

Case Study

A Rare Case Report: Diagnostic Challenge of Uncommon Site of Lymphoma Camouflaging as Cellulitis

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A B S T R A C T

Lymphomas are solid tumours of the immune system accounting for 14% of all head and neck malignancies. Non-Hodgkin lymphomas (NHL) are a heterogeneous group of lymphoproliferative disorders originating in B cells, T cells or natural killer T-cells. They have a vast array of clinical presentations and histological findings which can make diagnosis difficult. Lymph nodes, spleen, and liver are common sites of NHL. Extranodal soft tissue manifestation of NHL is a rare presentation of all lymphomas with a common occurrence in the geriatric population. Prevalent NHL is diffuse large B cell lymphoma which has significant clinical, morphological, and molecular heterogeneity leading to difficulty in timely diagnosis and treatment, and usually denotes an advanced stage. Here we throw light on primary skeletal muscle lymphoma-diffuse large B cell type, in an 81-year-old female patient mimicking cellulitis. It is vital to differentiate lymphoma from other entities to avoid perplexity in management.

Keywords: Primary Skeletal Lymphoma, Extranodal, Diffuse Large B Cell Lymphoma

Introduction

Lymphomas are a heterogeneous group of malignancies that present themselves as enlarged non-tender lymph nodes but may involve extranodal regions, commonly involving the gastrointestinal tract and head and neck. Extranodal presentation of non-Hodgkin's lymphoma is well recognised. Diffuse large B cell lymphoma (DLBCL) is a malignancy of B cells with a median age of occurrence in the seventh decade.¹ The most common sites of presentation are GI tract, testes, skin, lungs, bones, CNS, and respiratory system. Primary musculoskeletal origin is an unusual presentation accounting for less than 0.1 to 1.4% of all lymphomas,

commonly involving the extremities.²⁻⁴ Muscular lymphoma is most common in the thigh and upper arm muscles.⁴ The clinical presentations of extranodal DLBCL remain very diverse with heterogeneous histological appearances that may infrequently present a diagnostic challenge.⁵⁻⁷ Multimodal approach of pathologic and radiologic investigations may be needed for a definitive diagnosis.

Case History

An 81-year-old female patient presented to our outpatient department on September 2021 with swelling in her left arm for 3 weeks. It was insidious in onset, progressive, diffuse and associated with dull aching pain in the left

arm. She gave a history of intermittent high-grade fever, associated with chills and rigour for 1 month and low back ache for 10 years which was aggravated for the past one month. She is a known diabetic on irregular intake of oral hypoglycaemic agents and gave no history of any substance abuse. Family history was non-contributory. Erythema was not appreciated due to skin colour. The range of movements was restricted due to oedema and tenderness. Peripheral pulses were palpable. Clinically, a differential diagnosis of cellulitis was suspected in view of her poor diabetic status. The patient was admitted to the general ward and started on antibiotics empirically. Blood investigations were within the normal range except for her diabetic profile which was managed appropriately with insulin.

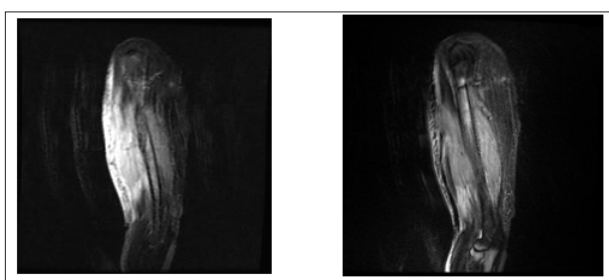


Figure 1. MRI-STIR - Hyper-intensity of Arm Muscles indicating Neoplastic Aetiology

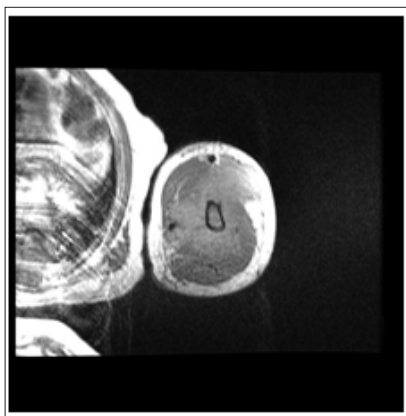


Figure 2. MRI-T1 - Anterolateral Muscle Infiltration

Radiological evaluation was done and diffuse subcutaneous inflammation was noted in the anterolateral muscular plane of the left arm in USG. MRI with CT screening of the left arm showed a large well-defined infiltrative mass lesion of 7 x 4 x 12 cm with isointensity in T1 and hyperintensity in T2 involving the intermuscular and intramuscular compartment of the anterolateral aspect (Figures 1 and 2). Lytic lesion in the 8th rib, adjacent 8th vertebral lamina, and an ill-defined left perirenal mass were reported in MRI. Bilateral and multiple small axillary lymph nodes were noted, with the largest node being 8 mm in the short axis. The diagnosis of lymphoma with skeletal metastases was reached after a diagnostic array that included a conflict between USG, which suggested cellulitis, and MRI with

