Eur Spine J (2005) 14: 613–618
DOI 10.1007/s00586-004-0866-2

CASE REPORT

Abderrahmane Hamlat
Stephan Saikali
Edouard-Marcel Gueye
Anne Le Strat
Beatrice Carsin-Nicol
Gilles Brassier

Primary liposarcoma of the thoracic spine: case report

Received: 26 July 2004
Revised: 15 November 2004
Accepted: 6 December 2004
Published online: 30 April 2005

A. Hamlat (✉) · E.-M. Gueye · G. Brassier
Department of Neurosurgery,
CHUR Pontchaillou,
Rue Henry Le Guilloux,
35000 Rennes, Cedex 2, France
E-mail: hamlat.abd@wanadoo.fr
Tel.: +33-02-99284278
Fax: +33-02-99284180

S. Saikali
Department of Pathology,
CHUR Pontchaillou,
Rue Henry Le Guilloux,
35000 Rennes, Cedex 2, France

A. Le Strat
Department of Physical Rehabilitation,
CHUR Pontchaillou,
Rue Henry Le Guilloux,
35000 Rennes, Cedex 2, France

B. Carsin-Nicol
Department of Radiology,
CHUR Pontchaillou,
Rue Henry Le Guilloux,
35000 Rennes, Cedex 2, France

Abstract Liposarcoma is a malignant tumor of soft tissue. The thoracic spine is an unusual location, even for metastasis, and to our knowledge, no case of primary pleomorphic liposarcoma of the vertebral body has been reported until now. A female patient presented with paraplegia. She had a previous medical history of mental depression, and complained of dorsal pain for three months following a road accident. Magnetic Resonance Imaging (MRI) revealed a collapse of T7-T8, and the diagnosis of plasmocytoma was made. She was treated with decompressive laminectomy and posterior instrumentation. Histological examination revealed a pleomorphic liposarcoma. She received a course of radiotherapy. At 13 months follow-up she developed pulmonary metastases and rib involvement. The spine is an unusual location for pleomorphic liposarcoma, even as metastasis. The differential diagnoses of this rare entity are discussed, as well as the criteria for diagnosing primary spinal liposarcoma. Although rare, our case demonstrates that liposarcoma should be considered in the differential diagnosis of spinal tumors.

Keywords Primary spinal tumor · Pleomorphic liposarcoma · Spinal cord compression · Differential diagnosis · Metastases

Introduction

Liposarcoma (LP) originates from primitive mesenchymal cells, rather than mature adipose tissue, and its diagnostic criteria have been well established [3, 4]. It is one of the more common soft tissue sarcomas [8, 9, 13, 19], and five histological types have been distinguished [4], pleomorphic liposarcoma (PLS) being the rarest of all subtypes [4, 8].

Pleomorphic liposarcoma is a high-grade sarcoma, occurring more frequently during the sixth and seventh decade of life, and both sexes are equally affected [17]. It has been described as occurring chiefly in the thigh or retroperitoneum [3, 8, 9, 17, 18, 20]. Although this tumor may be found in bone [19], primary involvement of the spine has not been reported, although a few cases have been described in the epidural space [16, 22], the foramen of T7 [2], and as metastasis [1, 13, 16, 18, 22].

Case report

This 45-year-old woman presented with an acute history of inability to move both lower limbs, and dorsal pain. In the three months prior to admission, she complained of intense dorsal pain following a road accident. The neurological and radiological examinations were normal at that time (Fig. 1A).

When she was admitted, neurological examination revealed symmetrical spastic paraplegia, and complete anesthesia with a sensory level at T4. Bilateral ankle clonus and extensor plantar responses were present. The physical examination, notably involving detailed clinical examination of the trunk and extremity soft tissues, gave no cause for concern.

The chest X-ray, biological assay and electrocardiogram were normal. X-Ray studies of the spine revealed marked collapse of T7-T8 vertebral bodies (Fig. 1B).

