International Journal of Advanced Research and Review

www.ijarr.in

PRIMARY LEIOMYOSARCOMA OF THE SACROILIAC JOINT: A CASE REPORT

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Abstract

A rare case of low-grade primary leiomyosarcoma of the sacrum is described in a young woman who was suffering from pain in the left sacroiliac region. An expansile lytic sacroiliac mass was observed on x-ray and conventional tomography. Magnetic resonance imaging revealed avidGd-DTPA enhancement in the entire mass suggesting its hypervascularity. Histopathology and immunohistochemical results of biopsy of the lesion confirmed the diagnosis. We report on the features of this rare tumor entity through imaging and diagnostic methods.

Keywords: Bone neoplasms, Sacrum, Leiomyosarcoma, MR imaging, Histopathology

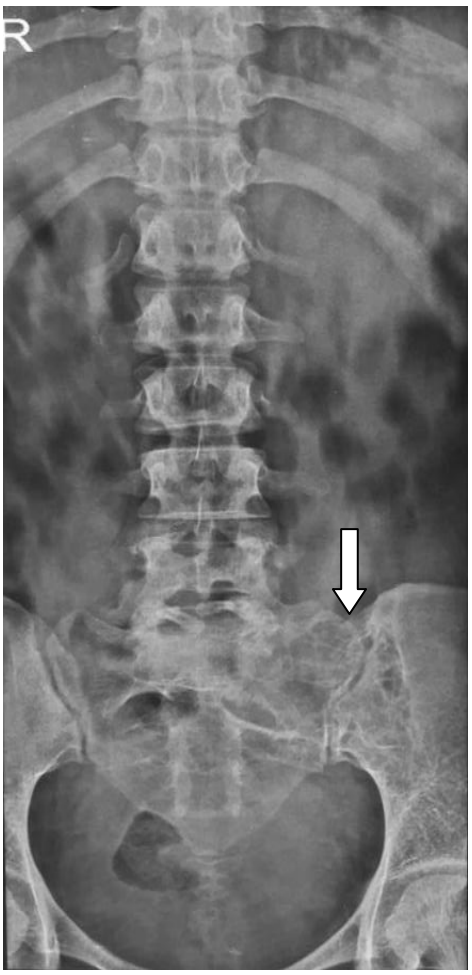
INTRODUCTION

Primary leiomyosarcoma of the bone is a rare primary osseous tumor that has not been extensively reported in literature. The diagnosis of leiomyosarcoma is primarily based on the histopathologic and immunohistochemical characteristics. PLB is a rare soft tissue sarcoma, accounting for <0.7% of all primary malignant bone tumors. It is most commonly seen in the long bones where as primary vertebral leiomyosarcoma is extremely rare. Here we report the case of young female with low grade primary leiomyosarcoma of the sacrum who was evaluated with several imagingmodalities like conventional radiography, CT and MRI.

