wjpmr, 2020,6(9), 197-204

SJIF Impact Factor: 5.922

WORLD JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.wjpmr.com

Review Article

ISSN 2455-3301

WJPMR

PRIMARY LYMPHOMAS OF BONE

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Article Received on 20/07/2020 **Article Revised on 10/08/2020** **Article Accepted on 30/08/2020**

ABSTRACT

Primary lymphomas of the bone are rare cancers that only affect the osseous structures without any lymph node involvement. These patients usually present with localized pain and swelling. Radiological studies may show an aggressive local osseous lesion. Treatment options also need to be aggressive, usually involving a combination of surgical and non-surgical options. We present a case of a 56 year old male who presented with progressive painful swelling in his right lower extremity.

KEYWORDS: Primary lymphoma of bone, Lymphoma, Magnetic Resonance Imaging.

INTRODUCTION

Primary lymphomas of bone is defined as when the disease affects one or more bones with or without involvement of local lymph nodes but with no evidence of disease in distant nodes or other extraosseous sites, i.e. Ann Arbor stage I and stage II disease,[1] a lymphoma with the above-mentioned pattern can be considered a primary lymphoma of bone, as well as when involvement of bone marrow is present at diagnosis, i.e. Ann Arbor stage IV.[2]

Case Report: This case is a 56-year-old male who presented with progressive painful swelling in his right lower leg of 10 months duration.

He underwent imaging studies, including plain radiographs, MRI (Magnetic Resonance Imaging), PET/CT (Positron Emission Tomography/Computed Tomography) and radionuclide bone scan. Plain radiographs demonstrated an ill defined lytic lesion involving the right tibial diaphysis with cortical breakthrough and small amount of periosteal reaction. The MRI studies demonstrated an intramedullary lesion involving right mid tibial shaft with cortical breakthrough and soft tissue extension. PET/CT images demonstrated increased FDG (fluorodeoxyglucose F 18) uptake involving the lesion. Nuclear Medicine Bone scan images demonstrated increased radiotracer uptake involving the lesion. No other lesions were present elsewhere in the body.

The tibial shaft lesion was biopsied. The histopathology revealed the bone lesion was lymphoma. Chemotherapy and surgical management are being planned for the patient.

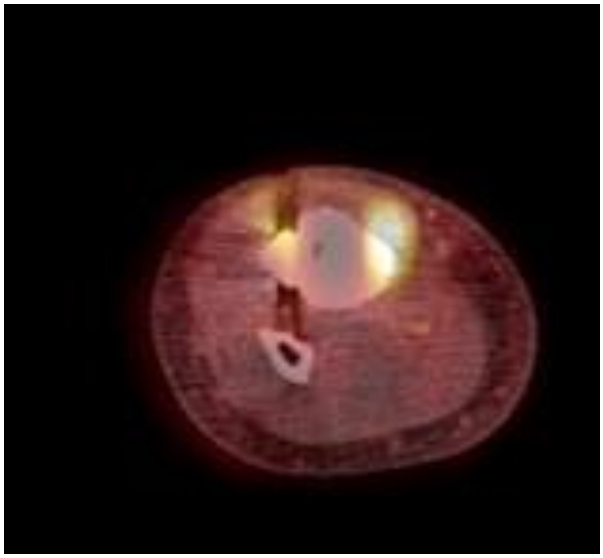


Figure 1a: Anteroposterior radiograph of right lower leg. A lytic lesion involving the tibial diaphysis with cortical breakthrough and small amount of periosteal reaction.

