REVIEW

Intramedullary lipoma mimicking syringomyelia: A case report and literature review

Article history:
Received: 26-09-2022
Revised: 22-02-2023
Accepted: 07-03-2023
Published: 15-03-2023

Jialu Wang[a], Penghao Liu[a], Zan Chen[a], Wanru Duan[b]

Abstract

Background: Intramedullary lipomas can sometimes be misdiagnosed as syringomyelia. In this report, we describe a case of intramedullary lipoma that presented with symptoms similar to syringomyelia and review the relevant literature.

Case presentation: A 35-year-old woman was admitted to the hospital due to hypoesthesia in her right limb, particularly in her leg. Spinal magnetic resonance imaging revealed a C5-T7 lesion and an L2-L3 lesion with hyperintensity in T1-weighted and T2-weighted image hypointensity in T1 fat suppression-weighted images. The lumbosacral lesion was resected, and the pathological results indicated cholesteatoma, while based on the clinical manifestation and imaging results, the cervicothoracic lesion was considered to be a lipoma and was observed without surgery. The patient's neurological function in the lower limb improved in 1 month after surgery.

Conclusion: Special caution should be exercised when attempting to differentiate between intramedullary lipoma and syringomyelia before surgery.

Keywords: Intramedullary lipoma; Syringomyelia; Surgical indications; differentiation.

BACKGROUND

Spinal lipoma is typically associated with spinal dysraphism and is considered a hamartoma. It is most commonly found in the lumbosacral spinal cord.[1] Intramedullary lipoma that is unrelated to spinal dysraphism is relatively uncommon, and is also known as true. In our case, the patient's numbness in the upper limb was subtle and the cervicothoracic lipoma was not removed. Her symptoms in both upper and lower limbs were relieved after the resection of the lumbosacral lesion due to the decompression of the spinal cord lipomas.[2]

The clinical manifestations of intramedullary lipoma typically include motor dysfunction, sensory hypoesthesia, and autonomic dysfunction of the corresponding spinal cord segment. The diagnosis mainly depends on spinal magnetic resonance imaging (MRI) and histopathology.[3, 4] Observation or surgical resection is considered according to the symptoms and locations.

Although intramedullary lipoma and syringomyelia may share similar clinical manifestations and imaging features, they require distinct treatment strategies. In this report, we describe a case of intramedullary lipoma that presented with symptoms similar to syringomyelia, and we review the relevant literature to discuss the differential diagnosis.

[a] Department of Neurosurgery, Xuanwu Hospital, Capital Medical University. Beijing, China. Lab of Spinal Cord Injury and Functional Reconstruction, China International Neuroscience Institute (CHINA-INI), Beijing, China.

[b] Department of Neurosurgery, Xuanwu Hospital, Capital Medical University. Beijing, China. Lab of Spinal Cord Injury and Functional Reconstruction, China International Neuroscience Institute (CHINA-INI), Beijing, China.
Corresponding Author: duanwanru@xwhosp.org

CASE PRESENTATION

A 35-year-old woman presented with a 6-month history of hypoesthesia in her limbs. The primary symptoms were numbness and discomfort below the right knee, which had worsened over the past month with increased numbness in her right calf. The occurring sequence of upper and lower limb symptoms was unknown, but the lower limb symptoms worsened first. Physical examination of the patient revealed a normal physiological curvature of the spine, without any notable kyphosis or scoliosis. There was no tenderness or percussion pain observed. The neurological examination revealed a muscle strength of level IV in the right upper and lower limbs, with no bladder disorders, bowel disorders, dermatological changes, or bulbar symptoms. The patient's limb reflexes were weakened, and no pathological signs were detected. There was no remarkable past medical history of trauma, diabetes, hypertension and infectious diseases, or family history. Cigarette smoking as well as alcohol and drug abuse were denied by the patient.

Preoperative MRI showed a C5-T7 lesion with homogeneous high signal intensity on T1-weighted (T1WI) images and T2-weighted (T2WI) images, with hypointensity on the T1 fat-suppression sequences (Figure 1A-C). The boundary of the lesion was clear, and no signs of pathological destruction or invasion were seen. A second L2-3 lesion with heterogeneous signals on T1WI, T2WI, and T1 fat-suppression sequences was also revealed on the spinal MRI (Figure 1D-G).

