Radiology Case Reports 16 (2021) 3016–3019

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Primary leiomyosarcoma of the distal tibia: A case report

Sachin Dheer, MD[a,*], *Pamela Gekas, DPM*[b], *Faith Schick, DPM*[b], *Homyar Karanjia, DPM*[b], *Nicholas Taweel, DPM*[b]

[a] *Department of Radiology, Thomas Jefferson University, 111 South 10th Street, Philadelphia, PA 19103*
[b] *Department of Podiatry, Rothman Institute, 925 Chestnut Street, Philadelphia, PA 19107*

ARTICLE INFO

Article history:
Received 22 June 2021
Revised 7 July 2021
Accepted 10 July 2021

ABSTRACT

The authors present an unusual case of a leiomyosarcoma of the distal tibia. Leiomyosarcoma tumors typically originate from smooth muscle tissue. It is rare for it to derive from bone and even rarer to be found in a bone of the lower limb. Given this extreme rarity in addition to nonspecific findings on plain film radiographs and magnetic resonance imaging (MRI), biopsy was needed in this case. It was only through immunochemistry staining that a definitive diagnosis was made. As such, this case is an illustrative example of an aggressive, though rare, primary lesion of the bone which should be considered in the differential diagnosis of a lytic intramedullary lesion. This case also highlights the need for careful evaluation of imaging features suggesting a potentially aggressive lesion requiring appropriate work up in a timely fashion.

Introduction

This case illustrates an uncommon, clinically significant, and initially incorrectly diagnosed, pathology in a middle-aged adult male presenting with non-specific, generalized ankle pain in the absence of trauma/injury. Initial radiographic evaluation, performed at the time of presentation, demonstrated a focal bone lesion; which, during subsequent evaluation, was found to represent an aggressive, malignant etiology requiring surgical excision.

Case report

A 47-year-old male, with no past medical history of malignancy or trauma presented with left ankle pain; specifically, pain at night at the level of the distal aspect of his left tibia. Radiographs (Figs. 1A and B) demonstrated an oval-shaped lytic area within the distal tibia with cortical thinning and erosion posteriorly, suggesting an aggressive lesion. Subsequent contrast-enhanced MRI showed evidence of a heterogeneously enhancing, intramedullary mass of the distal tib-

* Corresponding author.
E-mail address: sachdheer@yahoo.com (S. Dheer).
https://doi.org/10.1016/j.radcr.2021.07.020

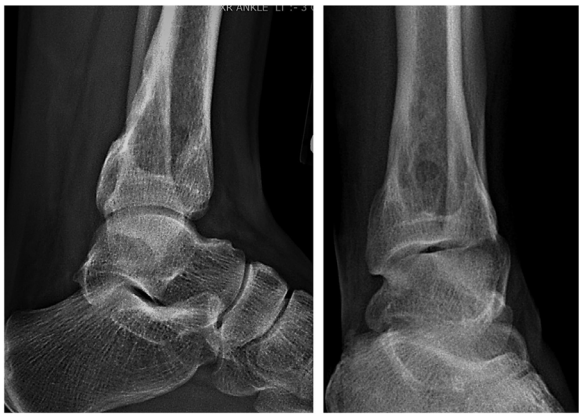

Fig. 1 – Lateral (a) and external oblique (b) radiographs of the left ankle demonstrate a poorly demarcated, lytic lesion centered within the medullary cavity of the distal tibia. There is cortical irregularity and thinning posteriorly, suggesting an aggressive etiology.

ial metadiaphysis measuring, in aggregate, 44 × 29 × 26 mm. MRI also revealed surrounding medullary cavity edema and enhancement with significant cortical erosion posteriorly. The patient was referred to a local tertiary care center for surgical biopsy which also necessitated open reduction internal fixation of the distal tibia. He had an unremarkable postoperative course, with full healing of his incision.

Final histologic analysis (Fig. 3) demonstrated spindled and pleomorphic epithelial cells with irregular hyperchromatic nuclei and amphophilic cytoplasm and immunohistochemical stains showed multifocal positivity for pan-keratin and CAM 5.2, along with diffuse nuclear positivity for CK-OSCAR and a positive vimentin stain with spindle cells. The tibial tumor was initially suspicious for a metastatic sarcomatous carcinoma due to some of the immunochemistry findings. With the presence of pleomorphic atypical cells, leiomyosarcoma could not be excluded. A subsequent sentinel node biopsy of the inguinal region and PET scan were both negative for metastasis.

Following evaluation by a dedicated bone pathologist and a national tumor board, the final pathologic diagnosis was primary leiomyosarcoma isolated to the left distal tibia. Because the lesion was a high grade, deep, non-metastatic tumor measuring less than 5 cm in size, it was graded as a Stage 2A as per the AJCC system. The patient underwent a below the knee amputation, felt to be necessary to obtain a sufficient wide margin excision of the tumor following the initial biopsy. He was subsequently treated with chemotherapy. A prosthesis for his lower extremity was fashioned post operatively. He completed his full course of chemotherapy and at two-year follow-up has had no signs of recurrence.