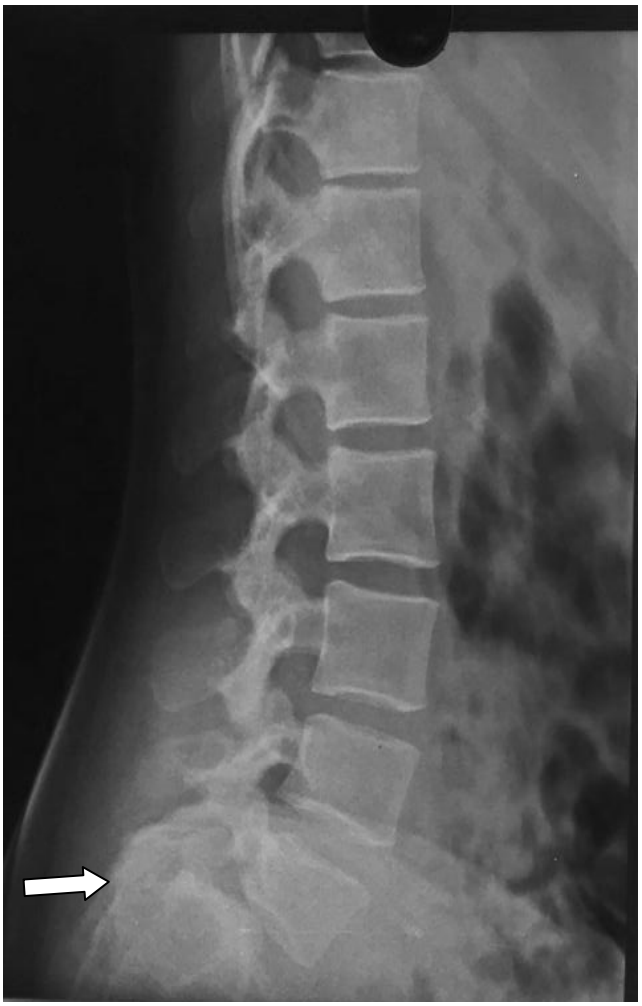
CASE REPORT

A 27-year-old female patient who presented with history of lower back ache radiating to left lower limb, numbness and walking difficulty since 1.5 years, with worsening of symptoms for the past one week. She had no history of spinal surgery or trauma in the past. No history of any comorbities.

Plain radiographs of the lumbo-sacral spine taken in AP and lateral views revealed the presence of a radiolucent lesion involving the iliac and sacral articular surface of left sacroiliac joint with internal septation and ill-defined cortical margins.



(Figure 1) Frontal radiograph of the LS spine demonstrating a lytic lucent lesion with few internal septations in the left hemisacrum extending across the sacroiliac joint.

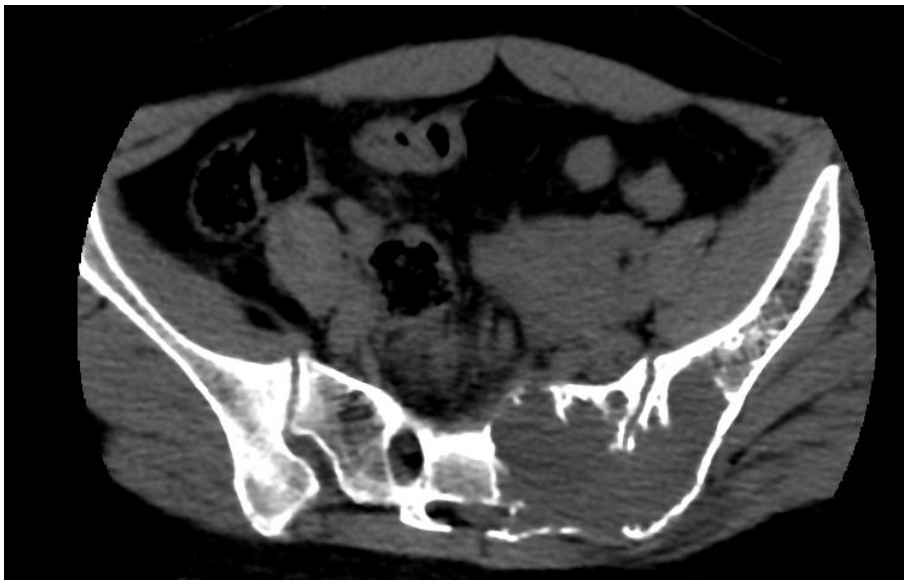


(Figure 2) Lateral radiograph of the lumbar and upper sacral vertebrae: subtle radiolucency noted in the S1 vertebral body with obscuration of the S1-S2 intervertebral disc. Focal cortical blurring and defect noted in the posterior cortex of S1 vertebrae with focal radio-opacity within the spinal canal. Anterolisthesis of L5 over S1 vertebrae also noted.

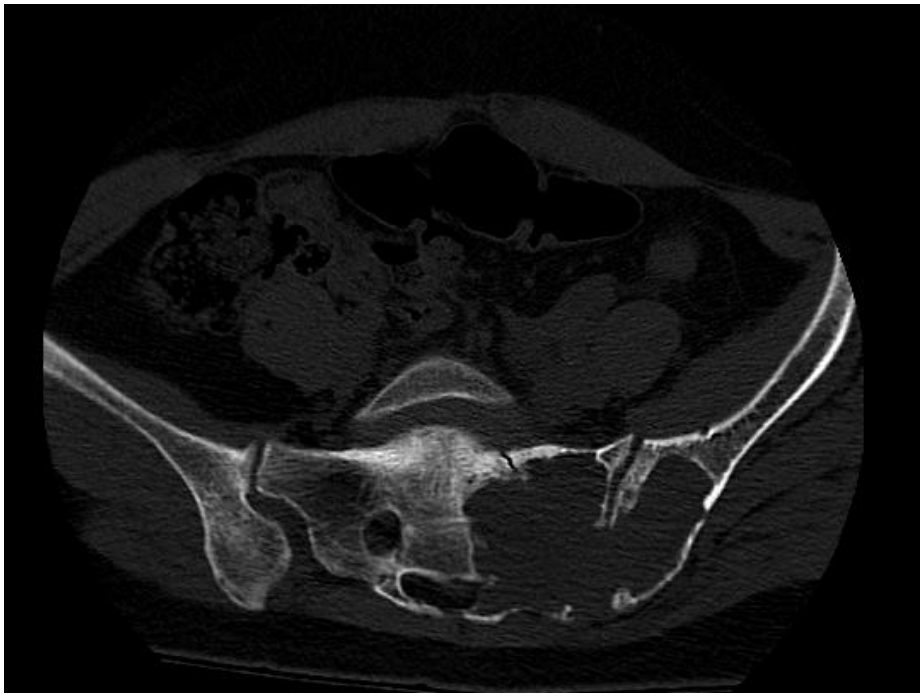
A CT study was performed on a 16-slice CT scanner (GE revolution) using the following parameters: slice thickness 5 mm; 120 kV; 130 mA; and matrix 512×512.

