

Paraplegia due to spinal epidural lipoma without spinal dysraphism in an adolescent patient: a case report

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ABSTRACT

We report the case of a rare lipoma arising in the epidural space of a 14-year-old boy without spinal dysraphism. Lipomas are rare in pediatric soft tissue tumors, accounting for only about 4% of cases. The incidence of an intraspinal epidural lipoma without spinal dysraphism is extremely rare in pediatric patients. In this case, the patient had progressive motor deficits in the lower extremities and difficulty in urination and defecation. Magnetic resonance imaging showed an extradural tumor compressing the spinal cord at the T3–T7 level. Because of the progressive neurological deficits, we performed an emergency surgery. The tumor was completely resected en bloc, and histopathology revealed mature adipose tissue with fibrous septa, diagnosed as atypical lipomatous tumor / well-differentiated liposarcoma. The patient fully recovered and there was no tumor recurrence for 6 years since the surgery. However, re-examination using fluorescence in situ hybridization after 6 years of surgery changed the diagnosis to lipoma as no amplification of murine double-minute type 2 oncogene was observed. In liposarcoma, histopathological diagnosis using fluorescence in situ hybridization is mandatory. Our case illustrates that immunohistochemical diagnosis alone can be misleading. Hence, prompt surgery is required for progressive neuropathy.

Keywords: spinal epidural tumor, spinal epidural lipoma, spinal epidural liposarcoma

Abbreviations:

MDM2: murine double-minute type 2

CDK4: cyclin-dependent kinase 4

WDL: well-differentiated liposarcoma

ALT: atypical lipomatous tumor

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INTRODUCTION

Although lipoma is a common form of soft-tissue tumor in adults, it is relatively infrequent in children, accounting for only about 4% of pediatric soft-tissue tumors.¹ While lipomas associated with spina bifida are well known, an intraspinal epidural lipoma without spinal dysraphism in a pediatric case is extremely rare. We herein report a rare lipoma arising in the epidural space of an adolescent patient without spinal dysraphism.

CASE PRESENTATION

The patient and his family were informed of and provided consent to the submission of the data of this case for publication. The patient was a 14-year-old boy with no medical or family history of neoplastic disease. Four months prior to admission, he experienced back pain and discomfort that persisted for about a month but did not seek medical attention because the pain improved spontaneously. One month before admission, the patient became aware of weakness in his lower limbs and visited his family doctor. However, X-ray and general blood testing showed no abnormal findings. The patient was admitted to our facility due to progression of muscle weakness in the lower limbs, which resulted in difficulty in maintaining a standing position and eventually, difficulty in urinating and defecating. At the time of admission, the patient's scores for the manual muscle testing of the lower limbs were 3 for iliopsoas, 3 for quadriceps, and 4 for others. No abnormalities were found in upper limb muscle strength. Sensory examination revealed numbness below the nipple and loss of pain perception. The patient became unable to urinate and required a urine guide. No anal sphincter contraction was observed.

X-ray showed no abnormalities. Magnetic resonance imaging (MRI) showed an epidural tumor at T3 to T7 with a low signal intensity on T1-weighted images (Fig. 1A) and an almost homogeneous high signal intensity on T2-weighted images (Fig. 1B). The spinal cord was strongly compressed posteriorly by the tumor. A part of the tumor was enhanced by contrast-enhanced MRI using gadolinium (Fig. 1C). The tumor had progressed to the bilateral vertebral foramina at the T4 level (Fig. 1D). No signal intensity changes were observed in the vertebrae. Computed tomography (CT) revealed no calcified lesions and no bone erosion (Fig. 2).