Discussion

Leiomyosarcoma is an extremely rare smooth muscle tumor most commonly associated with uterine, gastrointestinal, and/or retroperitoneal tissues. Primary onset in bone accounts for less than 0.1% of all of these tumors. These bone tumors are very aggressive in their growth and can appear radiographically with associated ill-defined cortical borders, fine trabeculations, irregular cortical destruction, and a “moth-eaten” appearance. Early symptoms can present with nonspecific pain and swelling, which can be easily misinterpreted for other conditions. For this reason, a diagnosis can be very difficult to obtain at a time when early detection is of paramount importance [1,2].

A study of 31 patients diagnosed with primary leiomyosarcoma in bone showed that 14 (45%) occurred in the distal femur, eight (26%) in the proximal tibia, and two (6%) in the proximal humerus. Only one patient was found to have leiomyosarcoma in the distal tibia [3].

Primary leiomyosarcoma of bone is histologically similar to leiomyosarcoma of soft tissue, both characterized by pleomorphic atypical spindle cells. Macroscopically the tumor appears as a fleshy, gray-white mass with areas of necrosis. It is very rare for this type of sarcoma to develop as a primary tumor in bone; but when it does, it primarily affects older individuals. Primary bone leiomyosarcoma has a much better survival rate than those metastasized to bone at the time of diagnosis. [4]

Radiographic findings for leiomyosarcoma typically show a permeative “moth-eaten”, appearance of bone with osteolysis and endosteal erosion, typically in long bones. Periosteal reaction and intralesional calcifications are typically absent. Given the osteolytic and erosive changes, pathological fractures occur in approximately 20% of cases. MRI is nonspecific for leiomyosarcoma because the findings are similar to many other types of osseous neoplasms. On MRI, the tumor has a hypointense T1 signal and an increased T2 signal. Definitive diagnosis is achieved via pathology and immunochemistry staining. Immunochemistry of smooth muscle actin (SMA), desmin, or vimentin, is an objective marker of smooth muscle origin [5-8].

The most commonly used scale for staging leiomyosarcoma is the American Joint Committee on Cancer system (AJCC). (Table 1) This scale uses an alpha numeric designation numbered from 1-4 with lettered subgroups grading tumors with increasing severity. Stage 1 or 2A, or low grade, have a 100% survival rate. Stages 2B, or intermediate grade, have a five year survival rate of 60%. The survival rate for Stage 3 or greater, which are classified as high grade, is no more than 2 years. Most primary leiomyosarcoma of bone are high grade in nature [8]. Antonescu found among 33 cases of primary leiomyosarcoma that 66% were classified as high grade, 21% were intermediate, and 12% were low grade. In the same study, they found a 51% chance of metastasis and a 60% survival rate at five years for patients with intermediate to high grade tumors. [9] Treatment has historically been resection of the tumor with wide margins. The tumor’s sensitivity to radiation is limited. As such, pre- and post-operative chemotherapy is advised [8]

The work-up of this case clearly illustrates the need for radiologists to escalate the levels of diagnostic evaluation in a timely fashion when findings are nonspecific, but indicative of an aggressive etiology. Only the lateral radiograph revealed a potentially aggressive, malignant etiology and the subsequent MRI certainly increased the sensitivity for more fully char-

