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Journal of Advanced Clinical Neurology Research

DOI: *doi.org/10.63721/25JACNR0104*

Paraplegia Revealing a Giant Nondysraphic Intramedullary Lipoma Mimicking a Dermoid Cyst: A Case Report and Review of the Literature

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Citation: Oumar Coulibaly, Kouassi Jean Marie Maurin Kisito Quénum, Daouda Sissoko, Mahamadou Dama, Drissa Kanikomo, et al. (2025) Paraplegia Revealing a Giant Nondysraphic Intramedullary Lipoma Mimicking a Dermoid cyst: A Case Report and Review of the Literature. J of Adv Clin Neu Res 1(2), 01-05. WMJ/JACNR-104

Abstract

Background: *Nondysraphic intramedullary lipomas are rare benign tumors and represent less than 1 % of all intramedullary tumors. They are slow-growing tumors which can cause progressive neurologic deficits due to local extension.*

Case Description: *A 38-year-old woman, with no significant medical history, was admitted within our department for 5 months history of thoracic back pain associated with progressive numbness of the lower limbs with bladder and bowel disturbance. Cervicothoracic spine MRI revealed a huge intramedullary lipoma extended from C6-T3. She underwent a total removal of the lesion. The post-operative course was usefulness after 12 months of physiotherapy.*

Conclusion: *Nondysraphic intramedullary lipomas are extremely rare. There is no consensus regarding the management of these lesions. Gross total resection can be an option for severe neurological compromised patients following by physiotherapy.*

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Submitted: 27.10.2025 **Accepted:** 01.11.2025 **Published:** 15.11.2025

Keywords : Intramedullary Lipoma, MRI, Surgery

Introduction

Spinal cord lipomas are rare benign lesions and constitute less than 2 % of all spinal cord tumors [1,2]. They are mostly associated with spinal dysraphism and are usually located in the lumbosacral region with subcutaneous lipoma in few cases [3-5]. Pure intramedullary lipomas not associated with spinal dysraphism are very rare [2,6]. These tumors grow slowly and can cause neurological deficits due to mass effect on the spinal cord. In this paper, we reported a nondysraphic spinal cord lipoma in a 38-year-old woman nourished by erroneous divine considerations, admitted paraplegic with bladder and bowel retention .

Case Report

A 38-year-old woman, with no significant medical history, was admitted within our department for 5 months history of thoracic back pain associated with progressive numbness of the lower limbs with bladder and bowel disturbance. She's suffering from asthma and had been delivered by cesarian just 01 month before admission. This woman came from her distant village, located far from Bamako and filled by false evil beliefs contributing to her diagnostic delay. In the interrogatory, no trauma or no fever was noted in her past. Neurological examination found a paraplegic patient (strength 2/5) with spastic reflex in both sides with bladder and bowel retention. MRI of the cervicothoracic spine in T1 and T2 weighted images with and without fat suppression were done in axial, sagittal and coronal planes with IV gadolinium administration. This MRI revealed from C6-T3 a huge intramedullary lesion measuring 9 x 2.5 x 2.3 cm causing enlargement of the spinal canal. On T1 and T2 weighted images, this lesion was mostly hyperintense with 02 compartments separated by a fine septum. This lesion was mostly hypointense after fat suppression and there was no enhancement with gadolinium (Figure.1).

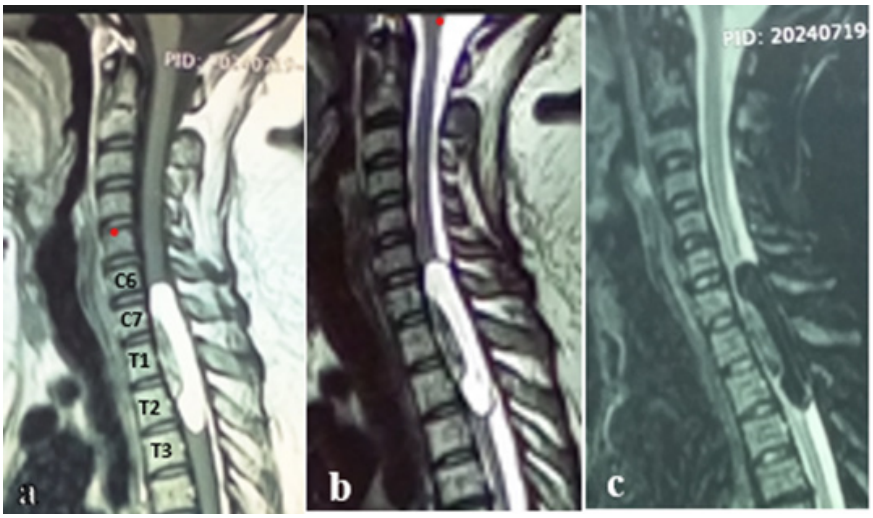

Figure 1: a,b) Sagittal T1 weighted and T2 weighted images showing a giant intramedullary hyperintensity lesion from C6-T3. c) Sagittal STIR : low signal intensity of the lesion.

