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Primary periosteal lymphoma—rare and unusual

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periosteum without affecting the adjacent medulla or the regional lymph nodes. No other lymphomatous foci were found in either the distant lymph nodes or viscera. This unusual presentation simulates the imaging appearance of surface lesions of bone, namely benign and malignant tumors, and departs from the typical appearance of primary lymphoma of bone. Therefore, this rare type of lymphoma should be considered in the differential diagnosis of surface bone lesions.

Keywords Lymphoma · Periosteum · Femur · MRI · Radiographs

Abstract We describe a primary periosteal lymphoma that involved only the

Introduction

Although lymphomas are considered to be tumors of lymph nodes, they may arise from other tissues. It was not until 1939 that Parker and Jackson described 17 cases of “primary reticulum cell sarcoma of bone” and established primary lymphoma of bone (PLB) as a clinical entity distinct from other round cell tumors, particularly Ewing’s sarcoma [1]. It is usually a large B-cell tumor [2, 3]. The peak incidence is in the fifth decade with a slight male preponderance. The presenting symptoms usually consist of localized bone pain (most commonly in a long bone) and occasionally a palpable mass. The vast majority of patients present with a localized disease and without systemic symptoms [4]. In most instances, the primary site of involvement is the appendicular skeleton predominating the lower extremity (i.e., distal femur, proximal tibia).

Radiographs typically demonstrate an aggressive pattern of lytic bone destruction with permeation or “moth-eaten” appearance and soft-tissue mass. The tumor is characterized by minimal cortical changes with an accompanying soft-tissue mass [5–7]. In intramedullary PLB, T1-weighted signal intensity ranges from isointense to hyperintense whereas T2-weighted characteristics of the tumor vary between hypointense, isointense, and hyperintense without correlation to intralesional fibrosis, maturity of fibrosis or intralesional vascularity. It is, therefore, not a simple reflection of the histologic findings of intralesional vascularity or fibrosis [7–9].

In the case presented, the tumor was excavating the superficial part of the periosteum without penetrating the medulla, simulating a surface tumor of bone that lacked mineralization.

Case history

A 27-year-old man experienced right knee pain after a mild trauma for which he did not receive treatment. Three months later, he again developed intermittent pain in the same knee, worse than the previous pain, and he sought orthopedic evaluation. According to the patient, MR imaging was done and was negative. Six months later, he had acute pain in the right knee, which prompted performing radiographs and MR imaging. Family history indicated that his elder brother had died from lymphoma a few years earlier. Physical examination of the right lower extremity revealed a subtle fullness in the lateral part of the distal shaft of femur with full range of motion of the right knee. There was no joint effusion or laxity of the knee, and the neurovascular examination was unremarkable. There was no lymph adenopathy or organomegaly. Radiography revealed a superficial erosion of the lateral cortex of the distal femur with ill-defined periosteal bone reaction (Fig. 1). Bone scan showed increased uptake at the site of the cortical erosion. MR imaging demonstrated a subperiosteal soft-tissue mass abutting the erosion with the medulla of the distal femur having normal signal intensities (Figs. 2, 3 and 4). The patient underwent an open biopsy. The histopathology showed morphology diagnostic of large-cell lymphoma (Figs. 5 and 6). The immunohistochemical stain for leukocyte common antigen (LCA) showed diffuse staining of all the cells (Fig. 7). The



Fig. 1 A–P view of the right knee reveals a focal superficial erosion in the lateral cortical margin of the metadiaphysis of the right femur (arrows). Note the ill-defined periosteal reaction



Fig. 2 A coronal T1-weighted image of the distal femur demonstrates a sharply defined elliptical soft-tissue periosteal mass, well outlined by the displaced fat (arrows). The mass is abutting the cortical saucerization and has an isointense signal to the muscles

diagnosis was periosteal large-cell lymphoma, B-cell type. The patient was scheduled for radiation treatment. Unfortunately we lost track of the patient, as he did not report to either the orthopedic or radiation therapy department.