A large lobulated eccentric expansile lytic lesion measuring 4.7x6.4x6.3cm (APxTRxCC) was noted, involving the left lateral aspect of upper sacral vertebrae extending into the lateral sacral foramina, left sacro-iliac joint, ilium and adjacent soft tissues. Few internal septations noted in the inferior aspect. The lesion is involving the left sacral ala and lateral masses of S2-4 vertebrae with narrow zone of transition. There were no fluid–fluid levels. No periosteal reaction/internal matrix mineralization.

Posteriorly the lesion is showing extension into the epidural space with indentation on the cauda equina and left sacral nerve roots.

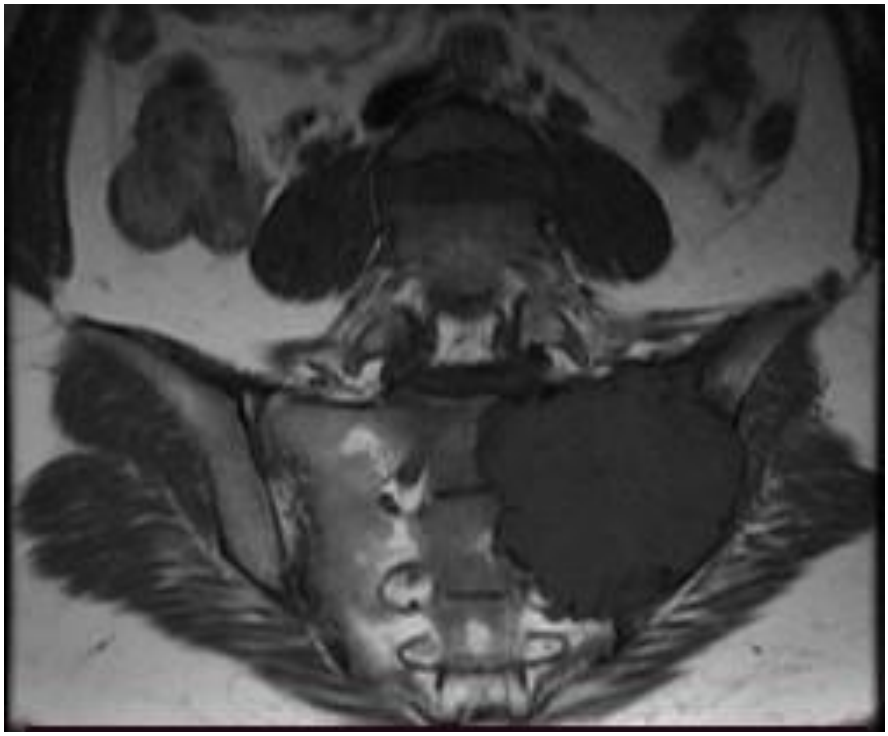


(Figure 3) Axial noncontrast CT scan shows a large expansile lytic sacral lesion (with attenuation similar to muscle) with extension across the sacro-iliac joint and involvement of lateral sacral foramina.

