

# Primary Lymphoma of Bone: A case report and brief review of literature

Mra Aye<sup>1\*</sup>, Jason Cabot<sup>2</sup>, Zulkimi Rosly<sup>3</sup> and Chiew Junloong<sup>3</sup>

<sup>1</sup>Department of Medicine, Melaka Manipal Medical College, Melaka 75150, Malaysia

<sup>2</sup>Cancer Center of Guam, 633 Gov. Carlos Camacho Rd., B5, Tamuning, Guam 96913, USA

<sup>3</sup>Jasin General Hospital, Jasin 77000, Malaysia

## Abstract

Skeletal manifestations of Non-Hodgkin Lymphoma (NHL) is unusual, occurring in less than 5% of NHL cases; and primary non-Hodgkin lymphoma of bone (PLB) is rare, usually diffuse large B-cell lymphoma. PLB generally presents with localised bone pain, or, less frequently, soft tissue swelling. Pathological fractures, well documented in soft tissue sarcomas, are unusual in PLB. We report a case of polystotic PLB that presented with involvement of left anterior tibia (with pathologic fracture) and left frontal skull.

## Introduction

Primary Non Hodgkin Lymphoma (NHL) of bone (primary bone lymphoma, PLB) a rare condition, accounting for less than 1-2% of adult NHL and less than 7-10% primary bone tumors [1-8], was first described by Oberling [9] in 1928 as reticulum cell sarcoma of bone. This was subsequently recognised as of lymphoid origin and renamed malignant lymphoma of bone [10,11]. Most primary lymphomas of bone are diffuse large B-cell lymphoma (DLBCL) subtype, the majority of cases are limited to bone, and occur in adults aged 45-60 years [1-5]. PLB may be difficult to diagnosis without a high level of suspicion [8]. Pain without antecedent trauma, unrelieved by rest is the most common presenting symptom [1,5-8]. PLB may present as monostotic or polystotic disease [3-8], often in long bones, with a slight male predominance. Bones most commonly involved are femur (29%), skull (11%) and tibia (10%) with variable radiographic manifestations [12].

PBL is rare compared to plasma cell neoplasms in bone, either plasmacytoma or myeloma. As bone marrow is the homing site of plasma cells there is circumstantial evidence that PBL represents post-germinal centre B-cells [13]. Although conventional radiology and other imaging modalities such as bone scan, CT and PET are not specific, MRI appears to be the most sensitive imaging technique for diagnosis of PBL [14-18].

## Case

A 46- year-old man with hypertension, gout, and chronic kidney disease was admitted for increasing painful swelling of anterior left tibia of two months duration causing ambulation difficulties. There was no fever or night sweats but poor appetite and significant weight loss. He had noted an asymptomatic, painless swelling of the left frontal region one year prior, but did not seek medical attention. This swelling subsequently rapidly increased in size, and was accompanied by painful swelling posterior and inferior to the left knee.

Examination revealed a thin, pale, weak, orientated male, with a non-tender soft, fluctuant mass of the left frontal region of skull

measuring 8 x 8 cm, fixed to the bone, a left preauricular lymph node 2 x 2 cm, firm, and non-tender. There was a 3 x 3 cm central mass around the umbilicus which was fixed and non-tender, multiple, bilateral palpable inguinal lymph nodes more prominent on the right, and gross bilateral pedal oedema. There was a swelling at the back of left tibia, about 10 cm below the knee, measuring 4x4 cm, non-tender to palpation, firm and with normal overlying skin. Two weeks later he developed massive abdominal distension, thought to be ascites, and gross, bilateral leg edema.

Plain X-ray of left knee showed a soft tissue swelling with moth eaten bone erosion of upper tibial posterior aspect with a pathological fracture. Plain X-ray skull revealed large erosive lesion of left frontal bone. Chest X-ray was normal with no rib erosions. Skeletal survey showed no lytic lesions except left frontal bone and tibia.

