The Spine Journal 15 (2015) e71–e75

Case Report

Primary pleomorphic liposarcoma of the thoracic epidural space: case report

P. David Halevi, MD[a], Fernando Ramirez-de-Noriega, MD[a], Yakov Fellig, MD[b], J. Moshe Gomori, MD[c], José E. Cohen, MD[a,c], Eyal Itshayek, MD[a,*]

[a]*Department of Neurosurgery, Hadassah-Hebrew University Medical Center, POB 12000, Jerusalem 91120, Israel*
[b]*Department of Pathology, Hadassah-Hebrew University Medical Center, POB 12000, Jerusalem 91120, Israel*
[c]*Department of Radiology, Hadassah-Hebrew University Medical Center, POB 12000, Jerusalem 91120, Israel*

Received 19 June 2015; revised 19 August 2015; accepted 25 August 2015

Abstract

BACKGROUND CONTEXT: Pleomorphic liposarcoma (PLS) is a rare malignant soft tissue tumor comprising 5%–15% of liposarcomas and characterized by high malignant potential. To our knowledge only three cases of this entity have been reported in the spine.
PURPOSE: We describe the only reported case of a purely epidural PLS with no macroscopic bone involvement at diagnosis.
STUDY DESIGN/SETTING: A case presenting clinical evidence that PLS may arise from the epidural fat is reported.
METHODS: The clinical presentation, management, and outcome in a case of primary PLS of the thoracic spine, and a review of the literature, are presented.
RESULTS: A 70-year-male presented with sudden onset lower extremity weakness, constipation, and back pain. Magnetic resonance imaging revealed an epidural lesion at T5 with noted mass effect compressing the spinal cord and extension to the T5–T6 foramen. Urgent decompressive laminectomy with gross total resection was performed. Histopathology revealed high-grade PLS. Adjunct radiotherapy was prescribed. The tumor recurred 3 months later. In spite of repeat surgery, additional radiation, and chemotherapy, the patient developed widespread metastases and succumbed to his disease 1 year after treatment began.
CONCLUSIONS: Spinal PLS is a rare entity, but nonetheless may arise from epidural fat and should be considered in the differential diagnosis of primary spinal cord lesions.

Keywords: Bone invasion; Epidural fat; Pleomorphic liposarcoma; Primary spinal tumor; Spinal cord compression; Spinal liposarcoma

Introduction

Liposarcoma is the most common soft tissue sarcoma in adults, accounting for 20% of all mesenchymal malignancies. The World Health Organization classifies malignant liposarcomas into four histologic subtypes: myxoid, mixed, pleomorphic, and dedifferentiated [1]. Pleomorphic liposarcoma (PLS) is a high-grade sarcoma that most commonly presents in the thigh and pelvis of older adults; 25% of cases present with metastatic disease, most commonly to the lungs, liver, and skeleton [2]. It is the rarest of the liposarcomas, with only 5%–15% meeting diagnostic criteria for this subgroup [3–5]. In patients with localized disease at diagnosis, there is a 5-year survival rate of approximately 60% [3,4,6–8]. Recurrent disease, no surgical resection, and positive resection margins are independent poor prognostic indicators [2].

Primary PLS of the spine is a particularly rare entity, with only three cases reported [7–9]. In all of the cases, the tumor presented with a significant involvement of the vertebral body

FDA device/drug status: Not applicable.

Author disclosures: ***PDH:*** Nothing to disclose. ***FRN:*** Nothing to disclose. ***YF:*** Nothing to disclose. ***JMG:*** Nothing to disclose. ***JEC:*** Nothing to disclose. ***EI:*** Other: MFast (B, outside the submitted work).

The authors did not receive financial support for the preparation of this case report and have no conflicts of interest.

* Corresponding author. Department of Neurosurgery, Hadassah-Hebrew University Medical Center, POB 12000, Jerusalem 91120, Israel. Tel.: +972 2 677 7092; fax: +972 2 641 6281.