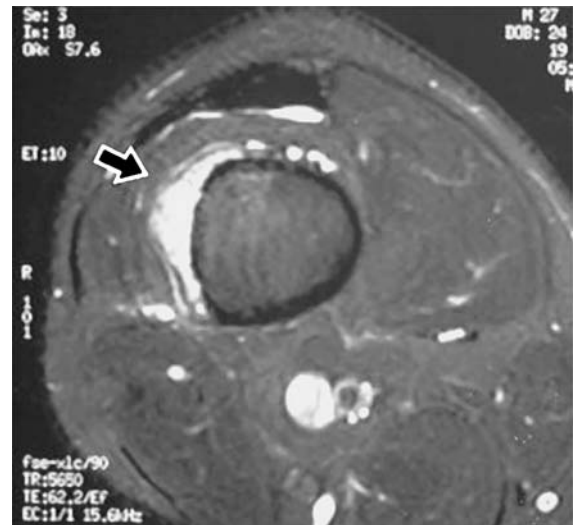


Fig. 3 An axial FSE fat-suppressed T2-weighted image reveals a sharply defined crescent-like periosteal soft-tissue mass abutting the cortical excavation. The mass has a homogeneous high signal intensity (arrow). The medulla has normal signal intensity

Fig. 4a,b Enhanced fat-suppressed T1-weighted image.

a Axial. **b** Coronal. Both cuts show enhancement of the periosteal mass with intralesional, multiple tiny areas of nonenhancement most likely representing infiltrating periosteal fibrous tissue. The medulla is not enhanced



Discussion

PLB accounts for 3% of primary bone tumors [10]. It is a rare disease that accounts for less than 2% of all lymphomas in adults [11] and 3–5% of all extranodal lymphomas [12]. In the pediatric population, it represents approximately 3–9% of lymphoma cases [13].

PLB commonly extends from the medulla through the cortex causing soft-tissue mass and periosteal bone formation [6, 7]. However, lymphoma strictly limited to the periosteum without involving the medulla is unusual and rare. Our case focally involved the metadiaphysis of

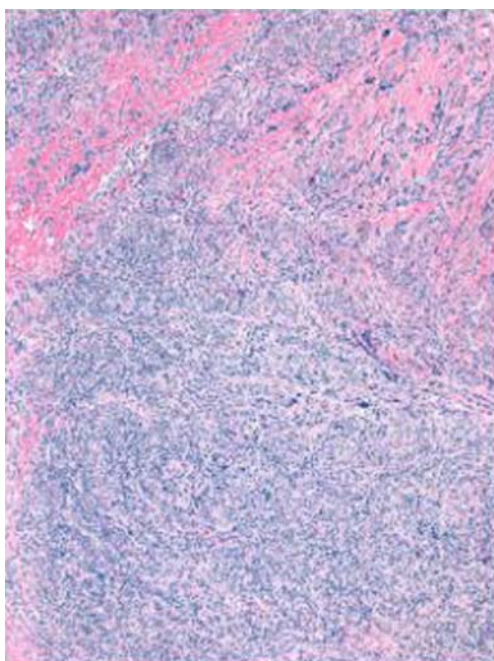


Fig. 5 Low-power view showing cellular tumor composed of cells with little cytoplasm with infiltrating periosteal fibrous tissue (*top*). The tumor infiltrates as sheets and individual tumor cells

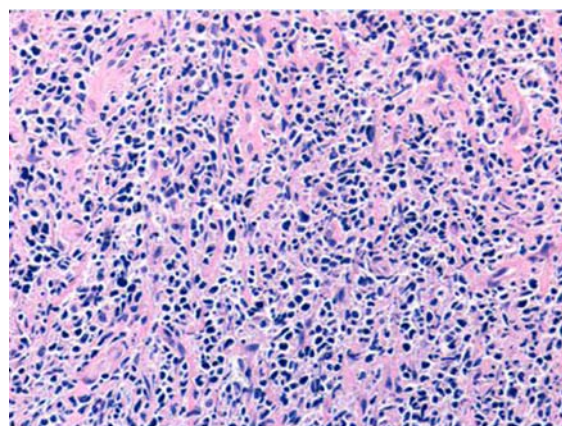


Fig. 6 Most tumor cells are larger than plump endothelial cells of tumor vessels and are admixed with smaller cells with twisted/cleaved lymphoid nuclei and small lymphatic cells. The morphology is diagnostic of large-cell lymphoma

