Primary Musculoskeletal Lymphoma

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KEYWORDS

• Extranodal lymphoma • Primary bone lymphoma • B-cell lymphoma
• Primary soft tissue lymphoma

KEY POINTS

- Primary musculoskeletal lymphoma is rare, almost invariably B cell, but often reveals imaging characteristics suggestive of the diagnosis.
- Primary osseous lymphoma shows aggressive bone destruction with soft tissue involvement, and intervening cortical destruction may be subtle.
- Primary soft tissue lymphoma typically demonstrates long cones of involvement, which may be multiple and intramuscular or intermuscular in location by MR imaging.
- PET with fluorodeoxyglucose fluorine 18 imaging reveals marked increased radionuclide uptake in areas of musculoskeletal lymphomatous involvement and is useful to evaluate disease extent, treatment effect, and recurrence.
- Initial biopsy of musculoskeletal lymphoma may suggest nondiagnostic tissue owing to discohesive and collapsed cells. However, immunohistochemical stains often remain positive, if performed.

MUSCULOSKELETAL LYMPHOMA

Introduction

Primary lymphoma of the musculoskeletal system is rare. Involvement may include the bone, subcutaneous tissue as well as muscle. Regional lymph nodes may also be affected. Imaging of primary musculoskeletal lymphoma often reveals suggestive intrinsic and morphologic features of this disease. Biopsy confirmation is vital to differentiate musculoskeletal lymphoma from sarcoma because treatment dramatically differs between these diseases. Musculoskeletal lymphoma is treated medically with overall excellent response as opposed to surgical treatment of sarcoma often supplemented with adjuvant chemotherapy and radiation therapy.

LYMPHOMA OF BONE

Primary lymphoma of bone accounts for less than 5% of malignant bone tumors.[1–3] The vast majority of cases are non-Hodgkin lymphoma. Primary Hodgkin lymphoma of the musculoskeletal system is exceedingly rare. Imaging features of skeletal lymphoma may be nonspecific and similar to other small round blue cell tumors. However, intrinsic characteristics and the morphologic appearance may reveal suggestive features.

Primary lymphoma of bone shows a wide age range of distribution with patients most commonly affected in the third and sixth through eighth decades.[1–4] There is a male predilection, with a large series by Mulligan and Kransdorf[1] showing a 1:8:1 ratio.[2,4] Long bones (71%) are affected more commonly than flat bones (22%).[1,2,4] The most commonly involved sites include the femur (33%), tibia (20%), humerus (13%), pelvis (11%), scapula (4%), clavicle (3%), ribs (3%), vertebral column (5%), foot (2%), radius (1%), and the patella (1%).[1,2,4] The specific most frequent locations are the distal femoral metadiaphysis (16%),

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proximal metaphysis of the tibia (12%), and the femoral midshaft (8%).[1,2,4] Ephiphyseal centered lesions accounted for only 5% of cases in the series by Mulligan and Kransdorf.[1,2,4,5]

The clinical presentation of primary osseous lymphoma is frequently insidious with intermittent pain persisting for several months. Additional symptoms include swelling, palpable mass, and systemic signs, such as weight loss and fever. Vertebral lesions can result in neurologic symptoms owing to paraspinal extension.

The radiographic appearance of primary musculoskeletal lymphoma is most commonly aggressive bone destruction seen in 70% of cases[1,2,4,6] (**Figs. 1–3**). The pattern of bone lysis

