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Case Report

Bone Leiomyosarcoma. Report of a Case

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Abstract

We present a 46-year-old male patient with a history of hypertension and cardiac arrhythmia who 9 months before going to the center began to present pain in the left knee that was relieved with analgesics, which is why he did not go to the doctor, but over time it began to intensify and present limitation of the mobility of the knee and impossibility for the support of the left lower limb because it failed. She went to the doctor, who indicated an X-ray, showing an extensive irregular lytic lesion and a diagnosis of Giant Cell Tumor (GCT); He was referred to our Department, where we performed imaging studies and a biopsy that reported primary leiomyosarcoma of the bone. The decision is made to perform supracondylar amputation. As it is an uncommon tumour that is difficult to treat, it is important to take advantage of the scientific and technical means that we have at our disposal that help us to reach a diagnosis of certainty.

Keywords: Bone Sarcoma; Tumor; Leiomyosarcoma; Treatment

Introduction

Primary leiomyosarcoma of the bone is a tumor that, due to its rarity, must be differentiated from metastatic leiomyosarcoma, especially those of uterine origin [1]. It predominates in males and is preferentially located in metaphyseal areas of the distal femur and proximal tibia in the form of purely lytic lesions [1,2].

Primary bone leiomyosarcoma is a rare tumor, with only 50 reported cases (constituting <0.1% of primary bone tumors) [3].

It can occur at any age, but is rare before the age of 20 [2-5]. The male-to-female ratio is 2:1. The most common location is the distal femur, proximal tibia, and proximal humerus [6-8]. It occurs more rarely in the pelvis, collarbone, ribs, and jaw. In this tumor, spindle cells predominate, showing smooth muscle differentiation [9,10].

We present a case of a 46-year-old man with primary leiomyosarcoma of the proximal metaphysis of the tibia, showing the clinical and radiological picture and treatment.

Presentation of the case:

- **Reason for consultation:** Pain in the left knee.
- **History of the Current Disease:** A 46-year-old male patient from rural areas, a construction worker, with a history of hypertension and cardiac arrhythmia, who has been treated with hypotensives and antiarrhythmics, who began to have pain in his left knee that was relieved with analgesics 9 months before going to the center, which is why he did not go to the doctor. But over time it began to intensify and present limitation of the mobility of the knee and impossibility for the support of the left lower limb because it failed. He went to the doctor, who indicated X-rays, showing extensive irregular lytic lesions and a diagnosis of Giant Cell Tumor (GCT); is referred to our Service. Once hospitalized in our service, with the patient's prior consent and acceptance, the corresponding studies and necessary surgical interventions are carried out.

Physical exam

Inspection

Patient who wanders with a claudicating gait at the expense of the left lower limb with the help of crutches.

Diffuse, smooth, ill-defined, blurred-edged volume increase on the lateral aspect of the upper third of the left leg extending to the anterior tibial tuberosity without discoloration changes with smooth, shiny skin (Figure 1).

Palpation

A firm, painful tumor that does not beat or fluctuate, with a slight increase in local temperature with undefined edges but a smooth surface, of approximately 6 cm, is palpable.

Hard and painful inguinal lymphadenopathy.

Joint mobility

Limitation for active and passive flexo-extension of the left knee.

Extension (-5 degrees) Flexion (-5 degrees up to 70 degrees).

Tone and trophism

Muscle atrophy of the left quadriceps and calves.

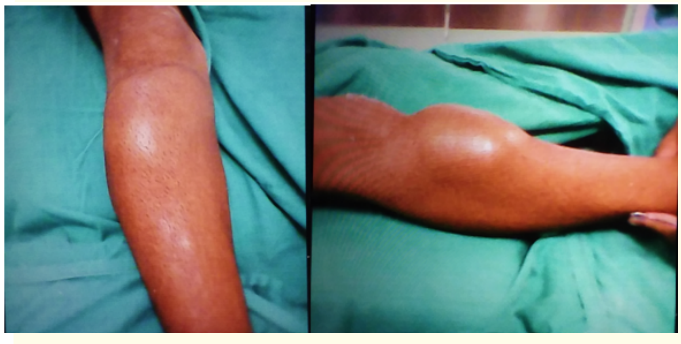

Figure 1: Left leg in anterior and lateral view.

