remains undiagnosed and untreated, especially in resource-limited settings. However, MRI proved to be essential in detecting the condition and therefore remains gold standard for diagnosis. It is a potentially treatable condition with early surgical intervention which proved to be beneficial in the index case due to prompt diagnosis by detailed MRI characteristic findings.

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