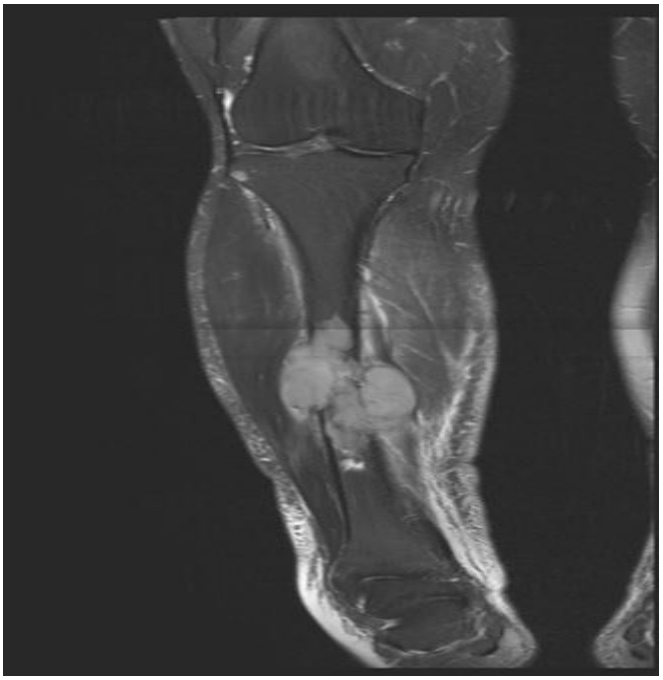


Figure 1b: Lateral radiograph of right lower leg. Lytic lesion of the tibial diaphysis demonstrates cortical breakthrough.

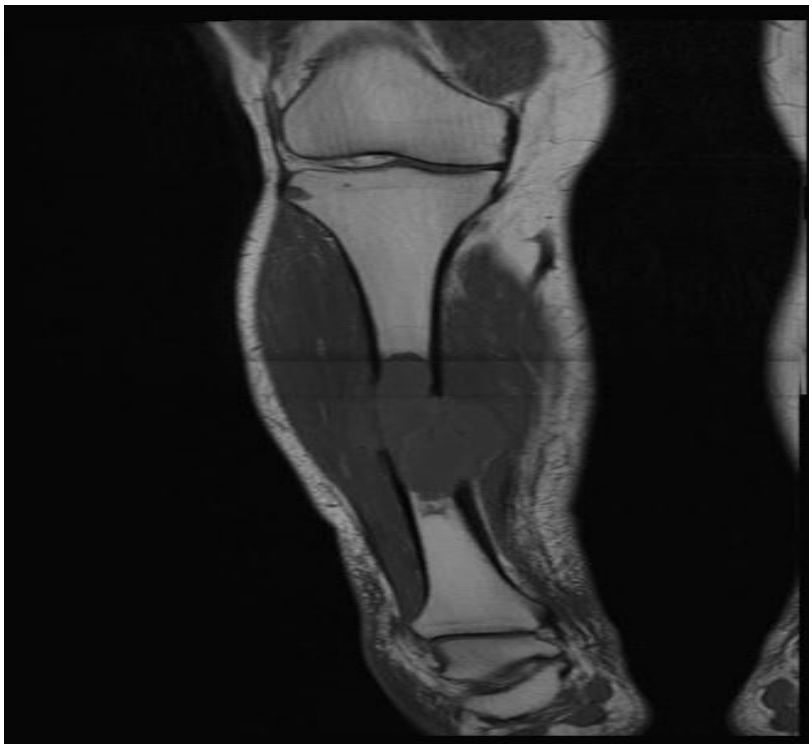


Figure 2a: Coronal T1 weighted MRI of right lower leg. A T1 hypointense lesion is seen in the diaphysis of the tibia with cortical breakthrough and associated soft tissue mass.