E-mail address: eyal.itshayek@gmail.com (E. Itshayek)

http://dx.doi.org/10.1016/j.spinee.2015.08.059
1529-9430/

at diagnosis. We present a case of a primary PLS in the thoracic spine that was purely epidural, extending for the full height of the T5 vertebral body at the right foramen. Bony involvement was not apparent on preoperative imaging or during the initial surgery. When the tumor recurred, involvement of the vertebral body was evident on magnetic resonance imaging (MRI), and microscopic bone invasion was found on histopathologic examination.

Case report

A 70-year-old man presented to the Emergency Department complaining of constipation. The patient was sent home after symptomatic treatment, only to return after 3 days with complaints of continued constipation, back pain, and sudden onset of progressive bilateral lower extremity weakness. Neurologic examination on admission was significant for +4/5 motor strength in the bilateral lower extremities, bilateral Babinski, and sensory level cut at T10 (American Spinal Injury Association [ASIA] Impairment Scale [AIS] D [10]). The patient underwent emergency MRI of the thoracic and lumbar spine, which revealed a hypointense solid, cystic, epidural nonenhancing mass. The mass was located in the right lateral aspect of the spinal canal at T5, spanning the discs at T4–T5 and T5–T6 with noted extension into the right foramen at T5–T6. No bone involvement was evident. The tumor measured approximately 2.5 cm×1 cm×1 cm, creating a prominent mass effect with marked cord compression (Fig. 1).

First surgery

The patient underwent emergency T4–T5 laminectomy. At surgery, the lesion was a white caseating mass in the spinal canal, emanating from the epidural space and expanding into the right T5–T6 foramen. No macroscopic bone involvement was observed. Gross total resection was achieved via piecemeal excision followed by T5–T6 foraminotomy. The patient tolerated the procedure well with no perioperative complications.

Pathologic diagnosis and clinical course

Histopathology revealed high-grade PLS (Fig. 2).

The patient's postoperative course was uneventful, and he made a complete neurologic recovery (ASIA E). MRI performed 1 month later showed no evidence of residual tumor. Positron emission tomography-computed tomography performed in the same period showed an area of increased uptake in the T4–T6 right paravertebral area with no evidence of other primary or systemic disease (Fig. 3).

The patient was referred for adjuvant external beam radiation therapy (EBRT); however, he was reluctant to begin treatment. He returned to the Emergency Department 3 months after his first surgery, after only one treatment, complaining of progressive lower limb weakness of 3 days' duration, difficulty initiating urination, numbness below the nipple line, and abdominal distention. On examination, the patient had