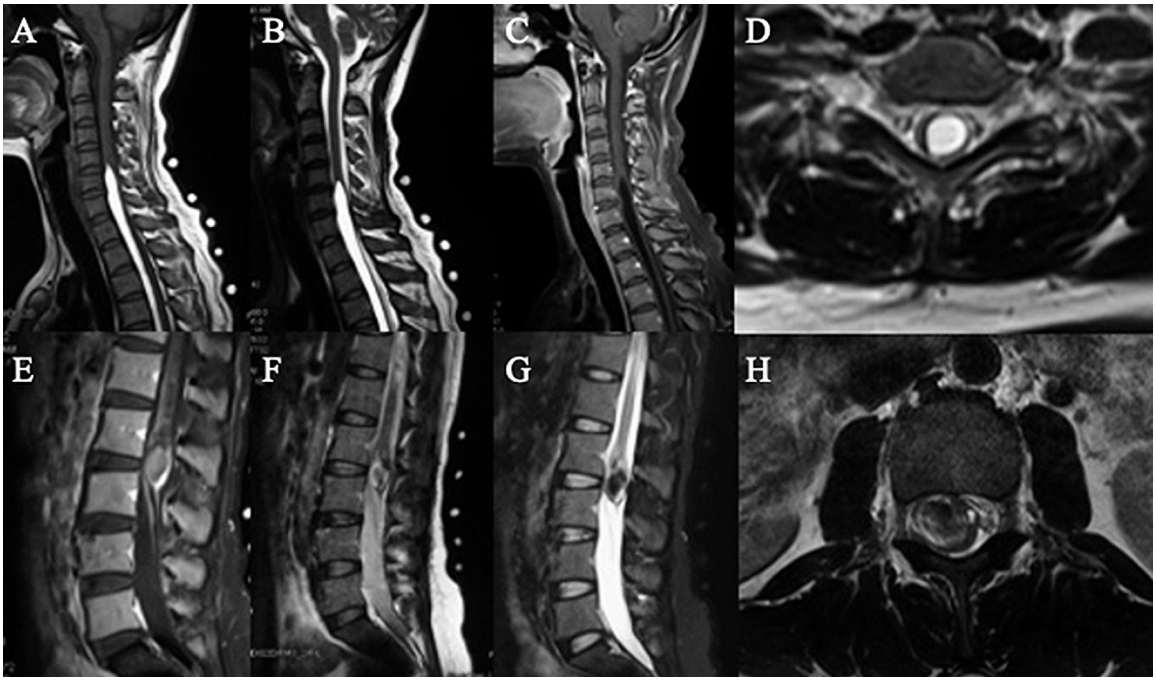

Figure 1. Preoperative spinal MRI of the cervicothoracic and lumbosacral lesions.

Cervical spinal MRI scan showed an intramedullary hyperintensed lesion evident on (A) T1WI and (B) T2WI images, and (C) intramedullary hypointensed lesion on T1 fat suppression-weighted images. Axial cervicothoracic MRI scan (D) showed a hyperintensed lesion on T1WI. Lumbosacral MRI scan showed in our case, the patient's numbness in the upper limb was subtle and the cervicothoracic lipoma was not removed. Her symptoms in both upper and lower limbs were relieved after the resection of the lumbosacral lesion due to the decompression of the spinal cord. Heterogeneously hyperintensed lesion evident on (E) T1WI and (F) T2WI images, and (G) hypointensed lesion on T1 fat suppression-weighted images. Axial lumbosacral MRI scan (H) showed a heterogeneously hyperintensed lesion on T1WI.

Based on the imaging results, cervical lipoma and lumbosacral cholesteatoma were suspected. Due to the major symptoms being in the right lower limb, we opted to perform intraspinal microsurgical resection of the lumbosacral tumors. Pathological

examination revealed keratomas and focal calcifications, confirming the diagnosis of cholesteatoma. One month after the operation, the numbness in the lower limbs had completely resolved, and the upper limb numbness was partially relieved. Spinal MRI showed no relapse of the lumbosacral lesion, and no progression or shrinkage was seen in the cervicothoracic lesion.

DISCUSSION AND CONCLUSIONS

Review and comparison of case reports

A cholesteatoma was found in the lumbosacral region, while a long-segment lipoma was located in the cervicothoracic region. Due to the similarity in clinical manifestations and imaging features between lipoma and syringomyelia, a misdiagnosis could occur. To our knowledge, this is the first reported case of an intramedullary lipoma mimicking syringomyelia.