Magnetic Resonance Imaging (MRI) showed collapse of T7-T8, which had narrowed the spinal canal and compressed the spinal cord (Fig. 2). Unfortunately, MRI with gadolinium infusion was not performed.

The diagnosis considered was a solitary plasmocytoma. An emergency decompressive laminectomy was performed at T7-T8. The tumor pushing the pedicle cortex outwards narrowed the spinal canal, and thus compressed the spinal cord, which was also compressed by the collapsed body. The main encapsulated bulk tumor pushing the cortex outward and clearly separated from the dura mater was easily stripped off. Neither the epidural fat tissue, nor the surrounding tissues were invaded. However, the bone was invaded and different biopsies were performed. Posterior instrumentation completed the surgical procedure.

The postoperative course was uneventful, with no neurological improvement.

The ENT, gynecological and gastroenterological examinations, breast-abdominal echography, and thoraco-abdominal CT scan were normal. A bone scan showed isolated hyper-fixation at the operative site (Fig. 3).

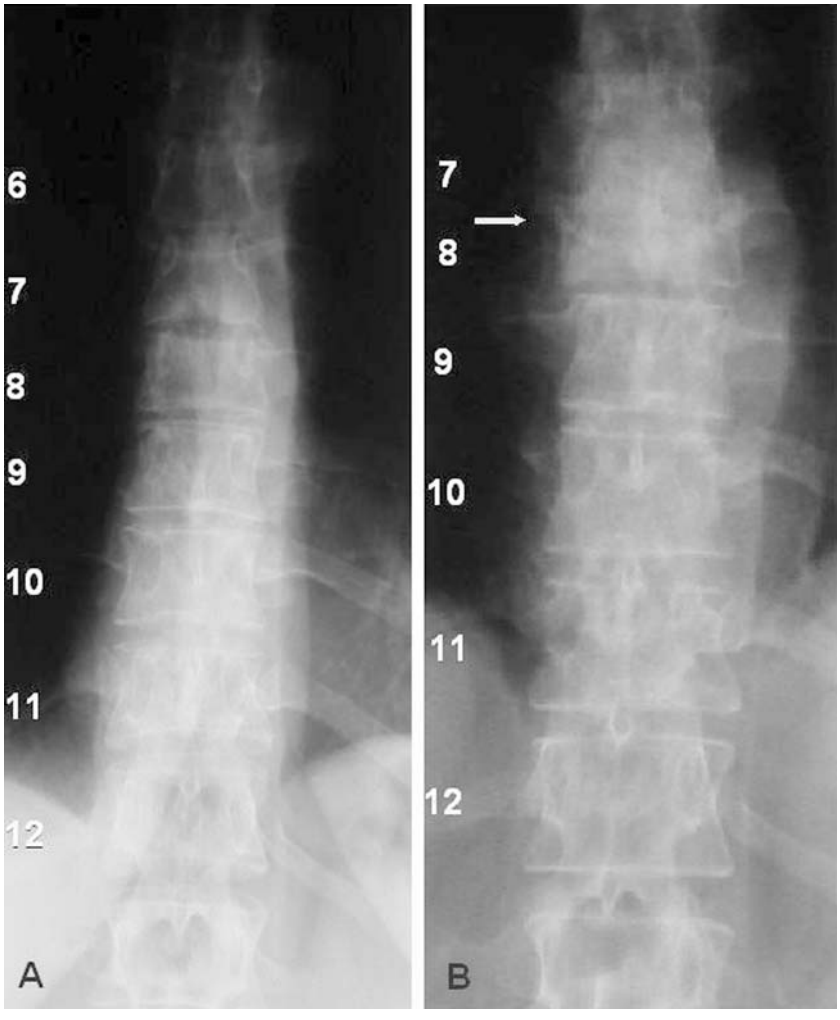

Fig. 1 Conventional radiographs (antero-posterior view) following trauma appear normal (**A**) and 3 months later reveal collapse of T8 (**B**)

