Turkish Archives of Pediatrics

CASE REPORT
DOI: 10.14744/TurkPediatriArs.2020.90187

Nondysraphic spinal intramedullary lipoma: A rare case and management

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What is already known on this topic?

- *Spinal intramedullary lipomas not associated with spinal dysraphism are very rare.*
- *The treatment of these pathologies is difficult and clear guidelines for management are still not well established.*

What this study adds on this topic?

- *Recommendation and supporting the approach of partial resection with follow-up.*
- *Aiming for total removal will result in a worse outcome.*

ABSTRACT

A 14-year-old female patient presented with symptoms of chronic mid and low back pain that radiated to both lower limbs for 5 months, with rapidly progressive lower limb weakness and urine retention. Radiologic evaluation revealed an intramedullary mass at the level of first to sixth thoracic vertebrae. The patient underwent surgery and intraoperative findings showed an intramedullary mass lesion composed of well-differentiated fat tissue. A postoperative histopathologic examination revealed mature adipose tissues consistent with lipoma. Post-operatively patient showed improvement in lower limb motor power and started an extensive rehabilitation program.

Keywords: Intramedullary lipoma, laminoplasty, spinal tumor

Introduction

Spinal intramedullary lipomas not associated with spinal dysrhaphism are very rare lesions with an incidence estimated as 0.45 to 0.6% (1, 2). The treatment of these pathologies is difficult and clear guidelines for management are still not well established. In this report, we present a rare case of nondysraphic spinal intramedullary lipoma, with a discussion of the clinical presentation and treatment options for this rare entity.

Case Presentations

A 14-year-old female patient was referred to the emergency service reporting chronic back pain radiating to both lower limbs for 5 months. She described progressive weakness in both lower limbs for 1 week. A neurological examination revealed lower limb weakness, with motor power of 0/5 on the left leg and 1/5 on the right leg. Paresthesia was found starting from the fourth thoracic (T4) level, with hyperreflexia in both lower limbs and decreased perianal sensation. Thoracic spine magnetic resonance imaging (MRI) revealed an intramedullary mass lesion seen at the level of T1-T6, which was hyper-intense on both T1 and T2-weighted images, with slight signal suppression on fat suppression sequences (Figure 1a and b).

The patient underwent surgery in the prone position and a midline skin incision at the level of the tumor was performed. Bone exposure was performed by en-block laminotomy from T1-T6 using a high-speed drill and Kerrison rongeur. The dura was opened and a posterior midline myelotomy was performed to expose the tumor. The intramedullary yellowish fatty and highly-vascularized mass was compressing the spinal cord internally and bulged extensively as the myelotomy was performed (Figure 1c). There was no definite cleavage between the tumor and spinal cord. Under the microscope and using low thermal coagulation

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Received: 25.12.2019
Accepted: 07.10.2020
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Cite this article as: Abuzayed B, Alawneh K, Al Qawasmeh M, Raffee L. Nondysraphic spinal intramedullary lipoma: A rare case and management. Turk Arch Pediatr 2021; 56(1): 85-7.

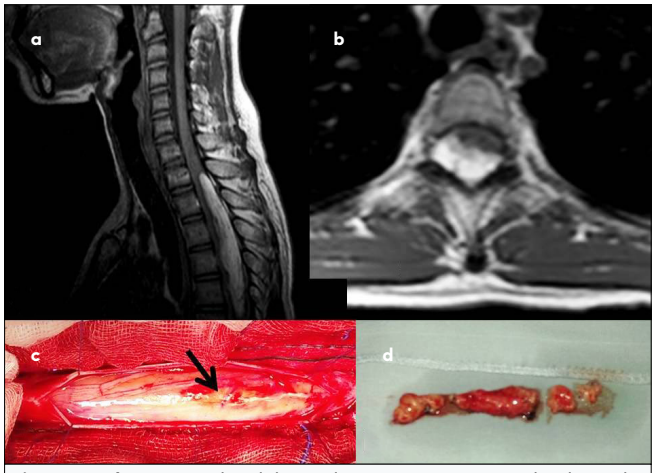

Figure 1. a-d. (a) Sagittal and (b) axial pre-operative T1-weighted spinal MRI demonstrating the intramedullary lipoma. (c) Intra-operative view after dural opening showing the yellowish lipoma (arrow) causing diffuse swelling of the spinal cord and bulging from the opening. (d) The internal part of the lipoma resected

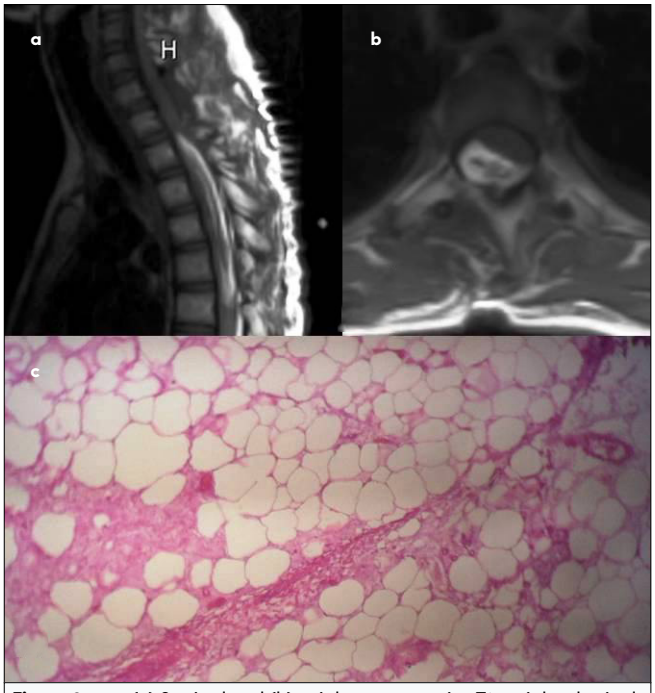

Figure 2. a-c. (a) Sagittal and (b) axial post-operative T1-weighted spinal MRI after 2 months demonstrating the reduced volume of the tumor and internal debulking. (c) A Photomicrograph shows mature adipose tissue separated by thin fibrous bands. No neuronal tissue was identified (X100 hematoxylin and eosin)

with bipolar and micro-instruments, internal debulking of the tumor was performed until reaching the firm outer portion of the tumor. The outer part was very adherent to the spinal cord, thus it was left in place (Figure 1d). Hemostasis was achieved, dural closure was performed primarily without the need for duroplasty and laminoplasty was performed using mini-plates and screws.

