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Nondysraphic Huge Cervical Intramedullary Lipoma: a Case Report and a Review of Literature

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Abstract

We report the case of a 14-year-old girl with huge cervical intramedullary lipoma, who presented with spastic quadriparesis. Her MRI revealed an intramedullary lipoma extending from the craniovertebral junction to the sixth cervical vertebra.

There was no spinal dysraphism. Subtotal excision of tumor with primary dural closure and laminoplasty was performed from C3 to C6, which produced an improvement in the patient's function.

Key Words: Cervical Lipoma, Intramedullary Spinal Tumor, Lipoma, Subtotal Resection.

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1- INTRODUCTION

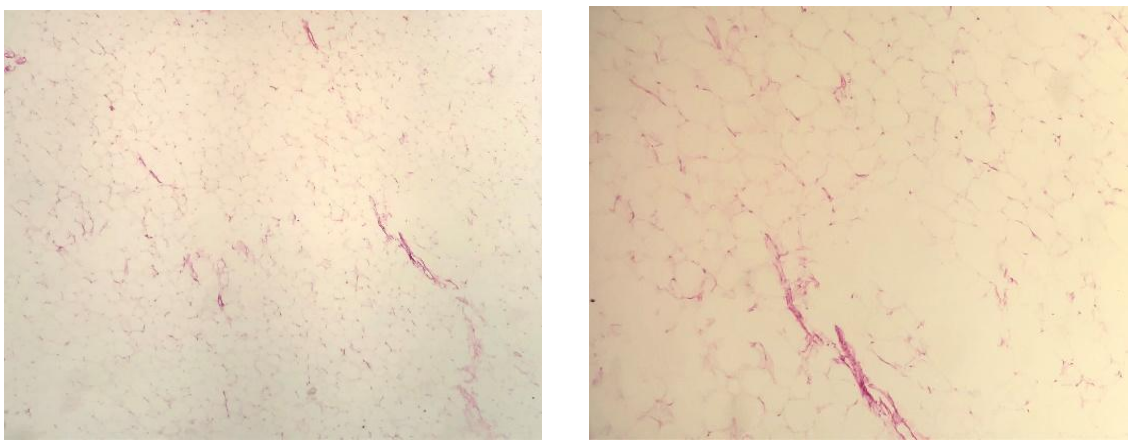
Spinal cord lipomas are rare lesions, accounting for less than 1% of all spinal cord tumors (1). Most isolated spinal cord lipomas are intradural extramedullary and true intramedullary lipomas are very rare (2, 3). Lipoma of spinal cord commonly occurs in association with spinal dysraphism (4). Nondysraphic spinal cord lipomas are very rare and usually present in the second or third decade of life (5). In this report we will describe a case of huge nondysraphic cervical intramedullary lipoma.

2- CASE REPORT

A 14-year-old girl presented with quadriparesis and numbness in the extremities for 12 months. The patient had no neck pain. She suffered from spastic quadriparesis. There were no bowel or bladder complaints. Neurologic examination revealed mild muscle weakness (4/5).

Plantar reflex was extended bilaterally. No clonus was detected. Muscular Atrophies of the upper and lower limbs with hyperreflexia were present. She had no evidence of spinal dysraphism.

Magnetic Resonance Imaging (MRI) showed a posteriorly placed cervical intramedullary mass lesion, extending from the craniovertebral junction (CVJ) continuously into the C6 level. The lesion was hyperintense on T1-weighted and T2-weighted sequences with cord compression (**Fig. 1A, B**).



(A) (B)

Fig. 1: A, B: Sagittal T1and T2 Weighted MRI showing intramedullary hyperintense lesion

Because the patient was quadriparetic, surgical decompression was performed. She was placed in a prone position and a posterior cervical midline incision was made from craniovertebral junction (CVJ) to C7. Laminectomy with a high speed drill was done from C1 to C6. After a midline durotomy, the lesion was exposed. Grossly, the tumor was yellow, ellipsoid and firm (**Fig. 2**). With microscopic evaluation, no clear plane of cleavage between the spinal cord and the lesion was seen; therefore, subtotal resection of tumor was performed. Then primary closure of the dura was done with fascial graft. Laminoplasty was performed from C3 to C6 with titanium plate and bone graft.

