Musculoskeletal Lymphoma: MRI findings of cases presenting as a bone lesion or soft tissue mass

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Learning Objectives

To assess the MR imaging findings of primary musculoskeletal lymphoma presenting clinically as either a bone lesion or a soft tissue mass.
Background

Lymphoma is a heterogeneous disease that can involve almost all types of extranodal tissue [1]. Although 20-30% of all non-Hodgkin's lymphoma manifests extranodally [2], primary involvement of the musculoskeletal system is rare.

Primary lymphoma of bone (PLB) accounts for less than 5% of primary bone tumours [3,4]. The vast majority of these cases are of the non-Hodgkin's type. Coley et al. outlined the diagnostic criteria of PLB in 1950 as a tumour presenting in an osseous site and involving bone marrow, with metastases only to regional lymph nodes, or with no evidence of disease elsewhere for at least 6 months. When multiple bones are involved without evidence of visceral or distant nodal disease, it is recognised as a subset of PLB known as primary multifocal osseous lymphoma (PMOL).

Involvement of the soft tissues by lymphoma is most often seen as part of a disseminated disease process or as contiguous extension from adjacent nodal or osseous deposits [2]. The least commonly encountered form is from an extranodal primary origin [6], with primary lymphoma of muscle (PLM) representing only 1.5% of cases of NHL and 0.3% of Hodgkin's lymphoma [7].

This is a retrospective review of the MR imaging of 23 cases of histologically confirmed primary musculoskeletal osseous and/or soft tissue lymphoma. The cases were predominantly tertiary referrals to an orthopaedic oncology unit that were presented at a weekly Statewide sarcoma multidisciplinary meeting.
Imaging Findings OR Procedure Details

PRIMARY LYMPHOMA OF BONE

1. Area of marrow replacement, usually poorly demarcated
2. Hypointense relative to normal marrow on T1
3. T2 hyperintense, may have a "mosaic" pattern
4. Diffuse contrast enhancement
5. May have minimal cortical disruption
6. May have an extraskeletal soft tissue component, either as a periosseous "cuff" or a mass

Osseous lymphoma manifests as an area of marrow replacement on MRI. This was almost invariably poorly demarcated in our study group (12 out of 13 patients, 92%). T1-weighted images showed lesions to be of low signal intensity (SI) relative to adjacent normal marrow (Fig. 1 on page 8).

The T2-weighted signal characteristics, however, were variable. The majority of osseous tumours were of heterogeneous SI, with 54% (7 out of 13) demonstrating what we have termed a "mosaic" pattern of marrow replacement. Four of the 7 cases had a mosaic pattern that was predominantly T2 hyperintense (Fig. 2 on page 8, Fig. 8 on page 14), with the remainder being of relatively mixed SI (Fig. 4 on page 10, Fig. 5 on page 11). Of the osseous tumours that did not have a mosaic pattern, the T2 SI was either heterogeneously (n=4) or homogeneously (n=2) hyperintense.

It has been proposed that areas of relatively low T2 SI within osseous lymphoma represents intralesional fibrous tissue [8,9]. It follows that areas of higher tumour SI may reflect immature fibrosis due to the presence of cellular elements, vessels and oedema [10]. White et al., however, was unable to demonstrate any correlation between the T2-weighted characteristics and the maturity of fibrosis on histological analysis.

Sampling error is a potential explanation for the controversy surrounding the MR appearance of fibrosis, with most lesions being diagnosed on the basis of a small sample (e.g. core biopsy). We have recently seen one case of lymphoma of the proximal femur
whereupon resection revealed extensive areas of necrosis; another possible explanation for the mosaic T2 pattern (Fig. 6 on page 12, Fig. 7 on page 13).

Post-contrast enhancement of the lymphomatous marrow was diffuse and heterogeneous in 91% of our cases (Fig. 9 on page 15), however, both homogeneous [2] and heterogeneous [12,13] enhancement patterns have been reported.