Studies

- Hematology and Chemistry in normal parameters.
- **X-ray:** Left tibia On the outer aspect of the epiphysometaphysodiaphyseal portion of the proximal third of the left tibia, a predominantly osteolytic mixed image is visualized that ruptures the antero-external cortical with infiltration to adjacent soft tissues (Figure 2). Diagnostic impression: Osteomyelitis of the left tibia, to rule out malignant bone tumor.
- **Chest X-rays:** No pleuropulmonary abnormalities.
- **Abdominal ultrasound:** Liver and spleen of normal size and echostructure, thin-walled vesicles, kidneys without lithiasis, inguinal lymphadenopathy.
- **Computed tomography (CT):** In the sections and reconstructions performed, the lesion described above is defined with disappearance of the anterior cortical lesion, with a mass of soft tissues that occupies the intramedullary portion of the segment and expands towards the anterior aspect of the leg. Rule out: Septic process and malignant bone tumor.

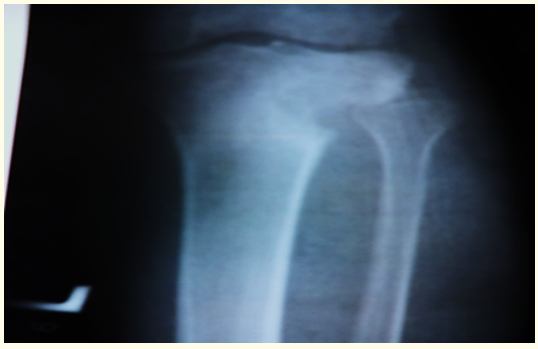

Figure 2: A predominantly osteolytic mixed image that ruptures the antero-external cortical of the proximal third of the left tibia.

With all the studies carried out, a collective discussion was carried out in the Peripheral Tumors Service of CCOI Frank País, with the Department of Radiology, Pathological Anatomy, Internal Medicine and Psychology and it was decided to perform a biopsy of the lesion (Figure 3), receiving the result of which as: Bone leiomyosarcoma

Biopsy: 16 B 367 (Frank Country Hospital).

Immunohistochemistry

- Consultation with the Pathological Anatomy Reference Center was performed, immunohistochemistry was performed and it was concluded as: Leiomyosarcoma that infiltrates soft tissues, CR 16-2130.
- Positive Vimentina.
- Alpha actin positive.
- K 167 more than 50% of tumor nuclei.

The case was consulted with the Oncology Service who recommended: supracondylar amputation of the left femur, which was performed at the level of the union of the middle third and less than ten centimeters above the upper pole of the patella, as it was a Bone Sarcoma (very rare), and with extension to soft tissues, the immediate post-surgical evolution was satisfactory.

Periodic follow-up is carried out by the Orthopaedic outpatient clinic with haematological and radiological check-ups, referral to

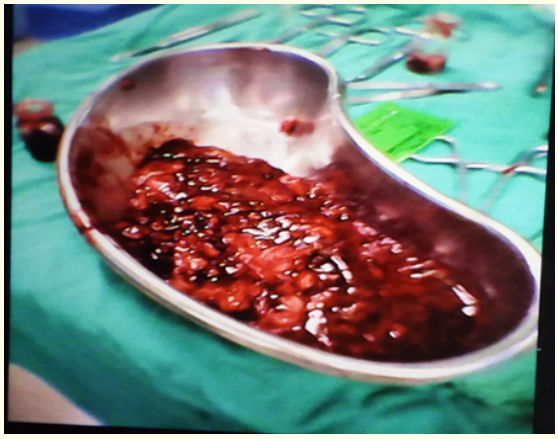

Figure 3: Material obtained during the biopsy.

the Rehabilitation consultation for evaluation, muscle strengthening, residual limb conditioning, prosthetic training and activities of daily living to achieve a prompt social reintegration. The Oncology Service did not recommend adjuvant treatment.