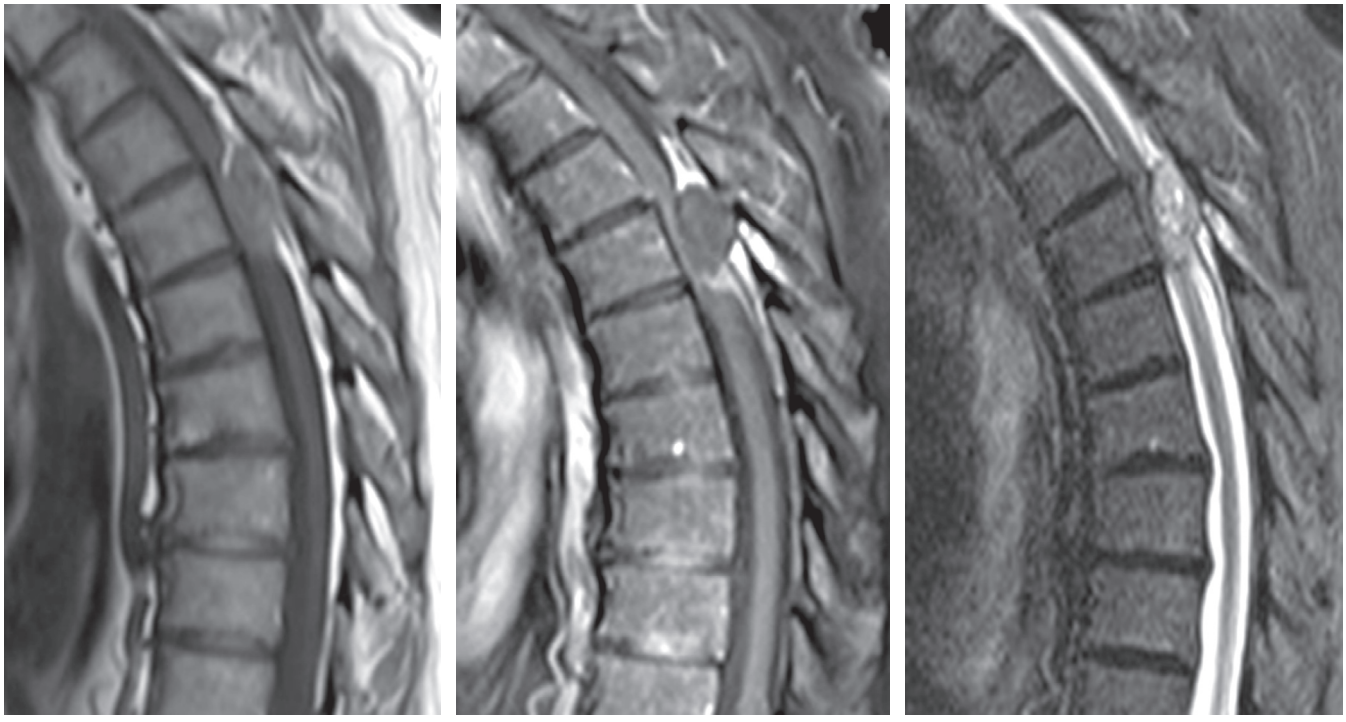

Fig. 1. Sagittal MRI in a 70-year-old man showing a posterior non-enhancing solid, cystic epidural tumor with cord compression spanning the discs at T4–T5 and T5–T6. (Left) T1-weighted; (Middle) Fat-suppressed, gadolinium-enhanced; and (Right) T2-weighted studies.

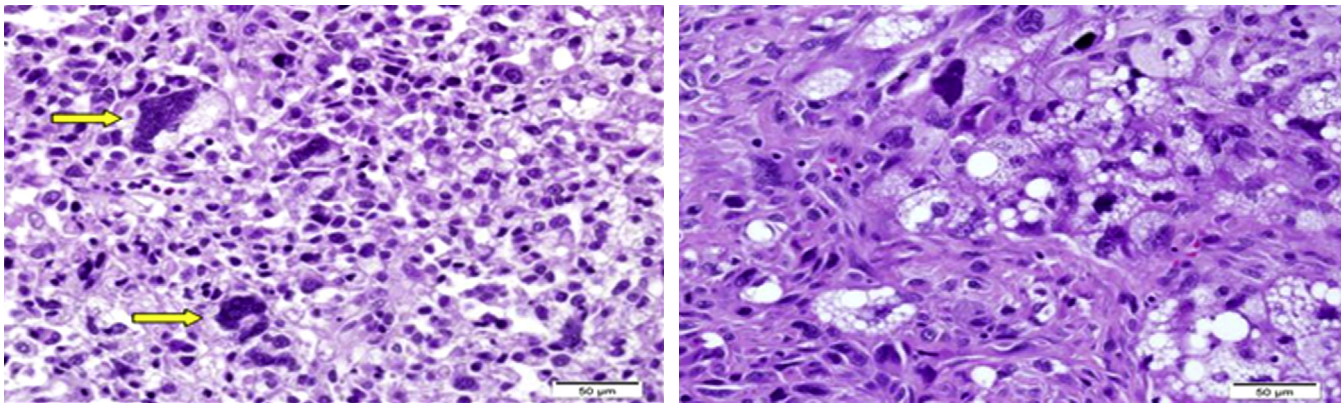

Fig. 2. Paraffin-embedded sections stained with hematoxylin and eosin (H&E) from the initial surgery demonstrated a malignant neoplasm composed of occasional pleomorphic lipoblasts (Left, arrows) in a background of high-grade non-lipogenic pleomorphic sarcoma, with areas of epithelioid features (Left) and spindle-cell features (Right), consistent with pleomorphic liposarcoma. (Original magnifications ×40.)

