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Lipoblastomatosis Extended into the Lumbar Spinal Canal in a Child: A Case Report

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Established Facts

- Lipoblastoma and lipoblastomatosis are rare benign mesenchymal adipose tumors that originate from embryonic white adipocytes and occur most commonly in infancy and early childhood.
- Lipoblastomas occur in the extremities and trunk, resulting in the fact that infiltration into the spinal canal has rarely been reported.
- Although lipoblastoma is benign with no potential for metastasis, it is prone to local recurrence.

Novel Insight

- We report here a rare case of lipoblastomatosis extending into the spinal canal.
- Little has been reported about lipoblastoma developing in the spinal canal and causing neurological symptoms due to compression.
- If you can find the difficulties in pathological diagnosis, molecular analysis could be used to differentiate between myxoid liposarcoma and lipoblastoma.

Keywords
Lipoblastoma · Lipoblastomatosis · Spinal canal

Abstract

Introduction: Lipoblastoma and lipoblastomatosis are rare benign mesenchymal adipose tumors that originate from embryonic white adipocytes and occur most commonly in infancy and early childhood. Lipoblastomas occur in the extremities and trunk, including the retroperitoneum and peritoneal cavity. Therefore, infiltration into the spinal canal has rarely been reported. ***Case Presentation:*** A 4-year-old girl presented to our clinic because of difficulty sitting on the floor with her legs straight. She also complained of enuresis and constipation for the past 6 months with persistent headaches and back pain evoked by body anteflexion. A magnetic resonance imaging revealed a massive lesion of the psoas major muscle, retroperitoneal, and subcutaneous spaces, extending into the spinal epidural space between L2 and S1. The patient underwent surgery which resulted in gross total removal of the tumor from the spinal canal. The mass was yellowish, soft, lobulated, fatty, and easily

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removed from the surrounding structures. Pathology confirmed the diagnosis of lipoblastoma. The postoperative course was uneventful, and the patient was discharged without any signs of neurological deficit. ***Conclusion:*** We herein discuss a rare case of lipoblastoma extending into the spinal canal, resulting in neurological symptoms. Although this tumor is benign with no potential for metastasis, it is prone to local recurrence. Therefore, close postoperative observation should be performed.
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Introduction

Rare benign mesenchymal adipose tumors derived from embryonic white fat cells, called lipoblastomas or lipoblastomatosis, are common in infancy and early childhood; nearly 90% of cases are diagnosed in the first year of life [1, 2]. Lipoblastoma is a rare tumor estimated to account for less than 19–30% of all pediatric adipose masses [3–5]. Although this tumor is benign and has no metastatic potential, it is prone to local recurrence [6, 7]. When locally invasive, it was originally described as lipoblastomatosis by Vellios et al. [2, 6]. Lipoblastomatous tumors have been reported in the extremities and trunk, including the retroperitoneum and peritoneal cavity. Therefore, infiltration into the spinal canal has rarely been reported. We describe a surgical case of lipoblastoma extending into the spinal canal in a 4-year-old girl.

Case Presentation

A 4-year-old girl presented to our clinic because of difficulty sitting on the floor with straight legs. She complained of persistent enuresis with constipation, which suggested a neurogenic rectal bladder disorder, although we did not perform an urodynamic study. She also complained of headaches and back pain evoked by body anteflexion for the past 6 months. Neurological examination revealed positive straight leg raising test bilaterally (positive Lasegue sign). Although she had difficulty with gait, no gross neurological abnormalities, either motor or sensory, were observed. Magnetic resonance imaging (MRI) revealed a 9 × 5 cm mass lesion in the right psoas major muscle, retroperitoneal, and subcutaneous spaces (Fig. 1a–e), which was lobulated and well circumscribed. In addition, the mass lesion was suspected to invade the spinal epidural space through the bilateral L2–S1 foramina. In particular, the right L4 foramen appeared

