Rare disease

Recurrent 'universal tumour' of the spinal cord

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Summary

Lipoma is popularly known as the 'universal tumour' because of its ubiquitous presence anywhere in the body. This is the first documented case of recurrent thoracic spinal cord intramedullary lipoma in a 44-year-old man, with a background of spinal dysraphism, which recurred 15 years after initial surgery. He was followed up every 2 years and currently presented with an 8-month history of progressive weakness in his lower limbs. An MRI of the spine confirmed recurrence of lipoma. He underwent redo laminectomy and partial resection and spinal cord decompression with duroplasty. Lipoma, although a low-grade tumour, can cause significant neurological deficits because of its location. Surgical exploration and removal of lipoma is recommended. However, to preserve the functionality of the spinal cord, one may resort to partial resection and aim for spinal cord decompression. The literature on spinal cord lipoma is reviewed and the aetiopathogenesis of this rare occurrence is described.

BACKGROUND

Spinal cord lipomas account for about 1% of all spinal neoplasms and are therefore rare spinal cord tumours.¹ True intramedullary spinal cord lipoma is extremely rare.² The aetiopathogenesis of spinal cord lipoma, however, is not fully understood. Several hypotheses have been postulated to explain the origin of the lipoma. The most common site of spinal cord lipoma has been found to be in the thoracic region. This is a case of recurring lipoma of the thoracic spinal cord, which required further surgical intervention 15 years after the initial surgery.

CASE PRESENTATION

A 44-year-old man, with a background of spinal dysraphism, presented with recurrence of an intramedullary spinal cord lipoma. Fifteen years ago, this patient presented with painless progressive weakness affecting the lower limbs, more on the left side. There was no history of trauma or infection and no significant medical history. Examination at the time revealed reduced power of 4 to 4+/5 bilaterally, increased tone, hyperreflexia with up-going planters as well as reduced sensation. These findings were found bilaterally in the lower limbs. An MRI scan at that stage revealed a lipoma at the levels of T9–T11. Following this, the patient underwent laminectomy and subtotal excision of the intramedullary spinal cord lipoma. He recovered well and regained much of his lower limb functions, with some residual weakness affecting the proximal power of the left lower limb.

He was followed up every 2 years as an outpatient to monitor any new change in his neurological function. A repeat routine MRI 2 years after surgery showed asymptomatic recurrence of the lipoma, now at the levels of T7–T9. Further MRI (figures 1–4) of the thoracic spine 9 years after surgery showed no significant change. It was over the past 8 months that symptoms began to recur and became progressively worse.

Examination on admission revealed wasting of muscles on the left quadriceps femoris. Power on the right side was 4+ to 5/5. The left side, however, showed reduced power throughout, ranging from 3 to 4/5 proximal to distal. Tone and reflexes were normal with up-going plantars. Sensory and dorsal column examination remained normal.

Figure 1 T1 weighted MRI demonstrating the intramedullary lipoma (black arrow).

Figure 2 Sagittal T2 weighted MRI with the lesion delineated by the arrows.

INVESTIGATIONS

An MRI of the thoracic spine was performed to investigate the worsening symptoms, which showed no significant size change from the previous MRI. A T1 and T2 weighted MRI was performed to confirm the diagnosis. A sagittal T1 and T2 weighted MRI revealed a hyperintense signal consistent with fat tissue. Axial T1 and T2 MRI suggested similar features.

Figure 3 Axial T2 weighted MRI with intramedullary lipoma (yellow arrow) and the deformed spinal cord adjacent to the lipoma (orange arrow).

Figure 4 STIR (short T1 inversion recovery—fat suppression) axial MRI demonstrating the hypointense lesion (orange arrow) and spinal cord (yellow arrow).

DIFFERENTIAL DIAGNOSIS

Leiomyolipoma, astrocytoma, ependymoma, meningioma, neurofibroma.

TREATMENT

The patient underwent a redo thoracic spinal laminectomy exploration and partial resection of the lipoma with spinal cord decompression. The lipoma was found to extend 5 cm craniocaudally and was found to occupy 70% of the spinal canal. Intraoperative fat tissue was noted and was found to be relatively less vascular in nature and densely adherent to the cord (figure 5). After decompression, a duroplasty was performed to demarcate the lipoma from the extradural tissue keeping in mind the possibility of further recurrence.