The patient woke up without any new neurologic deficits. Postoperatively, her numbness was relieved and she regained normal force in all limbs at the follow-up period. Lipomatous tissue was revealed in histopathologic review (**Fig 3A, B**).

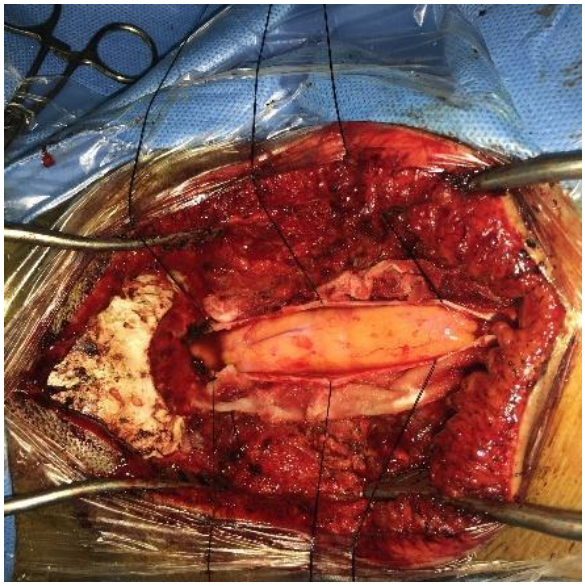
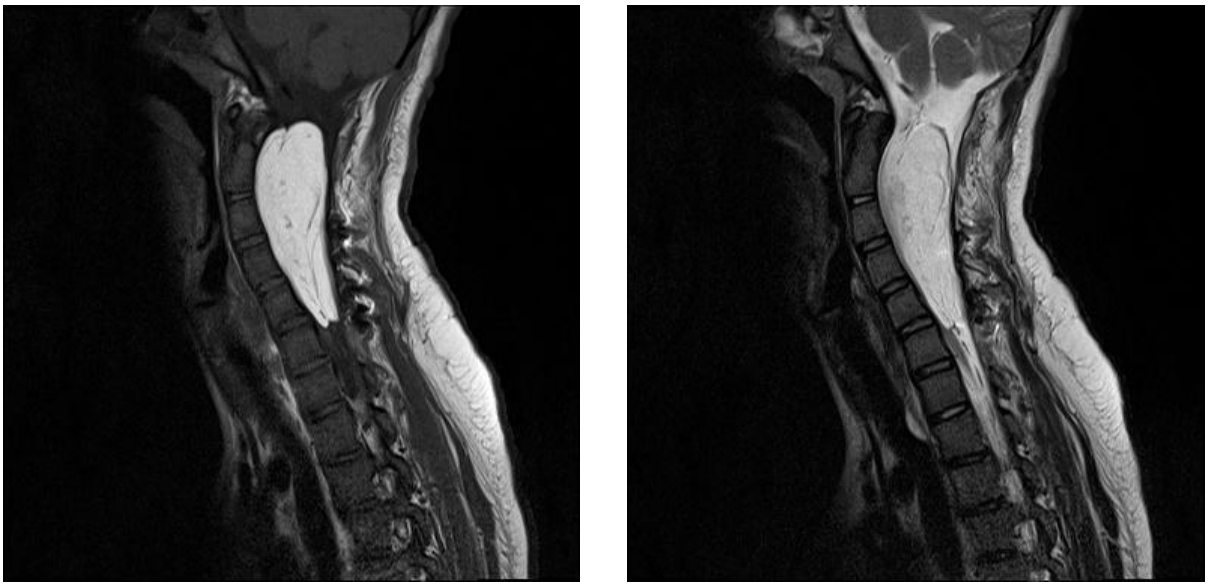


Fig. 2: Intraoperative view showed a yellowish tumor which completely filled the canal



(A) (B)

Fig. 3: A, B: Hematoxylin-eosin staining ×40(A) and ×100 (B) showing mature adipocyte proliferation

At the time of writing this article, the patient was symptom-free for about two years; the only complaints were about occasional spasms in legs. In a clinical examination, mild muscle atrophy in hands was presented. And Cervical X-ray demonstrated normal cervical lordosis in the follow-up period.