Extraosseous soft tissue components are commonly seen in patients with PLB and can occur in the absence of appreciable cortical destruction. It has been stated that the presence of intramedullary tumour with a "normal" appearing cortex and a substantial extraosseous soft tissue mass is likely to be lymphoma [3,4]. This feature has been attributed to tumour-mediated osteoclastic resorption and formation of cortical tunnels [14]. The MR imaging correlate is a permeative pattern of linear foci of intermediate to high T2 SI penetrating cortical bone. This pattern, however, is not specific to lymphoma and may be seen in other small round cell tumours, osteomyelitis, myeloma or metastases [15].

In our study, cortical abnormalities were present in 92% of cases of osseous lymphoma with 83% (10 out of 12) showing a permeative component to the cortical disruption (Fig. 10 on page 16). Complete cortical destruction was evident in 50% (Fig. 11 on page 17, Fig. 14 on page 20), and the vast majority of these cases had an associated extraosseous mass (Fig. 14 on page 20).

A "cuff" of periosseous soft tissue is commonly seen in PLB (6 out of 8 of our cases, 75%; mean depth 5mm). This soft tissue cuff was invariably isointense or iso- to slightly hyperintense on T1-weighted images and hyperintense on T2-weighted images. Periosseous soft tissue oedema was present in all of our cases. Enhancement of the soft tissue cuff was demonstrated in all cases where gadolinium was administered, helping distinguish extraosseous tumour from periosseous oedema (Fig. 10 on page 16).

Five of our 13 cases had lymphoma that probably arose in bone but also had a significant associated soft tissue mass. The mean volume of extraosseous tumour was 770cm³ (range 100-1497cm³). All 5 patients in this subgroup had similar signal characteristics of the soft tissue component, being iso- to slightly hyperintense on T1-weighted images and hyperintense on T2-weighted images. Enhancement of the soft tissue component was heterogeneous and diffuse in 4 patients (80%), and homogeneous in the fifth patient.

All cases with a soft tissue mass demonstrated either partial or complete encasement of adjacent periosseous structures (Fig. 12 on page 18). Intramuscular fat planes were preserved in two cases (Fig. 16 on page 22). Involvement of more than one anatomical compartment by tumour was seen in 4 patients (Fig. 16 on page 22, Fig. 17 on page...
Subcutaneous oedema was present in the same 4 cases, although one patient had deep venous thrombosis within a regional nodal mass proximal to the tumour that probably accounted for much of the oedema (Fig. 15 on page 21). Involvement of the adjacent skin was not a feature evident in this subgroup of patients, but has been reported in other studies [6].

PRIMARY SOFT TISSUE LYMPHOMA

1. Variable morphology, may be lobular or poorly defined
2. Iso- to slightly hyperintense to muscle on T1
3. T2 hyperintense
4. Diffuse contrast enhancement
5. Often associated with subcutaneous stranding
6. May involve more than one anatomical compartment
7. May encase adjacent vessels, nerves or tendons
8. Regional lymphadenopathy

Lymphoma primarily arising in skeletal muscle is very rare. It usually occurs in patients over the age of 60 with the thighs, chest and arms being the most frequently reported sites [9,16], as was reflected in our series. Clinically, patients present with a non-fatty intramuscular mass, whereby sarcoma is the most commonly suspected diagnosis [12]. Morphologically, however, lymphoma manifests as an infiltrative process rather than as a compartmental condition such as sarcoma. Involvement of multiple muscle compartments and encasement of adjacent vessels, nerves or tendons are features that have been associated with lymphomatous involvement of the soft tissues in other studies [12,17].

Of the 7 patients in our study with primary soft tissue lymphoma, 4 had well-defined lobular lesion morphology, while the tumour margins were poorly defined in 3 cases. Tumour SI was iso- to slightly hyperintense relative to skeletal muscle on T1-weighted imaging in all cases. All cases were hyperintense on T2-weighted imaging, being of heterogeneous SI in two cases (Fig. 20 on page 26). Peritumoural oedema was present in 4 cases (57%). Homogeneous enhancement was present in 5 cases following adminstration of intravenous contrast (71%).
In the 5 cases that were not limited to the subcutaneous fat, there was multicompartment involvement and tumour encasement of adjacent structures in all cases. Tumour completely surrounded the vessels in two cases, and partially encased neurovascular structures in three (Fig. 18 on page 24, Fig. 19 on page 25). The two cases of subcutaneous lymphoma presented as well-defined epitrochlear masses (Fig. 21 on page 27). Only one patient had skin involvement, clinically manifesting as a discharging mass. Regional lymphadenopathy was present in all but one patient (Fig. 24 on page 30).

