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Case Report

A rare case of thoracic lipomyelomeningocele in a young female: A case report ☆

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ABSTRACT

Thoracic lipomyelomeningocele is a rare type of congenital occult spinal dysraphism. It is characterized by lipomatous tissue connected to the dorsal spinal cord that protrudes through a spinal defect together with the meninges or spinal cord to form a posterior mass beneath the skin. Closed spinal dysraphism can present diagnostic challenges when resources are scarce and advanced imaging techniques like magnetic resonance imaging are not readily available. Here, we describe a case of thoracic lipomyelomeningocele, a type of closed spinal dysraphism in a young female presenting with gradually progressive weakness and tingling sensation in bilateral lower limbs over the last 6 months. On physical examination, she had a soft tissue swelling with dimpling over the dorsal spine and paraparesis. Magnetic resonance imaging of the spine revealed dorsal lipomyelomeningocele corresponding to D4-D7 vertebral levels with tethered spinal cord.

Introduction

Lipomyelomeningocele is a closed neural tube defect that occurs due to failure of primary neurulation and occurs in approximately 3-6 out of 100,000 live births [1]. It is characterized by a subcutaneous lipoma that is generally located in the lumbar or sacral region in the midline or slightly off the midline [2]. The subcutaneous lipoma extends via defects in the dura, vertebral neural arch, and lumbodorsal fascia before attaching to the tethered spinal cord. The majority also have other skin conditions, such as hairy nevus, skin dimples, and cutaneous hemangiomas associated with the lipoma. Since the fatty mass is clinically noticeable at birth, those who are affected are typically discovered before experiencing neurological problems, and almost half of the cases have been found to be neurologically intact at the time of their initial diagnosis [3]. Lipomas that are subcutaneously located often behave in a benign manner; nevertheless, depending on their location they may induce pressure symptoms [4]. Clinical manifestations could include soft tissue mass, sensorymotor loss, or bladder dysfunction [5]. The radiological features of lipomyelomeningocele can vary significantly depending on the size of the lipoma and meningocele, along with

Abbreviations: MRI, Magnetic resonance imaging; T1WI, T1-weighted image; T2WI, T2-weighted image.

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the direction of the neural placode. Magnetic resonance imaging (MRI) typically shows enlargement of the spinal canal with the cord and the dura extending dorsally through the spinal dysraphism [3]. Here, we report a case of a thoracic lipomyelomeningocele with tethered spinal cord in a 19-year-old female who had presented with progressive weakness and tingling sensation of bilateral lower limbs.

Case report

A 19-year-old female presented with a 6-month history of progressive weakness of bilateral lower limbs. She had a soft tissue swelling in her upper back, which she had noticed since childhood without associated neurological deficits. The condition, however, had not been considered important by her parents for seeking treatment. She sought medical attention due to progressive weakness and tingling sensation in her bilateral lower limbs. Physical examination revealed a compressible lump measuring approximately 5×4cm in her upper back with associated dimpling of skin in the swelling region. Her mental status and cranial nerve examination were normal. Baseline laboratory parameters were within normal range. There was no history of back pain, urinary complaint, and spinal or extremity deformity. Her sensory assessment and motor strength were normal. There was no evidence of saddle anesthesia, and the rectal tone was normal.

MRI spine revealed a cystic mass with lipomatous component adjacent to D4-D7 vertebrae, extending from the subcutaneous region to the cord through the defect in the neural arch (spina bifida) measuring approximately 13.6 mm in the D5 vertebra (Fig. 1A). Cystic component showing CSF signal intensity containing neural elements and a small fat component (Figs. 2 and 3) was noted extruding from the defect into the subcutaneous tissue plane. In addition, tethered cord reaching L4-L5 vertebral level was also noted (Fig. 2C). Furthermore, an enhancing intramedullary lesion (Fig. 4B) was identified at D6 vertebral level with syrinx proximal and distal to the lesion. Similarly, a non-enhancing CSF signal intensity intradural extramedullary cystic lesion adjacent to C4-C5 vertebral level was also noted (Fig. 2A and B). MR imaging findings were consistent with thoracic myelomeningocele with lipoma with an associated tethered cord. The patient was planned for tethered cord release surgery and excision of the lipoma.

