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# CLINICAL ARTICLE

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## Primary bone lymphoma: Imaging findings of a rare primary bone tumour

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### Abstract

Primary bone lymphoma (PBL) is a rare cause of primary bone malignancy and it is unusual for extranodal lymphoma to arise in the skeletal system. The imaging appearance is variable and the diagnosis is usually made on histology. We present the radiographic and CT features of primary bone lymphoma of the humerus in a young patient and discuss the variable imaging appearances of this condition.

### Key words:

primary bone lesion, skeletal, imaging, CT

### Introduction

Primary bone lymphoma (PBL) is defined as lymphoma occurring in the bone without evidence of distal nodal or extranodal tissue involvement.<sup>1,2</sup> It accounts for approximately 3% of all primary bone malignancies.<sup>3</sup> The vast majority of these are large B-cell non-Hodgkin lymphomas.<sup>4</sup> Alternative names for this malignancy include reticulum cell sarcoma and osteolymphoma.<sup>5</sup>

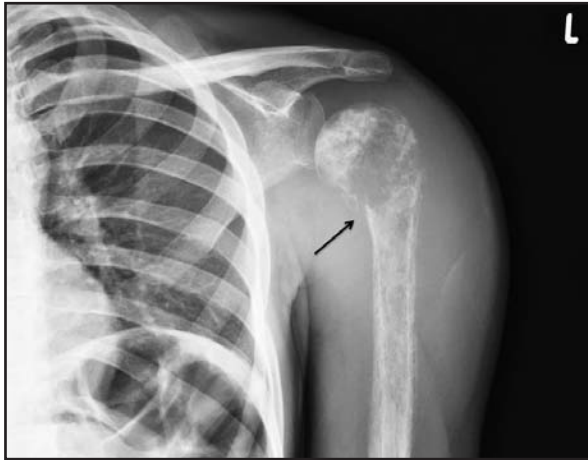
### Case report

A 22-year-old male patient presented with pain and swelling of his left shoulder for approximately three months. The pain had worsened over the two weeks prior to presentation. He had no history of night sweats or weight loss. On examination the shoulder was swollen with no evidence of infection. He also had a decreased range of movement around the shoulder joint.

Radiographs of the left shoulder demonstrated a large lytic lesion in the humeral head with surrounding permeative destruction extending into the epiphysis and diaphysis. A pathological fracture was noted through the surgical neck and a periosteal reaction was present (*Figure 1*).

A CT scan confirmed the osseous findings noted on the radiograph. It further demonstrated extension of the pathology into the surrounding muscles (*Figures 2a and 2b*). On a CT scan of the neck, chest and abdomen there was no evidence of metastatic disease or lymphadenopathy.

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**Figure 1.**

AP radiograph of the proximal left humerus including the shoulder joint demonstrates a lytic lesion in the metadiaphysis of the proximal humerus with a permeative pattern extending upwards into the epiphysis and downwards into the diaphysis. A lamellated periosteal reaction is present and a pathological fracture (arrow) is seen through the surgical neck of the humerus.

The full blood count, electrolytes and chemistry (calcium and magnesium) as well as the liver function tests were within normal limits. The patient's HIV status was also negative. Biopsy revealed a high grade large B-cell lymphoma.

## Discussion

Primary bone lymphoma is rare and the histology is indistinguishable from primary nodal or lymphoid tissue lymphomas. In order to be diagnosed as PBL the following criteria (known as Coley's criteria) should be met:<sup>1,2</sup>

- A primary focus in a single bone
- Positive histological diagnosis
- No evidence of distal soft tissue or distal nodal involvement

The concept of PBL with multifocal involvement has been suggested in publications.<sup>1,5</sup> However, this concept is controversial with some authors considering multifocal disease as being a disseminated disease, which excludes it from being classified as PBL. Supporting authors suggest that multifocal PBL has a predilection for bones around the knee which is uncommon in metastatic disease and therefore multifocal abnormalities in this region could be in keeping with primary multifocal osseous lymphoma.<sup>5</sup> There are also authors who have included patients with liver and splenic involvement without nodal involvement in their studies of PBL and who have used the expression of certain tumour markers as criteria for diagnosis. This lack of uniformity makes it difficult to assess the true incidence of the disease.<sup>6</sup>



**Figures 2a and 2b.**

Coronal CT scan of the proximal left humerus in bony and axial soft tissue windows demonstrating the focal lytic lesion with surrounding permeative pattern involving the epiphysis (including the articular surface) and diaphysis. Cortical breakthrough and the pathological fracture are clearly visualised. Soft tissue window shows extension into the surrounding soft tissue.

