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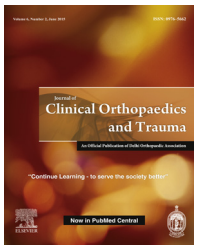

Case Report

Primary leiomyosarcoma of femur

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ABSTRACT

Primary leiomyosarcoma of the bone is exceedingly rare. In this case, we describe a middle-aged female with a primary leiomyosarcoma of the distal femur. The patient was treated by hip disarticulation. The patient continues to be disease-free at one-year follow-up.

1. Introduction

Malignant tumours comprising of spindle-shaped cells with distinctive smooth muscle cell features are called leiomyosarcomas (LMS).[1] Most of these tumours originate in the uterus, gastrointestinal tract, and retroperitoneum.[2] Primary bony LMS is a rare clinical entity. Majority of the cases are secondary deposits from a distant source. LMS of the bone occurs more commonly in middle-aged males.[2] Long bones are particularly involved, distal femur being the most common site.[3]

The first description of a primary bone leiomyosarcoma was given in 1965.[1] Since then, to the best of our knowledge, around 90 such cases have been reported. We present the clinical features, radiological findings and pathological description of a primary leiomyosarcoma originating from the distal femur in a middle-aged lady.

2. Case report

A 50-year-old female of Indian subcontinent presented to our department with a history of pain and swelling around the left

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knee for the last six months. The patient had a history of insignificant trauma to the left knee following which she noticed pain that gradually increased in severity. There was an associated swelling on the anterior aspect of distal thigh that gradually progressed in size. Over the course of the last month, the patient had become unable to bear weight on the affected extremity. These symptoms were associated with history of loss of weight and appetite. There was no other contributory family and past history.

Examination revealed a globular ill-defined swelling on the anterior aspect of distal thigh, extending over to the knee joint, around 10 cm × 10 cm × 5 cm in size. The skin over the swelling was stretched and shiny with dilated tortuous veins coursing over the swelling. There was marked wasting of muscles of the thigh. The swelling was warm and tender on palpation. Consistency varied from bony hard to firm. The patient had a flexion contracture of 30° at the knee. Patellar mobility was also restricted. There was no neurovascular deficit.

Plain radiographs revealed a large expansile lytic lesion originating in the distal femur that predominantly involved the epiphyseal and metaphyseal regions. Superiorly, it also involved the adjacent part of distal femoral diaphysis. A pathological fracture was observed at the metaphysio-diaphysial junction of the distal femur. The lesion had permeative, ill-defined margins along with evidence of cortical thinning. Gross disruption of anteromedial cortex and breach of the articular margin of the femur was noted with extension of the lesion into surrounding soft tissues (Fig. 1).

Multiplanar contrast-enhanced MR imaging was performed to obtain T1w, T2w and STIR images. The study revealed a large, well-marginated mass lesion involving the epiphyseal and metaphyseal region of distal femur with cortical disruption and extension of the lesion into adjacent soft tissue. The lesion appeared hypointense on T1w-images and heterogeneously hyperintense on T2w and STIR images. Heterogeneous enhancement of the lesion was also noted in the post-gadolinium sequences. Anteriorly, the lesion was observed to be abutting the quadriceps tendon. Inferiorly, destruction of articular surface was observed with extension of mass into the knee joint. Posteriorly, the mass was seen to be in close proximity to the popliteal neurovascular bundle, but without any evidence of neurovascular invasion. Superior extension of the lesion was noted as hypointensity on T2w and STIR images, and post-contrast enhancement of the marrow in femoral diaphysis. Subcutaneous tissue and skin along the anteromedial aspect of thigh was suspected to be involved since it revealed mild heterogeneous enhancement in the post-gadolinium sequences (Fig. 2). Skeletal survey, chest X-ray, abdominal sonography and a CT chest and abdomen revealed no other site of primary or metastatic disease.

A core biopsy revealed loosely cohesive clusters of cells with oval elongated nucleus, moderate amount of cytoplasm

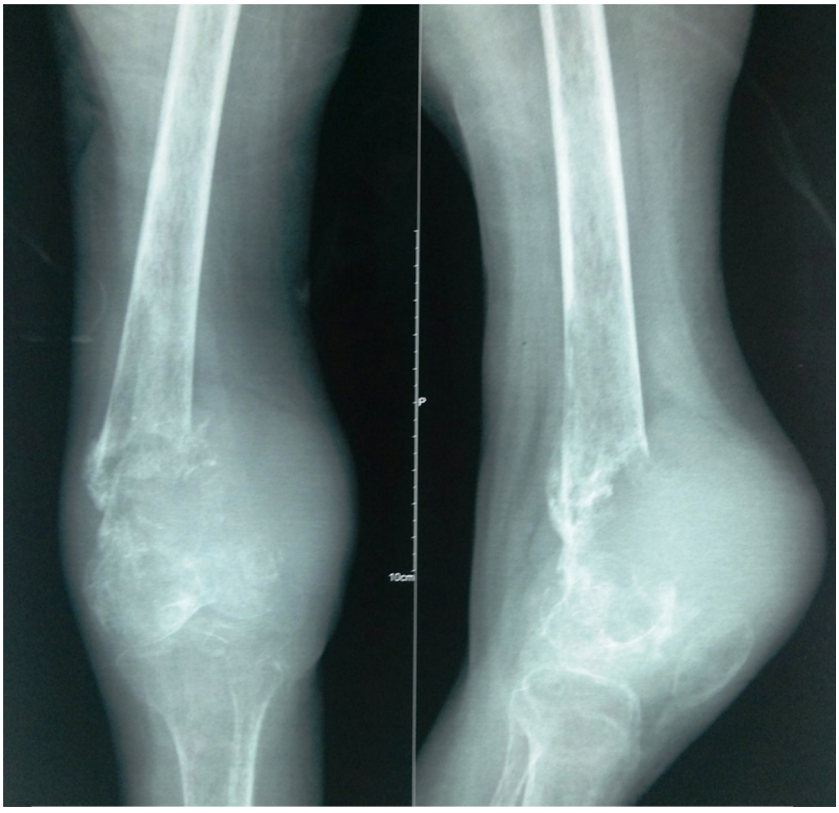

Fig. 1 – Plain radiographs showing an expansile lytic lesion with permeative ill-defined margins in the distal femur involving the epiphyseal and metaphyseal regions. A pathological fracture is seen at the metaphysio-diaphysial junction of the distal femur. Breach of the articular margin of the femur can be seen.