CT screening. Pathological diagnosis was mandatory for which incisional biopsy was done keeping in mind the oncologic principles. Histopathology showed sheets of large atypical cells with moderate pleomorphism, vesicular nuclei, prominent nucleoli with abundant cytoplasm and frequent mitoses (9/hpf) admixed with lymphocytes (Figure 3). Immunohistochemistry showed positivity for CD20 (Figure 4), CD45 and Ki67 (80%). Based on the above findings, B cell lymphoma - diffuse large B cell lymphoma NOS type.

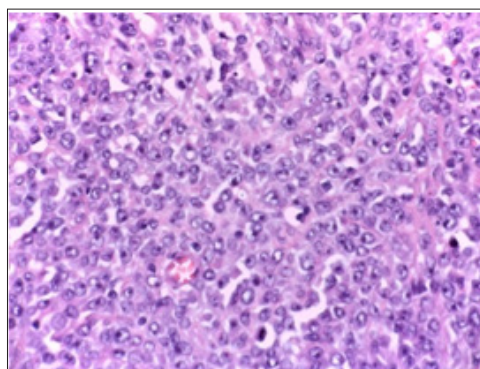


Figure 3. H&E Microscopic Section of Patients' Incisional Biopsy Specimen Stain

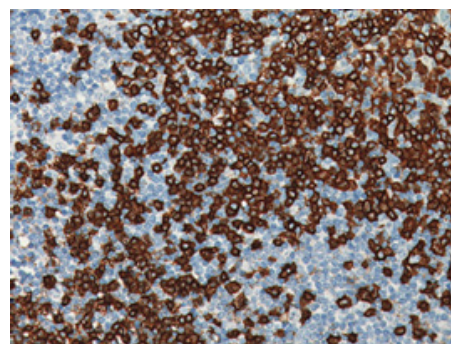


Figure 4. IHC with CD 20 Positivity

The patient was discharged as advised by the tumour board and was started on an R-CHOP regimen (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone). Four cycles were given once a month and PET/ CT imaging was done after four cycles which showed a reduction in tumour volume. The patient was symptomatically better and was followed up for 6 months. She was continued with 2 cycles of chemotherapy when she had a sudden deterioration of her general health with cardiovascular compromise following her sixth cycle of R-CHOP. She succumbed to death on 29th March 2022. Consent was obtained from the attenders for this case report.

Discussion

Extranodal DLBCL with primary skeletal muscle origin is rare which was first reported in the literature in 1984 by Kandel et al.⁷ The occurrence of primary extranodal non-Hodgkin

lymphoma (NHL) of soft tissue is rare, particularly in skeletal muscle.⁸ The hallmark of primary skeletal lymphoma is typically a rapidly growing mass or tissue infiltration with oedema of the affected limb with or without any redness or warmth, along with lymphadenopathy. B symptoms - fever, night sweats, significant weight loss, anorexia, fatigue. and neurological manifestations may be seen. DLBCL of not otherwise specified type is a diagnosis of exclusion and early diagnosis is essential for a better prognosis. It is an aggressive disease with a plunging overall survival rate even after initiating the treatment. Primary skeletal muscles of lower extremities are commonly involved wherein this case report the patient presented to us with upper extremity involvement. Infiltration of the muscle by lymphoma can mimic sarcoma, metastatic carcinoma, melanoma and osteosarcoma causing diagnostic difficulties for which radiological and pathological diagnosis is mandatory.

CT is useful in corroborating the MRI findings of skeletal muscle infiltration, and in addition, can be used for screening metastasis and nodal involvement. T1 and T2 weighted images in MRI constitute the most useful modality for the assessment of muscular lymphoma. The involved muscle shows enlargement, either with a discrete muscular mass or abnormal muscular signal intensity.⁴ PET/ CT serves as a useful tool for staging, and analysing the efficacy of treatment and excluding extranodal disease. Though vast radiological imaging is available for diagnosis, histological evidence is the gold standard for definitive diagnosis and further treatment planning. The primordial treatment option for DLBCL is curative or palliative immunochemotherapy. R-CHOP is the backbone regimen used for early-stage as well as disseminated disease. Further follow-up and the need for radiotherapy can be assessed. Recurrences after a complete response to treatment and relapse are common with insidious onset and aggressive pattern of spread.

Conclusion

This case of DLBCL warrants special handling with further research on prognostic and therapeutic evidence for early diagnosis, treatment and follow-up. Providing curative treatment is the primary goal but further advances are needed for the proper establishment of the disease pathology which will eventually crop up in future, thus reducing the mortality and morbidity rate.

Conflict of Interest: None

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