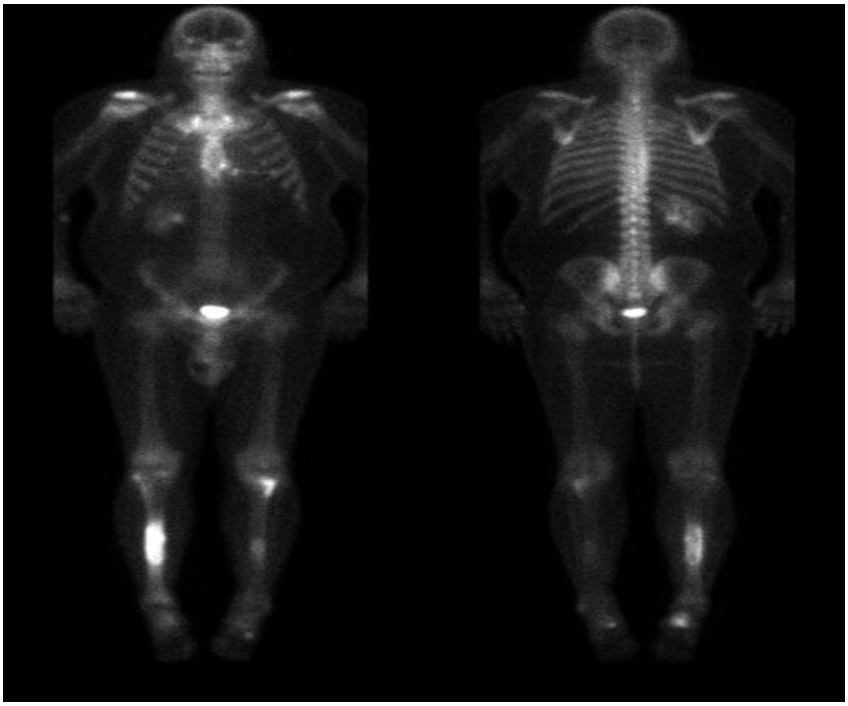


Figure 2b: Coronal T2 weighted fat saturated MRI of right lower leg. A T2 hyperintense lesion in the tibial diaphysis with associated soft tissue mass.

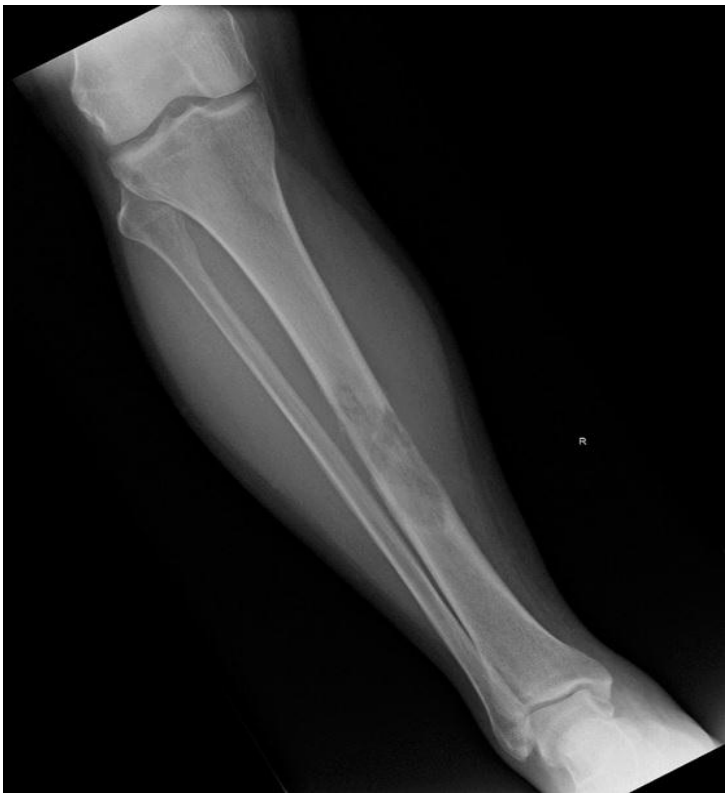


Figure 3: Radionuclide bone scan of whole body. Increased Technetium 99 radiotracer uptake is seen in the right tibial diaphyseal lesion. No other abnormal foci of increased Technetium 99 uptake is seen in the whole body.

