

Cervical Lipomyelomeningocele Presenting with Progressive Motor Deficit: A Case Report and Review of the Literature

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Established Facts

- Lipomyelomeningocele is the most common form of spina bifida occulta.
- Most often, it is located in the lumbosacral area.

Novel Insights

- Cervical spine is a very rare location for lipomyelomeningocele, which is asymptomatic at birth.
- If untreated, the cervical lesion can present with tethered cervical cord syndromes comprising pain and progressive weakness.
- Lipomyelomeningocele resection associated with cervical cord untethering is considered to improve the symptoms.

Keywords

Lipomyelomeningocele · Spinal dysraphism · Cervical cord · Neural tube defects

Abstract

Cervical lipomyelomeningocele is a very rare form of spina bifida occulta, which can cause some complications following tethered cord syndrome. We report a 10-year-old female with a history of progressive upper-extremity weakness, a very small soft-tissue mass at the posterior aspect of her neck, and evidence of lipomyelomeningocele in her radio-

logical study. The patient underwent laminectomy of C6 and C7 together with resection of lipomatous tissue attaching to the cord from superficial tissue and cord untethering, which resulted in progressive improvement of her weakness.

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Introduction

Lipomyelomeningocele (LMM) as the most common form of occult spinal dysraphism occurs in approximately 3–6 out of 100,000 live births. It comprises a subcuta-

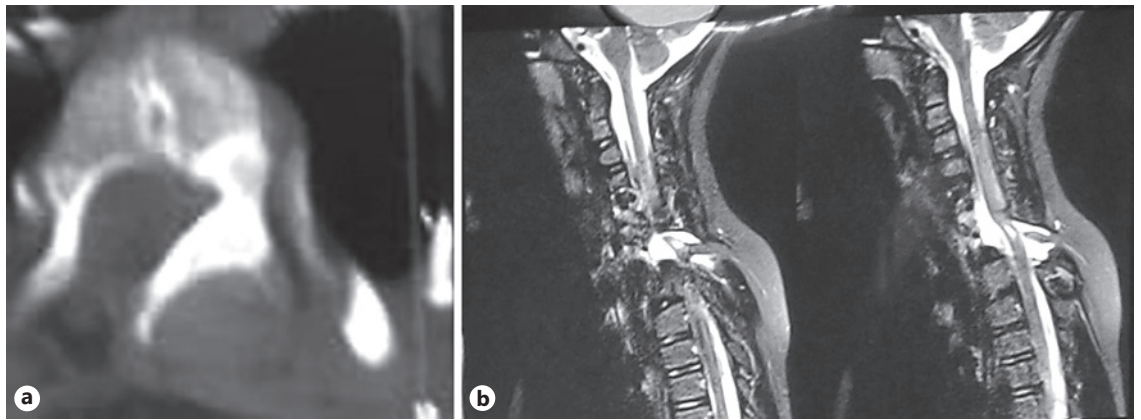


Fig. 1. **a** Axial view of the cervicothoracic CT scan, showing the spinous processes and lamina defect, especially on the right side. **b** Sagittal view of the cervicothoracic MRI of the patient, showing extension of the superficial mass to the dorsal aspect of the cord through the defect of the posterior spinal elements, suggestive of LMM.

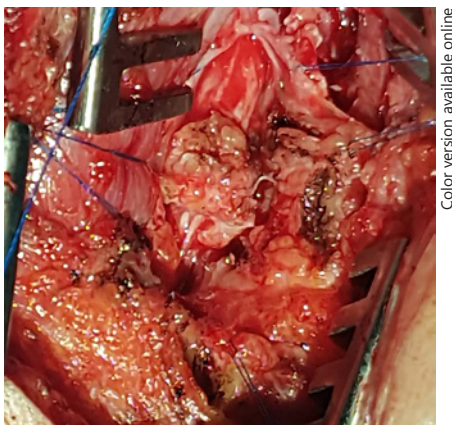


Fig. 2. Intraoperative photography showing fatty tissue attached to the spinal cord at the dorsal aspect of the neural placode.

neous fibro-fatty tissue extending to the intradural space that attaches to the cord. Most LMMs occur in the lumbosacral region, whereas a cervical location is very rare [1, 2]. In addition to soft-tissue mass skin lesions such as cutaneous hemangiomas, hairy nevi and skin dimples can be found at the same location [2]. Here, we report a cervical LMM in a 10-year-old female with tethered cord symptoms.

Case Presentation

A 10-year-old female was referred to our center with a 2-month history of progressive muscle weakness of the right hand. The patient had right-hand interosseous muscle atrophy and distal muscle force reduction (I/IV). The other extremities

seemed to be normal upon physical examination. Since her birth, there was only a small soft-tissue mass bulging at her midline cervical region that, however, had not been considered important so far by her parents. There was no other associated skin abnormality, cervical pain, urinary complaint, and spinal or extremity deformity.

The cervicothoracic computed tomography (CT) scan showed bony defect in the spinous process and lamina, especially on the right side at the C6–T1 levels (Fig. 1a). Spinal magnetic resonance imaging (MRI) revealed a cervical (C7) lipomatous mass extending from the subcutaneous region to the cord through the posterior element defect, suggestive of dorsal-type LMM (Fig. 1b). There was some cord retraction to the posterior associated with a small syrinx.

