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Case Report

Holocord Lipomatosis: The Rare Yet Deadly Conundrum

Jagadeesh A, Shivaprakash M, Sibhi Ganapathy*, Raykar R and Rao SAV

Department of Neurosurgery, St. Johns Medical College, Bengaluru, India

***Corresponding Author**: Sibhi Ganapathy, Department of Neurosurgery, St. Johns Medical College, Bengaluru, India.

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Abstract

Non-dysraphic intramedullary holocord lipomas are rare lesions, histologically benign neoplasms, constituting less than 1% of all intramedullary spinal tumors, and their presentation in the form of gradually worsening myelopathy, is nonspecific. We present a case of a 40 year old female patient, with a three year history of progressive low backache radiating to right lower limb upto the great toe and further developed tingling sensation associated with weakness of bilateral lower limbs (ASIA impairment scale grade D). MRI showed multiple multiloculated lipomatous nature of the tumor throughout the spinal cord with a homogenously hyperintense signal on T1-weighted images, a hyperintense signal in T2-weighted images, and signal suppression on fat suppression sequences with a conventional spin-echo protocol. Patient was further screened to rule out intra-cranial extension, lipomatous cystic lesions in the ovaries and any segmentation or fusion anomaly of the spine. Encapsulated neoplasm composed of lobules of mature adipocytes and thin fibrous septae was visualised on histo-pathological examination with no angiomatous or neuroglial elements. The lipoma was sub-totally resected with the aid of Carbon Dioxide Laser in a two-step debulking procedure over a span of one week. Multilevel microsurgical laminectomy and decompression with biopsy of intramedullary lesion and laminoplasty under neuromonitoring was performed. Post-operative prognosis is variable and dependant on pre-op deficits, extent of resection, etc. However, being a benign lesion, prognosis is promising as was proven in our patient eliciting significant improvement in power post operatively. Thus, management of holocord and longitudinally extensive intramedullary spinal cord tumors can be achieved with long-term preservation of neurological functions.

Keywords: *Holocord Lipomatosis; Intramedullary Spinal Tumors; Laminoplasty and Debulking*

Introduction

Intramedullary spinal cord lipomas are rare lesions, histologically benign neoplasms, constituting less than 1% of all intramedullary spinal tumors. Lipomas of the spinal cord are often a component of occult spinal dysraphic states and are diagnosed in neonates or infants based on skin markers. Nondysraphic intramedullary spinal cord lipomas are rare, and their presentation, in the form of gradually worsening myelopathy, is nonspecific [1-4]. The usual location is in the cervico-thoracic region, mostly extradural with an association with spinal dysraphism and confined to four or five segments of the cord. A sacral or lumbosacral defect that communicates with a subcutaneous lipoma is a common finding [3,4].

Case Report

A 40 year old female patient presented with a three year history of progressive low back ache, radiating to right lower limb upto the great toe. The pain was shooting in nature and was not relieved by conventional painkillers. She also developed tingling and sensation of

both lower limbs which was insidious in onset and progressive in nature. The pain was associated with stiffness of the lower limbs which initially manifested as loss of balance and frequent falls over uneven surfaces but progressed into tightness affecting her walking. No weakness of the limbs was experienced. The patient was still able to carry out her activities of daily living. No difficulty in passing urine or stools.

Neurological examination on admission showed lower limb weakness (ASIA impairment scale, lower extremities: grade D, upper extremities: grade E) and hyper-reflexic knee and ankle jerks on both sides with spasticity (modified Ashworth grade 2) in both lower limbs. There were no neuro-cutaneous stigmata, hair, dimple or mass.

Neuroimaging evaluation

MR spine showed multiple multiloculated T1/ T2 heterointense cystic structures noted throughout the spinal cord showing areas of fat suppression. At the cervical region extending from lower end of C4 to lower end of C7 vertebrae measuring 1.0 x 1.3 x 4.4 cm (AP x ML x CC). The lesion is noted to efface the thecal sac on the right side and posterior aspect with evidence of fat containing component in the posterior aspect. The upper end of D3 to lower end of D8 vertebrae measuring 1.1 x 1.5 x 11.4 cm (AP x Ml x CC) with fat containing component in the posterior aspect. An intradural extramedullary uniloculated cystic structure noted in the thoracic region extending from lower end of D9 to upper end of D11 vertebrae measuring 0.5 x 0.5 x 2.4 cm (AP x Ml x CC). The lesion is noted to efface the thecal sac on the posterior aspect. The lumbar region extending from upper end of L1 to mid vertebral body level of L2 It measures 1 x 1.8 x 3.7 cm (AP x Ml x CC) The lesion is noted to efface the thecal sac with evidence of fat containing component in the posterior aspect superiorly and holocord fat containing lesion inferiorly. In addition, evidence of a fat containing structure extending from lower end of L2 to L4- l5 intervertebral disc level. Smaller fat containing structures are also noted within the sacral canal (Figure 1).