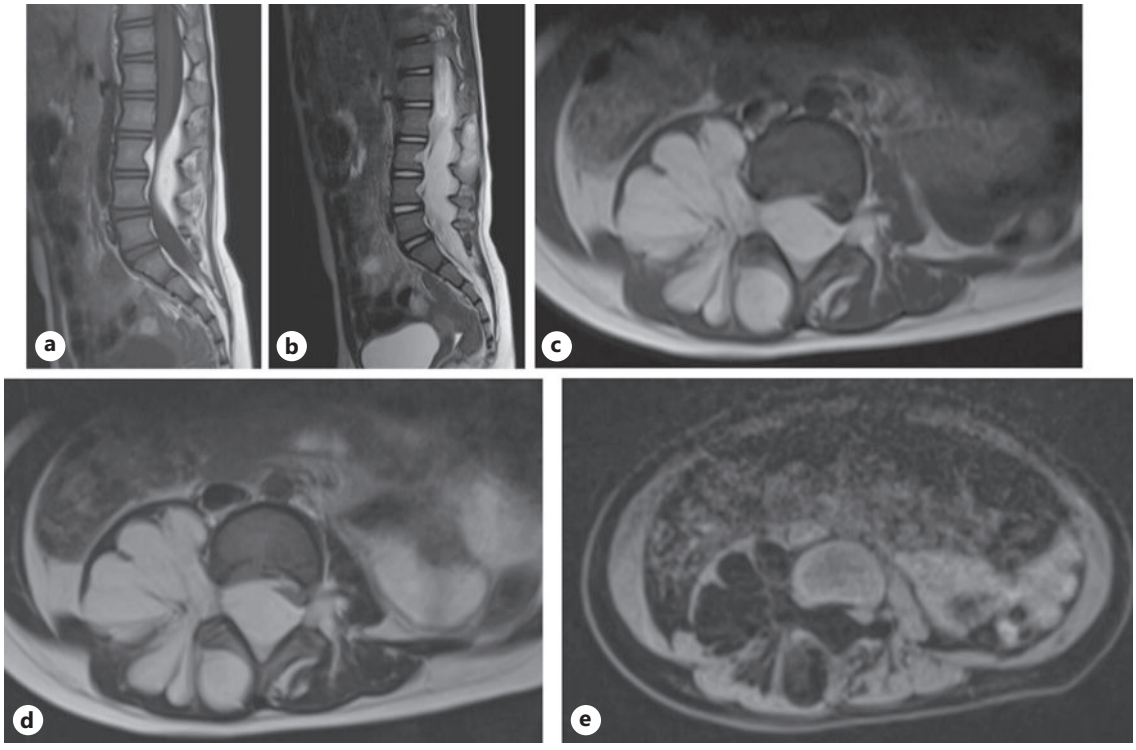

Fig. 1. Preoperative magnetic resonance images (MRIs), showing hyperintense tumor (9 × 5 cm) exceeding into the spinal canal from paraspinal muscle on T1-weighted sagittal (**a**), T2-weighted sagittal (**b**), T1-weighted axial (**c**), T2-weighted axial (**d**), and fat-suppression sequence (**e**).

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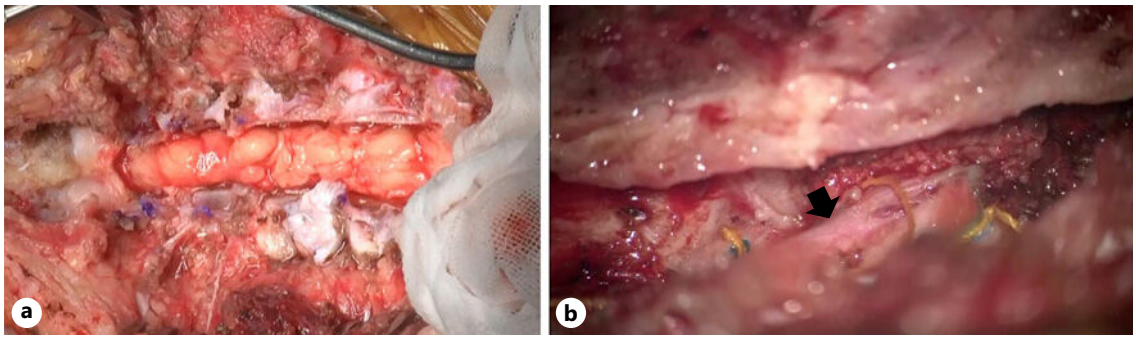

Fig. 2. Surgical photograph after laminotomies, showing yellowish, soft, lobular, and fatty tumor in the spinal canal (**a**), the tumor along the root of L4, and enlargement of the intervertebral foramen (**b**). Arrow: right spinal root of L4.

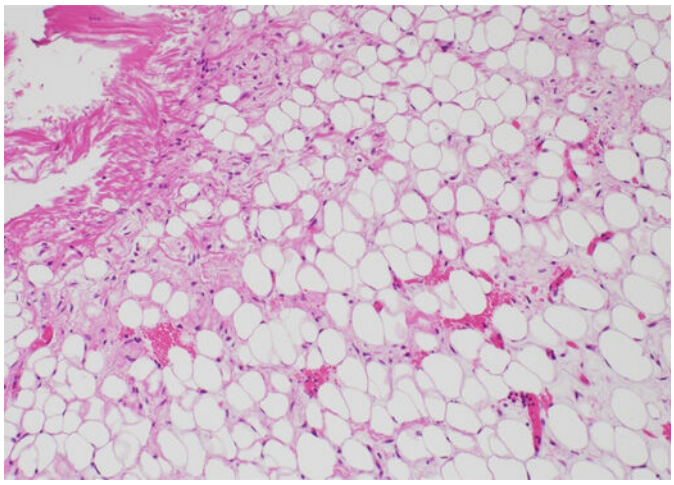

Fig. 3. Histopathological examination showed lobules of mature adipose cells, slight nuclear hyperplasia, spindle-shaped cells, and myxoid stroma, being confirmative findings of lipoblastoma.