The patient thereby underwent C5 laminectomy with excision of intradural extramedullary lesion along with excision and repair of spinal dysraphism at D4-D7 vertebral levels. Histopathology examination of the excised specimen from D4-D7 vertebral levels revealed keratinized squamous epithelium with the specimen showing bands of collagen fibrils, bundles of smooth muscles, mature adipose tissue and nerve fibers; the findings being compatible with lipomyelomeningocele. She was discharged on seventh post-operative day with no adverse events post-surgery.

Discussion

Lipomyelomeningocele is a rare congenital spinal anomaly that is more common in females with the prevalence of 3 to 6 per 100,000 live births. Lipomyelomeningocele falls into the category of closed spinal dysraphism which occurs because of a primary neurulation defect from nondisjunction, where mesenchymal tissue penetrates the neural placode and forms lipomatous tissue [1].

Those who are affected are often identified before developing neurological symptoms since the lipomatous mass is clinically visible at birth, and up to half of the instances have been documented to be neurologically intact at the time of initial diagnosis. Most of the patients also have other skin conditions, such as hairy nevus, skin dimples, and cutaneous hemangiomas that are associated with lipoma [3].

The patient in our case was a young female who presented with a midline soft tissue mass over her upper back since her birth. The condition, however, had not been considered important by her parents for seeking treatment which was the reason for her delayed presentation.

The conus medullaris can end anywhere between the T12 and the L2-L3 interspace. A conus that is abnormal and lower than the L2-L3 interspace is referred to as "tethered" as in our

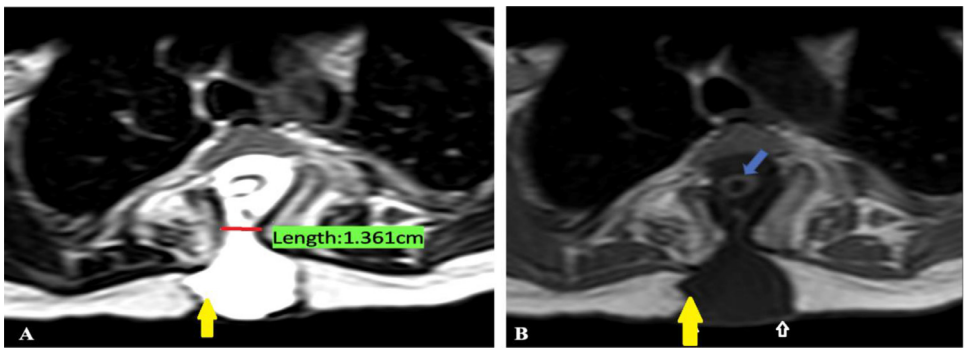

Fig. 1 – Axial MRI images (Figure 1A-T2 weighted and Figure 1B-T1 weighted) demonstrate a bony defect in the neural arch of D5 vertebra measuring approximately 13.6 mm (red horizontal line in Figure 1A). A cystic protrusion showing high signal intensity on T2 weighted image (yellow arrow in Figure 1A) and low signal intensity on T1 weighted image (yellow arrow in Figure 1B) is also noted. The overlying skin is intact (white arrow in Figure 1B). In addition. there is dilatation of the central canal of the spinal cord (blue arrow in Figure 1B).

