

# Precursor B-Lymphoblastic Lymphoma Presenting as a Solitary Bone Tumor Mimicking Osteomyelitis on MRI

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## Citation

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## Abstract

We describe a 22 year-old man with the rare diagnosis of precursor b-lymphoblastic lymphoma involving only the proximal tibia without evidence of leukemia. He presented with right knee pain and swelling of approximately three months duration and nonspecific radiologic findings and was initially treated for osteomyelitis of unknown etiology. Following histologic diagnosis, the patient received surgical excision of the lesion followed by chemotherapy and radiation. The patient was alive without evidence of disease at nine months follow-up.

## INTRODUCTION

Primary precursor B-lymphoblastic lymphoma (B-LBL) of bone is an uncommon neoplasm and accounts for less than 10% of cases of lymphoblastic lymphoma (LBL) and less than 1% of primary bone tumors. It usually involves extranodal sites in young adults and occurs most often in the skin [3]. B-LBL presenting as a solitary bone tumor without involvement of blood, bone marrow, or other sites is extremely rare and less than 10 cases of this type have been reported in the English language literature [3,6]. To the best of our knowledge, only 3 cases of B-LBL involving only the tibia have been reported and this represents the fourth case [6]. B-LBL often presents with few specific radiologic and laboratory findings and thus, can be difficult to diagnose and is often not considered in the patient presenting with bone pain.

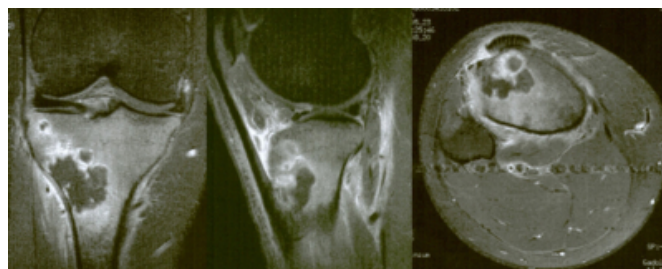
## CASE REPORT

A 22-year-old man began to have spontaneous pain and swelling of the right knee in May 2002. Suspecting a tendonitis at the insertion of the patellar tendon, he was treated at an outside hospital with non-steroidal anti-inflammatory medications. This treatment relieved some, but not all of the symptoms. Three months later, the patient presented to the clinic in our department complaining of pain involving the right proximal tibia, including the musculature in this area, when he did any weight-bearing activities. Also, he reported occasionally feeling pain at the region of interest, even while at rest. He had Magnetic resonance

(MR) images from an outside hospital that showed an irregular, lytic lesion involving the proximal tibia. (Fig. 1)

## Figure 1

Figure 1A-C: MRI; coronal T1 (TR 748 TE 14ms), sagittal T1 (TR 687 TE 12ms) and transversal T1 (TR 440 TE 14ms) after i.v.-administration of Gadolinium-DTPA (Magnevist, Schering, Berlin) and fat saturation showing proximal right tibia with an irregular, lytic lesion with fluid chambers and intense periosteal reaction



The physical examination at this time revealed some pain to palpation at the right, proximal, lateral tibia without any other signs of inflammation. The examination of the right knee joint was unremarkable. Significant laboratory studies included: leukocytes  $4.6 \times 10^3/\mu\text{l}$  (normal  $4.5 - 11 \times 10^3/\mu\text{l}$ ), c-reactive protein 2,2 mg/dl (normal 0 – 1.0 mg/dl), and erythrocyte sedimentation rate (ESR) 31/60mm (normal 1-13 mm/hr). A bone scintigram was performed which demonstrated accumulation in all three phases at the region of interest, consistent with acute inflammation. No other foci of involvement were noted (Fig. 2).