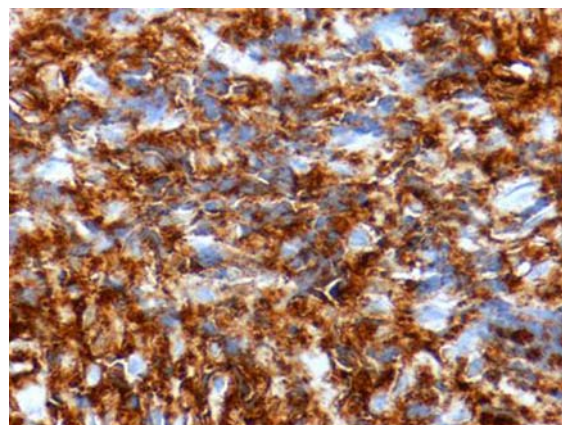


Fig. 7 Immunohistochemical stain for leukocyte common antigen (LCA) shows diffuse staining of all cells

the distal shaft of the femur causing superficial cortical erosion and ill-defined periosteal reaction. The malignant infiltrative process did not involve the adjacent medulla.

There is no typifying appearance of MR imaging of bone lymphoma, which ranges from hypointense to isointense signal on T1-weighted images, and mainly heterogeneous hypointense signal on T2-weighted images, with fewer cases showing either isointense or hyperintense signal on T2-weighted images [6, 8]. Our case showed an isointense signal on T1-weighted images and homogeneous hyperintense signal on T2-weighted images (Figs. 2 and 3). Although the tumor enhanced with contrast (Fig. 4), the enhancement was not homogeneous, with intralesional tiny areas of nonenhancement, which may have been due to infiltration of the cellular tumor by periosteal fibrous tissue (Fig. 5). The medulla showed normal signal intensity on T1- and T2-weighted images and did not enhance with contrast.

Although primary periosteal lymphoma was not mentioned as an entity in the literature in the large series of PLB, a recent report appeared in the literature describing a periosteal lymphoma of the distal femur [14]. Our case shared with the reported case the site of the lesion and the extent of the soft-tissue mass. However, the patient was younger, a 20-year-old man, the periosteal reaction was more evident, and MRI signals were slightly hyperintense on T1- and T2-weighted images.

The histopathology of the open biopsy in our case was typical for primary periosteal lymphoma, diffuse large B-cell type. On the other hand, the radiologic diagnosis is difficult as this tumor is extremely rare, and it also simulates other surface lesions of bone exhibiting sub-

periosteal mass. The notable absence of matrix mineralization in primary periosteal lymphoma helps to distinguish the tumor from surface tumors with matrix mineralization, such as periosteal chondrosarcoma, osteosarcoma, chondroma, and osteblastoma [15–18], but can not help separate primary periosteal lymphoma from periosteal masses not exhibiting matrix mineralization such as periosteal ganglion [19], hemangioma [20], solid aneurysmal bone cyst [21, 22], cortical metastases causing external excavation of the cortex [23], and periosteal Ewing's sarcoma [24, 25].

The radiologic appearance of our case simulates to a great extent periosteal Ewing's sarcoma. However, the age of our patient, 27 years, is far older than the mean age of periosteal Ewing's sarcoma, which is 14 years [24].

The treatment of PLB is either by radiation therapy, chemotherapy or combined modality therapy. A study of the long-term results of combined modality therapy in PLB was performed in Massachusetts General Hospital. Comparing the data of the study with previous experience in patients treated with local measures only, the authors concluded that patients treated with combined radiation therapy and chemotherapy had a favorable outcome, which appeared superior to radiation therapy alone [26]. The same conclusion came from a similar study performed in M.D. Anderson Cancer Center [11].

As malignant bone tumors limited to the periosteum, such as periosteal osteosarcoma, chondrosarcoma, and Ewing's sarcoma, have far better prognosis than their medullary counterparts, we predict that primary periosteal lymphoma may prove to have a better prognosis than the typical PLB.

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