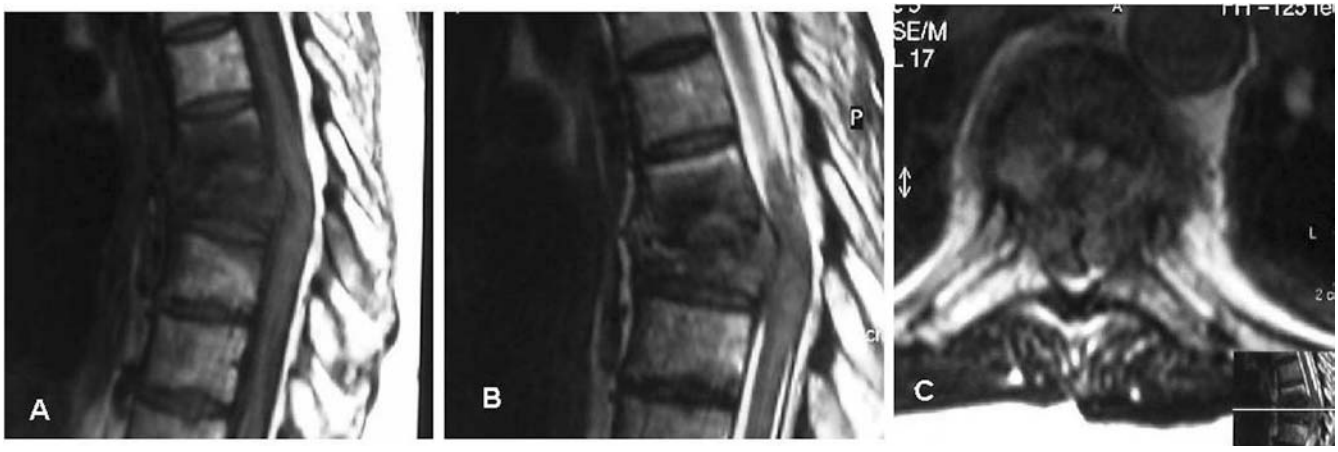

Fig. 2 **A** MRI sagittal T1-weighted imaging showing the collapsing vertebral body of T8 with marked spinal cord compression; T7 is also pathologic and both bodies show a diffuse hyposignal. The spinal cord is compressed by the bulging of the posterior part of T8. **B** MRI sagittal T2-weighted image with no saturated fat demonstrating the widening of the spinal canal. There is partial preservation of the fatty marrow in the superior plate of T7 and the vertebral bodies appear to be hypo-intense. **C** MRI axial T2-weighted image showing the bilateral narrowing of the spinal canal and the spinal cord compressed by the postero-lateral aspects of the tumor. The surrounding tissues are not invaded

The patient was referred to a rehabilitation center. Postoperative radiotherapy was performed delivering 45 Gy, although the patient was still paraplegic. At 13 months follow-up, the patient complained of chest pain and left paravertebral mass. Clinical examination revealed a left paraspinous mass, and detailed physical examination was normal. A thoracic CT scan disclosed left extension of the liposarcoma, which involved the seventh left rib, and pulmonary metastases (Fig. 4). New abdominal and pelvic CT scans were normal. She underwent pulmonary and rib irradiation. Six months after the second irradiation she is still alive with no other complaints but gradual physical deterioration.