The heterogenous portion was also important . Regarding these radiological data, the diagnosis of intramedullary lipoma or intramedullary dermoid cyst were suggested. Under general anesthesia, she was admitted for surgery only under per-operative fluoroscope to focus on the surgical era. There was no neurophysiological monitoring. On prone position, a dorsal midline incision was made. After laminectomy from C6 to T3 , the dura was opened and permitted to have a total removal of this lesion. The tumor was posterior, under pia and intramedullary, and the cord was pushed anteriorly. Per operatively, a soft yellowish compact tumor with no additional structures like hair or teeth was seen . Some radicelles emerged through this tumor. A progressive debulking permitted to carry out all the mass. Regarding all these data, the diagnosis of intramedullary lipoma was clear. Histopathological examination of the specimen revealed lobes of mature fat cells confirming the diagnosis of intramedullary spinal cord lipoma (Figure.2).

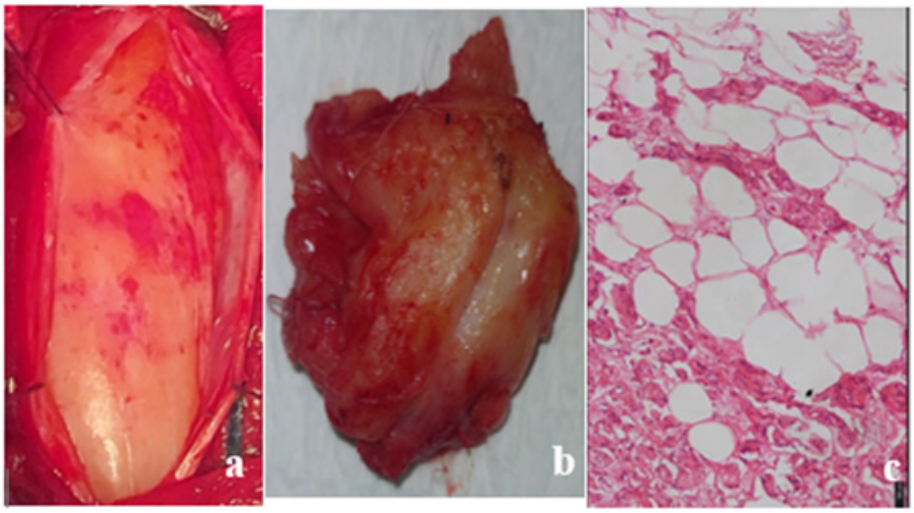

Figure 2: a) Operative view showing a fatty mass underneath the dura. b) Gross total resection of the lesion. c) Histopathological examination showed an encapsulated tissue composed of lobules of mature adipose tissues separated by fibrovascular septa (x 400).

Postoperatively, the patient neurological status worsed and was addressed to physiotherapy for rehabilitation. Postoperative MRI was satisfactory (Fig.3) and the patient recovered her preoperative status after 12 months of physiotherapy.