Syringomyelia occurs due to an abnormal accumulation of cerebrospinal fluid in the central canal or spinal cord,[5] and is often associated with Chiari malformation, tumors, and other diseases.[6] Spinal MRI can be used to distinguish it from other conditions. Lipoma is characterized by hyperintensity on both T1WI and T2WI images, whereas syringomyelia appears hypointense on T1WI images and hyperintense on T2WI images. Both conditions appear hypointense on fat suppression sequences. In the present case, an MRI scan revealed a long intramedullary lesion at the level of C5-T7, which is a relatively rare finding and can be easily misdiagnosed as syringomyelia.

There have been reported syringomyelia cases mimicking intramedullary lipoma[7, 8] and an intramedullary lipoma case combined with syringomyelia[9] (Table 1). In patients with intramedullary tumors, syringomyelia can occur either above or below the tumor. There were two cases reporting that the syringomyelia contained fat droplets, which was named fatty syringomyelia.[7, 8] According to Goel and colleagues, excess fluid, fat tissue, and even bone tissue in the spinal cord may be associated with spinal cord instability, which represents the body's self-protective mechanism for the spinal cord.[7] The fatty syringomyelia and lipoma were alike in MRI appearance. In one reported case of syringomyelia secondary to lipoma, the patient had severe tethered cord syndrome, which required decompression and tumor resection.[9]

In addition to syringomyelia, other diseases such as Intramedullary dermoid, lipomatous differentiation of neoplasm, subacute hemorrhage, and encephalocraniocutaneous lipomatosis should also be distinguished from lipoma. Spinal MRI is important in the differential diagnosis, and in cases with ambiguous imaging appearances, biopsy or tumor resection might be warranted.

Controversies on the treatment

Intramedullary lipoma and syringomyelia require different treatment strategies. In the case of syringomyelia and fatty syringomyelia, the underlying lesions should be removed, which can lead to a decrease in the size of the syrinx and eventual disappearance within one year.[7] If the tumor is not removable or in cases with acute symptoms, decompression should be performed.[7] The use of surgical treatment for patients with intramedullary lipoma is still considered controversial in the medical community. Pang *et al.* believed that to prevent the deterioration of neurological function, surgery for intramedullary lipoma should be performed as early as possible.[10] And Moghaddam *et al.* also suggested early release before the appearance of irreversible neurological symptoms.[11] However, Roujeau *et al.* reported that the improvement of neurological function after surgery for intramedullary lipoma was not optimal, and there was a high risk of complications.[12]

In our opinion, surgical resection of lipoma should depend mostly on the severity of the related symptoms. It has a low proliferation rate, and rarely causes acute symptoms. Therefore, preventive surgery for intramedullary lipoma is not generally recommended. If symptoms occur, surgical resection and decompression should be performed to relieve spinal cord compression and alleviate the patient's condition. Subtotal resection is acceptable if the lipoma is difficult to be completely removed without progressive harm to the spinal cord. Although there is no significant difference in recurrence rate between the two groups, the probability of neurological complications after total resection of intramedullary lipoma is higher.[13] In our case, the patient's numbness in the upper limb was subtle and the cervicothoracic lipoma was not removed. Her symptoms in both upper and lower limbs were relieved after the resection of the lumbosacral lesion due to the decompression of the spinal cord.

Author, year	No. of cases	Age, sex	Clinical manifestations	Location of lesion	Imaging features	Treatment	Late outcome
Mahdi *et al.* (2022)[8]	1	27, F	Progressive neck pain and dysesthesia and tingling in the shoulders and upper limbs for 1.5 years.	Syringomyelia at C1-C5; Pilocytic astrocytoma at C5-T2	MRI showed low signal on T1w, high signal occupying lesions on T2w at C1-5; T1w and T2w showed high signal occupying lesions at C5-T2	Tumor resection under the microscope	Neurological dysfunction was significantly improved and syringomyelia was resolved
Goel *et al.* (2020)[7]	1	32, M	Burning pain over the entire right hand and right side of the chest and back for 2 years	Atlantoaxial dislocation; Syringomyelia at C5-C7	MRI T1w and T2w showed high signal occupying lesions at C5-7	Atlantoaxial lateral mass distraction fixation	Neurological dysfunction was not completely improved but syringomyelia was resolved
Luca *et al.* (2019)[9]	1	4, M	Urinary incontinence associated with paraparesis, which quickly progressed and prevented ambulation.	Syringomyelia at T2-T7 and L5-S4; Lipoma at S3-S4	MRI showed low signal on T1w and high signal on T2w at T2-T7, L5-S4T1. MRI showed high signal occupying lesions at S3-S4	Spinal cord decompression and surgical tumor resection	Neurological dysfunction was significantly improved and syringomyelia was resolved