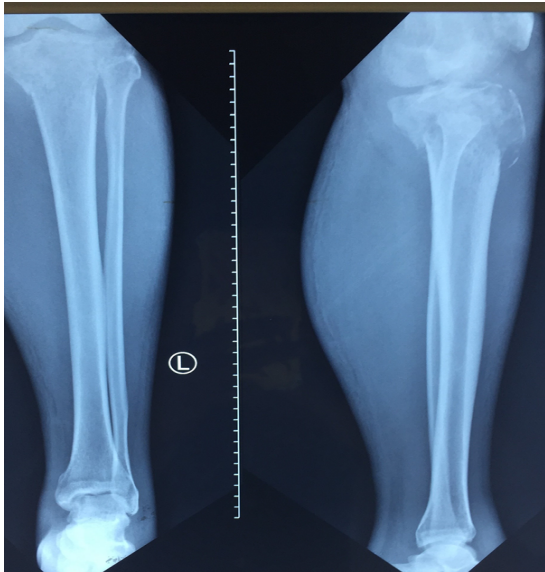
USG Doppler of legs demonstrated no deep vein thrombosis and USG abdomen, a large lobulated heterogeneously hypoechoic mass in the right lower abdomen measuring 8.4 x 8.7 x 13.4 cm with similar smaller masses around this mass with minimal ascites. Liver, spleen, gall bladder and pancreas were normal and there was renal parenchymal disease.

CT and Contrast Enhanced CT (CECT) of left fibula and tibia revealed lytic bone lesion involving the proximal left tibia with pathological fracture of left proximal tibia posteriorly (Figures 1 and 2), with a soft tissue component measuring 9 x 10.9 x 10.5 cm, extending superiorly into the popliteal fossa and erosions noted in distal left femur. CECT brain revealed a large enhancing mass in the left frontal

**Correspondence to:** Mra Aye, Department of Medicine, Melaka Manipal Medical College, Melaka 75150, Malaysia, Tel: +60-1955-45052; E-mail: mraaye@hotmail.com

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**Figure 1.** CT of left tibia and fibula.



**Figure 2.** Contrast enhanced CT of left tibia.

region measuring 8.7 x 6.7 x 7.8 cm extending from the scalp to the extradural space (Figure 3). There were no focal brain parenchymal lesions and normal grey/white matter differentiation. CT of abdomen and pelvis revealed a large lobulated mass anterior to the lumbar spine extending from the level of 3<sup>rd</sup> lumbar vertebrae superiorly and down to the right iliac region measuring 13.3 x 10.9 x 19.8 cm (Figure 4), with no clear plane between the mass and right psoas muscle, and encasing the common iliac arteries, right iliac vessels and inferior vena cava and a dilated right ureter traced down to the mass with mild right hydro nephrosis. Multiple enlarged nodes were seen in the left iliac and inguinal regions and lateral to the mass. Features of bilateral renal parenchymal disease were noted and there was minimal ascites.

#### Urine was negative for light chains.

Laboratory exams revealed anaemia {Hb 10.3 g/L (13-17)} with normal white blood cells and platelets, ESR was markedly elevated {98mm/1st hr (0-22 mm)}, blood urea 18.4 mmol/L (3.2-8.2) with creatinine 471 umol/L (82-115), corrected serum calcium 2.92 mmol/L

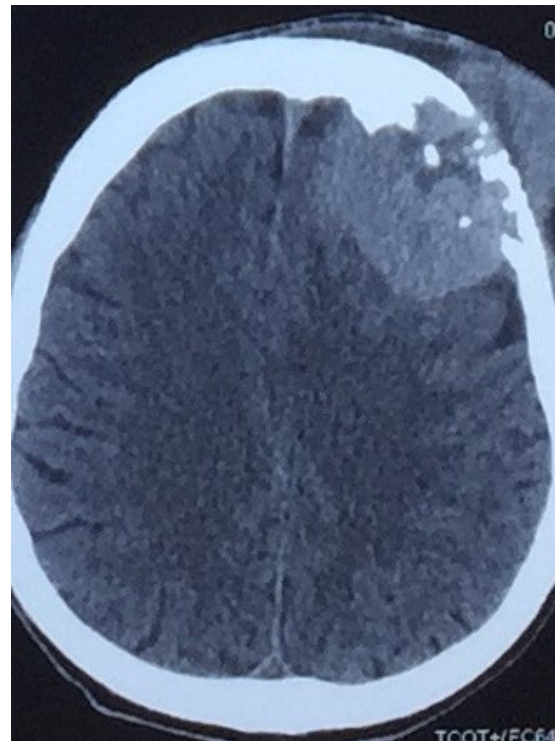
(2.16-2.50), uric acid 806 mmol/L (208.4-428.4). Serum phosphate, magnesium, liver function test and coagulation profile were within normal range.