to be the main invasion pathway, resulting in a tumor mass that grew significantly on the right side, pushing the dural sac to the left side. MRI showed high intensity on T1WI and T2WI, with hypointensity on the fat-suppression sequence (Fig. 1e) and no contrast enhancement. Computed tomography showed a low-density mass lesion and enlarged intervertebral foramina at L2/3, L3/4, and L4/5, although all laminae were intact. Based on these findings, lipoblastoma was the most likely diagnosis. The patient underwent resection of the tumor in the spinal canal with bulbocavernosus reflex and spinal evoked potential monitoring; the bulbocavernosus reflex and evoked potentials were unchanged throughout the surgical procedure. Laminotomies from L1–L5 achieved gross total tumor removal in the spinal canal. Tumors in the retroperitoneum and paraspinal muscles were left in place because there were no physical signs and symptoms related to the tumor. The tumor was yellowish, soft, lobulated, and fatty and could be easily removed from the surrounding structures. The attachment of the tumor to the dura was weak, as was the usual spinal epidural fat. However, the surgeon observed the right L4 foramen enlarged and tight adhesion between the tumor and the spinal root, suggesting invasion through the L4 foramen (Fig. 2a, b). Ultrasonography confirmed the absence of intradural invasion, and the laminae were fixed with absorbable plates. All spinal roots were preserved. Pathological examination showed lobular growth of mature adipocytes, slight nuclear hyperplasia, spindle-shaped cells separating the lobules, and some mucinous interstitium (Fig. 3). Based on these findings, we diagnosed the tumor as lipoblastoma.

The postoperative course was uneventful; she was discharged from the hospital with improvement of her preoperative signs and symptoms. She also had normal continence at discharge. Postoperative MRI showed no residual tumor in the spinal canal (Fig. 4a, b). However, because the residual tumor in the right psoas muscle or subcutaneously was at risk of local regrowth, we decided to follow the patient closely with MRI.

Discussion

We report a 4-year-old child with a massive lipoblastoma extending into the lumbar spinal canal. Lipoblastoma is a rare benign tumor that occurs predominantly in infants and children under the age of three with a male predominance [1, 2, 5, 8, 9]. Although benign, this tumor can grow rapidly, compressing surrounding structures and causing symptoms by mass effect. This is quite different from dysraphic lipomas, such as the conus type of spinal lipoma, which always reside in the dural sac, adhere to the conus medullaris, and encase some spinal roots, resulting in the fact that even large lipoblastomas may have a good prognosis. Gross total removal of the tumors is considered the gold standard treatment for lipoblastoma due to a 9–25% chance of recurrence, although metastasis and/or malignant transformation have not been reported [3, 6–9]. Two types of tumors have been reported: lipoblastoma is a localized and circumscribed type with a tendency to occur superficially and mimic lipoma. Another type of tumor is

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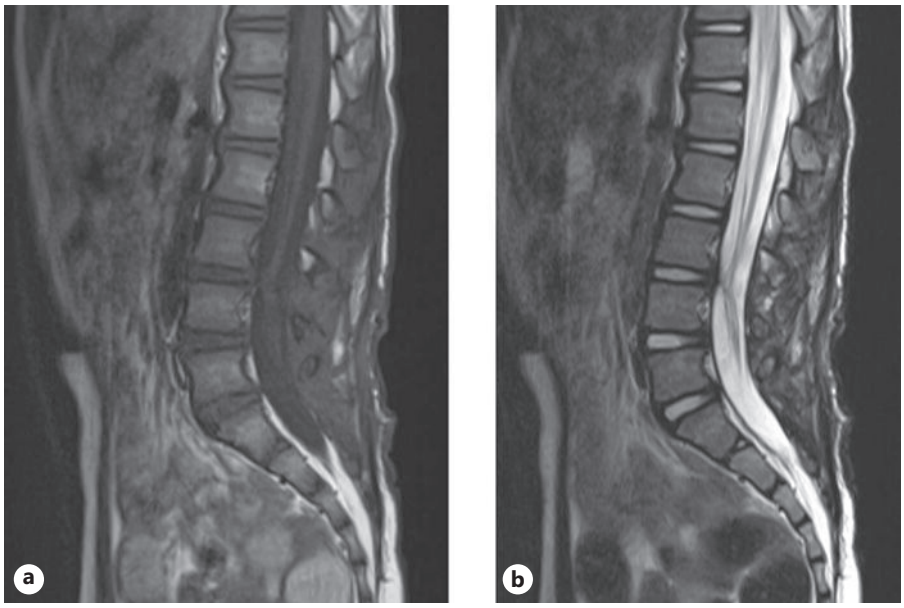

