Primary Leiomyosarcoma of the Hip Bone: A Case Report and Literature Review

Francisco De Assis Serra Baima Filho[1]

Abstract

Introduction: Leiomyosarcoma is a rare smooth muscle mesenchymal neoplasm. The primary bone form is the rarest subtype. It mainly affects middle aged and female people. The most common complaint is the presence of a mass accompanied by pain. Diagnostic imaging and biopsy are required. The gold standard treatment is resection with oncologic margin.

Case Report: A 20-year-old female had a history of tumor mass and left hip pain beginning in September 2018.The patient showed the radiographic examination, osteolytic lesion of the left hip, and computed tomography scan of the abdomen with a solid expansive process measuring 15.8 cm left hip destruction and absence of organ damage. Biopsy and immunohistochemical result of leiomyosarcoma. Neoadjuvant chemotherapy was not effective. Thus, we opted for tumor resection with internal hemipelvectomy Type I.

Conclusion: Primary bone leiomyosarcoma is a rare malignant neoplasm which only shows tumoral resection with oncological margins as a curative treatment, thus the need for early diagnosis to allow adequate resection and lower patient morbidity.

Keywords: Leiomyosarcoma, case reports, rare diseases, literature review.

Introduction

Leiomyosarcoma is a rare mesenchymal malignant neoplasm of smooth muscle with an incidence of 0.7/100,000 [1]. It often occurs in the uterus, retroperitoneum, subcutaneous tissue, gastrointestinal tract, and in medium and large veins [2]. Leiomyosarcoma is divided into three groups based on the clinic and prognostic: Gastrointestinal and uterine, cutaneous, and somatic. In the somatic group, it occurs most often in the retroperitoneum, soft parts of the extremities, veins, and rarely in bones [3].

Primary bone leiomyosarcoma (PBL) was first described by Evans and Sanerkinin 1965 [4] and most commonly involved bones close to the knee [5, 6]. It tends to affect middle-aged people and is slightly more common in females than in males [7].

The most common complaint is the presence of a mass accompanied by moderate pain. Other symptoms are related to tumor location [7]. Diagnosis requires imaging and biopsy.

The most common radiographic sign is a solitary osteolytic lesion with cortical destruction and no bone production [8]. The computed tomography (CT) scan usually shows a well-defined osteolytic mass [2]. On Magnetic resonance imaging (MRI),Leiomyosarcoma appear iso-to high-intensity onT1 and low-to high-intensity on T2, and with homogeneous or heterogeneous enhancement. However, these findings are nonspecific and radiological features have many differential diagnosis including lymphoma, plasmacytoma, other small round cell tumors including sarcoma, metastatic tumors, and even benign tumors including chordoma[2].

A histopathological examination typically shows smooth muscle differentiation and smooth muscle actin positive spindle cells and other muscle tumor cell markers such as desmin and h-caldesmon, in addition to the absence of osteoid or chondroid matrix [8].

The main differential diagnoses are dermatofibrosarcoma, bone fibrosarcoma, myxofibrosarcoma, lymphoma, myeloma, and metastatic carcinoma [2, 7].

The gold standard treatment is oncologic margin resection. Chemotherapy and radiotherapy do not yet have a definite role and further studies are needed [3, 7, 8].

This paper reports a case of a 20-year-old female patient with primary leiomyosarcoma of the hip bone, describing the patient's diagnosis, treatment, and evolution, as well as a literature review.

Case Report

Patient MOS, female, 20-year-old, had a history of tumor mass and left hip pain beginning in September 2018. The first visit was in January 2019. On that day, the patient arrived on a stretcher as she felt pain from strong intensity to the left hip mobilization. She arrived with a biopsy and immunohistochemical result of leiomyosarcoma (positive for 1A4 and negative for CD34, TLE, and S100).

The patient showed the radiographic examination, osteolytic lesion of the left hip, and CT scan of the abdomen with a solid expansive process measuring 15.8 cm left hip destruction and absence of organ damage. Chest CT showed no signs of metastasis.

The patient underwent two sessions of neoadjuvant chemotherapy and it was noticed that the tumor increased in volume. A new total abdomen CT showing increased

[1]Department of Orthopedic Oncology, Maranhão Institute of Oncology, Aldenora Bello, São Luís, Brazil.