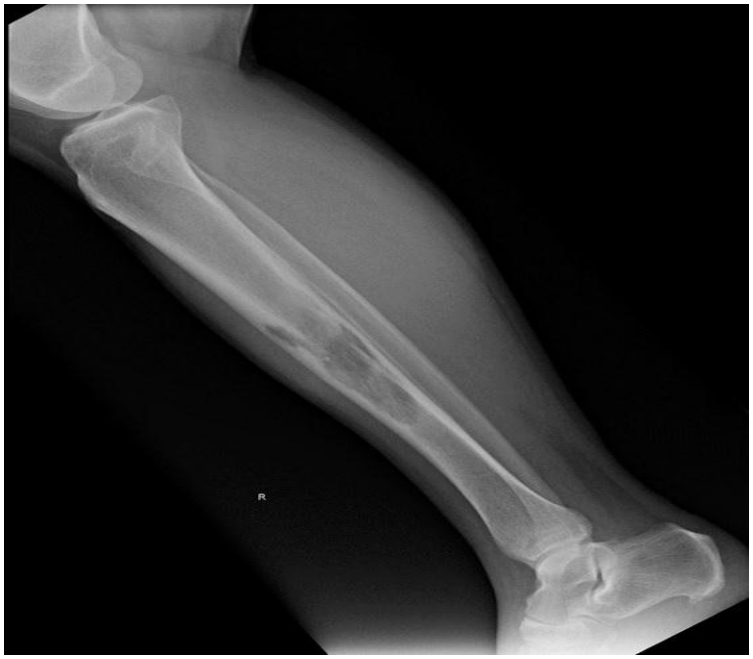
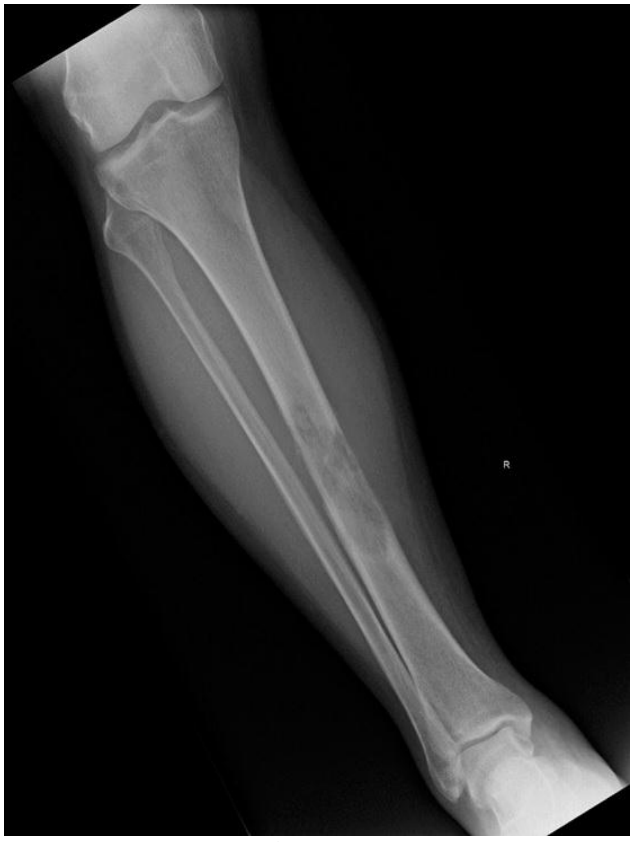


Figure 4: Axial fused PET/CT axial images of the right lower leg. Increased F 18 FDG uptake is seen in the right tibial diaphysis.



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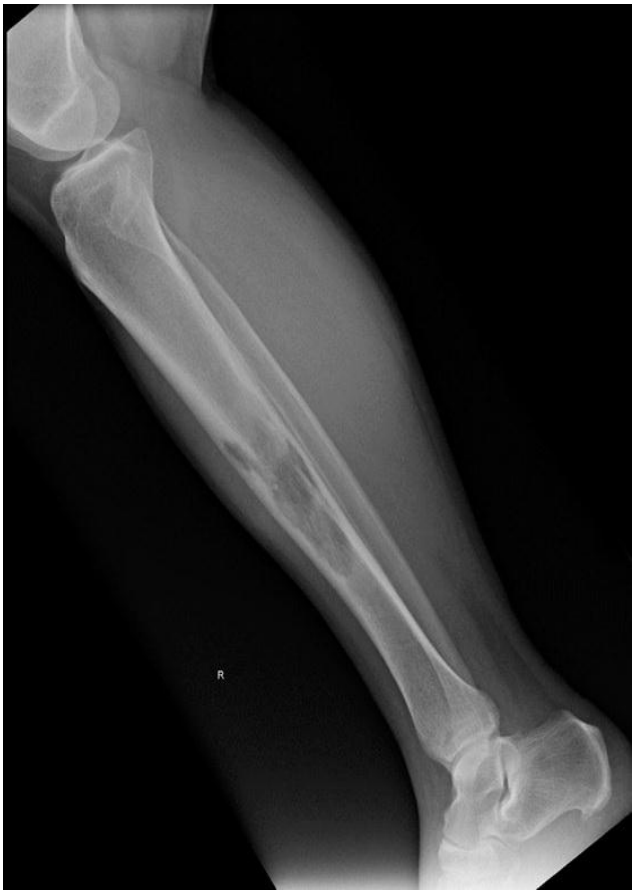