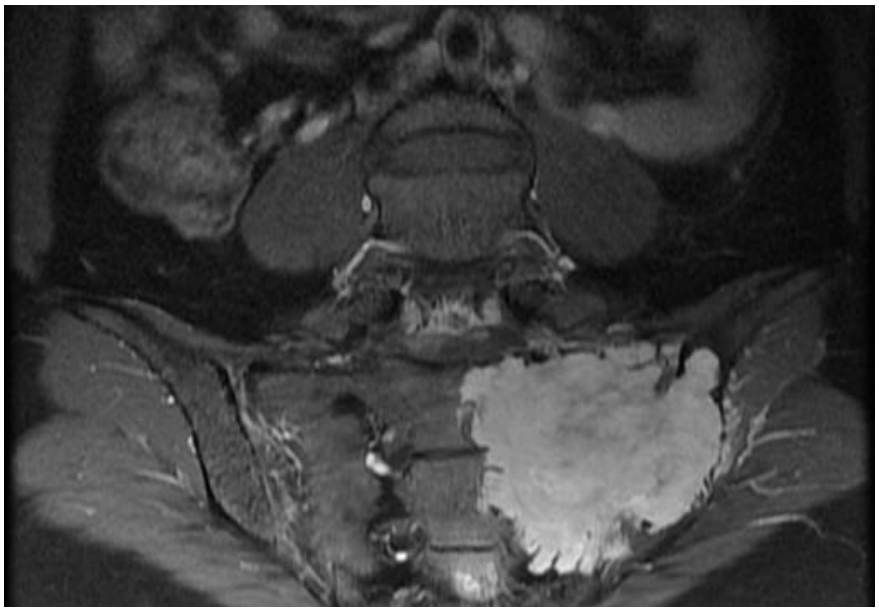


(Figure 4) Axial noncontrast CT scan (bone window) shows a large expansile lytic sacral lesion with extension across the sacro-iliac joint, involvement of lateral sacral foramina and cortical destruction.

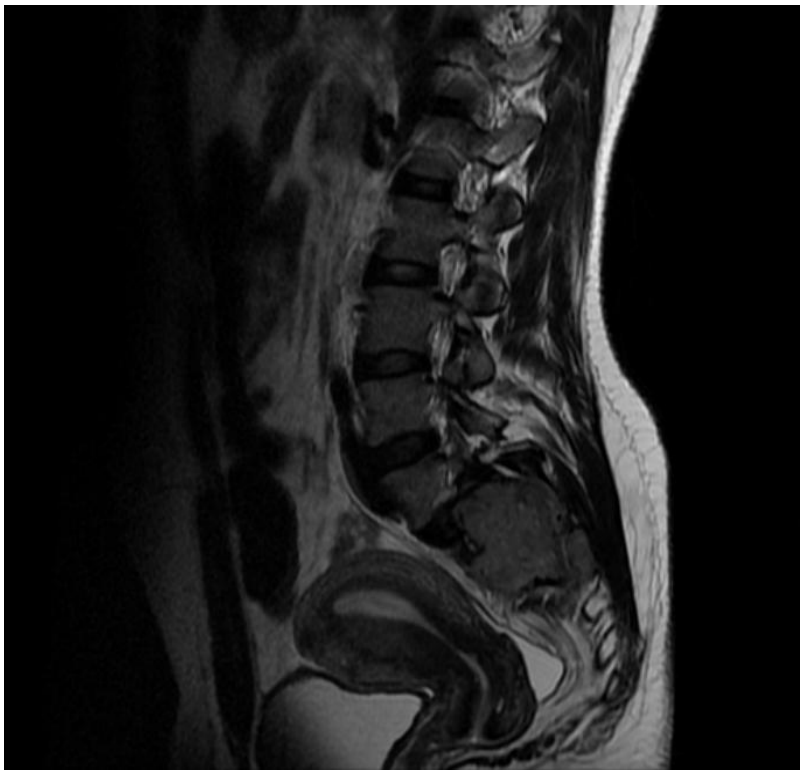
Magnetic resonance images were obtained on a 1.5T scanner (GE SIGNA HDXT). The signal intensity of the lesion on T1-weighted images (TR/TE=620/9.6ms, 256x160 matrix size, 22-cm field of view, 4-mm slice thickness, 128-s acquisition time) was is intense to muscle.