Address of Correspondence
Dr. Francisco De Assis Serra Baima Filho,
R. Seroa da Mota, 23 - Apeadouro, São Luís - MA, 65031-630, Brazil.
E-mail: assisbaima@gmail.com

Dr. Francisco De Assis Serra Baima Filho

© 2020 by Journal of Bone and Soft Tissue Tumors | Available on www.jbstjournal.com | doi:10.13107/jbst.2019.v05i03.016
This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (http://creativecommons.org/licenses/by-nc/3.0) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

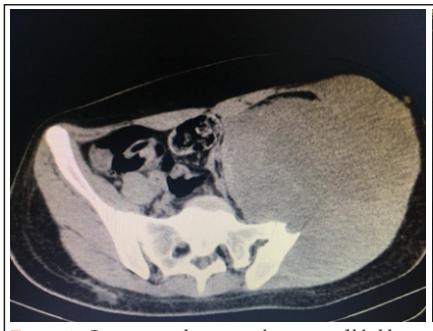

Figure 1: Computerized tomography image of bladder in axial view showing a large tumoral mass.

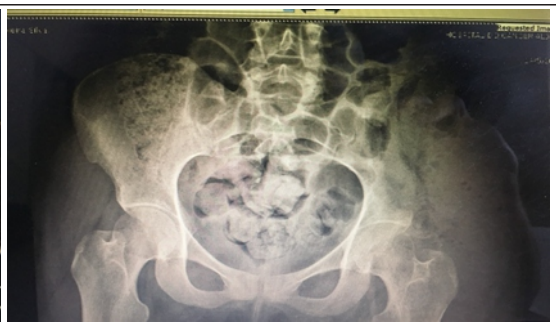

Figure 2: Radiography of bladder in immediate postoperative anteroposterior view.

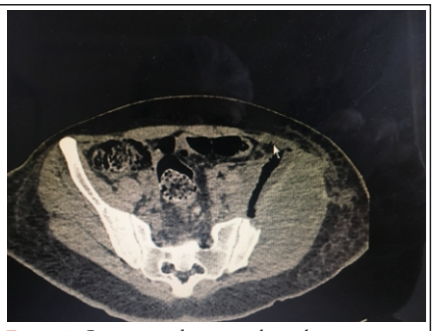

Figure 3: Computerized tomography with post-operative axial cut in the bladder.

tumor mass (18 cm × 16.6 cm) (Fig. 1). Thus, we opted for tumor resection with internal hemipelvectomy Type I (Fig. 2 and 3).

The tumor was whitish and had an irregular, elastic-firm outer surface, measuring 19 cm × 16 cm × 9 cm, and weighing 1.880 kg (Fig. 4). The result of the histology was moderately undifferentiated Grade I leiomyosarcoma, without necrosis, focally compromised surgical margins.

The patient evolved well and is currently being programmed for radiotherapy due to margin contamination. The patient was instructed to sit down and perform exercises to gain range of motion in the left knee (with the help of the physiotherapy team) as tolerated from the fourth post-operative day (when discharged from the intensive care unit).

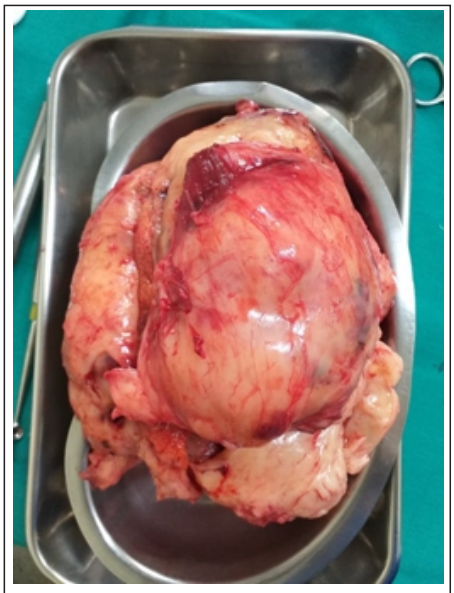

Figure 4: Presentation of the large tumor volume.

Discussion

PBL is a rare malignant tumor originating from smooth muscle cells. It is more common in middle-aged people and a little more frequent in females. According to the study by Mori et al., the most affected bone is the femur (50%), tibia (18.7%), and pelvic bones (14.6%) [3]. The case in the article is a 20-year-old patient with a tumor located in the pelvic bone (left hip).