3- DISCUSSION

As previously mentioned, spinal cord lipomas are rare lesions, accounting for less than 1% of all spinal cord tumors (1).

A review of literature demonstrated cases with cervical involvement before our case (**Table 1**).

Table-1: Literature review of Cervical Intramedullary Lipoma

Author, year	Age–sex	Symptoms	Level of Lesion	Outcome
Rapaport, 1982 (6)	4m,M	Quadriparesis and hypotonia	Post fossa–cervical cord	Died
	10m,F	Quadriparesis and hypotonia	Post fossa–cervical cord	Functional improvement
White ,1983 (7)	11y,F	Quadriparesis	Post fossa–T2	No change
Wood ,1985 (8)	5m,M	Spastic paraparesis, Hypotonic both arms	Post fossa–T2	No change
	15y,F	Back and leg pain	C3-T6	Functional improvement
Mori ,1986 (9)	7y,M	Flaccid quadriparesis	Posterior Fossa–C7	No change
McGillicuddy ,1987 (10)	24y,M	Right hemiparesis	FM–C3	Functional improvement
Fan ,1989	18y,M	Quadriparesis	FM–C5	Functional improvement
Ammornmarn ,1992 (11)	18m,M	Apnea	Post fossa–cervical cord	No change
Donati ,1992 (12)	2y10m	Right hemiparesis	Post fossa–cervical cord	No change
Crols ,1993 (13)	17y,M	Spastic quadriparesis, Bilateral pes equinovarus	CVJ–C4	Functional improvement
Naim-Ur-Rahman ,1994 (14)	9m,F	Floppy infant syndrome, spastic quadriparesis	CVJ–T5	No change
Wilson, 1996 (15)	2.5m,M	Mild increase in tone of the left upper extremity	Post fossa–cervical cord	Functional improvement
Chaskis ,1997 (16)	3m,F	Progressive quadriparesis	4th Ventricle–C7	No change
Kogler,1998 (17)	2y,M	Hydrocephalus	Post fossa–C1–T4	Functional improvement
Kai ,2003 (18)	8m,F	Limb paresis and dysphagia	CVJ–C7	Functional improvement
Kim, 2003 (19)	6m,F	Left hemiparesis	foramen magnum–T1	Functional improvement
	12y,M	Left hemiparesis	C6–T11 & L1–2	No change
Le Feuvre. 2004 (20)	18y,F	Quadriparesis, bladder problems	Medulla–C6	Functional improvement
Bhatoe, 2005 (21)	6y,M	Quadriparesis	foramen magnum–C4	No change
	43y.M	Quadriparesis	C4–C6	Functional improvement
	37y,M	Quadriparesis	C6-T3	No change
	40y,M	Quadriparesis	C6-T2	No change
	24y,M	Quadriparesis	C7–T4	Functional improvement