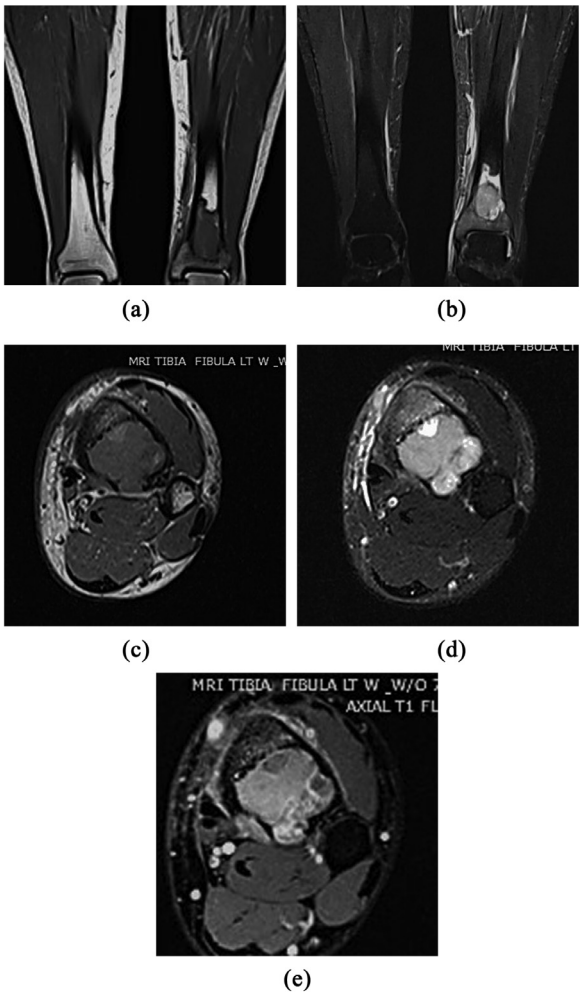

Fig. 2 – Contrast enhanced MRI of the left tibia fibula. Coronal T1 weighted, large field of view of both lower extremities (a) demonstrates a cellular, intermediate signal lesion in the medullary cavity of the distal left tibia; corresponding T2 weighted, large field of view (b) image demonstrates surrounding marrow edema, suggestive of an infiltrative, aggressive lesion. Dedicated, small field of view, pre-contrast, axial T1 (c) and T2 weighted, fat saturated (d) images of the left lower extremity reveal pronounced endosteal scalloping/erosion and frank cortical breakthrough posteriorly, indicative of an aggressive neoplasm. A post-contrast, axial, T1 weighted, fat saturated image demonstrates non-specific, heterogeneous enhancement, consistent with neoplasm.

acterizing the aggressive imaging features, including marrow edema and extensive cortical erosion/breakthough.

This case illustrates the need for the radiologist to carefully evaluate and appreciate nonspecific lesions with potentially aggressive imaging features. The case also provides experiential evidence with regard to an extremely rare, though clinically important intra-osseous tumor the lower extremity: primary leiomyosarcoma of bone.

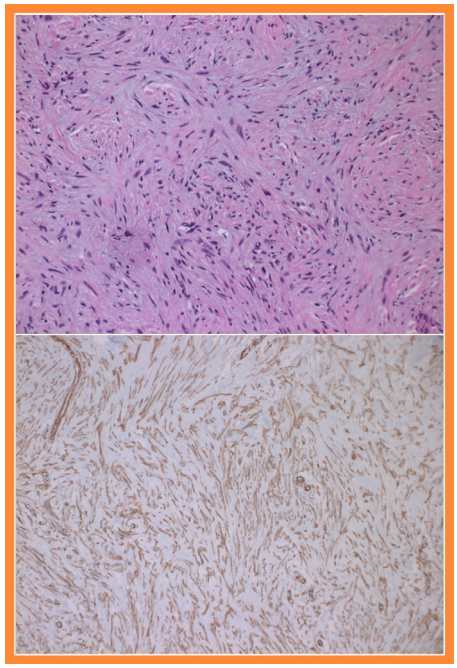

Fig. 3 – Top panel demonstrates spindle-shaped and pleomorphic epithelial cells with irregular hyperchromatic nuclei and amphophilic cytoplasm. The immunohistochemical stains, bottom panel, show diffuse nuclear positivity for CK-OSCAR and a positive vimentin stain with spindle cells. The findings are consistent with sarcomatous neoplasm with prominent leiomyomatous features.

Competing interest

None of the authors have any conflict of interest with regard to the publication of this case report.

Case reports

We, the authors, have made multiple attempts to obtain publication consent from the patient. However, our efforts have been in unsuccessful. We confirm that there is no personal identifying information contained within the entirety of the submitted manuscript. Furthermore, as this is a retrospective case report, no institutional review board approval was necessary. Therefore, we request that this letter be accepted in lieu of a completed patient consent form.

REFERENCES

[1] Sarcohelp.org Leiomyosarcoma Cancer. 2015. http://sarcomahelp.org/leiomyosarcoma.html#tpm1_1
[2] Mascard E, Gaspar N, Brugieres L, Glorion C, Pannier S, Gomez-Brouchet A. Malignant Tumours of the foot and ankle. Effort Open Reviews 2017;5(2):261–71.

[3] Brewer P, Sumathi V, Grimer RJ, Carter SR, Tillman RM, Abudu A, et al. Primary Leiomyosarcoma of Bone: Analysis of Prognosis. Sarcoma. 2012:4 Volpages. doi:10.1155/2012/636849.
[4] Sundaram M, Akduman I, White LM, McDonald DJ, Kandel R, Janney C. Primary Leiomyosarcoma of Bone. AJR 1999;172:771–6.
[5] Delsmann B, Pfahler M, Nerlich A, Refior HJ. Primary Leiomyosarcoma Affecting the Ankle Joint. Foot Ankle Int 1996;17(7):420–4 Jul.
[6] Kitay A, Rybak L, Desai P, Villalobos CE, Wittig JC. Primary Leiomyosarcoma of the Proximal Tibia: Case Report and Review of the Literature. Bulletin of the NYU Hospital for Joint Diseases 2008;66(1):49–53.
[7] Singh D, Kumar R, Kamau GG, Mmopelwa T, Mhembelo DK. Primary Leiomyosarcoma of the first metatarsal bone: A case report. SA Orthop J. Autumn 2017;1(16):45–8.
[8] Hanafy Marwan. Primary Leiomyosarcoma of the distal fibula: A case report and review of the literature. Ortho Rev (Pavia) 2017;9(4):7236 Dec 14.
[9] Atonescu CR, Eriandson RA, Huvos AG. Primary leiomyosarcoma of bone: a clinicopathologic, immunohistochemical, and ultastructural study of 33 patients and a literature review. Am J Surg Pathol 1997(11):1281–94 Nov; 21.