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Case Report

Late diagnosis of dorsolumbar lipomyelomeningocele with tethered cord in a middle aged adult: A case report from Nepal☆

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ABSTRACT

Closed spinal dysraphism can present with diagnostic issues in settings with limited resources, when knowledge of the disorder and specialized radiological studies, such as magnetic resonance imaging (MRI), may not be readily available. Undiagnosed cases can develop serious neurological deficits. Here, we describe a case of dorsolumbar lipomyelomeningocele, a type of closed spinal dysraphism, presenting in a middle aged with paraplegia complicated by bed sores. A 38-year-old female with no significant past medical history experienced gradually progressive weakness of bilateral lower limbs over 9 years. On physical examination, patient had a soft swelling with hairy tuft over the lumbar spine, paraplegia, grade III bed sore over the gluteal region, and sensory loss below L1 sensory level. Her bowel and bladder sensation were decreased. The soft tissue swelling over her back was not evaluated appropriately before this presentation. MRI of the spine revealed dorsolumbar lipomyelomeningocele with tethered spinal cord.

Introduction

Lipomyelomeningocele is a type of closed spinal dysraphism typically presenting at birth as a subcutaneous lipoma over the lower back and contiguous with a neural defect [1]. It is inherently associated with a tethered cord that causes neurological deficits during infancy or as late as childhood [2]. The prevalence is 3-6 per 100,000 live births [1]. Initial clinical diagnosis of lipomyelomeningocele during infancy might be difficult because almost half of affected infants have a normal neurological examination [1]. Radiological assessment should be performed when cutaneous manifestation of closed spinal dysraphism is present as the condition is potentially treatable

Abbreviations: MRI, magnetic resonance imaging; T1WI, T1-weighted images; T2WI, T2-weighted images.

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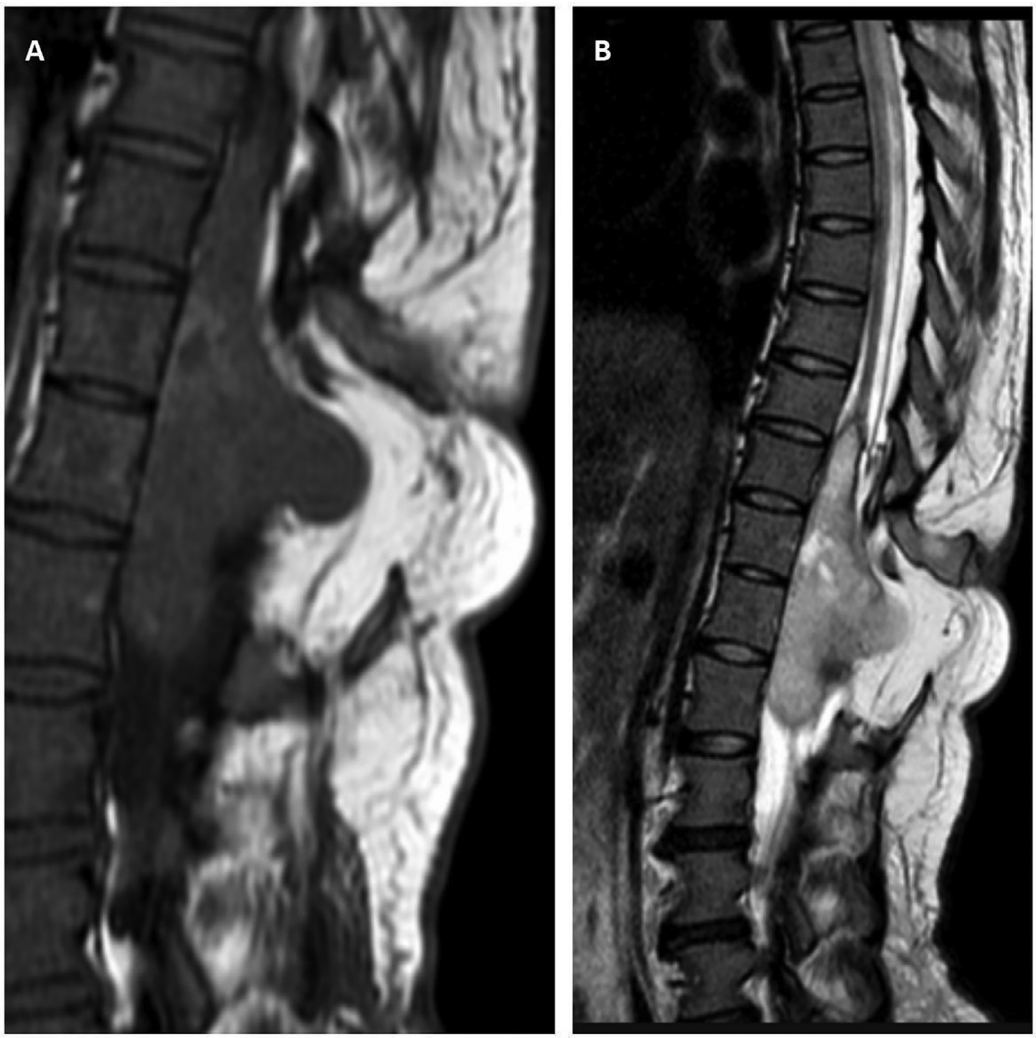