OUTCOME AND FOLLOW-UP

Postoperatively, this patient's neurological state started to improve. Repeat examination of the lower limb a few days after the operation revealed a normal tone and sensation. Reflexes were normal bilaterally, although with the persistence of upgoing planter response. Some improvement was noted in muscle power. Overall, the patient had a good postoperative outcome.

DISCUSSION

Lipomas can occur anywhere in the spinal cord, but frequently they are found localised at the lower thoracic and lumbosacral levels.¹ Spinal cord lipoma is a fatty tumour

Figure 5 Intraoperative picture of the intramedullary lipoma (the black arrows indicate the lipoma, the grey arrows indicate the normal spinal cord, and the blue arrows represent thickened dura mater and scar tissue).

within the spinal cord. Intramedullary spinal cord lipoma is a benign lesion that can present in childhood or later in life, in the third or fourth decade for example. The aetiology of lipoma is not well understood as yet however. Several hypotheses have been postulated on the aetiology of this condition. One of them is the 'developmental error theory'³ which states that adipocytes become misplaced during neural tube development, and cause development of a lipoma in later life. Another hypothesis states that metaplasia of connective tissue in the spine causes the formation of lipoma.⁴ Neural crest cells that fail to prevent mesenchymal cells from forming adipocytes are another possible cause of this occurrence.⁵

Symptoms of lipoma relate to mass effect at the area where they occur as well as to compressive myelopathy.⁶ Spinal cord lipomas account for about 1% of all spinal neoplasms.¹⁻⁷ True intramedullary spinal cord lipoma is extremely rare.² The distribution of lipoma within the spinal cord has been shown to be thoracic in 32% of cases, cervico-thoracic in 24% and purely cervical in 13% of reported cases.⁸ Depending on the level of occurrence, symptoms and signs include sensory disturbances, weakness, change in muscle tone and reflexes and difficulty in emptying the bowel and bladder. Surgical intervention may be required to resolve the symptoms caused by these lesions; no other intervention is as successful as surgical excision recommended currently.

When investigating these lesions as part of the work-up before intervention, both CT and MRI scans can reveal the fat component of the lipoma, but MRI is superior to CT in demonstrating the relationship of the lipoma with surrounding normal neural tissue.⁹⁻¹⁰ Thus, MRI allows for more effective planning of the operative procedure and peri-operative and post-operative care as described in our case.

While reversal of neurological deficits is the ultimate goal of surgery in these cases, the choice between subtotal and complete excision of the tumour is not always straightforward. Complete resection can sometimes be achieved that allows for preservation of neurological function and seems to be curative, as there have been no previous reports of recurrence or metastasis.¹¹ Subtotal excision is often preferred, however, as complete excision carries increased risk of postoperative morbidity as compared to subtotal excision. This was the scenario in our patient and complete resection was not feasible secondary to dense adhesion to the spinal cord. Subtotal removal of lipoma allows the neurological grade to improve postoperatively.² Evidence suggests that decompression with or without duroplasty often provides satisfactory clinical outcome with long-lasting symptom-free survival.¹¹⁻¹³ Duroplasty was performed in our case to delineate the scar tissue from the tumour tissue. A chance of recurrence of lipoma does exist. The difficulty in surgical treatment arises because these lesions do not often have clear-cut margins, making it hard to perform an exact excision while avoiding damage to the

surrounding spinal cord structures.¹⁴ For more apparent reasons, it is vital to preserve neurological function, even if this means incomplete resection at the time of surgery as described above in our case. One study has shown that progression-free survival for total resection was 82.8% at 16 years, compared with 34.6% for subtotal resection at 0.5 years.¹⁵

Learning points

- Intramedullary spinal cord lipoma is a rare condition and, after the initial resection, recurrence must be anticipated.
- Regular follow-up with MRI scan is recommended accompanied by a thorough neurological examination.
- Recurrent intramedullary spinal cord lipoma is best managed by spinal surgery with the aim of resecting the tumour partially or fully depending on the morphology and location in the spinal cord.

Competing interests None.

Patient consent Obtained.

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