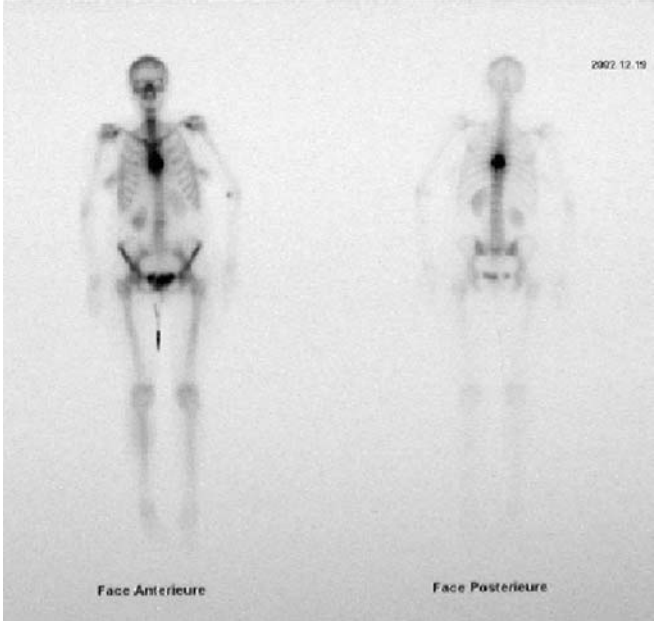

Fig. 3 Bone scan showing an isolated site of fixation at the tumoral site

Histopathological studies were performed on large surgical samples which were fixed in 10% formalin. Paraffin-embedded sections were stained with hematoxylin-eosin-safran and Gordon-Sweet silver stain.

The tumor was densely cellular, with no specific architectural structure. Cells were large, polymorphous and often poorly demarcated with abundant eosinophilic cytoplasm. The nucleus was round to ovular with a granular chromatin and small nucleolus. Anisocytosis and anisocaryosis were detected with a high mitotic index (mitotic count of 15 mitoses in 10 high-power fields) (Fig. 5).

Other tumoral fields presented adipocytic differentiation with clearly recognizable lipoblasts. These cells were well-demarcated with abundant multi-vacuolated cytoplasm, and large irregular nuclei with dense chromatin and prominent nucleoli. Cells were sometimes scalloped by the cytoplasmic vacuoles which were typically small and abundant in a few giant tumor cells (Fig. 6).

The stroma was hemorrhagic and the fibrohyalin had no mucinous component. Necrosis represented 10% of all the tumor samples examined.

Immunohistochemical work-up involved a streptavidin-biotin peroxydase technique: cells expressed vimentin. Staining for S100 protein, Desmin, Myogenin, Smooth muscle actin, Caldesmon, mdm2, cdk4, CD68, HMB45, EMA and KL1 markers was negative.

Thus the diagnosis was pleomorphic liposarcoma determined as grade III according to the FNCLCC method and grade 2 according to the NCI grading system.

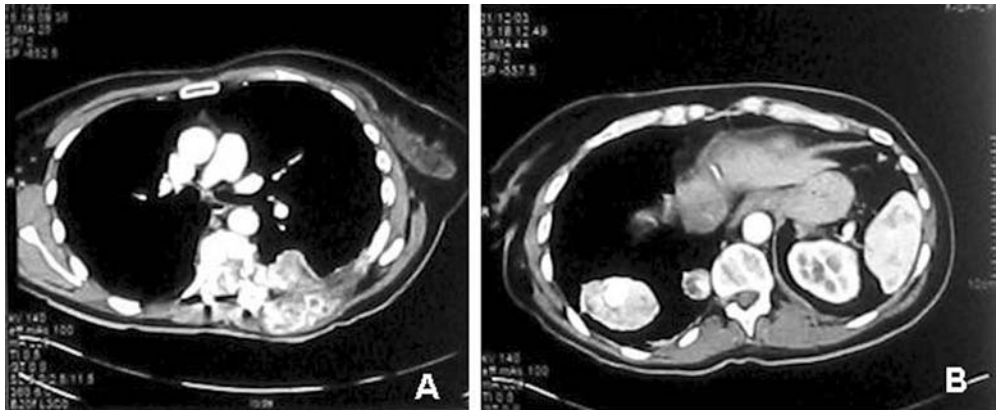

Fig. 4 **A** CT scan showing extension of the liposarcoma to the left paraspinous muscles with rib involvement, and **B** pulmonary metastases