Fig. 1 Preoperative magnetic resonance images

Fig. 1a: Sagittal view of T1-weighted image

Fig. 1b: T2-weighted image

Fig. 1c: Gadolinium-enhanced image

Fig. 1d: Trans view of T2-weighted image at the T4 level

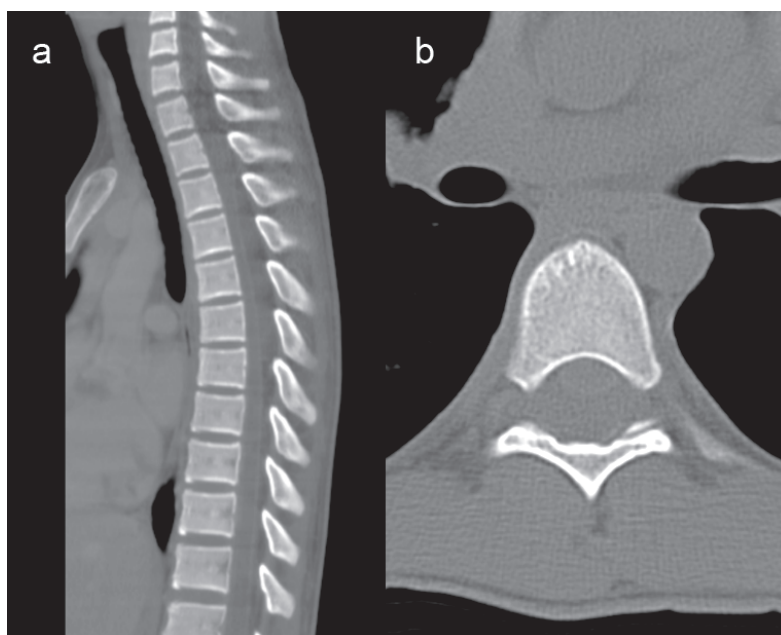


Fig. 2 Preoperative computed tomography images

Fig. 2a: Sagittal view

Fig. 2b: Axial view

Since no well-known disease that occurs in the thoracic epidural has been reported in teenagers, differential diagnosis was considered. Prior to surgery, we suspected neoplastic disease (especially malignant neoplasm) or cystic disease based on the clinical course and imaging findings. However, because of his progressive paralysis, emergency surgery was performed. The surgery included laminectomy at the T3–T7 levels and tumor removal with the aid of microscopy. Intraoperative

ultrasonography showed a clear border between the tumor and surrounding tissue and poor blood flow inside the tumor. Since little adhesion to the surrounding tissue was observed, the tumor was completely resected en bloc, including the portion that had advanced into the intervertebral foramen. No continuity with the dura or nerve roots was observed nor was any traffic seen in the extradural arachnoid cysts. The lamina was then recapped to prevent the progression of kyphosis.² Histopathological analysis of the resected tumor showed that it consisted of mature adipose tissue with fibrous septa, and some lipoblasts were observed. Several hyperchromatic cells were identified, and nuclear atypia was observed (Fig. 3). Thus, a fatty tumor was suspected on differential diagnosis. Since the majority of the tumor was composed of adipose tissue and atypical cells, and lipoblasts were only observed in a small area, the possibility of pleomorphic liposarcoma and myxoid liposarcoma was excluded. Immunohistochemical staining result was negative for murine double-minute type 2 (MDM2) but positive for cyclin-dependent kinase 4 (CDK4). Based on these results, it was determined that the tumor was pathologically suspicious for atypical lipomatous tumor (ALT)/well-differentiated liposarcoma (WDL).

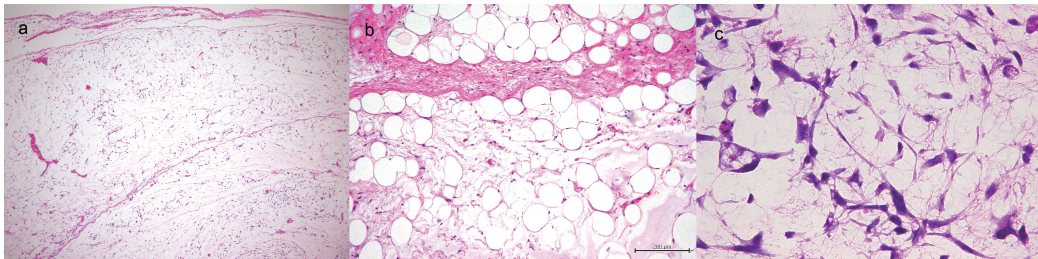


Fig. 3 Microscopic image of the tumor tissue

Fig. 3a-b: The presence of mature adipose tissue with fibrous septa in the tumor.