She underwent laminectomy of C6–C7, which had some congenital laminar defect. The lipomatous tissue was attached to the cervical cord, as in dorsal-type LMM. The lipoma adhering to the spinal cord was resected from the surface to near the spinal cord, and cord tethering was released (Fig. 2). The cord was attached to the lipoma, on the right side more than on the left. The neural placode with a thin layer of lipoma remaining on it was closed with a pial suture. The primary motor-evoked potential was 900 mV during surgery on the right side and 100 mV on the left side. After resection of the lipoma and cord untethering, motor-evoked potential changed to 200 mV on both sides and showed better electrophysiological discharges intraoperatively. Her postoperative period was uneventful. Muscle force gradually improved within a few months after surgery.

Discussion

LMM is a rare congenital spinal abnormality that affects females more than males [2]. Contrary to open neural tube defect, LMM pathogenesis is not known very well and might not be affected by prenatal folic acid.

However, a higher incidence of LMM in mothers belonging to younger and older age groups (bimodal distribution curve) besides previously reported familial cases of LMM suggest a possible inherited nature and a genetic role [3, 4].

Most LMM patients are neurologically asymptomatic on gross physical examination. As the patient's age increases, compression and tethering of the cord continue. Some symptoms such as urinary frequency or incontinence, bowel problems, neurogenic bladder, constipation, leg length disparity, foot deformity, gait abnormality, back and leg pain, muscle weakness, scoliosis, and spasticity may develop subsequent to spinal cord stretching and perfusion disturbance [4–6].

While the clinical and paraclinical manifestations of lumbar and sacral LMM are well described, cervical LMM is extremely rare, and to the best of our knowledge, only a few cases of cervical LMM in the pediatric age group have been reported so far [7, 8]. One of these cases was a girl with lateral thoracic LMM reported to have progressive soft-tissue mass swelling at the level between the left Th9 and Th12 paraspinal region [5]. Gürkanlar and Gonul [8] reported a 22-year-old individual with normal neurological examination who had a posterior cervical mass in the C6 region managed surgically after LMM diagnosis. A right cervical LMM in a 1-day-old neonate has been described before that was concurrent with multiple anomalies and cervical diplomyelia [7].

MRI has an essential role in the diagnosis and treatment of LMM. MRI findings include expansion of the spinal canal and subarachnoid space, dorsal extension of the cord, and dura through the spinal dysraphism. A deformed, stretched, and segmental neural placode is a common finding that is rotated towards the lipoma, mostly on one side. Meninges herniation also occurred on the opposite side. The spinal roots on the side of the lipoma were shorter than on the opposite side where the meninges herniate and emerge closer to the neural foramina [5].

Conservative management is preferable for asymptomatic cases of LMM. Although progressive symptomatic patients should be managed surgically [9]. Surgical goals include removal of the adipose mass with preservation of neural elements, release of the tethered spinal cord, and prevention of retethering [10].

Selection of early- or late-time surgical intervention is still a controversial issue. More literature suggests early surgery before any neurological dysfunction [11, 12]. However, Cochrane et al. [13] suggest that based on the

morphology of the malformation, delayed surgical intervention until the appearance of dysfunction has a better outcome.

Another factor that can affect the timing of surgery is morbidities of LMM. In case of lumbar and sacral LMM, bowel and bladder dysfunction constitute the highest morbidities, and appearance of neurogenic bladder or any new changes in bowel or bladder dysfunction should lead to surgical intervention, otherwise any delay may result in outcome worsening [14]. Outcome is dependent on the preoperative function of the patient. Results of the case series regarding the outcome of LMM revealed that sensory and motor deficits may benefit from surgical intervention, but preoperative urodynamic status is a primary predictor for the postoperative outcome of normal bladder function [13–15].

However, due to the extreme rarity of cervical LMM, appropriate timing for surgical treatment in cervical LMM is unknown. Some authors suggest that the recovery of motor and sensory deficits happens more frequently than the recovery of bowel and bladder function [6]. Cord untethering and releasing all associated adhesions are mandatory for the completion of surgery. If there is an obvious soft-tissue mass in the posterior cervical area, MRI screening is helpful to discover the tethered cord.

In the current study, we present a rare case of cervical LMM with no noticeable mass lesion since birth. Due to the lack of an apparent cervical mass, no previous neuroimaging study was performed before the onset of neurological symptoms. The patient presented progressive interosseous muscle atrophy and distal muscle weakness of the right hand due to cord tethering by LMM at the cervical level. The patient's symptoms were treated successfully with cord untethering.

Conclusion

Cervical LMM is an extremely rare congenital lesion. If cervical mass is evident, MRI can delineate the tethered cord and associated intradural lipoma. If no soft-tissue mass is seen, the diagnosis may defer to the time when neurological symptoms are presented. Surgical management is helpful to eliminate the symptoms caused by LMM mass effect and cord retraction.

Statement of Ethics

This study was approved by the Faculty of Medical Sciences Ethics Committee, Iran University of Medical Sciences.

Disclosure Statement

The authors have no conflicts of interest to declare.

Author Contributions

Farideh Nejat and Syed Vahid Heidari designed the project and supervised the accuracy and integrity of all stages of manuscript preparation. Sina Asaadi, Ehsan Farhadi, and Hasan Ghandehari contributed to patient management, data interpretation, and writing the discussion of the manuscript. Mehdi Soulat, Karen Bavand, and Fatemeh Abbasi contributed to writing the manuscript. Reza Mollahoseini developed the final version of the manuscript. All authors were involved in the management of the patient from admission to follow-up, worked on the draft, and revised it together.

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