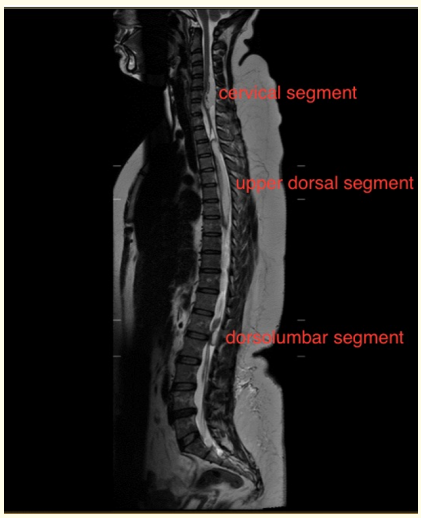

Figure 1: *Showing an MRI of the whole spine with lipomatosis of the cervical, dorsal and lumbar segments.*

MRI brain screening was done to rule out any intracranial extension. Abdominal ultrasound ruled out lipomatous cystic lesions in the ovaries. Plain radiography of spinal regions was performed to exclude any segmentation or fusion anomaly.

Management strategy and surgical planning

Due to the extensive nature of the disease and the probability of giving devastating disabilities to a patient who was largely intact, a conservative approach to the disease was taken. The segments found most responsible for her disease was taken into consideration and dealt with in a manner as to confer the least neurological deficit to the patient. There was also considerable thought given to stability of the spine which would be affected by long level laminectomies which would generally be required to access the lesion. Due to the extensive nature of the disease as well as two localised discontinuous segments identified as being responsible for her disease, it was decided to stage the procedure. We opted to decompress the D4-D7 intramedullary segment first before attempting the dorsolumbar junction. The cervical region, although much more involved radiologically was spared due to the absence of upper limb symptoms and normal chest expansion and single breath count.

One of the methods used to minimise damage to viable neural tissue was by using carbon dioxide laser, which selectively destroyed fat globules. This was especially employed in the intramedullary lesion, ensuring the best possible functional result for the patient. Other adjuncts used in the process included neuro-invasive monitoring systems to monitor Somatosensory evoke potentials (SSEP) and Motor Evoke Potentials (MEP) to ensure salvage of the long motor tracts of the cord during intramedullary resection of the lipoma.

Surgical procedure

The patient was positioned prone after the induction of relaxant free anaesthesia, on bolsters and monitoring electrodes were placed in her lower limbs. A D4-D7 en-bloc laminectomy along with a midline durotomy was done in standard fashion. The lesion was seen emanating from the fattened and engorged spinal cord (Figure 2a). A myelotomy was performed under NIM guidance over the adipose tissue and the tissue was followed into the cord. CO_2 laser was then used to destroy the adipose tissue and decompression with biopsy of intramedullary lesion was done (Figure 2b). Dura was closed in a watertight fashion and the laminoplasty bone fragments were replaced *in situ* and held together in position with implants to ensure stability and prevent kyphosis. The wound was closed in standard fashion and the anaesthesia was reversed.

The patient was shifted to the ICU for monitoring. She at this time developed spasticity of grade 2 in both lower limbs with a weakness of 3/5 in the left lower limb. Steroids and physiotherapy were started along with Baclofen. The spasticity improved well on POD3. On POD4 of the first surgery she was taken up again for a D12, L1, L2 laminectomy and decompression of conus lipoma under General Anaesthesia. Here the lipoma was seen extending form the conus into the cauda equina. NIM monitoring, especially around the anal verge proved vital in preserving bladder and bowel functions for the patient post operatively. Microsurgical decompression of the lesion was carried out followed by again a water tight duroplasty (Figure 2c and 2d).

The patient tolerated the procedure well and was ambulated after POD3 of the second surgery. Her weakness improved well, although she did have residual spasticity which was managed by oral medications. Sensory symptoms such as radicular pain and paraesthesia were present initially for 3 days after the second surgery, but gradually reseeded on response to medications. The patient was discharged after 2 weeks of physiotherapy, ambulant with minimal support with manageable spasticity and paraesthesia. No bladder dysfunction was present at the time of discharge. She is expected to regularly follow up in the outpatient clinic for further symptoms and to monitor disease progression.

Histopathological findings

All tumor samples were received as multiple bits of pale, fatty tissue. On microscopic inspection, the sections showed sheets of adipose tissue. No angiomatous or neuroglial elements were visualized in any of the tumors. The diagnosis of lipoma was confirmed (Figure 3a and 3b).