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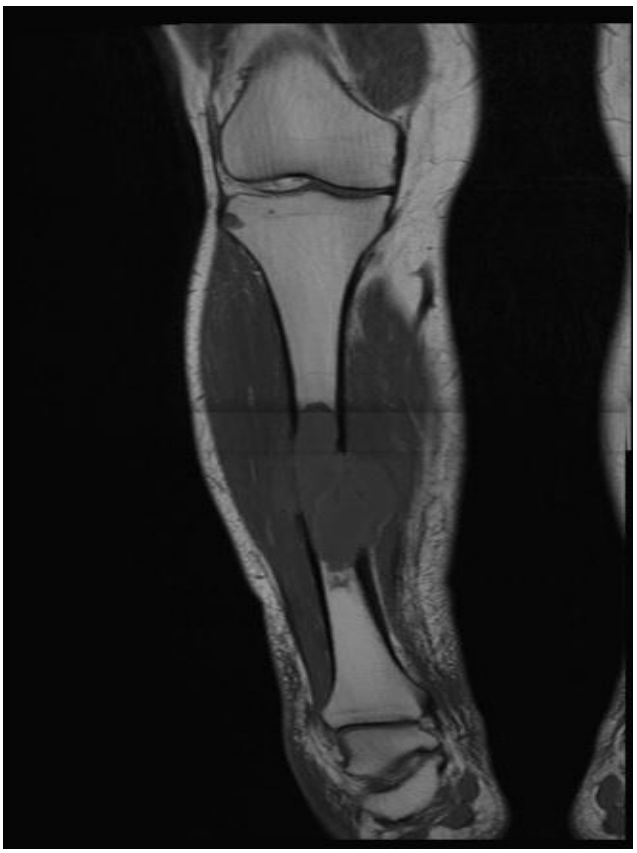

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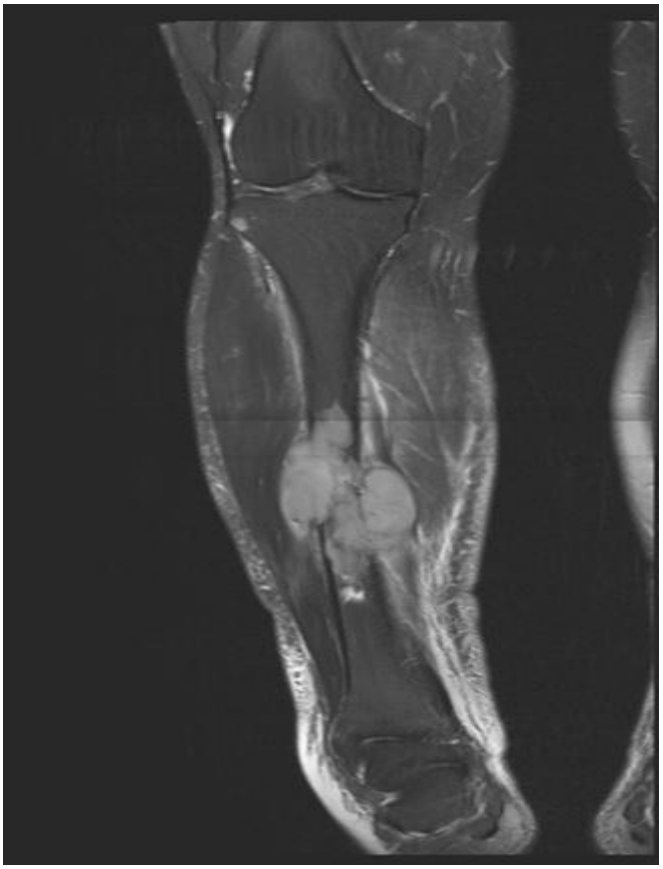

Figure 2b:

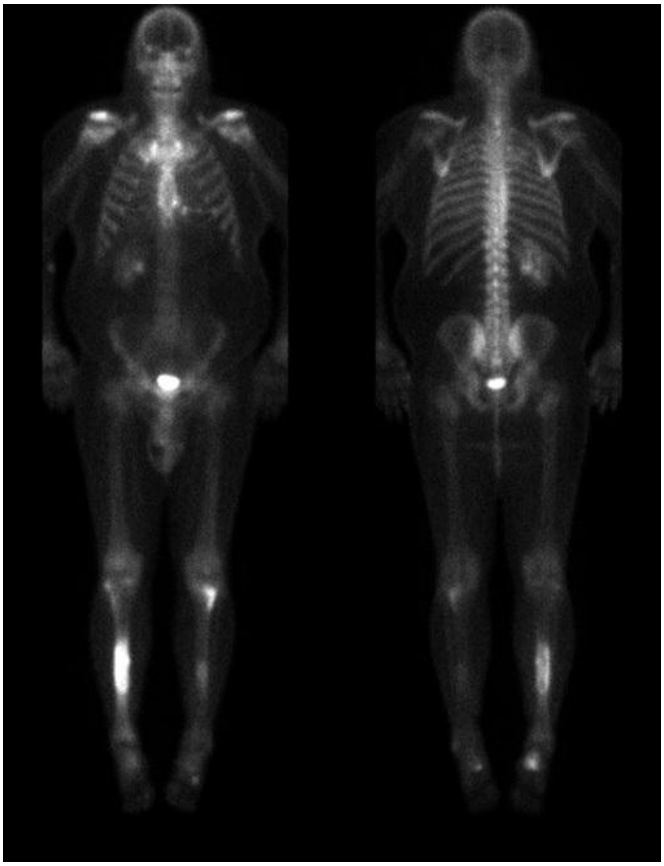

Figure 3:

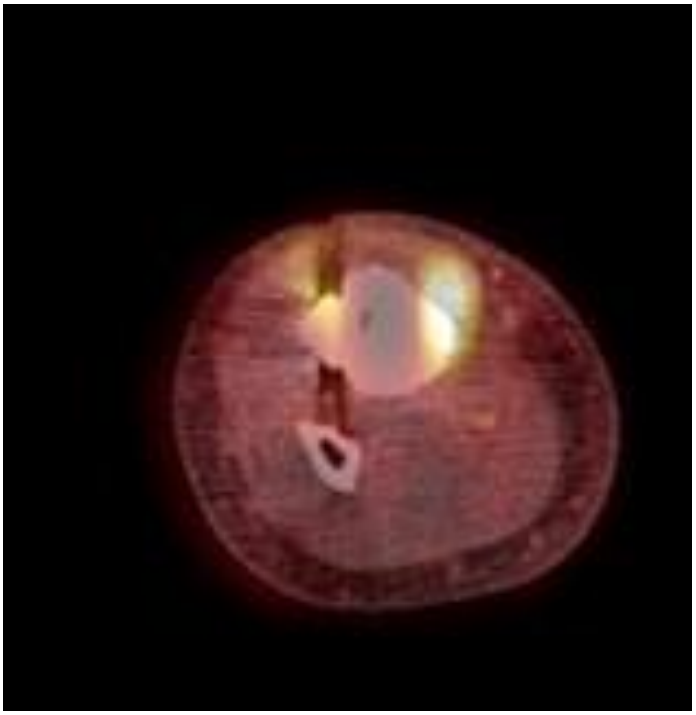

Figure 4:

DISCUSSION

Primary lymphomas of bone is considered as when the disease affects one or more bones with or without involvement of local lymph nodes, but with no evidence of disease in distant nodes or other extraosseous sites, i.e. Ann Arbor stage I and stage II disease.[1] According to the Ann Arbor staging system, our patient had stage 1 lymphoma.[2]

Primary bone lymphoma is a rare malignant entity, accounting for 2% of all bone tumors and approximately 5% of all extranodal lymphomas.[3] The lesion can be seen at any age, although most patients are over 30 years of age with the peak incidence in the 6th decades of life. Males are affected twice more often than females.[4] Femur, tibia, pelvic bones are most commonly involved: together they make up to 50% of cases. Other locations include long bones of upper limbs (20%); ribs, mandible and scapula comprise the remaining 30%.[5]

The most common symptom of primary bone lymphomas is localized pain and swelling.[5]

On radiographs and computed tomography (CT) scans, primary bone lymphoma usually presents as an aggressive lytic lesion with ill-defined margins. Less often, a sclerotic or mixed bone lesion may be seen.[6] Cortical breakthrough or a pathologic fracture not uncommon. On magnetic resonance imaging (MRI), the lesion is usually hypointense on T1 weighted images (T1WI), hyperintense on fat-suppressed T2-weighted images (T2WI), and show heterogeneous enhancement on post contrast fat-suppressed T1WI. Cortical breakthrough and soft tissue extension when present are well demonstrated on MRI.[7,8] On F18-PET/CT, the bone lesion usually shows avid radiotracer uptake.[7,9]

Treatment options include surgery, chemotherapy, radiation therapy, or more often, a combination thereof. Surgical treatment in primary bone lymphomas should aim to restore function and eliminate pain, and thus minimizing potential delays in chemotherapy initiation.[3] Pathologic fractures often require internal fixation to minimize deformity. Overall survival of patients affected by primary bone lymphomas was 74.2 % at five years.[3] Persisting radiological abnormalities after treatment is a common finding in bone lymphomas and does not always mean that the patient is not in remission.[10]

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