Clinically, patients may present with pain, swelling, palpable mass, weight loss and fever.<sup>1,5</sup> Neurological problems related to the spinal cord may be the presenting symptoms in patients with vertebral involvement.<sup>6,7</sup> PBL occurs across a broad age group ranging from 18 months to 86 years although it is rare under the age of 10 years and has peak prevalence in the sixth and seventh decades.<sup>1,5</sup> It occurs slightly more commonly in males and affects portions of the bone with persistent haematopoietic marrow.<sup>8</sup> The femur is the most common site followed by the pelvis, humerus, skull and tibia.<sup>2</sup> Vertebral involvement has also been described. The mandible has been described as a rare location.<sup>3</sup>

PBL has a widely variable imaging appearance that has been divided into certain radiographic patterns by Krishan *et al.*<sup>5</sup> in their review of 20 cases of proven primary bone lymphoma. The 'lytic destructive pattern' is described as the most common pattern, reported in more than 70% of cases. This pattern may be described as 'permeative', 'moth-eaten' or 'focal lytic' with well-defined margins. Cortical breakthrough and pathological fractures are described in 25% of cases and a lamellated or interrupted periosteal reaction has been described in 60% of cases. Sequestra have also been demonstrated by CT in cases of PBL. An associated soft tissue mass usually heralds a poorer prognosis.

Our patient demonstrated all the findings described in this pattern with a large focal lytic lesion, a permeative pattern extending into the diaphysis, a lamellated periosteal reaction, cortical breakthrough and a pathological fracture. An associated soft tissue mass was also seen.

The second pattern, described as a 'blastic sclerotic pattern', is more common in metastatic lymphoma to the bone. Hodgkin's lymphoma, the rarer type of primary bone lymphoma, may occasionally present with this pattern but even in Hodgkin's lymphoma the lytic pattern predominates.

Subtle or 'near normal' radiographic findings may also be a presenting feature of PBL. Symptomatic patients with this pattern require a more sensitive modality of investigation, such as bone scintigraphy or MRI that may show striking abnormalities.

CT may be more sensitive than plain film radiography for soft tissue infiltration, cortical breakthrough and pathological fractures but its main role is in ruling out the presence of nodal disease or other soft tissue involvement in the chest and abdomen.

MRI is useful to assess the extent of bone marrow and soft tissue involvement.<sup>2</sup> T1-weighted pulse sequences are best for demonstrating marrow involvement. Lesions are usually low signal intensity on T1-weighted images and high signal intensity on T2-weighted images with areas of enhancement within the lesion on post-contrast imaging.<sup>5</sup>

Bone scintigraphy is useful for demonstrating multifocal osseous involvement but the role of positron emission tomography has not been well established.<sup>2</sup> Biopsy remains essential to confirm the diagnosis.

Imaging is also important during and after treatment and recognition of the treatment related changes is vital. Mengiardi *et al.* reported the imaging changes in 25 patients during and after the completion of therapy.<sup>4</sup> CT may show unstructured new bone formation up to six months after the start of therapy. Some patients may initially show extensive osteolysis before new bone formation. From 6 to 12 months after the start of therapy the bone develops a more structured appearance but up to 33 months post-therapy no patient demonstrated return to totally normal bone structure.<sup>4</sup>

Follow-up MRI imaging showed tumour volumes to decrease rapidly in the first three months after the start of therapy with disappearance of the soft tissue component within months of starting treatment. Residual bone marrow signal abnormalities were seen up to 25 months after the start of therapy and these may be as a result of necrosis and inflammation or residual tumour. Therefore post-treatment imaging findings may not be specific in ruling out active disease.<sup>4</sup>

PBL may be treated with a combination of chemotherapy and radiotherapy, or chemotherapy alone. The advances in newer chemotherapeutic agents have improved the prognosis of PBL so that it has a far better survival rate than other primary malignant bone tumours.

## Conclusion

PBL is a rare malignant bone tumour with a highly variable radiographic appearance. The prognosis, however, is much better than for other malignant bone tumours, thus a high index of suspicion should be maintained for this disease in patients over the age of 30 years presenting with solitary bone lesions.

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