Fig. 4. Postoperative MRI, T1-weighted sagittal (**a**), T2-weighted sagittal (**b**), revealing that the intraspinal tumor was completely resected and the tumor was left in paraspinal muscles.

lipoblastomatosis, which is a diffuse type with infiltrative growth that tends to occur in deeper tissues and has a higher recurrence rate [2]. Chung and Enzinger [6] described 72% incidence in the extremities in 35 patients. Other reported locations include the chest wall [8, 10], mediastinum [1, 9], retroperitoneum [1, 9, 11], head and neck [9], etc. However, little has been mentioned about lipoblastomas developing in the spinal canal. Takebayashi et al. [12] reported a 3-year-old girl with a dumbbell-shaped lipoblastoma extending into the cervical spine and recommended early surgical intervention to avoid neurological deficits. Spinal dumbbell lipoblastomas occurring in the epidural space of the entire spinal canal have been reported, resulting in paresis due to compression of the thecal sac [13, 14]. In our case, we used a single-stage posterior approach with laminotomy followed by fixation of the laminae with absorbable plates to prevent spinal deformity. If the tumor is not expected to be strongly adherent to the surrounding normal tissue preoperatively, especially the ventral tissue, it can be resected through a posterior approach (Fig. 4). Laminotomy should be the standard in growing children to prevent instability and deformity resulting from laminectomy [15]. Radical excision of lipoblastoma is the gold standard, but it is not always possible to achieve because there are some strategic and technical difficulties in performing the surgical procedure without compromising function. We should be conscious that, despite the size of the tumor, it is a benign tumor. The residual extraspinal tumor should be carefully monitored for at least 3–5 years because of the possibility of regrowth and reinvasion into the spinal canal.

Lipomas and liposarcomas are in the differential. Lipomas are slow-growing, and the presence of thin fibrous septa on MRI is key to differentiation, but of course, the exact diagnosis is made through histopathological examination. In lipomas, mature adipocytes proliferate subcutaneously in a nodular fashion and are characterized by separated adipose lobules. Their adipose lobules are larger than normal adipose tissue. The adipocytes are not abnormally large or small, and there are no atypical lipoblasts. There is usually no recurrence after resection, but incomplete resection may lead to recurrence. On the other hand, lipoblastomas are lesions that show increased numbers of lobulated adipocytes surrounded by fibrous connective tissue, with mucinous deposits in the interstitium and various forms of increased vascularity. The lesions are composed of a mixture of mature adipocytes and immature lipoblasts of various morphologies, ranging from star-shaped and spindle-shaped to annular. Areas of increased mature adipocytes resemble normal lipomas. Although the definitive pathology of lipoblastoma differs from that of lipoma as noted above, lipoblastoma more closely reflects many of the features of lipoma, including early onset, the ability to mature into a simple lipoma, a mature adipocyte-dominated cellular composition, and a benign course.

Liposarcoma occurs in the lower limbs and retroperitoneum and is often seen as a slow-growing, painless mass. Because lipoblastoma with a mucus-rich substrate is similar to the pathological findings of myxoid liposarcoma, deep consideration should be taken when diagnosing

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lipoblastoma [16]. In the present case, the preoperative diagnosis was lipoblastoma based on imaging and neurological examination, and few other diagnoses were suspected. Molecular analysis may help make an accurate diagnosis when histological evaluation alone is not diagnostic. Lipoblastomas are known to have rearrangements in 8q11–13 affecting the PLAG1 gene [16]. Therefore, the molecular analysis could be used to differentiate between myxoid liposarcoma and lipoblastoma.

In conclusion, we report a rare case of lipoblastoma extending into the spinal canal with successful gross total removal of the tumor in the spinal canal. However, close postoperative observation should be performed because of a higher recurrence rate.

Statement of Ethics

Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the parent/legal guardian of the patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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Author Contributions

Conception, design, acquisition of data, and drafting the article: Marina Saga. Critically revising the article and study supervision: Akira Yamaura and Tadashi Miyagawa. Revision of submitted version of the manuscript: all authors.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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