Fig. 1 – (A & B) T1- and T2-weighted MRI images in sagittal plane demonstrate dorsal bony defect at the D10-L1 level. There is a protrusion of the neural placode outside the spinal canal limited externally by the skin and subcutaneous tissue. Few patchy T1/T2 high signal intensity areas are noted within the protrusion (lipomatous elements). The neural placode lipoma interface lies outside the spinal canal.

if detected early [1]. MRI is the investigation of choice for diagnosing spinal dysraphism and for planning surgical management [3].

Case report

A 38-year-old female with no prior past medical history presented to the hospital with paraplegia and pressure ulcer. She had soft tissue swelling over her lower back since infancy but there was no weakness or other neurological deficit during childhood or adolescence. She developed gradually progressive weakness of bilateral lower limbs noticeable since her late 20s. She had paraplegia and decreased sensation of bilateral lower limbs complicated by pressure ulcers at the time of presentation. She also complained of decreased bowel and bladder sensations. There was no family history of similar illness.

On clinical examination, her mental status assessment and cranial nerve examination were normal. There was loss of sensation to all modalities below L1 sensory level and complete loss of motor strength. The rectal tone was decreased. She had a soft tissue swelling in her lumbar region measuring approximately 15 cm × 10 cm, with skin dimpling and a tuft of hair. There were grade III pressure ulcers over her buttocks. Rest of the systemic examinations were normal.

On laboratory investigation, her hemoglobin was 10.1 g/dl, and serum albumin was 3 g/dl. Other lab parameters were within normal range. MRI of the lumbar spine revealed dorsal bony defect at D10-L1 vertebrae with protrusion of the neural placode (primitive neural tissue) outside the spinal canal with T1/T2 high signal intensity in the protrusion, suppressed on STIR sequences indicative of lipomatous element. The protrusion was limited externally by the skin and subcutaneous tissue (Fig. 1). There was low lying conus medullaris with tethering of spinal cord reaching L4-L5 intervertebral level (Fig. 2).

