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Case Report

Primary leiomyosarcoma of the distal tibia: A case report

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\section*{A B S T R A C T}
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 immunohistochemistry 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.

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\section*{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.

\section*{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-
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 permissive “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 immunohistochemistry staining. Immunohistochemistry 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.

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.

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