Table 1. Reported cases of syringomyelia similar to intramedullary lipoma and intramedullary lipoma combined with syringomyelia

https://doi.org/10.37819/hb.2.326

In this case, we reported the first case of intramedullary lipoma mimicking syringomyelia. This highlights the importance of distinguishing between intramedullary lipoma and syringomyelia, as although they may present with similar clinical symptoms and imaging findings, their management differs significantly. Misdiagnosis can lead to iatrogenic injury and may compromise the quality of life of the patient after surgery. Therefore, it is crucial to pay close attention to differentiating between the two conditions to ensure optimal patient outcomes.

Ethics approval and consent to participate

No consent was necessary for this study.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images and videos.

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Competing interests

The authors declare that there are no conflicts of interest.

Funding

The "Young Talents" Program, supported by Beijing Municipal Hospital Administration (No. QML20210801).

Beijing Natural Science Foundation Grant (No. L212039).

Authors' contributions

JW wrote the manuscript. PL reviewed and edited the manuscript. CZ, ZC, and WD participated in the management of the patient. WD was the corresponding author of this manuscript.

REFERENCES

1. Fleming, K. L., Davidson, L., Gonzalez-Gomez, I. & McComb, J. G. Nondysraphic pediatric intramedullary spinal cord lipomas: report of 5 cases. *Journal of Neurosurgery. Pediatrics* 5, 172-178, doi:10.3171/2009.9.peds09238 (2010).
2. Love, J. G., Daly, D. D. & Harris, L. E. Tight filum terminale. Report of condition in three siblings. *Jama* 176, 31-33, doi:10.1001/jama.1961.03040140033009 (1961).
3. Abul-Kasim, K., Thurnher, M. M., McKeever, P. & Sundgren, P. C. Intradural spinal tumors: current classification and MRI features. *Neuroradiology* 50, 301-314, doi:10.1007/s00234-007-0345-7 (2008).
4. Abuzayed, B., Alawneh, K., Al Qawasmeh, M. & Raffee, L. Nondysraphic spinal intramedullary lipoma: A rare case and management. *Turkish Archives of Pediatrics* 56, 85-87, doi:10.14744/TurkPediatriArs.2020.90187 (2021).
5. Guinto G, A. M., Ar´echiga N, *et al*. Different types of syringomyelia and their management: part I. *Contemp Spine Surg* 11, 1-7 (2010).
6. Flint, G. Syringomyelia: diagnosis and management. *Practical Neurology* 21, 403-411, doi:10.1136/practneurol-2021-002994 (2021).
7. Goel, A. *et al*. Central atlantoaxial instability as a cause of syringomyelia mimic intramedullary lipoma. *Journal of Craniovertebral Junction & Spine* 11, 152-154, doi:10.4103/jcvjs.JCVJS_65_20 (2020).
8 Arjipour, M., Ownagh, F. & Eftekharian, M. Fatty syringomyelia mimicking lipoma with spinal cord pilocytic astrocytoma. *The Neuroradiology Journal*, 19714009221140535, doi:10.1177/19714009221140535 (2022).
9. Massimi, L. *et al*. Acute presentations of intradural lipomas: case reports and a review of the literature. *BMC Neurology* 19, 189, doi:10.1186/s12883-019-1413-4 (2019).
10. Pang, D. Commentary to the article: asymptomatic lumbosacral lipomas--a natural history study, by Wykes V, Desai D, and Thompson DNP. *Child's Nervous System: ChNS: Official Journal of the International Society for Pediatric Neurosurgery* 28, 1741-1742, doi:10.1007/s00381-012-1808-7 (2012).
11. Moghaddam, A. M., Tanriöver, N., Ulu, M. O., Muhammedrezai, S. & Akar, Z. Cervical intradural lipoma with associated hemivertebra

formation at C6 level: a case report. *Turkish Neurosurgery* 18, 187-190 (2008).
12. Roujeau, T., James, S., Forin, V. & Zerah, M. Results of the prophylactic surgery of lumbosacral lipomas: the pendulum of management? *Child's nervous system: ChNS: Official Journal of the International Society for Pediatric Neurosurgery* 33, 561-562, doi:10.1007/s00381-017-3345-x (2017).
13. Bhatoe, H. S. et al. Nondysraphic intramedullary spinal cord lipomas: a review. *Neurosurgical Focus* 18, Ecp1, doi:10.3171/foc.2005.18.2.10 (2005).