Fig. 3c: Several hyperchromatic cells present in the tumor tissue. Nuclear atypia was observed.

(Hematoxylin and eosin staining; original magnification: 40 \times , 100 \times , and 400 \times for images a, b, and c, respectively).

We tentatively followed up with the patient as a case of ALT/WDL that could be completely removed. This was because although the aggressive clinical course of the tumor suggested that it was a fatty tumor with malignant potential, the intraoperative findings suggested that it was a benign fatty tumor. However, since the tumor had been completely removed, no additional treatment was required for either of the tumors. Therefore, there was little need to distinguish between the two, and we followed up the patient closely. Postoperatively, the patient's motor paralysis and sensory deficits quickly recovered, and he was able to begin gait training. Bladder and bowel functions gradually recovered. No perioperative complications developed. The patient regained the ability to walk unassisted and was discharged at 3 weeks after surgery. Thereafter, the patient fully recovered to perform daily and sports activities without re-exacerbation of symptoms. An MRI scan at 6 years after surgery showed no recurrence of the tumor (Fig. 4). A case review pointed out that the pathological findings by immunohistochemistry were not definitive; thus, we decided to revisit the diagnosis. Reexamination by fluorescence in situ hybridization (FISH) was performed 6 years after surgery, and the diagnosis was changed to lipoma since no amplification of MDM2 was observed (Fig. 5).