To date, there have been no metastatic lesions, and the patient is incorporated into the usual activities of his life using orthopedic orthoses (prostheses).

Discussion

Primary bone leiomyosarcoma is extremely rare [1,7-11]. Approximately 90 cases have been recorded since it was first described in 1965 [8-12]. Many cases considered primary bone diseases, upon further investigation, are revealed as bone metastasis or invasion from a neighboring soft tissue lesion. Most of the bone leiomyosarcomas recorded so far have been located in the metaphysis of long bones [13-16]. These lesions are thought to originate from smooth muscle cells in the wall of intraosseous blood vessels or pluripotent mesenchymal cells [17,18]. The histology is the same as that of soft tissue leiomyosarcoma. The gender distribution of these tumors has been equally or slightly predominant in males [19]. Normally, its radiographic appearance is a radiolucent lesion in the metaphysis of a long bone, although the tumor has also been described in other locations [20].

A permeative appearance is characteristic. There are no specific radiographic features that allow a leiomyosarcoma to be diagnosed solely by radiography.

Given the low incidence of these tumors and the need for a multidisciplinary team, it is recommended that treatment be carried out in a specialized center with experience in sarcoma care. At our center, treatment planning begins with a multidisciplinary review of the patient's history, all available radiographic images, and Anatomical Pathology results. Next, a treatment plan is defined based on the information provided by the pathologist, the Radiology service and the consulting oncologist.

The largest current series reviewed demonstrated no difference between wide surgical resection and resection plus radiation therapy and/or chemotherapy in the treatment of primary bone leiomyosarcoma [20-22].

Differential diagnosis is mainly in the radiological [9,20,21] and pathological aspects [20-23].

From a radiological point of view, we must rule out:

- Fibrosarcoma, Malignant Fibrous Histiocytoma and Lymphoma. Because of its aggressive pattern of bone destruction.
- Solitary metastases in older patients.
- Ewing sarcoma and Langerhans cell granuloma. It appears most often in young patients.

For histological differentiation we must take into account:

- The immunohistochemical study is conclusive [20,21].
- Metastasis of an extraskeletal leiomyosarcoma. In this case, clinical information is crucial.
- Fibrosarcoma and malignant fibrous histiocytoma. In the event that there are areas in a wheat ear or storiform pattern.
- Osteosarcoma. Because there are sometimes broad bands of hyaline collagen that resemble the osteoid.

Conclusions

Leiomyosarcoma is an aggressive sarcoma that can occur in several locations. Although there have been advances in treatment protocols, leiomyosarcoma remains one of the most difficult soft tissue sarcomas to treat. Accurate diagnosis, triage, and multidisciplinary treatment by medical professionals with expertise in these tumors are essential for a favorable outcome.

The scarcity of these tumors makes it difficult to carry out definitive studies. For this reason, it is important to take advantage of the scientific-technical means that we have at our disposal that help us to reach a diagnosis of certainty.

Accurate diagnosis, triage, and multidisciplinary treatment are essential to achieve a favorable outcome.

Better survival is achieved when radical treatment or wide resections are performed.

Conflict of Interest

The authors of this paper certify that there are no conflicts of interest in the realization and review of this topic.

An informed consent form was signed by the patient and her family member after verbal explanation by the attending physician and the Peripheral Tumours Service staff.

Bibliography

1. Gustafson P., *et al*. "Soft tissue leiomyosarcoma: a population-based epidemiologic and prognostic study of 48 patients, including cellular DNA content". *Cancer* 7 (1992).