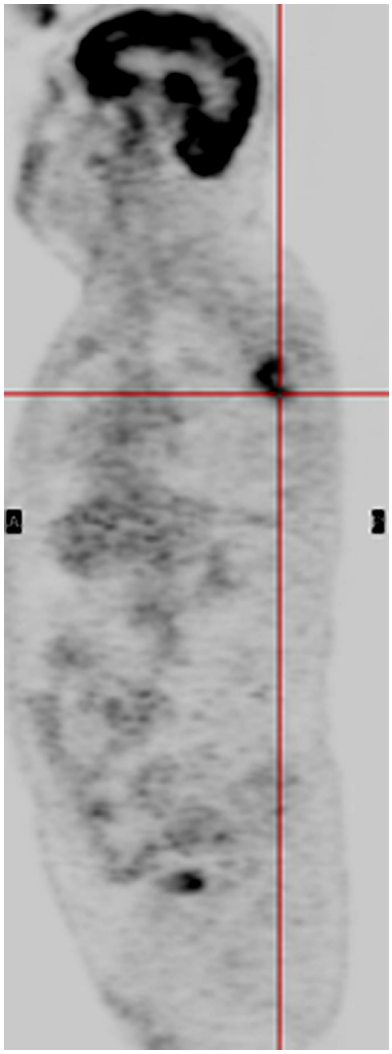

Fig. 3. Lateral fludeoxyglucose (18F) positron emision tomography (^{18}F-FDG PET) performed after the first surgery shows FDG uptake at T5 with no evidence of systemic disease.

bilateral lower limb weakness (proximal 3/5 distal +4/5) and sensory level at T5–T6 (ASIA D). Urgent MRI revealed an extensive recurrent epidural mass at the surgical site. There was compression of the posterior and right lateral aspects of the cord extending from T3 to T6, most prominently at T4–T5, with infiltrating enhancing pathology now involving the T5 vertebral body (Fig. 4).

Second surgery, histopathology, and clinical course

The patient underwent emergency partial T5 vertebrectomy, right T4 pediculectomy, and resection of the head of rib 5. No solid space-occupying lesion was identified, but a white caseating mass was resected from the paraspinal space. T2–T9 posterior fixation was performed with pedicle screws and rod bars, and the T5 vertebral body was reconstructed with methyl methacrylate. The postoperative course was unremarkable with partial neurologic recovery (ASIA D). Histopathology again revealed PLS. The tumor was infiltrating native bone with areas of dedifferentiation with osteoid formation (Fig. 5).

Surgery was followed by radiotherapy, but 5 months later the patient returned to the Emergency Department with complaints of urinary retention, loss of bowel control, and worsening paraparesis, which was more significant on the left side (ASIA D). Magnetic resonance imaging confirmed recurrence of the tumor at the edge of the previous resection. The patient underwent emergency T7–T8 laminectomy with removal of a white caseating lesion similar to the tumor seen at the first surgeries. Histopathology revealed PLS with extensive necrosis, which was possibly treatment-related.

Widespread metastatic disease was detected following another episode of cord compression at T1-T2, 9 months after the first surgery. The patient was treated with a second session of radiation and chemotherapy. A year following the initial diagnosis of PLS, he succumbed to metastatic disease.