**Figure 2**

Figure 2: Bone scintigram (99mTc-DPD) demonstrating acute inflammation at the region of interest of the right proximal tibia with no other foci of involvement and degenerative findings at the left ankle



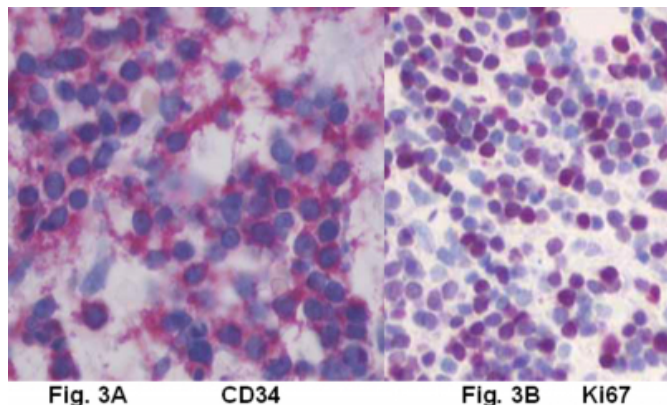
Due to the morphology on the MR images, osteomyelitis of the right proximal tibia was suspected. A debridement with application of local antibiotics was done. No microorganisms were isolated from cultures of the debrided material.

Grossly, the debrided material consisted of multiple fragments measuring 5.5 cm in aggregate. Microscopically, the lesion showed highly concentrated, lymphoid infiltration, areas of necrosis, and foci of bone destruction and remodeling. The lymphoid infiltrations were partially follicular and focally accompanied by foam cells.

Immunohistochemical stains were positive for CD10, CD20, CD34, CD45, CD79a, CD99 (MIC 2), bcl-2, and terminal deoxynucleotidyl transferase (TdT). High proliferation activity (greater than 95%) was demonstrated with Ki67 (Fig. 3). Based on these results, the diagnosis of precursor B-lymphoblastic lymphoma was made.

### Figure 3

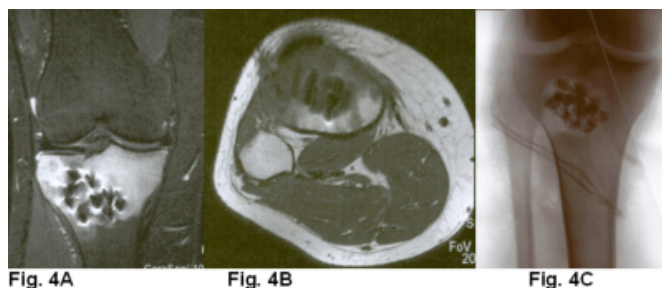
Figure 3A-B: histology showing positive immunohistochemical stains for CD 34 and Ki 67



During the staging procedure, an extensive evaluation was undertaken, including bone marrow aspiration and cerebrospinal fluid (CSF) evaluation. Chest and abdominal computed tomography (CT) studies showed single enlarged lymph nodes in the left cranial inguina measuring up to 2.0 x 0.9 cm and, slightly more caudal on the right side, measuring up to 2.3 x 0.8 cm. No other evidence of lymphadenopathy or other focal lesion was found. A post-operative positron emission tomography (PET) one month later showed no evidence of lymphomatous involvement beyond the vicinity of the right proximal tibia. The postoperative MRI and plain film at 2 months did not show any involvement of soft tissue (Fig. 4).

### Figure 4

Figure 4A-C: MRI; coronal T2 (TR 4540 TE 26ms), transversal T1 (TR 541 TE 15ms) with extensively diminished signal due to metal artifact and x-ray AP showing postoperative status of proximal right tibia without any involvement of soft tissue

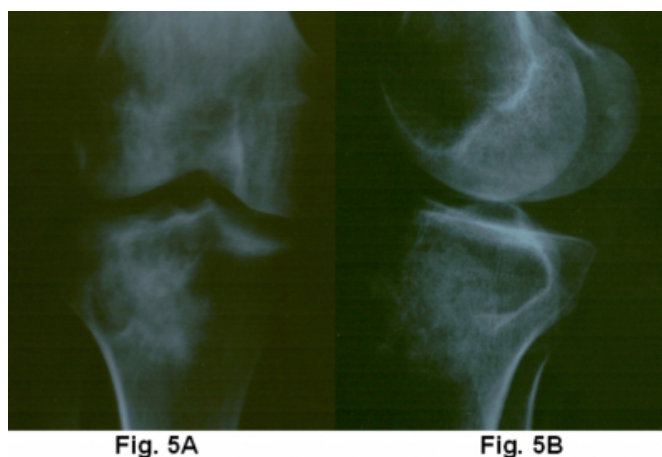


Three months later, 6 cycles of adjuvant VACOP-B-chemotherapy were completed. This was followed by involved field radiation (40-45 Gy) of the right proximal tibia.