Author, year	Age–sex	Symptoms	Level of Lesion	Outcome
	31y,M	Quadriparesis	C6–T1	No change
	23y,F	Quadriparesis	C6–T6	Functional improvement
	30y,M	Quadriparesis	FM–C2	No change
	30y,F	Quadriparesis	C7–T4	No change
	27y,M	Quadriparesis	C6–T4	No change
Naim-Ur-Rahman ,2006 (22)	3y,F	Quadriparesis, opisthotonus, respiratory difficulty	CVJ–T3	Functional improvement
Muthusubramanian ,2008 (4)	16y,F	Quadriparesis, neck pain	foramen magnum–C6 & conus medullaris	Functional improvement
Chagla ,2008 (3)	17y,M	Spastic quadriparesis	CVJ–C6	Functional improvement
Mengual ,2009(23)	16m,M	Hemiparesis, gait difficulty, and headaches	Post fossa–T2	No change
Sanli, 2010 (24)	12y,F	Progressive quadriparesis, gait disturbance	Post fossa–T2	No change
Fleming, 2010 (25)	2m,M	Hemiparesis	C2–T8	Functional improvement
	13m,F	Cervical subcutaneous mass	CVJ–C7	No change
	2y9m,F	Quadriparesis	C7–T7	Functional improvement
	3y,F	Hemiparesis	C5–T8	Functional improvement
Ahmed ,2015 (5)	31y,F	Numbness of hands	C6-7	No change
Meher , 2017(26)	30,M	Quadriparesis	CVJ -C4	Functional improvement
Severino , 2017(27)	39,M	Quadriparesis	C5-T2	Functional improvement
Present 2021	14y,F	Quadriparesis and numbness	CVJ–C6	Functional improvement

As a result of the increased fat deposition in metabolically normal fat cells, these lesions generally enlarge over time and cause various symptoms (28). Fifty-five percent of intradural lipomas become symptomatic in the second and third decades (29). It is uncertain that these lesions are true neoplasms or hamartomas. There are reports from surgically resected nondysraphic lipoma, recurred due to hyperplasia, which support its being a true neoplasm. Dysraphic lipomas are believed to be hamartomas (22).

Spinal cord lipomas are more common in the pediatric population, possibly due to their embryonal origin (5). Patients with intramedullary spinal cord lipoma have a prolonged clinical course that is punctuated by a decline in neurological functions shortly before presentation. (30)

Depending on the location and involvement of neural structures, presented symptoms are various; they include spinal pain, dysaesthetic sensory changes, gait difficulties, weakness, incontinence, growth retardation, dysphagia, and respiratory difficulty (5, 31).

Histologically, intramedullary lipomas consist of mature adipocytes that lie in fibrous connective tissue (31). Striated muscle, bone, epithelial derivatives, and neural tissue may also be found within these lesions (5). Intramedullary lipomas have the potential to change in size as the body fat content changes (32).

MRI is the best imaging modality, because of the significant delineation of the lesion and the surrounding neural structures, soft tissue, and subcutaneous planes. Complete screening of neuraxis is mandatory in all cases of spinal dysraphism and in intramedullary lipomatous lesions, whenever suspected. Fat is hyperintense on T1W with no increased contrast uptake and relatively hyperintense on T2W. Although other pathologies can have similar characteristics on MRI, a fat suppression sequence can confirm the presence of adipose tissue and help to distinguish a lipoma from blood or calcifications (4, 33, 34).

There are several theories on the pathogenesis of spinal cord lipomas. Most theories believe that the development of dysraphic spinal cord lipomas is predominantly due to an adipocyte migration problem during the embryonic development (19). The pathogenesis of nondysraphic spinal cord lipomas is still remained unclear.

Treatment of intramedullary lipoma is controversial. Selection of the treatment option is based on the clinical scenario. If the lipoma is found incidentally, observation may be sufficient (5). Treatment of symptomatic patients involves decompression and resection (30). Surgical debulking of lipomas prior to permanent spinal cord injury may prevent irreversible neurological deficits (3). The goal of surgery is to prevent progression of neurological deficits and stabilize the patient's clinical course (30).

Gross total resection may prove to be technically challenging because most of these lesions are adherent to the normal tissue of the cord and do not have a true plane for dissection (25). Trying to separate the tumor from adjacent spinal cord will compromise the vascularity of the cord. Conservative intratumoral internal decompression can provide long-term symptom control and aggressive attempts in tumor excision must be avoided (3, 26).

4- CONCLUSION

In this article we report a rare case of huge nondysraphic cervical spinal cord lipoma. Surgical debulking of lipomas must be performed prior to permanent neurological deficit. Adequate decompression with subtotal resection is an effective surgery with favorable outcome.

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