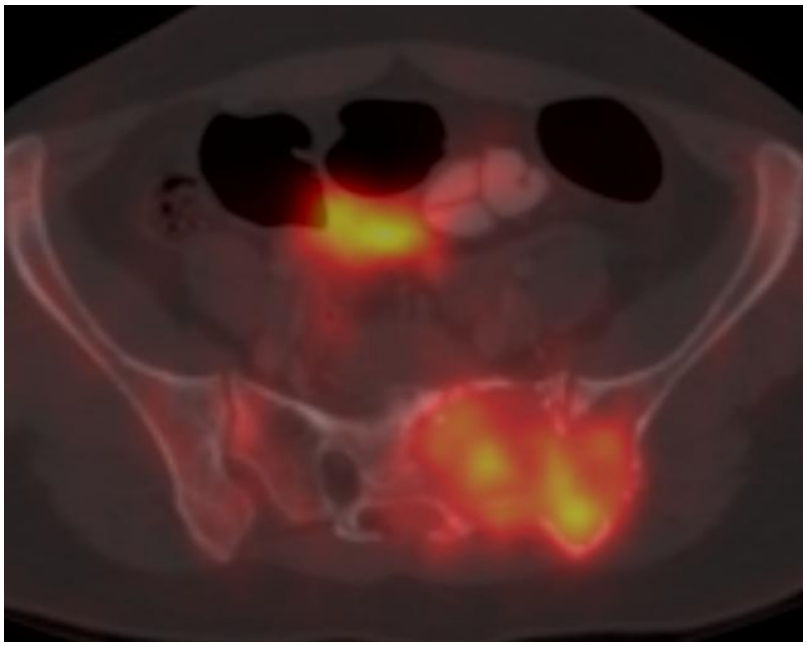
(Figure 5) Coronal-oblique T1-weighted image shows an isointense tumor with respect to adjacent muscle.



(Figure 6) Coronal-oblique T1-weighted image following gadolinium administration reveals intense enhancement of the tumor. Invasion of the upper left sacral foramina seen.



On T2-weighted images (TR/TE=3600/108 ms, 384x224 matrix size, 28-cm field of view, 4-mm slice thickness, 123-s acquisition time), the mass was mostly hyperintense to skeletal muscle (Figure 7)with few tiny fluid intensity foci within.



(Figure 8) PET-CT scan showing the presence of intense FDG avid (SUVmax 7.2) expansile lytic lesion with soft tissue involving the sacrum and adjoining left iliac bone with extensions. Mildly FDG avid (SUV max 4.3) mesenteric lymph node seen in the right lumbar region.

Trucut biopsy was done for this patient, which revealed the presence of poorly differentiated neoplasm. Immunohistochemical analysis was done which show edneoplastic cells positive for desmin, ER/PR, SMA and negative for CK, myogenin, synaptophysin. Ki67 was 10-15%. Overall features were in favor of leiomyosarcoma.

DISCUSSION

The first case of primary osseous leiomyosarcoma was described by D M Evans, N G Sanerkin in a case study done in a 73-year old patient in 1965, who was diagnosed with primary leiomyosarcoma of tibia(1). An extraossseous source of leiomyosarcoma (bony metastasis from the uterus or digestive tract) must be ruled out prior to coming to the diagnosis of PLB. In our patient, abdominal and pelvic sonography was done to rule out the presence of any other primary pathology.

Leiomyosacroma is a malignantmesenchymal soft tissue tumor arising from the smooth muscle cells and is considered as the malignant counterpart of leiomyoma. It is characterized by smooth muscle differentiation in the absence of malignant osteoid formation. It can occur anywhere with smooth muscles and the most common locations includes uterus (accounting for nearly 1/3 rd cases), retroperitoneum (most common non-uterine site), stomach, esophagus, small intestine and vascular structures. Primary osseous leiomyosarcoma is a rare entity accounting for less than 1% of primary bone tumors (2). Low grade leiomyosarcomas are extremely rare, accounting for less than 20%.(3)

Lesser than 100 cases of primary leiomyosarcoma of the bone have been reported around the world. Primary leiomyosarcoma involving vertebrae is extremely rare and only 11 cases has been reported in the literature. Among the 11 patients, 2 patients had tumor in cervical vertebrae, 6 in thoracic, 2 in lumbar and 2 in sacral vertebrae. (4) Out of 11 reported cases, 3 were male and 9 were female patients.

In a case review series done by Miura (5) in which he evaluated 136 cases of PLB from 1944-1999, the age range of patients was 9 to 87 with a mean age of 49.1. No gender predilection was observed in the study.

Most common sites of PLB is mainly in the bones of the lower extremities, predominantly around the knee joint (distal femur and proximal tibia). The most common clinical presentation is pain. Swelling, pathological fractures and soft tissue involvement are other associated findings.

Cell of origin of PLB is controversial. Prominent post contrast enhancement in the patients of PLB was suggestive of vascular smooth muscular origin.(6) Berlin et al. (7) performed angiography in six patients with PLB. out of 6 patients, 4 showed hypervascular tumors. Hypertrophied arteries which supplied these lesions were demonstrated in these patients.

