

Leiomyosarcoma of the Left Femur: A Case Report and Review of the Literature

Hind Ameer^{1*}, Motsatsa M. Gedeon¹, S. Barkich¹, N. Oumghare¹, M. Darfaoui¹, A. Alomrani¹, M. Khouchani¹

¹Radiation Oncology Department, Mohammed VI Teaching Hospital, Marrakech, Morocco

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*Corresponding author: Hind Ameer

Radiation Oncology Department, Mohammed VI Teaching Hospital, Marrakech, Morocco

Abstract

Case Report

Soft tissue sarcomas of the limbs are rare tumors with a poor prognosis. Their diagnosis is difficult and its histological confirmation. Surgery represents the main therapeutic weapon while radiotherapy allows better local control. Chemotherapy is reserved for high-grade tumors with high metastatic potential. Our work is based on a study of a rare case of metastatic leiomyosarcoma of the thigh in a 49-year-old patient.

Keywords: Soft tissue sarcomas, tumors, Surgery, therapeutic weapon, radiotherapy.

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INTRODUCTION

Leiomyosarcoma (LMS) is a rare, aggressive mesenchymal tumor with smooth muscle differentiation. LMS is one of the most common histological subtypes of soft tissue sarcoma, occurring most frequently in the extremities, retroperitoneum or uterus. SML often demonstrates aggressive tumor biology, with a higher risk of developing distant metastatic disease than most histological types of sarcoma.

OBSERVATION

The patient was a 49-year-old housewife with no specific pathological history who presented with a left thigh mass that had been developing for 6 months following a minor trauma that caused a fracture of the lower extremity of the femur on a pathological bone. An MRI of the left femur was performed, which revealed a tumoral mass in the lower third of the metaphyseal left femur measuring 97x67x87mm with T1 hypersignal, T2 hypersignal and diffusion. It infiltrates the medullary bone medially. Externally, it infiltrates the neighbouring soft tissues and extends towards the sub-quadricipital cul-de-sac. It arrives close to the vascular pericle without endoluminal extension. Respect for joint spaces and cartilage.

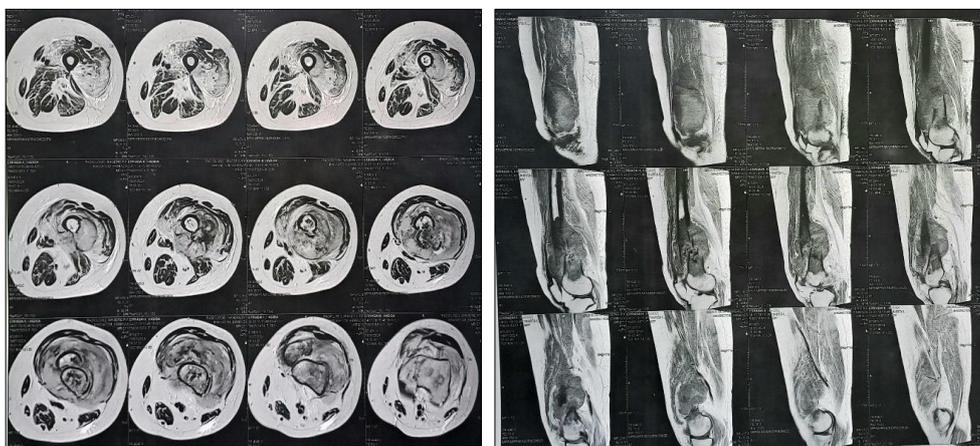


Figure 1: Microphotographs of the tumor

Then completed by a Thoraco-abdomino-pelvic CT scan, which showed the presence of pulmonary micronodules, mediastinal and axillary adenopathies of secondary origin. Right scapular lytic mass associated with stepped osteo-condensing lesions of secondary origin. Bilateral adrenal nodules of secondary origin.

The patient underwent surgery with placement of an external fixator and biopsy of the mass.

Anapath: morphological appearance of a mesenchymal proliferation with spindle cells and moderate cell density, immunohistochemical appearance in favour of a leiomyosarcoma.