Discussion

PLS represents the rarest form of liposarcoma [4, 9], and three main morphological aspects have been described: (1) High-grade pleomorphic MFH-like sarcoma, (2) Cellular spindle cell neoplasm and (3) PLS characterized by epithelioid morphology [3]. The difficulty in diagnosing PLS lies in its exceedingly varied histological appearance [19]. Diagnostic criteria therefore rely on the histological recognition of multi-vacuolated lipoblasts in a high-grade pleomorphic sarcoma [17]. Nonetheless, although a lipoblast is the hallmark of any type of malignant adipocytic lesion, it is not sufficient for diagnosing liposarcoma as there are a number of benign adipocytic lesions, and its absence does not rule out liposarcoma [4].

Primary tumors of the spine are relatively infrequent lesions compared with metastatic disease, multiple myeloma and lymphoma [10]. Their presentation is variable and insidious before the signs of spinal cord compression are detected, at which point the diagnosis becomes obvious. Therefore, MRI is one of the most valuable modalities for anatomic evaluation of spinal tumors. Although radiological features of spinal liposarcomas have rarely been studied [16, 17], no pathognomonic images have been described for any imaging modality elsewhere in the body [7, 12, 14, 20].

The differential diagnosis of PLS includes any high-grade pleomorphic sarcoma. These represent an important group of tumors with different histological features. Although this grouping itself is obsolete as regards information about the nature and biological behavior of a given neoplasm, it is useful for discussing differential diagnoses in this case. Routine light microscopy may be common and helpful in ultimately distinguishing them.

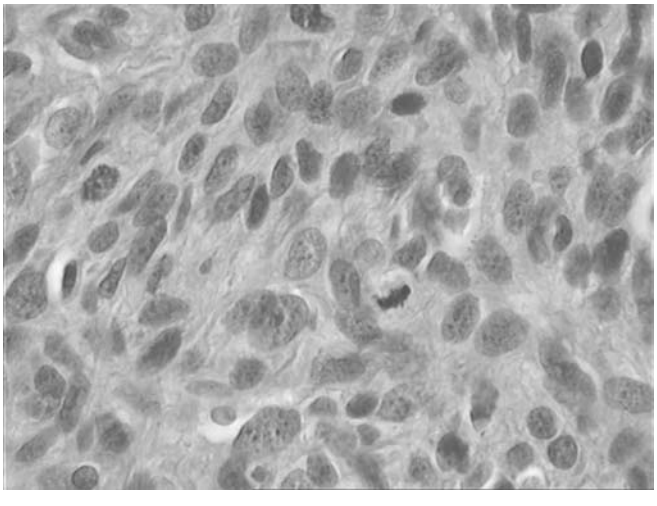

Fig. 5 Most tumoral cells are densely packed and poorly demarcated. Cytoplasm is eosinophilic with voluminous nuclei. Mitoses are numerous (HES; 1000×)

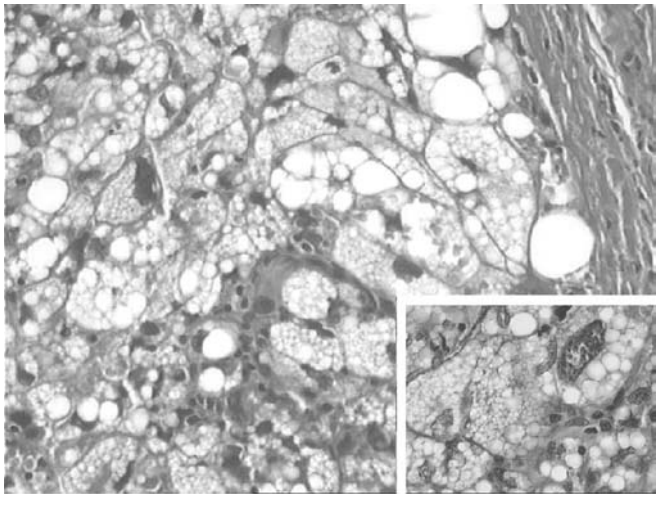

Fig. 6 Lipoblastic cells appear well-demarcated with abundant clear and multi-vacuolated cytoplasm. Nuclei are often scalloped (HES; 400×). Right inset: With high magnification, the cytoplasmic vacuoles are typically small and abundant with multiple grape-like vacuoles (HES; 1000×)