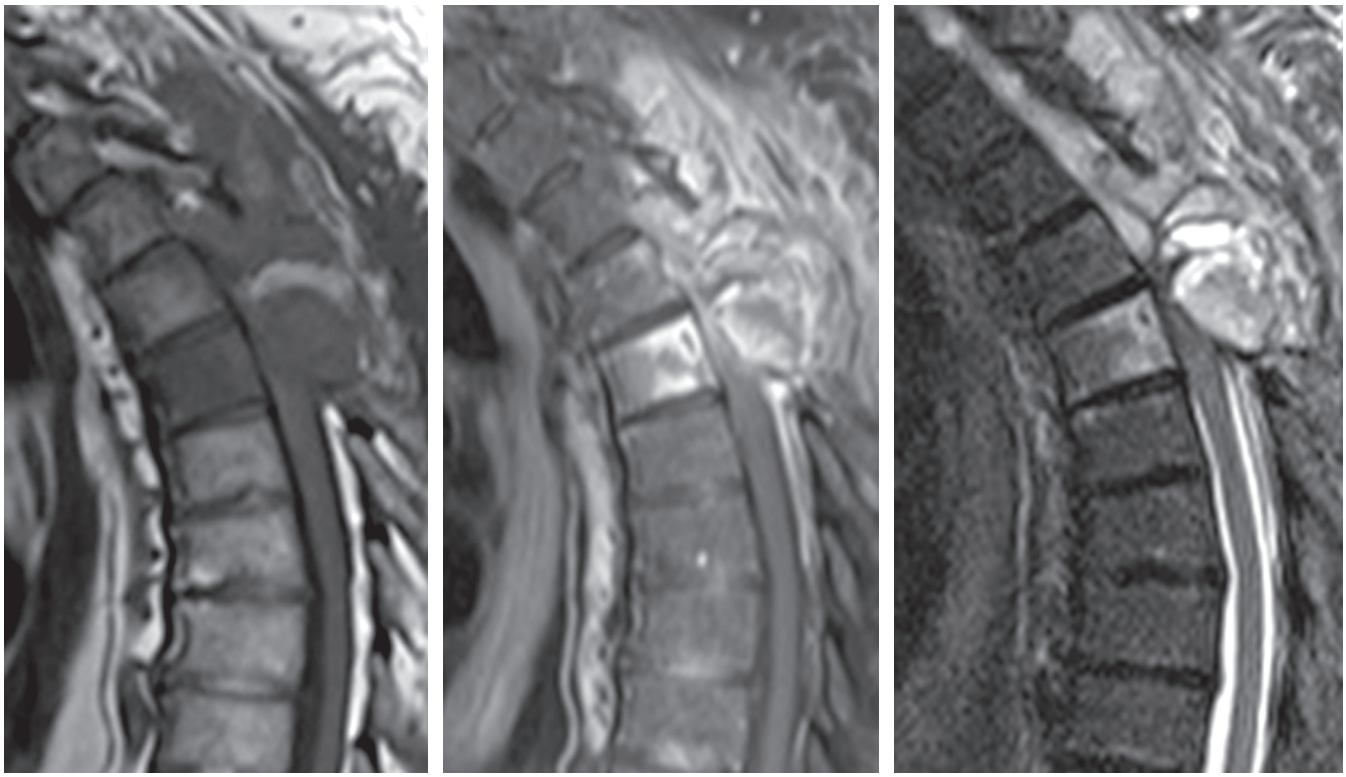

Fig. 4. Sagittal MRI in the same patient 3 months after diagnosis shows recurrence of the posterior epidural tumor at the site of the first surgery, now with anterior and posterior bone involvement and severe cord compression with rostral edema. Note heterogenous enhancement. (Left) T1-weighted; (Middle) fat-suppressed, gadolinium-enhanced; and (Right) T2-weighted studies.

Discussion

We present a rare case of primary spinal PLS in a 70-year-old man. The case is unique in that there was no radiologic or macroscopic involvement of the vertebral body at initial presentation.