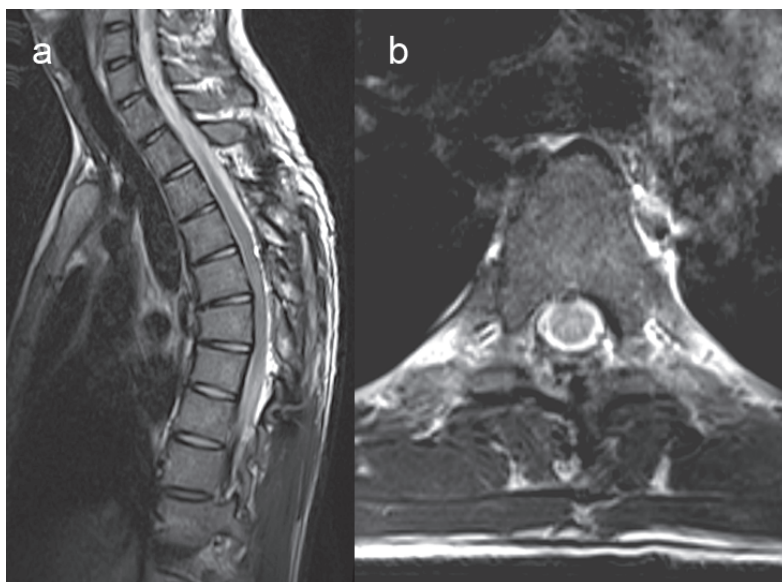


Fig. 4 Magnetic resonance images 6 years after surgery

Fig. 4a: Sagittal view of T2-weighted image

Fig. 4b: Trans view of T2-weighted image

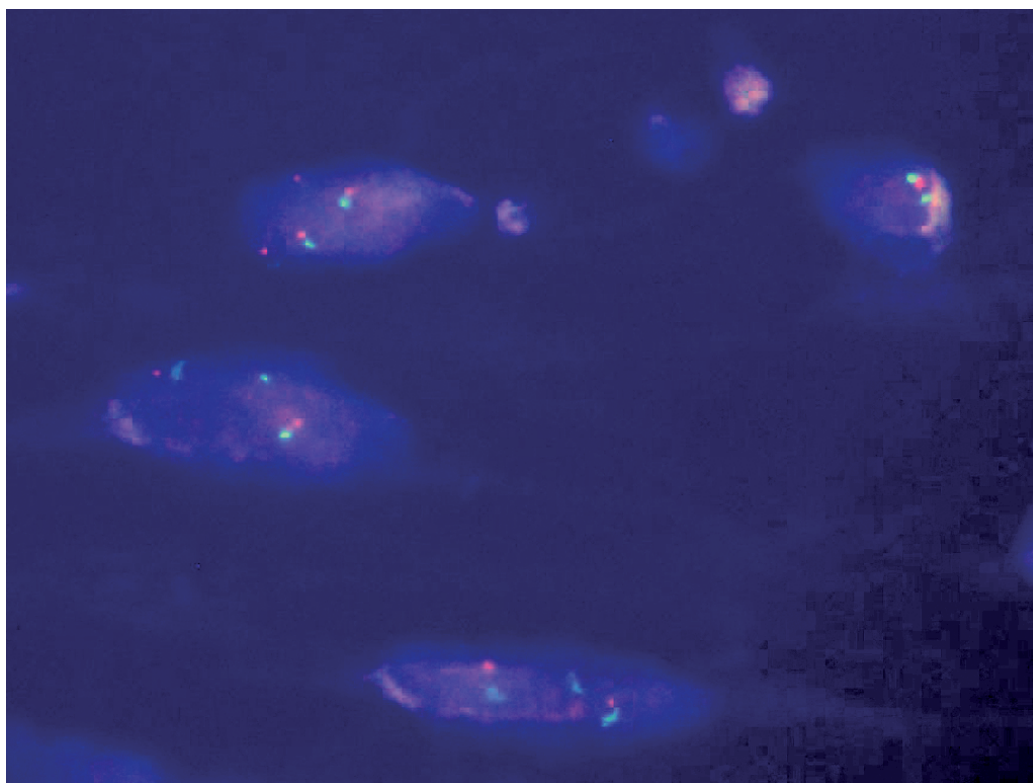


Fig. 5 Bicolor fluorescent in situ hybridization analysis

DISCUSSION

We report the first adolescent case of paraplegia due to an intraspinal epidural lipoma without spinal dysraphism. Because of progressive paralysis, emergency en bloc tumor resection was performed. Although the initial histopathological diagnosis was ALT/WDL, no additional postoperative treatment was performed because the en bloc resection was successful. Complete recovery of paralysis was achieved, and then the patient had a good course with no recurrence for 6 years. Fortunately, we had the opportunity to review the histopathological diagnosis and change the diagnosis to lipoma.

An epidural tumor was suspected preoperatively based on the clinical course and image findings. The differential diagnosis of epidural soft-tissue tumors include nerve sheath tumor, synovial cyst, arachnoid cyst, soft-tissue sarcoma, cavernoma, hamartoma, angioliipoma, hemangiopericytoma, and calcified pseudotumor.³ Hematomas and abscesses are also differential diseases. In this case, the shape was atypical for a nerve sheath tumor, and synovial cyst was not suspected considering the CT findings. Furthermore, the lack of contrast effect was not consistent with cavernoma. Abscess was negative based on blood test findings. An arachnoid cyst was one of the diseases we suspected in this case, along with soft-tissue sarcoma. Spinal angioliipomas are rare epidural tumors, but several case reports have described them as causing paraplegia.^{4,6} Angioliipoma could not be ruled out in this case because the MRI findings varied depending on the ratio of fat to vascular components.⁷