MULTIFOCAL MUSCULOSKELETAL LYMPHOMA

Primary multifocal osseous lymphoma is a rare subset of PLB in which there are lymphomatous deposits in multiple bones without evidence of visceral or distant nodal disease for at least 6 months following presentation [18]. Clinical manifestations are similar to those of PLB with most lesions occurring around the knee [18]. PMOL tends to involve the vertebrae more often than PLB [3], as seen in our two cases. The femur, pelvis, sternum and clavicle were the other sites of involvement. Tumour deposits were of similar signal intensity those with single site osseous disease, i.e. T1 hypointensity and T2 hyperintensity relative to normal marrow (Fig. 26 on page 32, Fig. 27 on page 33). Regional lymphadenopathy was present in both patients.

The third patient with multifocal disease presented with an elbow lump, seen as a poorly defined subcutaneous mass on MR imaging (Fig. 28 on page 34). PET revealed multiple FDG-avid lesions within the subcutaneous tissues of both lower limbs (Fig. 29 on page 35), without evidence of visceral or distant nodal disease.
Fig. 1: 34-year-old male with NHL of the proximal tibia. Axial T1-weighted image shows homogeneous low signal intensity relative to normal marrow.
Fig. 2: Same patient as Figs. 1 & 3. Coronal fat-suppressed T2-weighted image reveals a predominantly hyperintense mosaic pattern outlined by serpiginous rims of higher SI.
**Fig. 3:** Same patient as Figs. 1 & 2. Coronal fat-suppressed T1 post-contrast image shows geographic non-enhancing areas with avid serpiginous rim enhancement superimposed on a background of diffuse marrow enhancement.
Fig. 4: 25-year-old female with NHL of the proximal humerus. Coronal fat-suppressed T2-weighted image shows a mosaic pattern of marrow replacement of mixed signal intensities.
Fig. 5: Same patient as Fig. 4. Axial fat-suppressed PD-weighted image reveals focal destruction of the posterior cortex with a narrow cuff of periosseous soft tissue, perilesional soft tissue oedema and a small glenohumeral joint effusion.
**Fig. 6:** 52-year-old male with a pathological fracture of the femoral neck. Coronal fat-suppressed T2-weighted image shows heterogeneous marrow replacement, predominantly hyperintense.
Fig. 7: Macroscopic specimen of the femoral head from the patient in Fig. 6. Histology revealed areas of necrosis and a lymphocytic infiltrate within oedematous marrow. There were degenerative/reparative changes related to fracture but no significant fibrosis was seen. FISH confirmed follicular B-cell NHL.
**Fig. 8:** 18-year-old male with NHL of the distal tibia. Coronal fat-suppressed T2-weighted image shows a predominantly hyperintense mosaic pattern of marrow replacement.
Fig. 9: Same patient as Figs. 8 & 10. Coronal fat-suppressed T1 post-contrast image shows heterogeneous marrow enhancement and a small amount of enhancing extraosseous soft tissue.
**Fig. 10:** Same patient as Figs. 8 & 9. Axial fat-suppressed T1 post-contrast image shows a permeative cortical pattern with focal destruction posteromedially and a heterogeneously enhancing cuff of extraosseous soft tissue.
Fig. 11: 74-year-old male with NHL of the calcaneus. Sagittal T1-weighted image shows predominantly hypointense marrow replacement with complete cortical destruction and tumour extension into the subtalar joint.
Fig. 12: Same patient as Figs. 11 & 13. Axial fat-suppressed T2-weighted image reveals a mosaic pattern of marrow replacement and a large homogeneously hyperintense extraosseous component, encasing the medial and lateral tendon groups.
Fig. 13: Same patient as Figs. 11 & 12. Axial fat-suppressed T1 post-contrast image shows diffuse heterogeneous enhancement of both the osseous and extraosseous components.
Fig. 14: 83-year-old female who presented with a rapidly enlarging thigh mass. Axial T1 weighted image shows complete cortical destruction and a large extraosseous component.
**Fig. 15:** Same patient as Fig. 14. Coronal STIR image depicts a long segment of heterogeneous marrow replacement within the distal femur with multicompartmental soft tissue involvement. There is an ipsilateral inguinal/iliac nodal mass. Extensive subcutaneous and skin oedema is probably the result of external iliac vein thrombosis (not shown), although lymphomatous infiltration could also be present.