In the early postoperative period, the patient showed improvement of lower limb motor power (left 1/5, right 2/5) and normal urinary function. Post-operative MRI showed significant internal debulging of the tumor with rim-like remnants in the outer part of the tumor cavity (Figure 2a and b). A histopathologic examination revealed mature adipocytes with fibrous tissue and few vascular channels with no signs of malignancy, with final diagnosis of lipoma (Figure 2c). Written informed consent was given by the patient's parents.

Discussion

Spinal lipomas not associated with spinal dysraphism are considered as tumors, thus termed 'true' lipomas (3). Their incidence ranges between 0.45-0.6%, mostly showing symptoms in the third decade with no sex predominance (4). They generally have a growth pattern of extending along several segments of the spinal cord, mostly in the thoracic levels (4).

Histologically, lipomas are composed of highly vascularized lobulated fatty tissue. The origin of these lesions is unclear. It is suggested that they originate from mesenchymal cells migrating from the ectoderm before the closure of the neural tube or embryonic crests of fat cells' inclusion during the embryonic period (4). These mesenchymal cells differentiate into adipose tissue rather than the normal evolving to dura mater (5). This theory of a mal-developmental process at the stage of germ cell layers cleavage indicates that intramedullary lipomas are non-neoplastic tissue; accordingly, they are considered true hamartomas that behave according to the general fat metabolism of the patient (6). The metabolic characteristics of both congenital intraspinal lipoma cells and normal adipocytes are identical with the capability of growth and regression. Lipoma cells can be affected by excessive weight gain, or treatment with corticosteroids, with consequent systemic fat accumulation, corresponding to the slow evolution with periods of clinical worsening interspersed with periods of remission (5, 6).

The clinical presentation depends on the location of the tumor. The most common symptoms are sensory deficits and gait disturbances (7). Other possible presenting symptoms include localized pain and/or motor deficits in the extremities, Brown-Sequard syndrome, and signs of long tract involvement such as clonus, hyperreflexia and Babinski (8, 9). The symptoms may be long-standing and show slow progression (7, 10). In our case, the patient presented with chronic back pain for 5 months, that radiated to her both lower limbs and associated with numbness and weakness in both lower limbs started progressively in the last 1 week.

Diagnosis can only be confirmed using MRI with the classic radiologic appearance of fat tissue of increased signal intensity in both T1- and T2-weighted images because intramedullary lipomas present with nonspecific symptoms of compromise of the spinal cord (2, 9).

The main goal of surgery is the decompression of the spinal cord. Symptomatic patients could have clinical improvement following decompression, which can be achieved through bony decompression only, partial tumor debulking or duraplasty (2, 11). There is no difference between subtotal removal and total removal in terms of risk of recurrence; however, total removal showed a higher risk of postoperative neurologic dysfunction (12). For this reason, it is widely accepted that complete removal of a spinal cord lipoma is not recommended when it is adherent to and/or invading the spinal cord without a clear surgical

plan (11). A 44% improvement rate is reported for symptomatic patients after decompressive surgery, and complete resolution of symptoms is reported 14.2% (12). However, 6% showed deterioration after surgery (12). These results show that surgery for spinal lipomas is safe and influences the improvement of the symptoms; however, it seldom results in complete cure of symptomatic patients. For this reason, early untethering for any kind of symptomatic spinal lipoma is advocated (2). In our case, despite the late presentation of the patient and the progressive weakness in both lower limbs, surgery provided a good outcome in terms of pain relief and improvement regarding motor weakness.

Continuous intraoperative electrophysiologic monitoring is a useful method for performing a more safe resection with maximum tissue removal and has been shown to result in much better long-term progression-free survival than partial resection (13). An ultrasonic aspirator was used to reduce the tumor mass volume. It is better tolerated by spinal cord tissue surrounding the lesion than bipolar cautery due to heat conduction (1, 11). However, in cases when the tumor is fibrotic and hard, CUSA may have a limited effect (11). In such cases, the use of a carbon dioxide (CO_2) laser can provide gentle resection. CO_2 lasers act by vaporizing the tissue by ablation (11). The emitted energy is concentrated and focused, providing precise tumor reduction without disrupting the connecting neural structures (11). It is applied over the exposed surface of the tumor, reducing it layer by layer until the liponeural interface is exposed (11). In our case, the outer part of the tumor was fibrotic and hard, and without a clear surgical plan between the lipoma and the neural tissue. For this reason, we used electrocautery at low sittings in a safe manner using the hard outer part of the tumor as a clear debulging limit.

Informed Consent: Written informed consent was obtained from patient's parents who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – B.A.; Design – B.A.; Supervision – K.A.; Funding – B.A.; Materials – M.A.; Data Collection and/or Processing – L.R.; Analysis and/or Interpretation – L.R.; Literature Review – K.A., M.A.; Writing – B.A.; Critical Review – K.A.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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