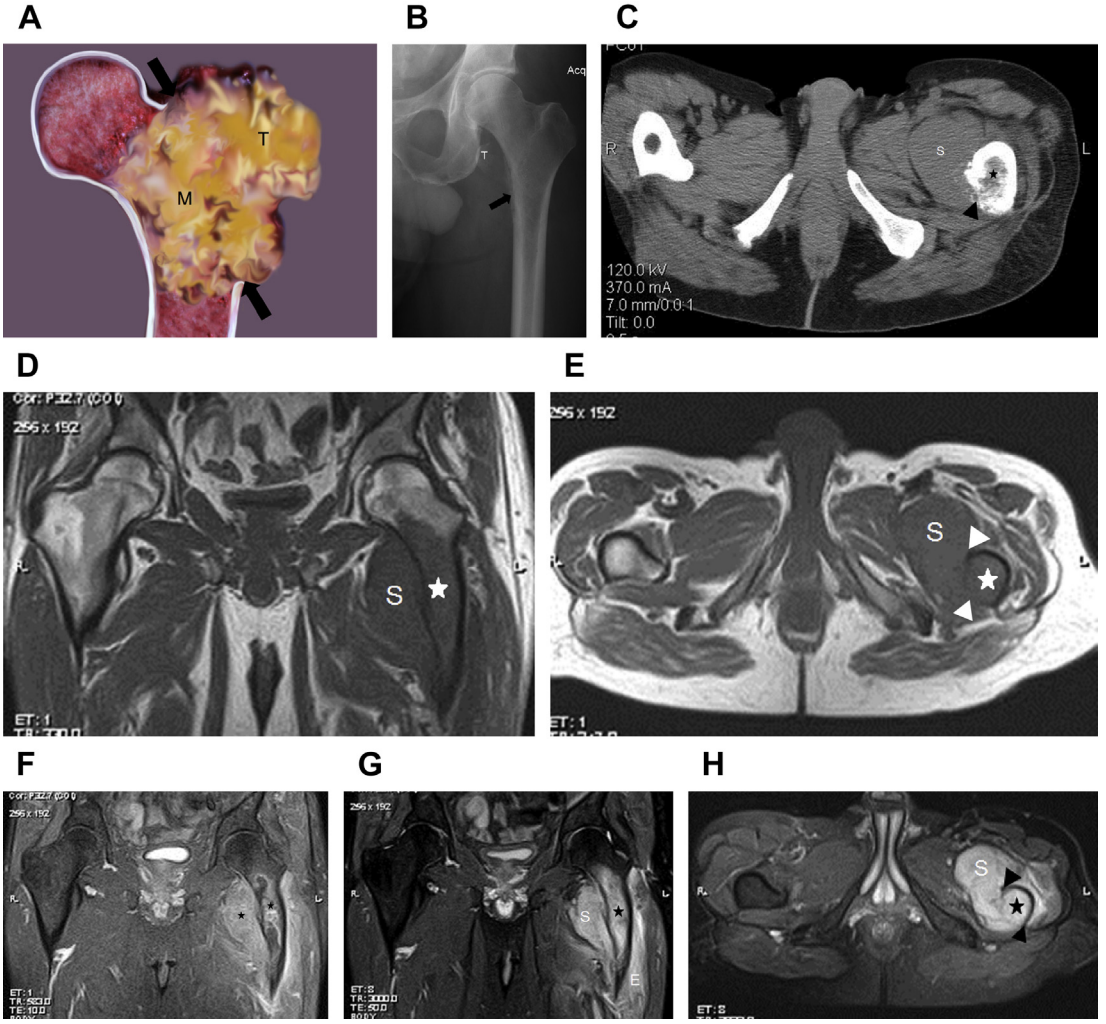

Fig. 1. Lymphoma of the proximal femur with pathologic fracture and associated soft tissue mass. (*A*) Pictorial representation of intramedullary lymphoma (M) with cortical destruction (*arrows*) and direct extensive into the soft tissue (T). (*B*) Frontal radiograph of the right hip shows aggressive bone destruction with cortical permeation medially (*arrow*) and avulsion of the lesser trochanter (T). (*C*) Axial CT demonstrates marrow involvement (*star*) with cortical destruction (*arrowhead*) and soft tissue extension (S). The soft tissue component has similar attenuation to muscle. (*D*, *E*) Coronal and axial T1-weighted (TR = 330; echo time [TE] = 10) MR images show marrow replacement (*star*) and soft tissue mass (S) in direct continuity with intervening cortical destruction (*arrowheads*). (*F*) Postcontrast fat-suppressed T1-weighted (TR = 583; TE = 10) MR image reveals mild diffuse enhancement of both the intraosseous and the soft tissue components (*stars*). (*G*, *H*) Coronal and axial fat-suppressed T2-weighted (TR = 3000; TE = 50) MR images demonstrate similar features with the marrow replacement (*star*) and soft tissue mass (S) and continuity through cortical destruction (*arrowheads*). The signal of the lymphomatous involvement is high intensity. There is surrounding soft tissue edema (E). ([*A*] *From* Murphey MD, Senchak LT, Mambalam PK, et al. From the radiologic pathology archives: Ewing sarcoma family of tumors: radiologic-pathologic correlation. Radiographics 2013;33(3):803–31; with permission.)