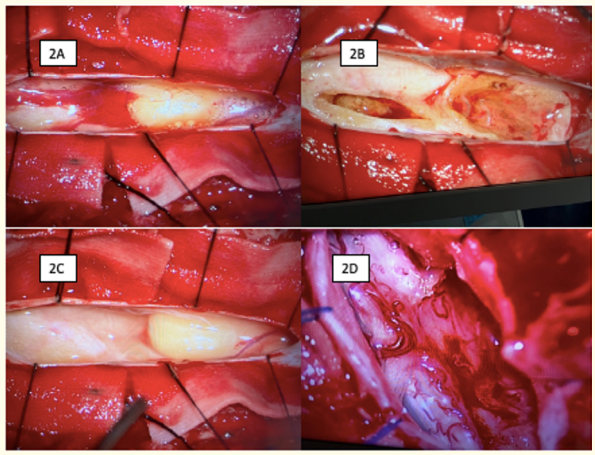

Figure 2: *Showing the intraoperative pictures of the lesion. 2A shows the dorsal lesion once the dura was opened, 2B shows coagulated fat and excised sections of the lesions after laser ablation., 2C shows the lumbar lipomatosis preop and 2D shows it post resection with nerve roots freely visible.*

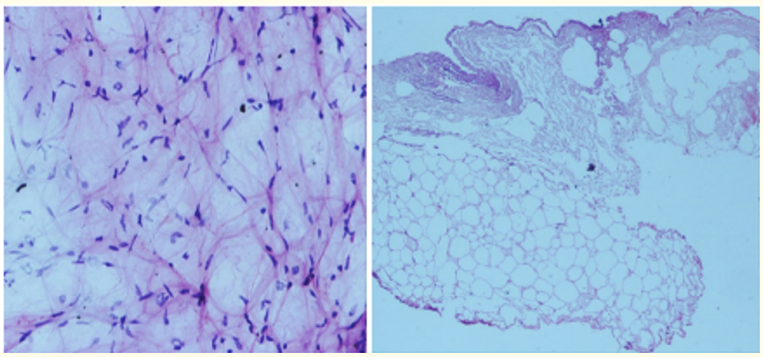

Figure 3: *The resected specimens were stained and viewed under microscope showing fat globules and adipocytes.*

Discussion

Intramedullary lipoma is an entity that is not commonly observed. The clinical manifestations include paraparesis, spasticity, gait difficulties, urinary incontinence, sensory disturbances, and localized pain. MRI is the gold standard modality in the diagnosis of these lesions. It shows the lipomatous nature of the tumor confirmed by a homogenously hyperintense signal on T1-weighted images, a hyperintense signal in T2weighted images and signal suppression on fat suppression sequences [2,4].

Several hypotheses are proposed as the origin of the intramedullary lipoma. The first is 'developmental error theory'. Inclusion of the misplaced adipocytes during the formation of the neural tube causes growth of lipoma in the spinal cord. This theory also explains the dorsal location of lipoma which causes a flattening of the cord. The second hypothesis is the 'metaplasia theory'. Connective tissue meta-

plasia may lead to the deposition of fat within the dura. The third is the 'hamartomatous origin theory'. None of these theories explain the exact origin of a spinal lipoma but the first hypothesis is widely acknowledged [3]. A slow ascending spastic monoparesis or paraparesis is a common presenting symptom when it affects the cervical or thoracic region. The clinical course of intramedullary spinal lipoma is characteristic. Initial symptom progression is slow and late deterioration is rapid [5,6].

Histologically, spinal intramedullary lipomas consist of lobulated fatty tissue separated by connective tissue septae. So, if they are asymptomatic, they can be managed without any surgical intervention [3,9]. However, if symptoms progress as was the case in our patient, surgical debulking is recommended and decompression would be sufficient as was seen in our literature review. Furthermore, the infiltrative nature of these lipomas complicates a gross total removal. Therefore, the operative principle is decompression before symptom progression.

Conclusion

Intramedullary lipomas are often focal, and only a few cases of extensive cervico-thoraco-lumbar intramedullary lipomas have been reported [6]. Patients are usually asymptomatic at presentation or show progressive neurological deterioration. MRI shows lipomatous nature of the tumor associated with a homogenously hyperintense signal on T1-weighted images, a hyperintense signal in T2-weighted images, and signal suppression on fat suppression sequences with a conventional spin-echo protocol [7]. As a Histologically benign tumor, it accounts to < 1% of all spinal cord tumors [8]. Management of holocord and longitudinally extensive intramedullary spinal cord tumors can be achieved with long-term preservation of neurological functions [9].

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