A Tru Cut biopsy of the tibial mass was performed and the pathological fracture immobilized. Biopsy revealed diffuse large B cell lymphoma, with diffusely positive CD20 and MUM1; and negative CD3, CD10, CD138, BCL6 and CKAE1/AE3. Ki- 67 was 80%.

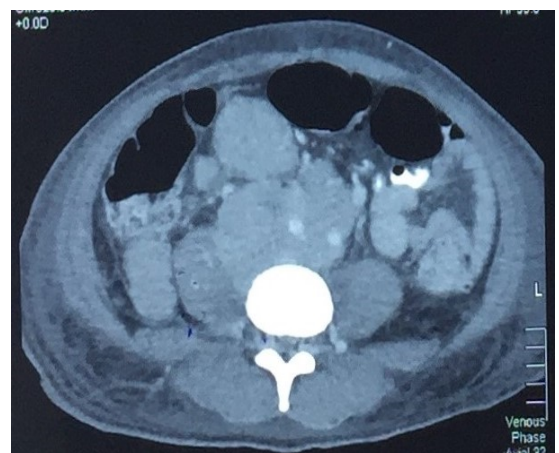
The patient was referred to the haematology department for further management but expired one month later while receiving chemotherapy.

#### Discussion

Originally provisional diagnosis of multi-osseous plasmacytoma was suspected, but changed to aggressive lymphoma when the huge



**Figure 3.** Contrast enhanced CT brain scan.



**Figure 4.** CT abdomen.

lobulated abdominal mass was seen on abdominal USG. It is felt our patient had PBL and bone involvement as a primary and not secondary event since the disease began in bones, with subsequent pathological fractures, and visceral masses becoming evident only after a time lapse of one year.

Literature review indicates skeletal involvement with NHL is less than 5%, and in all cases, bony involvement followed some time after presentation of the primary lesion [13]. Disseminated NHL presenting with pathological fracture is unusual [18,19]. Our patient appeared to have a left frontal swelling at onset, followed some months later by a tibial tumour with fracture, both with osseous erosions. There was also a large retroperitoneal mass which infiltrated widely.

Our case had many attributes of multifocal PBL such as age, painless bone swelling for considerable time without systemic symptoms, diffuse large B cell lymphoma in histology [1-5] and presence of common radiological features of PBL on CT scan: permeative osteolytic moth eaten lesions in proximal tibia with cortical break and fractures on CECT (contrast enhanced CT) of left tibia [14] (Figures 1 and 2). MRI has specific features of PBL [14-18] but our patient succumbed before the due date of MRI.

Although the original criteria of PBL by Coley [20] described solitary bone involvement, a larger group was described by Ostrowski et al. [21] who classified osseous lymphoma into four groups: Group 1, solitary primary bone lymphoma; Group 2 involving more than one bone without nodal or visceral disease; Groups 3 and 4 with distant nodal or visceral disease. The difficulty with multifocal PBL is distinguishing cases with primary osseous sites and extension (groups 3 and 4), from disseminated NHL and secondary osseous involvement. Multifocal osseous PBL, like solitary PBL, has a predilection for bones about the knee [14]. The combination of radiologic abnormalities of proximal tibia, distal femur, and skull is very uncommon in osseous involvement of disseminated NHL [22]. Overall, multifocal PBL exhibits a significantly better prognosis compared to patients with advanced-stage diffuse large B-cell NHL [23].

## Conclusion

We report a case of diffuse large B cell lymphoma of bone originating in multi-osseous sites. PBL is rare and diagnosis might be delayed or missed if it is not considered. It is reported to have a better prognosis than disseminated lymphoma if treated early and should be in the list of the differential diagnoses of bone swelling with bone fracture.

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