2. Fleming ID., *et al*. "AJCC Cancer staging manual, 5ta ed". Philadelphia: Lippincott-Raven; (1997).

3. Enneking WF., *et al*. "A system for the surgical staging of musculoskeletal sarcoma". *Clinical Orthopaedics and Related Research* 153 (1980): 1.

4. Jensen ML., *et al*. "Intradermal and subcutaneous leiomyosarcoma: a clinicopathologic and immunohistochemical study". *Journal of Cutaneous Pathology* 23 (1996): 458.

5. Fletcher CDM., *et al*. "Correlation between clinicopathological features and karyotype in spindle cell sarcomas: a report of 130 cases from the CHAP study group". *American Journal of Pathology* 154 (1999): 6.

6. Rubin BP and Fletcher CDM. "Myxoid leiomyosarcoma of soft tissue, an under recognized variant". *American Journal of Surgical Pathology* 24.7 (2000): 927-936.

7. Templeton K. "Leiomyosarcoma of bone". *American Journal of Orthopaedics (Belle Mead NJ)* 34.5 (2005): 249-251.

8. Evans DM and Sanerkin NG. "Primary leiomyosarcoma of bone". *Journal of Pathology and Bacteriology* 90 (1965): 348-350.

9. Berlin O., *et al*. "Primary leiomyosarcoma of bone. A clinical, radiographic, pathologic-anatomic, and prognostic study of 16 cases". *Skeletal Radiology* 16.5 (1987): 364-376.

10. Antonescu CR., *et al*. "Primary leiomyosarcoma of bone: a clinicopathologic, immunohistochemical, and ultrastructural study of 33 patients and a literature review". *American Journal of Surgical Pathology* 21.11 (1997): 1281-1294.

11. PRIMARY BONE LEIOMYOSARCOMA: Dr. Jesús Mª Elizalde Eguinoa and Dr. María Victoria Martínez Galarreta.

12. Perez Martin A., *et al*. "Leiomyosarcoma of the humerus: a case of non-specific shoulder pain". *SEMERGEN* 32.1 (2006): 15.

13. Caron J., *et al*. "Leiomyosarcoma of the distal femur after anterior cruciate ligament reconstruction". *Clinical Orthopaedics and Related Research* 412 (2004): 24.

14. Eady J., *et al*. "Primary leiomyosarcoma of bone. A case report and review of the literature". *Journal of Bone and Joint Surgery* 69.2 (1987): 4.

15. Evans D and Sanerkin N. "Primary leiomyosarcoma of bone". *Journal of Pathology* 90 (1965): 348-350.

16. Goto T., *et al*. "Primary leiomyosarcoma of the femur". *Journal of Orthopaedic Science* 7.2 (2002): 12.

17. Jundt G., *et al*. "Primary leiomyosarcoma of bone: report of eight cases". *Human Pathology* 25.11 (1994): 14.

18. Khoddami M., *et al*. "Primary leiomyosarcoma of bone: report of seven cases and review of the literature". *Archives of Pathology and Laboratory Medicine* 120.7 (1999): 671-675.

19. Lopez-Barea F., *et al*. "Primary epithelioid leiomyosarcoma of bone. Case report and literature review". *Virchows Archiv* 434.4 (1999): 367-371.

20. Myers J., *et al*. "Leiomyosarcoma of bone. A clinicopathologic, immunohistochemical, and ultrastructural study of five cases". *Cancer* 67 (1991): 1051-1056.

21. Young M and Freemont A. "Primary leiomyosarcoma of bone". *Histopathology* 19 (1999): 257-262.

22. Carrasco CAL., *et al*. "MRI and PET-CT findings in metastatic primary bone leiomyosarcoma". *Med Sur* 18.1 (2011): 12.

23. Penna W., *et al*. "Primary leiomyosarcoma of bone". *Revista Brasileira de Ortopedia* 34.3 (1999): 11.