The negativity of both Smooth muscle actin and caldesmon excluded the diagnosis of leiomyosarcoma while pleomorphic cell lipoma and well-differentiated liposarcoma were ruled out owing to negativity for both mdm2 and cdk4.

Although PLS has a non-specific immunohistochemical profile [17], special stains may be required to identify the presence of intracellular fat, exclude other substances such as mucine or glycogen, and eliminate other diagnoses [8, 9]. Like other sarcomas, PLS expresses vimentin, while S100 protein, smooth muscle actin, cytokeratins AE1/AE3 and CAM5.2 have a variable expression [17]. Desmin is generally absent and either Epithelial Membrane Antigen (EMA) or CD34 is negative [17]. Morphological features and positivity for Vimentin eliminate the hypothesis of hemopathy such as "signet-ring" lymphoma or clear cell myeloma.

According to SPRINGFIELD, it is difficult to differentiate PLS from malignant fibrous histiocytoma (MFH) [20]. MFH is rarely seen in the spine, only nine cases have been described [21], and lipoblasts are not histological features of this entity [20], so this diagnosis was ruled out. Clear cell sarcoma consistently exhibits immuno-reactivity for S100 protein [5].

Giant cell tumor (GCT) of the bone rarely affects the spine, and occurs even less frequently above the sacrum [11]. Generally, GCT is an expanding osteolytic lesion which destroys bone and invades the surrounding structures [23]. GCT usually stains with CD 68 and CD 45.

Metastatic diseases represent an important heterogeneous group of entities in providing a complete differential diagnosis. In this specific case, we should exclude metastases of liposarcoma. They occur in 22–45.8% of cases, often several years after treatment of the primary tumor [1, 19]. The most frequent metastatic site is the lung [8, 18] as was the case in our patient. Distant metastases are rarely evident when the original diagnosis is made [6], and occurrence as an initial presentation of a soft tissue liposarcoma is unusual [13].

SCHWARTZ and co-workers defined criteria for differentiating true primary liposarcoma of bone from metastasis and involvement from surrounding tissue as follows: "*the tumor should be a histological proved liposarcoma which arose within bone, and metastasis has been ruled out*" [19]. Our case represents a primary vertebral LPS owing to the histological features consistent with this diagnosis and the lesion which developed within the vertebrae. The preoperative and postoperative examinations produced no evidence that the tumor was metastatic from an original tumor elsewhere. In addition, when bone invasion occurs from soft tissue liposarcoma, deep penetration is unusual [19], and most metastases are predominantly located on one side of the midline with a frequent pre-vertebral soft tissue extension [15].

Owing to the histological diagnosis, the lack of neurological improvement and the poor prognosis, only radiotherapy was additionally administered. In our patient, despite irradiation, the aggressiveness of this lesion makes long term prognosis uncertain as the median survival time for patients with PLS is less than three - years [17].

Conclusion

The spine is an unusual location for pleomorphic liposarcoma, even as metastasis. Although the radiological appearance of the tumor is somewhat nonspecific, liposarcoma should be considered in the differential diagnosis of primary spinal tumors.

Acknowledgments We are grateful to Abed El-Ghalem for his help and advice. The authors would like to thank Prof. J. M. Coindre for his help in reviewing the slices and confirming the diagnosis, and Ms T. Westcott for correcting the English.