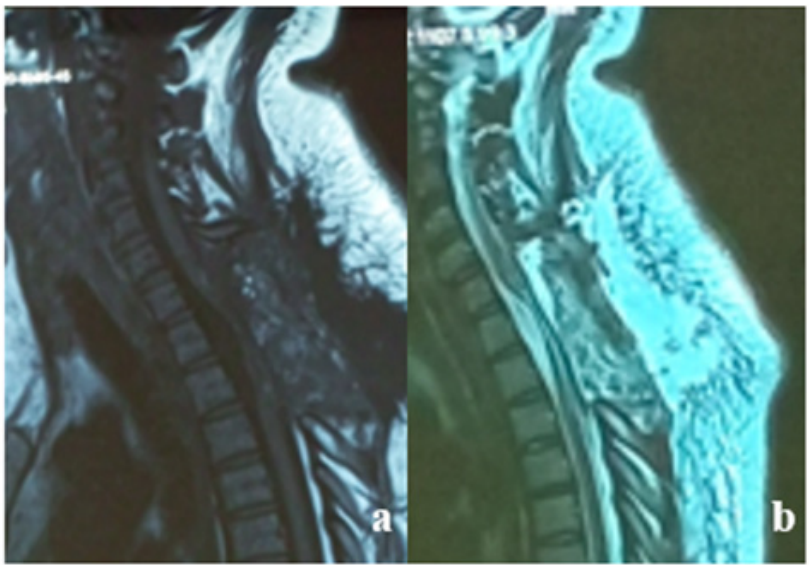

Figure 3: Postoperative MRI sagittal view in T1W (a) and T2 W(b) images showing a gross total resection of the lesion.

Discussion

Spinal cord lipomas are classified into those with spinal dysraphism and those without spinal dysraphism [7]. The first group of these tumors occurs mostly in pediatric ages and are generally associated with spinal dysraphism such spina bifida or lipomeningocele, and are located at the lumbosacral junction [8,9]. The second group is less frequently observed, and tends to occur principally in the adult and are located in cervicothoracic junction and thoracic regions [10,5,11]. True intramedullary spinal cord lipomas without spinal dysraphism are extremely rare and accounting less than 1 % of intramedullary tumors [10,5,9] . Young people are more often affected in the second to fifth decade of life and there is no gender distribution according to this tumor [3,9] . In 1876, Gowers was the first to describe an intraspinal lipoma [2,9]. Up to date, about less than 186 cases of intramedullary spinal cord lipoma had been reported in the literature as scattered, single case reports [10,6].

The tumor's symptoms depend on its location in the spinal cord. The most common symptoms are sensory deficits , gait disturbances , or local back pain in the region of the tumor [7,9]. These symptoms were also seen in our case. Intramedullary lipomas tend to grow slowly with a long-term static clinical course, but followed by an abrupt decline in neurological function shortly before admission, as seen in our case. The rapid clinical progression in our patient led her to paraplegia that is considered as a high neurological dysfunction and classed grade IV by Lee M. et al. and adapted from McCormick PC. et al. [2,12]. It seems that severe deficits are not the result of mass effect but they are caused by replacing normal neural tissue with fat tissue [2,13]. MRI with fat saturation is the gold standard to explore spinal cord lipoma. On MRI , in most cases fatty structures are hyperintense in both T1 and T2 sequences and will be suppressed using fat-suppression sequences [14,9]. In our case, on MRI this tumor had the same radiological characteristics of those described in the literature [6,8,9]. The main differential diagnosis cited , was also an intramedullary dermoid cyst. This differential diagnosis was only discarded after histological examination. The adequate management of these lesions is controversial. Symptomatic patients could be operated, while asymptomatic patients with local sign could be observed. The main goal of this surgery is the decompression of the spinal cord to stop neurologic deterioration [10,6]. Gross total, subtotal, or partial resection is practiced depending on the scenario. In our case, regarding the late presentation of the patient and her clinical status at admission, we opted for total removal. Symptomatic patients could have clinical improvement following decompression, which can be achieved through bony decompression only, partial tumor debulking or duraplasty [14,15]. For better postoperative neurologic outcomes, early surgical intervention should be considered. However, total removal showed a higher risk of postoperative neurologic dysfunction : 6 % of patients showed deterioration after surgery [6,10,15]. In the literature, there is no difference between subtotal removal and total removal in terms of risk of recurrence [1,4]. Because of the significant risk of neurologic deterioration after surgery, preventive surgery is not recommended for asymptomatic patients [2,9,10]. Patients in whom the intramedullary spinal cord lipoma is only partially resected should be informed that substantial weight gain or pregnancy may aggravate their symptoms [8]. However, few cases of excellent result in total or near total removal have been reported [16]. Prognosis depends on the extent of the lesion and clinical neurological status before surgery[17-19].

Conclusion

Nondysraphic intramedullary lipomas are extremely rare. There is no consensus regarding the management of these lesions. Gross total resection can be an option for severe neurological compromised patients following by physiotherapy. No surgery is needed for asymptomatic patients.

Acknowledgment: We are grateful to Mrs. Barry Najah for her contribution.

Funding Declaration: No.

Conflicts of Interest: There are no conflicts of interest.

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