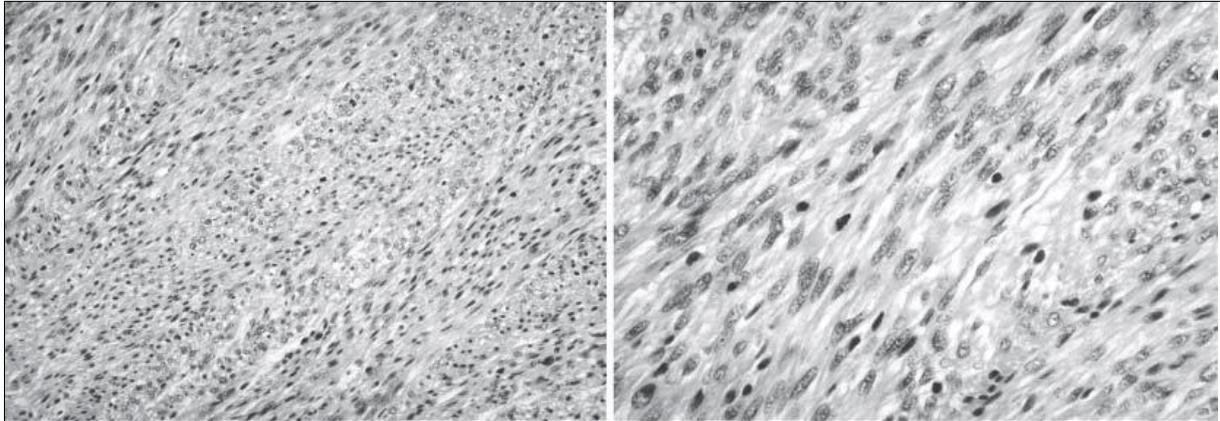


Figure 2: Spindle cell nuclei are centrally located and blunt-ended. Mitosis can be observed. A H&E 60.

She then performed a PET SCAN, which showed a locally advanced, intensely FDG avid, left femoral meta-physo-diaphyseal lytic tissue process, in favour of the neoplasm known at this level, associated with: a left scapular lytic mass, also intensely hypermetabolic. Mediastino-hair and right axillary lymph node involvement. Bilateral hypermetabolic pulmonary nodules. Bilateral adrenal and splenic hypermetabolism, and hypermetabolic muscular and bone foci in favor of metabolically active secondary dissemination.

The patient received analgesic radiotherapy at a dose of on the femoral and scapular lesions, and is currently undergoing palliative chemotherapy of the Gemcitabine-Docetaxel type, Biphosphonates for the prevention of other bone fractures and a level 2 analgesic treatment which has enabled pain to be controlled.

Palliative radiotherapy for pain relief using a 30gy, 10-fraction regimen showed clear improvement, with a reduction in pain.

Initial doses of Docetaxel- Gemcitabine chemotherapy were effective and well tolerated.

On this basis, we intend to continue the same treatment with palliative care and close monitoring.

DISCUSSION

Leiomyosarcoma is a malignant tumor that usually develops in the uterus or gastrointestinal tract. It also occurs in soft tissue and accounts for between 5 and 10% of all malignant soft tissue tumors. In contrast, primary leiomyosarcoma of the bone is rare. Berlin *et al.*,

estimated the incidence of this disease in Sweden at 0.09 cases per million inhabitants per year. In 1944, Carmody *et al.*, [4], reported for the first time a case of leiomyosarcoma arising in the mandible. Later, in 1965, Evans and Sanerkin⁶ reported the first case of extragnathic leiomyosarcoma of the bone. Since then, over 120 cases of primary bone leiomyosarcoma have been reported. However, their existence is still questioned or not accepted.

The age distribution is fairly even, from the first to the eighth decade.

There appears to be no predilection for gender. The long tubular bones of the extremities are the most common anatomical locations, with a predilection for the femur and tibia [1-8].