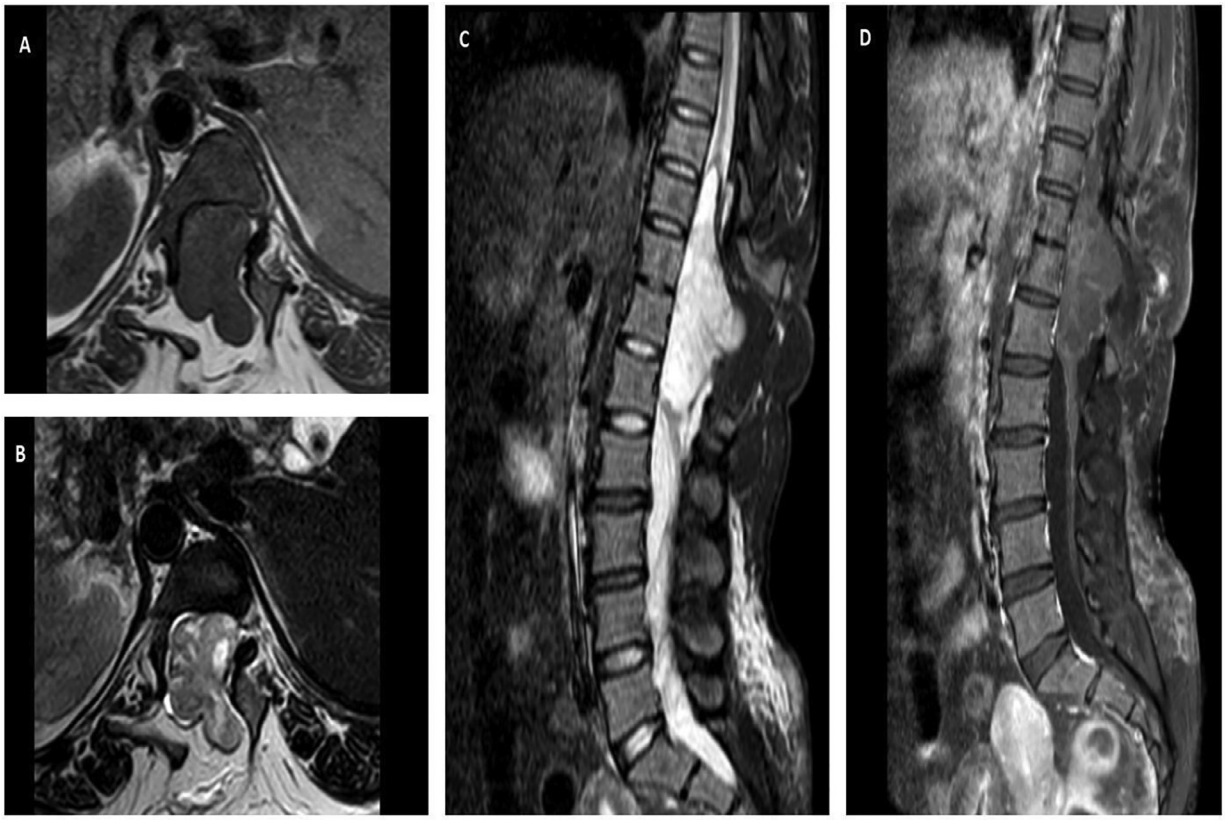

Fig. 2 – (A & B) T1- and T2-weighted MRI images in axial plane shows dorsal bony defect with splaying of the neural arch. There is a protrusion of neural tissue through the bony defect into the subcutaneous tissue plane limited externally by intact skin. Few patchy areas of T1 and T2 high signal intensity areas are noted within the herniated neural tissue. (C) Sagittal STIR MRI images show suppression of high signal areas demonstrated in T1WI and T2WI (lipomatous elements). (D0 Post-contrast MRI images in the sagittal plane shows no enhancing areas within the protrusion and rest of the spinal axis.

However, there was no evidence of herniation of cerebellar tonsils or syrinx (Fig. 3).

Based on the MRI findings, she was diagnosed with dorsolumbar lipomyelomeningocele associated with tethered spinal cord reaching up to the L4-L5 vertebral level. The patient opted conservative management of her newly diagnosed condition due to financial constraints. The grade III pressure ulcer was treated with debridement and flap covering.

Discussion

Lipomyelomeningocele is a type of closed spinal dysraphism typically presenting at birth as a fatty mass over lower back contiguous with a neural defect [1]. The prevalence is 3-6 per 100,000 live births and is more common in females [1]. Since the fatty mass is clinically noticeable at birth, those who are affected are typically discovered before experiencing neurological problems, and up to 48% of cases have been reported to be neurologically intact at the time of the initial diagnosis [2]. High-risk lumbosacral cutaneous manifestation for closed spinal dysraphism include atypical dimples, lumbar masses, large pedunculated lesions, raised lumbar hemangiomas, dermal sinus tract, subcutaneous lipoma, caudal appendage, midline pedunculated swelling, sacral agenesis, and extremely unusual hair patterns. The presence of more than one lumbosacral cutaneous manifestation predicts a considerably higher probability of closed spinal dysraphism than the presence of a single one. All individuals with a lumbosacral cutaneous manifestation should have a spinal ultrasound performed since different lumbosacral cutaneous manifestations have differential risks of closed spinal dysraphism [4]. The patient in our case was a female with soft tissue mass and a hairy tuft over her lower back at the midline since birth. However, the patient was not assessed promptly because of resource limitation, the absence of screening ultrasounds as well as advanced imaging techniques like CT scan/MRI scan and a lack of information regarding the presentation of closed spinal dysraphism.

The conus medullaris can terminate anywhere between the T12 and the L2-L3 interspace due to the different rates of growth for the spinal cord and musculoskeletal components. A "tethered conus" is a conus that is aberrant and below the L2-L3 interspace [5]. Tethered cord is inherently linked to lipomyelomeningocele because the lipoma attaches the cord to the neighboring dura and soft tissue. Lipomyelomeningocele can present with neurological deficits secondary to a tethered cord during infancy or as late as childhood [2]. In line with this finding our case had teth-