Histologically, a large amount of mature adipose tissue components were observed in the tumor, and the differential diagnosis included lipoma, spinal epidural lipomatosis (SEL), and liposarcoma. Although the MRI findings of lipomas are generally identical to those of an adipose tissue, approximately 13% of lipomas contain a non-fatty component.⁸ Therefore, lipoma could not be ruled out. SEL is a disease that is caused due to an excessive growth of adipose tissue in the epidural area, and case reports with paraplegia due to SEL have been published.^{9,10} Since the patient was not under steroids, which is a risk factor for SEL, and the lesion did not include the entire epidural, but only a portion of the thoracic spine, the possibility of SEL was considered too low. Liposarcoma is one of the most common malignant soft-tissue tumors in adults,¹¹ but it is rare in children, accounting for only about 2% of all pediatric soft-tissue sarcomas.^{12,13} Liposarcoma is more heterogeneous than lipoma on MRI images, containing more non-fatty elements, and in some cases, no fat at all.¹

In this case, the initial pathological diagnosis was ALT/WDL, but the diagnosis was later changed to lipoma. Before submitting the current report, the pathologist mentioned the necessity of performing FISH instead of immunohistochemistry in diagnosing ALT/WDL. After performing FISH, the initial diagnosis of ALT/WDL was changed to lipoma. Distinguishing deep lipomas from ALT/WDL can be problematic. In ALT/WDL, *MDM2* and *CDK4* are amplified, and these genes are detected by immunohistochemical staining and FISH to differentiate between ALT/WDL and lipoma. At the time of initial diagnosis of this case, immunostaining for *MDM2* and *CDK4* was thought to be effective in diagnosing ALT/WDL.¹⁴ However, the uncertainty of immunostaining has since been pointed out; immunostaining for *MDM2* and *CDK4* for ALT/WDL had a sensitivity of 45% and 41% and specificity of 98% and 92%, respectively.¹⁵ Furthermore, in some cases, even benign lipomatous lesions were positive, in which case the false-positive staining was comparable in intensity to ALT/WDL.¹⁵ For this reason, the determination of *MDM2* gene amplification by FISH is now the gold standard for the diagnosis for ATL/WDL.¹⁵⁻¹⁷ Since cells with nuclear atypia were found in this case, histopathological diagnosis using immunostaining for *MDM2* and *CDK4* was performed. The tumor cells were negative for *MDM2* but positive for *CDK4*, which was diagnosed as ALT/WDL. However, we later had the opportunity to perform

additional studies using FISH for *MDM2*. Unexpectedly, no amplification of *MDM2* was found; therefore, we rejected the initial ALT/WDL diagnosis and reconfirmed the diagnosis as lipoma.

Spinal epidural lipoma without spinal dysraphism is an uncommon condition and has been estimated to account for 0.4%–0.8% of intraspinal tumors.¹⁸ Although no cases of lipoma in adolescents have been retrieved, several adult cases have been reported. Meisheri et al¹⁹ reported a case of epidural lipoma at T4–T9 in a 20-year-old man who underwent laminectomy and tumor removal 7 days after onset but did not recover from neurological deficits. Noia et al²⁰ described the case of a 46-year-old man who developed a thoracic vertebral fracture with the same level of lipoma at T5–T6. His spinal cord was compressed by the lipoma due to kyphosis caused by the fracture, which resulted in neurological deficits. He underwent laminectomy, tumor removal, and fixation to improve his symptoms. Wiedemayer et al²¹ described a lipoma at T2–T10 in a 48-year-old man 3 years after symptom onset. He underwent unilateral laminectomy and tumor removal. Reviewing these case series revealed that the severity of myelopathy was strongly associated with neurological recovery and prompt surgical decompression is mandatory.

Our report describes the features and the various evaluations and verifications implemented to diagnose a rare spinal lipoma in an adolescent patient. Histopathological diagnosis using FISH is mandatory in liposarcoma, considering diagnosis using only immunohistochemistry can be misleading. Prompt en bloc tumor resection after neurological symptoms is crucial for a sufficient neurological outcome.

DISCLOSURE STATEMENT

The authors state that they have no conflict of interest.

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