Finally, in February 2003 (6 months post-operatively), the filling of the defect with spongiosa from the left dorsal iliac crest could be performed. Orthopaedic follow-ups at seven and nine months showed a patient without complaints or clinical findings. On radiographic studies at both times, constant stable bone with zones of sclerosis was found (Fig. 5A-B).

### Figure 5

Figure 5A-B: x-ray; AP and lateral of proximal right tibia presenting constant stable bone with zones of sclerosis two month after the filling of the postoperative defect with spongiosa from the left dorsal iliac crest



## DISCUSSION

B-LBL presenting as a solitary bone tumor is very uncommon. While lymphoblastic lymphoma (LL) comprises less than 5% of adult non-Hodgkin lymphomas in the United

States, approximately 90% of these cases are the more common T-cell type. [2,3]. In one series of 25 cases of B-LBL, 5 (20%) of the patients had bone lesions and, of those, only 2 patients (8%) had solitary bone involvement [3].

There have been only three reported cases of B-LBL involving only the tibia [6]. The criteria for a diagnosis of primary lymphoma of bone has been established by Guidici et al: (a) on presentation, the patient demonstrates a primary focus in a single bone; (b) histological proof is obtained from the skeletal focus (not from a metastasis); (c) the onset of symptoms of the primary skeletal lesion precedes the appearance of distant metastases by at least 6 months [1]. These criteria were fulfilled in this case.

Patients with solitary or multifocal B-LBL generally present with localized bone pain. [2,3,6]. This finding is similar to patients with any primary lymphoma of bone (PLB), where the presenting feature is bone pain in 60-100% of patients [4,5]. The mean age of patients with B-LBL is 20 years [3], which is significantly older than the three patients with involvement of the tibia only whose mean age was 6.3 years [6]. B-LBL and PLB can, as in this case, present with a relatively long clinical prodrome, from months to over one year, and an initially benign radiographic appearance [1,6].

Patients with B-LBL and PLB have a widely varied clinical presentation. Cases have been reported as mimicking chondroblastoma [1], chronic synovitis [8], monoarthritis [7] Ewing's sarcoma [6], fibrous dysplasia [3], and osteomyelitis[2]. Although many imaging tests are usually done when attempting to diagnose a lesion of this type, they may be non-contributory. Plain radiographs and CT scan are usually non-specific and demonstrate a lytic or sclerotic bone lesion [2,6,8]. Early use of MRI may be helpful, and will display high signal intensity on T2-weighted images. In most cases of this type, systemic symptoms are usually absent and there are no clinical signs pointing to malignant lymphoma [8]. Immunophenotypically, the neoplastic cells of B-LBL express TdT and B-cell antigens such as CD10, CD19, CD22, and CD79a [6]. The combination of TdT, CD79a, and CD10 positivity is diagnostic of B-LBL [2].

Indications for operative involvement by orthopedic surgeons in bone lymphoma most commonly fall into 1 of 3 categories: diagnosis, treatment of pathologic or impending pathologic fractures, and decompression of spinal canal compromise. At this time, no clear role for surgical

debulking procedures or resection exists for PLB. In cases where PLB is suspected, the surgeon must obtain fresh tissue for immunophenotyping, cytogenetic, and molecular studies, where indicated [4].

Patients with solitary, primary B-LBL of bone appear to respond well to aggressive chemotherapy [3], so it is essential to diagnose this lesion prior to leukemic spread or metastasis. The three patients with solitary B-LBL of the tibia all received a high-dose multiagent chemotherapy and were disease free at more than 1, 11, and 12 years, respectively [6]. Thus, in a patient with osteomyelitis-like symptoms and bone pain refractory to medical treatment, a thorough diagnostic work-up should be undertaken and unusual neoplasms such as this hematopoietic malignancy should be considered in the differential diagnosis.

## CORRESPONDENCE TO

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