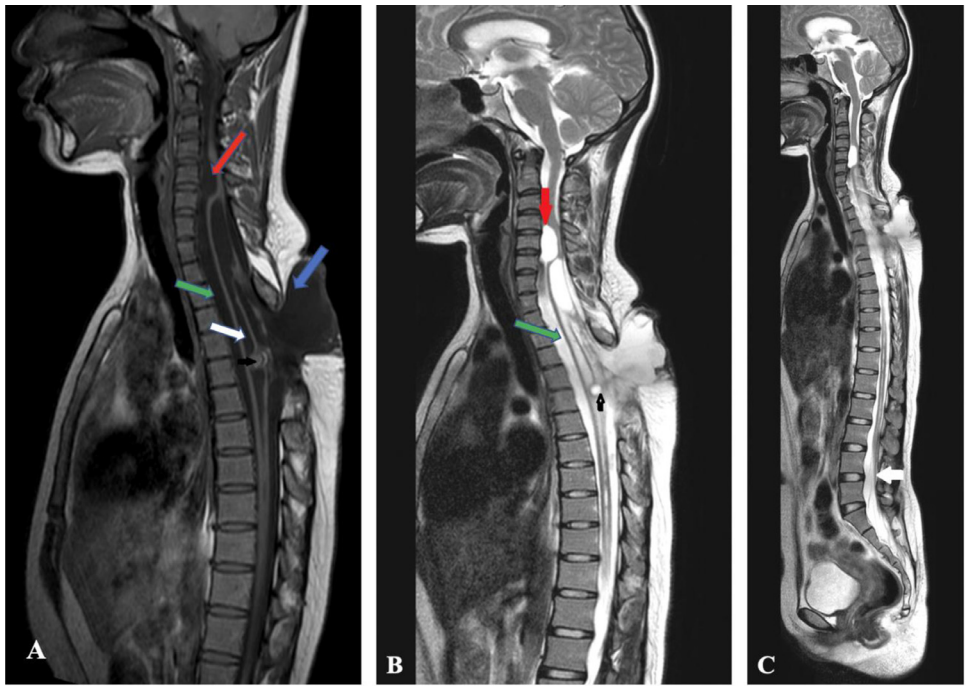

Fig. 2 – Sagittal MRI images (Figure 2A-T1 weighted image), (Figures 2B, 2C-T2 weighted images) demonstrate CSF-filled cystic protrusion through the neural arch of D5 vertebra which also contains neural elements (blue arrow in Figure 2A). There is an intradural extramedullary CSF-signal intensity lesion adjacent to the C4 and C5 vertebral levels (red arrow in Figures 2A,2B) which has caused indentation in the adjacent spinal cord. In addition, there is dilatation of anterior subarachnoid space spanning from the level of C6 vertebral body to D7 vertebral body (green arrow in figure 2B). A tiny lesion is noted in the central canal of spinal cord at D6 vertebral body level demonstrating intermediate signal intensity on T1 weighted image (black arrow in Figure 2A) and high signal intensity on T2 weighted image (black arrow in Figure 2B) . Furthermore, there is dilatation of central canal of spinal cord (syrinx) spanning from the level of C6 vertebral body to L3 vertebral body (white arrow in Figure 2A). Tethered spinal cord reaching L4-L5 vertebral level is also noted (white arrow in Figure 2C).

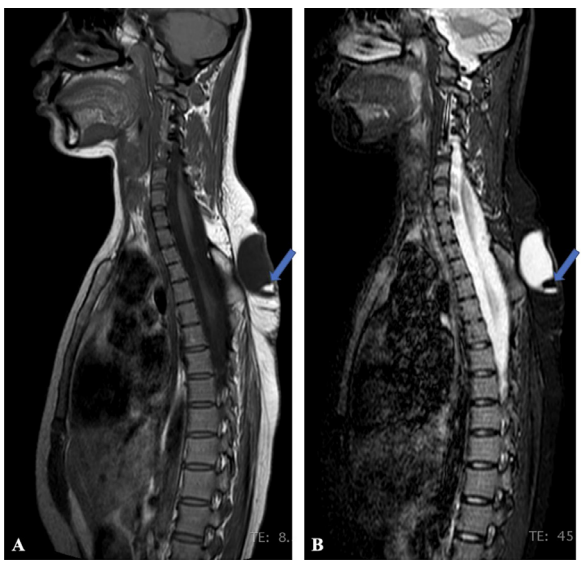

Fig. 3 – MRI sagittal images (Figure 3A-T1 weighted, Figure 3B-STIR sequence) demonstrate a high signal intensity area in the inferior aspect of the cystic protrusion (blue arrow in Figure 3A) which is suppressed in STIR sequence (blue arrow in Figure 3B), suggestive of fat component.