With the literature review, it is clear that the diagnosis requires imaging (especially MRI) associated with histopathological and immunohistochemical examination. The main differential diagnoses are dermatofibrosarcoma, bone fibrosarcoma, myxofibrosarcoma, lymphoma, myeloma, and metastatic carcinoma [2,7].

In the study by Rekhi et al., the most common symptom was pain and, less frequently, edema and pathological fracture. The duration of symptoms before diagnosis ranged from3 months to 3 years [4]. In the present report, the symptoms were the presence of tumor and pain and 3 months passed before there was a diagnosis.

The gold standard treatment is surgical resection. Chemotherapy (neoadjuvant and adjuvant) and radiotherapy do not have a definite role yet and need further study. However, as the study by Potsi et al. cites, if resection has contaminated margins, radiotherapy is indicated [5]. As we have seen in the reported case, the patient underwent two sessions of neoadjuvant chemotherapy, but the tumor increased in volume.

Leiomyosarcoma has a poor prognosis with a 35% survival rate. The most important prognostic factors associated with reduced survival rates are the presence of metastasis at diagnosis, tumor grade, age >40 years, tumor size >8 cm, presence of pathological fracture, amputation, contaminated resection margins, and poor response to preoperative chemotherapy. Therefore, radical surgery with negative margins represents the only curative option [3,4,7,8].

Local recurrence is relatively uncommon, while metastasis may occur in the early stages of the disease. The most frequent metastasis site is the lungs. Metastasis may occur to other bones, especially the axial skeleton. Less frequent to the liver, adrenal gland, kidney, lymph nodes, brain and skin [2,8].

Once metastasis is diagnosed and chemotherapy represents the main therapeutic option [8].

Conclusion

PBLis a rare malignant neoplasm which only shows tumoral resection with oncological margins as a curative treatment, thus the need for early diagnosis to allow adequate resection and lower patient morbidity. More studies are needed regarding chemotherapy and radiotherapy.

References

1. Gordon RW, Tirumani SH, Kurra V, Shinagare AB, Jagannathan JP, Hornick JL, et al. MRI, MDCT features, and clinical outcome of extremity leiomyosarcomas: Experience in 47 patients. Skeletal Radiol2014;43:615-22.

2. Tahara K, Yamashita K, Hiwatashi A, Togao O, Kikuchi K, Endo M, et al. MR imaging findings of a leiomyosarcoma of the thoracic spine: A case report. Clin Neuroradiol 2016;26:229-33.

3. Mori T, Nakayama R, Endo M, Hiraga H, Tomita M, Fukase N, et al. Forty-eight cases of leiomyosarcoma of bone in Japan: A multicenter study from the Japanese musculoskeletal oncology group. J Surg Oncol 2016;114:495-500.

4. Rekhi B, Kaur A, Puri A, Desai S, Jambhekar NA. Primary leiomyosarcoma of bone--a clinicopathologic study of 8 uncommon cases with immunohistochemical analysis and clinical outcomes. Ann Diagn

Pathol2011;15:147-56.

5. Potsi M, Stavrinou P, Patsinakidis N, Hatzibougias D, Foroglou N, Karayanopoulou G, et al. Primary osseous leiomyosarcoma of the spine: A rare entity--case report and review of the literature. J Neurol Surg A Cent Eur Neurosurg2012;73:238-42.

6. Yang Y, Ma L, Li L, Liu H. Primary leiomyosarcoma of the spine: A case report and literature review. Medicine (Baltimore) 2017;96:e6227.

7. Zumárraga JP, Arouca MM, Baptista AM, Caiero MT, Rubio DE, Camargo OP. Primary leiomyosarcoma of bone: Clinicopathologic and prognostic factors analysis in a single institution. Acta Ortopédica Bras 2019;27:152-5.

8. Recine F, Bongiovanni A, Casadei R, Pieri F, Riva N, De Vita A, et al. Primary leiomyosarcoma of the bone: A case report and a review of the literature. Medicine (Baltimore) 2017;96:e8545.

Conflict of Interest: NIL
Source of Support: NIL

How to Cite this Article

De Assis Serra Baima Filho F | Primary Leiomyosarcoma of the Hip Bone: A Case Report and Literature Review | Journal of Bone and Soft Tissue Tumors | Sep-Dec 2019;5(3): 5-7.