As a neoplastic entity, PLS is known to have a very aggressive clinical course with overall median survival in patients with metastatic disease of 9.1 months and with a 1-year disease specific survival rate of 45% [2].

In general, primary tumors of the spine have a low incidence when compared with the rate of diagnoses of other neoplastic spinal lesions such as metastatic disease, multiple myeloma, lymphoma, and other primary spine tumors [11]. Primary spinal PLS is particularly rare; to our knowledge, only three cases have been reported in the literature [7–9]. Hamlat et al. [8] describe a case of primary liposarcoma of the thoracic spine in a 45-year-old woman who presented with a lesion involving the T7–T8 vertebrae and pedicle. Compression fracture was seen on the initial imaging workup. The

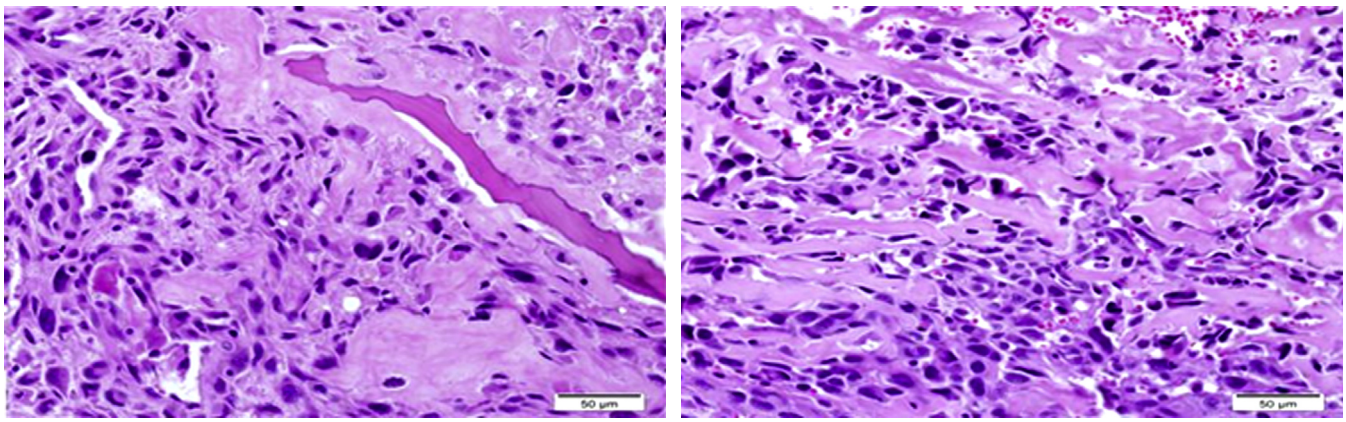

Fig. 5. Paraffin-embedded sections stained with H&E from the second surgery in this patient. The neoplasm infiltrated and destroyed native bone (Left) and manifested areas of dedifferentiation with osteoid formation (Right), consistent with a diagnosis of pleomorphic liposarcoma. (Original magnifications ×40.)

lesion remained confined to the bone with no involvement of the epidural fat, dura, or surrounding tissue. Lmejjati et al. [9] presented a similar case in the lumbar spine in a 45-year-old male. The lesion invaded the L4–L5 vertebral body with involvement of the roots of the intradural area surrounding the cauda equina. More recently, Barra de Moraes et al. [7] described the management of a 60-year-old woman who presented with an extensive lytic lesion in the L4 vertebral body that invaded the medullary canal and neural foramens with nerve root compression.

Our case is unique, as it is the only case that describes a primary liposarcoma presenting as a primarily epidural tumor with no evidence of bone invasion on preoperative imaging or macroscopic invasion at the first surgery. The tumor in our case most likely arose from epidural fat. At the first surgery we achieved macroscopic gross total resection; nevertheless, the tumor recurred in the initial location, most likely from a microscopic cellular remnant. Whereas surgery is rarely curative because microscopic epidural spread is likely to have occurred even when only an isolated lesion is noted on imaging, timely intervention may provide the important benefit of preserving ambulation and sphincter control.