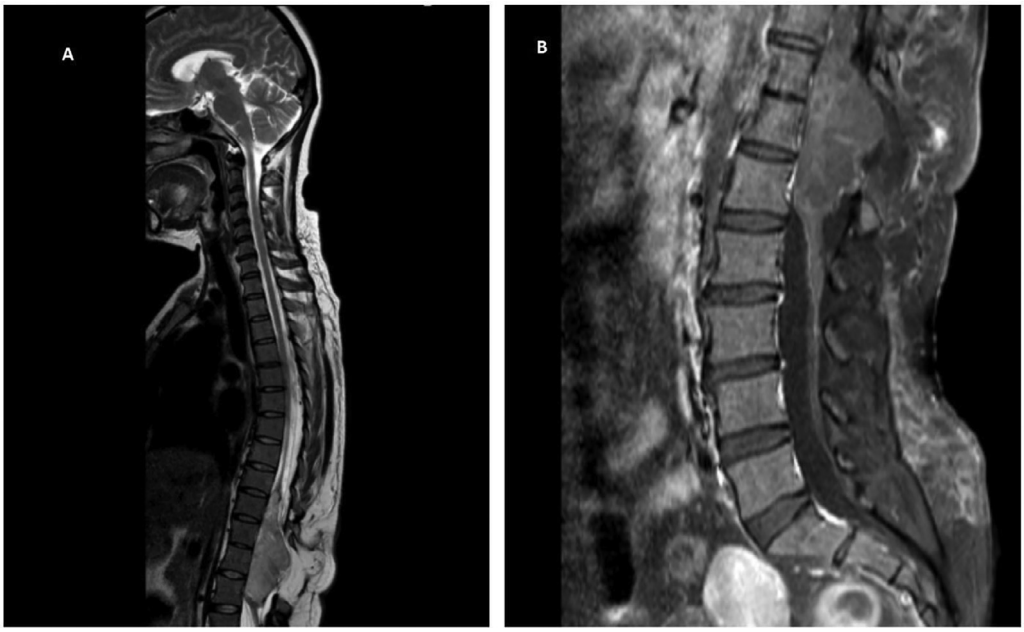

Fig. 3 – (A) T2-weighted MRI image in sagittal plane of the spine shows dorsal bony defect at D10-L1 vertebral level. No evidence of herniation of cerebellar tonsils or syrinx. (B) T1 fat-suppressed post-contrast MRI images in sagittal plane shows low lying conus medullaris with tethering of spinal cord reaching up to L4-L5 intervertebral level. No evidence of enhancing lesion/s in the visualized spinal cord.

ered spinal cord secondary to lipomyelomeningocele, however, she developed progressive neurological deficits starting in her late twenties. Neurological function loss has been reported to increase with age and is thought to be caused by increasing spinal cord stretch caused by axial growth spurts [2]. As the disease progresses, it can cause frequent urinary tract infections, neurogenic bladder and bowel incontinence or constipation, motor and sensory loss, limb length disparity, foot deformities, gait abnormalities, scoliosis, stiffness, and back and leg pain [2]. In our case, the patient's symptoms started in her late twenties and progressed gradually. At the time of diagnosis, there was paraplegia, loss of sensation to all modalities below L1 sensory level, and loss of bowel and bladder control. The lower limbs were spastic on both sides.

Lipomyelomeningoceles are characterized by a subcutaneous fatty mass above the intergluteal crease that usually extends asymmetrically into one buttock. Because of the enlargement of the underlying subarachnoid spaces, the neural placode-lipoma interface is located outside the spinal canal, resulting in a posterior meningocele [6]. MRI scan revealed similar findings in our case. Lipomyelomeningocele can be associated with other pathologies such as Chiari malformation type 1 (13%), spina bifida (14.4%), split cord malformations (3.1%), related dermal sinuses (3.1%), dermoid or epidermoid cysts (3.1%), diastematomyelia (3.1%), terminal hydromyelia (3.1%), anal stenosis (1.0%), and Down syndrome (1.0%) [3]. There were no associated findings in our case.

The condition is potentially treatable if detected early. In patients with progressive neurologic deficit, surgical detethering is indicated [1]. Bowel and bladder dysfunction is one of the most common morbidities of lipomyelomeningocele; hence the appearance of these symptoms should prompt immediate surgical intervention. Delayed surgery have been linked to worse outcomes [2]. In our case, neurosurgical intervention was deferred as the patient chose non-surgical management due to financial constraints.

Conclusion

Lipomyelomeningocele is a rare condition that, if untreated, can have serious consequences. The cutaneous signs of this condition must continue to be discussed with primary care practitioners to promote an early diagnosis and referral to expert services. Furthermore, lipomyelomeningocele can present as neurological deficits in adults, therefore clinicians should suspect the diagnosis even in adults. The case study also demonstrates the diagnostic challenges clinicians may face in areas with limited resources when radiological imaging is not always available.

Patient consent

Consent from the patient was taken on written form for case report and using the MRI images in any journal after explaining in her own language.

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