There are no specific symptoms of this disease, swelling or pain being the usual symptom. Typical radiographic findings are poorly circumscribed osteolytic lesions involving cortical and medullary bone [14]. Plain radiographs of this disease show aggressive-looking bone destruction with one or more of the following features: osteolysis, permeation, endosteal erosion, and fine periosteal reaction [1-16]. Our patient presented with solitary spinal cord lesions associated with soft tissue extensions. In the long bones, the metaphysis was mainly affected. As the bone lesion has no matrix, the radiographic differential diagnosis includes the following malignant lesions: malignant fibrous histiocytoma, fibrosarcoma and osteolytic osteosarcoma [12]. CT is useful for assessing cortical penetration and wide extension of the tumour into adjacent soft tissue, which is not apparent. Magnetic resonance imaging confirmed the presence of an

intramedullary lesion with soft-tissue extension. The tumor is iso-intense in relation to the muscle on T1-weighted images, and shows high signal intensity heterogeneity on T2-weighted images. MRI is useful for assessing lesion extension, particularly in the case of a primary bone leiomyosarcoma.

Histologically, bone leiomyosarcoma is a spindle cell sarcoma. The spindle-shaped tumor cells form bundles or fascicles with an intertwining pattern [1-16]. The treatment of choice is wide excision of the tumor with a tumor-free surgical margin and adequate normal tissue covering the tumor, followed by reconstruction with a stent.

As this tumour often metastasizes to the lung [2-12], careful observation is required after surgery. The efficacy of chemotherapy in improving survival has yet to be demonstrated.

CONCLUSION

Bone leiomyosarcoma can be primary or secondary; the primary variant is very rare, accounting for a very small percentage (around 0.7%) of all primary malignant bone tumors, according to the literature. Very few cases are reported in the literature, and the management of this type of tumor is controversial, particularly with regard to chemotherapy and radiotherapy.

REFERENCES

1. Antonescu, C. R., Erlandson, R. A., & Huvos, A. G. (1997). Primary leiomyosarcoma of bone: a clinicopathologic, immunohistochemical, and ultrastructural study of 33 patients and a literature review. *The American journal of surgical pathology*, 21(11), 1281-1294.
2. Berlin, Ö., Angervall, L., Kindblom, L. G., Berlin, I. C., & Stener, B. (1987). Primary leiomyosarcoma of bone: a clinical, radiographic, pathologic-anatomic, and prognostic study of 16 cases. *Skeletal radiology*, 16, 364-376.
3. BOUTSELIS, J. G., & ULLERY, J. C. (1962). Sarcoma of the uterus. *Obstetrics & Gynecology*, 20(1), 23-35.
4. Carmody, T. E., Janney, H. M., & Husesman, H. A. (1944). Leiomyosarcoma of the mandible: report of case. *The Journal of the American Dental Association*, 31(15), 1110-1113.
5. Enzinger, F. M., & Weiss, S. W. (1995). Soft tissue tumors, 3rd ed. St. Louis : *Mosby*, 491-510.
6. Evans, D. M. D., & Sanerkin, N. G. (1965). Primary leiomyosarcoma of bone. *J Pathol Bacteriol*, 90, 348-50.
7. Fornasier, V. L., & Paley, D. (1983). Leiomyosarcoma in bone: primary or secondary? A case report and review of the literature. *Skeletal radiology*, 10, 147-153.
8. Huvos, A. G. (1991). Bone tumors. Diagnosis, treatment, and prognosis, 2e éd. Philadelphie : *W. B. Saunders*, 756-62.
9. Jundt, G., Moll, C., Nidecker, A., Schilt, R., & Remagen, W. (1994). Primary leiomyosarcoma of bone: report of eight cases. *Human pathology*, 25(11), 1205-1212.
10. Khoddami, M., Bedard, Y. C., Bell, R. S., & Kandel, R. A. (1996). Primary leiomyosarcoma of bone: report of seven cases and review of the literature. *Archives of pathology & laboratory medicine*, 120(7), 671-675.
11. Marymont, J. V., & Clanton, T. O. (1990). Leiomyosarcome de l'os cal Foot Ankle, 10, 239-42.
12. Myers, J. L., Arocho, J., Bernreuter, W. Leiomyosarcome de l'os.