case where the conus has reached up to L4-L5 level. Because the cord is connected to the nearby dura and soft tissue via the lipoma, the tethered cord is fundamentally associated with lipomyelomeningocele [6].

The patient in our case had a lipomyelomeningocele-related tethered spinal cord who gradually showed neurological impairments over the previous 6 months which is likely due to enhancing nodule in central canal of spinal cord at D6 vertebral level and developing syrinx. Impairment in neurological function has been reported to increase with age and is thought to be caused by increased spinal cord stretch caused by axial growth spurts [4]. A common presentation of lipomyelomeningocele is a subcutaneous fatty mass in the lumbosacral region [3] above the intergluteal crease unlike in our case where the location is in thoracic spinal segment and a similar presentation has not yet been reported. In addition, there were lesions in spinal axis in the intramedullary and intradural extramedullary location, the association of these lesions with the anomaly could not be obvious from the literature.

For asymptomatic cases of lipomyelomeningocele, conservative management is preferred. Surgical detethering is recommended for patients with neurologic deficits that worsen over time. Patients with progressive symptoms should undergo surgery. The objectives of surgery include releasing the

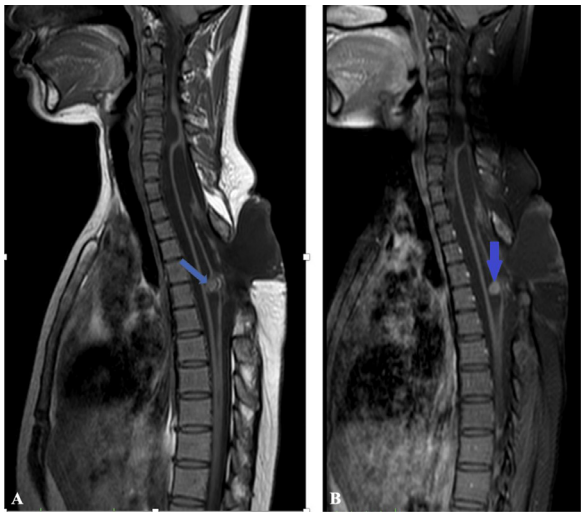

Fig. 4 – MRI Sagittal images (Figure 4A-Pre-contrast T1 weighted image, Figure 4B-fat saturated T1 post-contrast image) demonstrate low signal intensity lesion in the central canal of the spinal cord corresponding to D6 vertebral body (blue arrow in Figure 4A) which shows significant enhancement on post-contrast image (blue arrow in Figure 4B). However, no other enhancing lesion/s was noted in the rest of the visualized spinal axis.

spinal cord from tethering, removing the adipose mass while protecting neuronal components, and avoiding retethering [2].

Conclusion

Dorsal lipomyelomeningocele is an extremely rare congenital lesion. Although infrequent, lipomyelomeningocele may present with neurological deficits in adults. Therefore, all young patients with paraparesis or quadriparesis should have a clinical examination to look for cutaneous signs of spinal dysraphism for early diagnosis and management.

Author contributions

1. **Dr. Suraj Sharma**—Analyzed and interpreted the patient data and contributor in writing the manuscript.
2. **Dr. Hensan Khadka**—Provided guidance, observed and supervised the entire authorship process. Major contributor in writing and organizing the manuscript.
3. **Dr. Sajiva Aryal**—Contributed in writing the manuscript and organization of images in the manuscript.

Patient consent

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

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