Radiographically, Leiomyosarcoma of the bone are mostly osteolytic lesion with geographic, moth eaten orpermeative type of bone destruction. Geographic osteolytic lesions and surrounding sclerosis are commonly seen with low grade lesions (8).

Both CT and MRI are useful for assessing the tumor extension, soft tissue involvement and associated findings like zone of transition, perilesional edema and periosteal reaction. Yamaguchi et al (9) and Gold et al (10) studied on how useful MRI was in detecting the intertrabecular-infiltrating lesions. Lo et al. [11] in 1995 described a primary leiomyosarcoma of the spine, with T1 intermediate and T2 hyperintense signals. Sundaram et al. [12] summarized the findings of MR images in 12 patients with osseous leiomyosarcoma. All the lesions showed isointensesignal intensities to muscle on T1W images. On T2-weighted images, isointense signal was seen in two, iso-to-hyperintense signal in two and hyperintense signal in eight patients. In our patient, the lesion was showing T1 isointense and predominant T2 hyperintense signals.

Our patient came with complaints of lower back ache and imaging features revealed the presence of lytic lesion involving the sacrum. It is difficult to demystify sacral lesions purely on the basis of imaging features, certain entities like patient age, sex and certain specific features like fluid-fluid levels and calcifications aid in differentiating different lesions. the final diagnosis is determined with the help of biopsy. Most common malignancy involving the sacrum is metastasis. In our patient we excluded the possibility of secondary leiomyosarcoma from uterus with the help of pelvic ultrasonography. Most common primary malignancy of the sacrum is chordoma and second most common is giant cell tumor. Sacral chordoma is midline lesions commonly seen in middle aged men whereas giant cell tumors are eccentric and typically seen in young women (2^{nd} to 4^{th} decade).

Giant cell tumors are locally aggressive and tends to cross sacroiliac joint. They are vascular tumors and show avid post contrast enhancement. In view of similar imaging features (lytic expansile lesion crossing sacroiliac joint with avid enhancement) seen in our case, we came to the conclusion that giant cell tumor could be a possible diagnosis. On account of few patchy intra-lesional areas of diffusion restriction, lymphoma was given as a differential diagnosis.

In young, Ewing's sarcoma and osteosarcoma can occur but they present with aggressive imaging features like permeative or moth eaten lytic bone destruction, periosteal reaction and cortical destruction.

Lymphoma, myeloma and metastasis are other osteolytic lesions which can involve the sacrum. Primary lymphoma of bone is rare, accounting for less than 5% of malignant bone tumors. Mulligan et all (13) done a case series on 237 patients with primary bone lymphomas and concluded that long bone are more commonly affected than flat bones (71% in long bone v/s 22% in flat bones). Out of 237 tumors, only 2 were located in the sacrum. In our patient, metastaticlymphomatous involvement of the sacrum was ruled out because of absence of palpable enlarged lymph nodes and organomegaly. Metastasis and myeloma are extremely rare, in patients of less than 30 years of age.

Electron microscopy is the most useful investigation in differentiating PLB from other types of spindle cell tumors (since most of them have similar imaging features). Macroscopically PLB appears as a fleshy, greyish white intramedullary bone tumor with areas of necrosis.

Microscopically, the features are similar to leiomyosarcoma of any other site. Typical electron microscopic features are elongated pleomorphic spindle cells which are separated by collagen, thin cytoplasmic filaments forming dense elongated condensations, and pinocytoticvesicles. The tumor is devoid of osteioid/chondroid matrix.

Immunohistochemically, the tumor cells show positive staining for muscle markers such as smooth muscle actin (SMA), muscle specific actin (MSA or HHF35) , desmin and myosin.

Our patient displayed histologic and immunohistochemical features of smooth muscle tumor. The size and aggressive nature of the lesion was suggestive of high-grade leiomyosarcoma. However Ki 67 index was 10-15% indicative of low-grade tumor.

In conclusion, we report a rare case of low grade leiomyosacroma of bone. The age of the patient and location of the lesion were uncommon. There is considerable overlap in imaging findings of sarcomas although histological and immunohistochemical analysis serve as useful tools in differentiating these neoplasms. We are hoping that our case study would aid in widening the diagnostic spectrum of the disease.

ACKNOWLEDGEMENT
We would like to express our gratitude to everyone at the department of Radiodiagnosis in KMCT medical college, Kozhikode for their constant support and guidance. The authors would also like to thank Dr. Sathi, Department of pathology and Dr. Thanseer, Department of nuclear medicine for their constant support.

CONFLICT OF INTEREST: none

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