In our case it is possible that the purely epidural nature of the lesion allowed the patient to make a complete neurologic recovery following the first surgery, as his symptoms resulted from an epidural cord compression and there was no direct involvement of nerve tissue. One can speculate whether there would have been an increase in his progression-free and overall survival had he undergone radiation therapy immediately after surgery as recommended, rather than delaying treatment for 3 months.

PLS should be included in the differential diagnosis in cases of tumors causing epidural cord compression. Widening the differential diagnosis allows the surgeon to discuss the option of a more aggressive resection and possible stabilization of the spine in the event that PLS or a similarly aggressive tumor is found at surgery. Thus, during surgery, when the surgeon encounters unexpected pathology, he will have the option to achieve a wider, more oncological resection.

In conclusion, PLS is a rare but aggressive entity that may originate from the epidural fat. It should be included in the differential of an isolated lesion in the epidural space, even when bone involvement is not noted on MRI investigation.

Acknowledgment

We wish to thank Shifra Fraifeld, a senior medical writer and research associate in the Departments of Neurosurgery and Radiology, for her editorial contribution and writing assistance during the preparation of this manuscript.

References

[1] Ghadimi MP, Liu P, Peng T, Bolshakov S, Young ED, Torres KE, et al. Pleomorphic liposarcoma: clinical observations and molecular variables. Cancer 2011;117:5359–69.

[2] Dalal KM, Kattan MW, Antonescu CR, Brennan MF, Singer S. Subtype specific prognostic nomogram for patients with primary liposarcoma of the retroperitoneum, extremity, or trunk. Ann Surg 2006;244:381–91.

[3] Gebhard S, Coindre JM, Michels JJ, Terrier P, Bertrand G, Trassard M, et al. Pleomorphic liposarcoma: clinicopathologic, immunohistochemical, and follow-up analysis of 63 cases: a study from the French Federation of Cancer Centers Sarcoma Group. Am J Surg Pathol 2002;26:601–16.

[4] Downes KA, Goldblum JR, Montgomery EA, Fisher C. Pleomorphic liposarcoma: a clinicopathologic analysis of 19 cases. Mod Pathol 2001;14:179–84.

[5] Fiore M, Grosso F, Lo Vullo S, Pennacchioli E, Stacchiotti S, Ferrari A, et al. Myxoid/round cell and pleomorphic liposarcomas: prognostic factors and survival in a series of patients treated at a single institution. Cancer 2007;109:2522–31.

[6] Hornick JL, Bosenberg MW, Mentzel T, McMenamin ME, Oliveira AM, Fletcher CD. Pleomorphic liposarcoma: clinicopathologic analysis of 57 cases. Am J Surg Pathol 2004;28:1257–67.

[7] Barra de Moraes F, Cardoso ALP, Tristão NA, Pimenta WE, Daher S, de Souza Carneiro S, et al. Primary liposarcoma of the lumbar spine: case report. Rev Bras Ortop 2012;47:124–9.

[8] Hamlat A, Saikali S, Gueye EM, Le Strat A, Carsin-Nicol B, Brassier G. Primary liposarcoma of the thoracic spine: case report. Eur Spine J 2005;14:613–18.

[9] Lmejjati M, Loqa C, Haddi M, Hakkou M, BenAli SA. Primary liposarcoma of the lumbar spine. Joint Bone Spine 2008;75:482–5.

[10] Kirshblum SC, Burns SP, Biering-Sorensen F, Donovan W, Graves DE, Jha A, et al. International standards for neurological classification of spinal cord injury (revised 2011). J Spinal Cord Med 2011;34:535–46.

[11] Hsu W, Kosztowski TA, Zaidi HA, Dorsi M, Gokaslan ZL, Wolinsky JP. Multidisciplinary management of primary tumors of the vertebral column. Curr Treat Options Oncol 2009;10:107–25.