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Fig. 16: 81-year-old female with NHL of the proximal calf. Axial fat-suppressed T2-weighted image shows a mixed mosaic pattern of marrow replacement. There is focal cortical destruction anterolaterally and involvement of the proximal tibiofibular joint and adjacent fibula. An infiltrating soft tissue component involves the anterior, lateral and deep posterior compartments of the calf, with preservation of intramuscular fat planes. The anterior tibial artery was partially encased by tumour (not shown).
**Fig. 17:** 60-year-old female with NHL of the arm. Coronal fat-suppressed T1 post-contrast image shows abnormal heterogeneous marrow enhancement within the proximal humeral shaft and a large, heterogeneously enhancing soft tissue component involving multiple compartments. There is axillary lymphadenopathy.
Fig. 18: Same patient as Fig. 17. Axial fat-suppressed T1 post-contrast image shows tumour encasement of the long head of biceps tendon and brachial artery, with soft tissue oedema and pathological axillary lymph nodes.
Fig. 19: 60-year-old male with NHL of the chest wall. Axial STIR image shows a lobular, heterogeneously hyperintense mass partially encasing the brachial plexus.
**Fig. 20:** Same patient as Fig. 19. Coronal STIR image shows a heterogeneous lobular mass extending into the axilla.

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Fig. 21: 68-year-old male with Hodgkin’s lymphoma. Axial T1-weighted image shows a well-defined subcutaneous lesion of homogeneous signal intensity.
Fig. 22: Same patient as Fig. 21. Axial fat-suppressed T2-weighted image. Staging revealed an ipsilateral axillary lymph node that was considered pathological.
Fig. 23: 56-year-old female presenting with an anterior thigh mass. Axial fat-suppressed T2-weighted image shows a poorly defined subcutaneous mass with adjacent oedema, extending through the fascia to partially encase the superficial femoral vessels and involve the anterior margin of vastus intermedius.
Fig. 24: Same patient as Fig. 23. Sagittal fat-suppressed T1 post-contrast image reveals inguinal and internal iliac lymphadenopathy. Marginal zone NHL was diagnosed following ultrasound-guided core biopsy of the thigh mass.
Fig. 25: Same patient as Figs. 23 & 24. Axial CT image from a subsequent FDG-PET/CT shows one of the two pleural-based nodules, presumed to be secondary lymphomatous deposits.
Fig. 26: 87-year-old male presenting with anterior chest pain and swelling. Sagittal T1-weighted image shows marrow replacement within the sternum and the C3 vertebral body (top).
Fig. 27: Same patient as Fig. 26. Coronal T2-weighted images shows marrow replacement within the sternal manubrium, sternal body and medial left clavicle.
Fig. 28: 58-year-old male presenting with a mass near the elbow. Axial fat-suppressed T2-weighted image shows a poorly defined subcutaneous mass.
**Fig. 29:** Same patient as Fig. 28. PET reveals multiple FDG-avid subcutaneous deposits. Histology confirmed marginal zone NHL.
Conclusion

The MRI features of primary lymphoma of bone include heterogeneity on T2-weighted imaging (often as a mosaic pattern), a periosseous cuff of soft tissue or a more substantial soft tissue mass and cortical disruption that is often disproportionate to the extent of extraosseous tumour.

Features often seen in cases of soft tissue lymphoma include relative homogeneity on T1- and T2-weighted imaging, multicompartiment involvement, encasement of neurovascular structures and subcutaneous stranding.
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References