References

1. Bogoch ER, English E, Perrin RG, Tator CH (1983) Successful surgical decompression of spinal extradural metastases of liposarcoma. Spine 8:228–234
2. Boriani S, Weinstein JN, Biagini R (1997) Spine update. Primary bone tumors of the spine. Terminology and surgical staging. Spine 22:1036–1044
3. Dei Tos AP (2000) Liposarcoma: new entities and evolving concepts. Ann Diagn Pathol 4:252–266
4. Dei Tos AP (2001) Lipomatous tumor. Curr Diagn Pathol 7:8–16
5. Eckardt JJ, Pritchard DJ, Soule EH (1983) Clear cell sarcoma. A clinicopathologic study of 27 cases. Cancer 52:1482–1488
6. Edland RW (1968) Liposarcoma. A retrospective study of fifteen cases, a review of the literature and a discussion of radiosensitivity. Am J Roentgenol 103:778–791
7. Ehara S, Rosenberg AE, Kattapuram SV (1995) Atypical lipomas, liposarcomas and other fat-containing sarcomas; CT analysis of fat element. Clin Imaging 19:50–53
8. Enzinger FM, Weiss SW (1988) In soft tissue tumors, 2nd edn, C.V. Mosby, St. Louis, pp. 346–382
9. Enzinger FM, Winslow DJ (1962) Liposarcoma. A study of 103 cases. Virchows Arch 335:367–388
10. Flemming DJ, Murphey MD, Carmichael BB, Bernard SA (2000) Primary tumors of the spine. Semin Musculoskelet Radiol 4:299–320

11. Garcia-Bravo A, Sanchez-Enriquez J, Mendez-Suarez JL, Melian-Suarez A, Miranda-Calderin G (2002) Secondary tetraplegia due to giant-cell tumors of the cervical spine. Neurochirurgie 48:527–532
12. Jelinek JS, Kransdorf MJ, Shmookler BM, Aboulafia AJ, Malawer MM (1993) Liposarcoma of the extremities: MR and CT findings in the histologic subtypes. Radiology 186:455–459
13. Kirollos R, Koutsoubelis G, Ross S, Al Sarraj S (1996) An unusual case of spinal metastasis from a liposarcoma. Eur J Surg Oncol 22:303–305
14. Kransdorf MJ, Murphey MD (1997) Lipomatous tumors. In: Kransdorf MJ, Murphey MD (eds) Imaging of soft tissue tumors: WB Saunders Company, London, pp. 57–101
15. Laredo JD, El Quessar A, Bossard P, Vuillemin-Bodaghi V (2001) Vertebral tumors and pseudotumors. Radiol Clin North Am 39:137–163
16. Ogose A, Hotta T, Inoue Y, Sakata S, Takano R, Yamamura S (2001) Myxoid liposarcoma metastatic to the epidural space without bone involvement. Report of two cases. Jpn J Clin Oncol 31:447–449
17. Oliveira AM, Nascimento AG (2001) Pleiomorphic liposarcoma. Semi Diagn Pathol 18:274–285
18. Reszel PA, Soule EH, Coventry MB (1996) Liposarcoma of the extremities and limb girdles. J Bone Joint Surg Am 48:229–244
19. Schwartz A, Shuster M, Becker SM (1970) Liposarcoma of bone. Report of a case and review of the literature. J Bone Joint Surg Am 52:171–177
20. Springfield D (1993) Liposarcoma. Clin Orthop 289:50–57
21. Sturm PF, Abramvitz J, Wagner C, Ferguson R, Walker S (1992) Malignant fibrous histiocytoma of the spine. Spine 17:975–977
22. Turanli S, Özer H, Özyürekoglu T, Cakiroglu E (2000) Liposarcoma in the epidural space. Spine 25:1733–1735
23. Verhagen WI, Bartels RH, Schaafsma HE, Rob de Jong TH (1998) A giant cell tumor of the sacrum or a soft tissue giant cell tumor? A case report. Spine 23:1609–1611