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Case Report

Primary Skeletal Muscle Lymphoma: A Case Report and Literature Review

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ABSTRACT

Background
Lymphomatous involvement of the muscle is a rarely seen disorder, constituting 0.1-1.4% of all extranodal lymphomas. It may be seen as the intramuscular involvement of disseminated disease, as an extension from adjacent bone or lymph nodes, or as primary extranodal lymphoma. Primary lymphoma of the skeletal muscle mainly consists of B-cell non-Hodgkin lymphoma (NHL) (>95%).

Case Presentation
A 50-year-old male patient presented with progressive swelling and pain in his right flank area over a period of one year. Ultrasound (US) was done, and it showed diffuse enlargement of pelvic and limb muscles with hypoechoic regions that irregularly infiltrated normal muscle tissue. An incisional biopsy was done, and the histopathologic diagnosis was diffuse large B-cell lymphoma (DLBCL).

Conclusion
Primary intramuscular lymphoma is extremely rare. Nevertheless, it should be considered a differential diagnosis in patients presenting with intramuscular tumors.

Keywords
Extranodal lymphoma; Muscle lymphoma; Intramuscular mass; non-Hodgkin lymphoma (NHL).

INTRODUCTION

Lymphoma is a heterogeneous group of malignant tumors that result from the clonal proliferation of lymphoid cells. They are classified as Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL) based on histopathologic features, or nodal and extra-nodal based on site of origin. NHL can be B-cell or T-cell in origin.[1,2]

Primary extranodal lymphoma can arise from tissues throughout the body other than lymph nodes. Extranodal involvement is relatively common in the NHL and is present in up to 30% of cases. Involvement of the muscle, however, is much less frequent, accounting for 0.1-1.4% of all extranodal lymphomas and 1.2-2.0% of all malignant muscle tumors, and mainly consists of B-cell non-Hodgkin type (NHL) (> 95%).[3-5]

The most frequently affected structures are the thigh, upper extremities, calf, and pelvis.[2] Even a definitive diagnosis is established through histopathological and immunohistochemistry diagnostic imaging, notably ultrasound (US) and magnetic resonance imaging (MRI), which play a role in diagnosis.[2,3,5]

Here we present a rare case of primary intramuscular lymphoma, presenting as multiple intramuscular masses.

CASE PRESENTATION

A 50-year-old male patient presented to our hospital with multiple painful lumps that started from the left lower quadrant of the abdominal wall and progressively involved his right flank area over a period of one year. He has unquantified significant weight loss, night sweats, and loss of appetite. In the past 2 months, he started to feel a burning kind of pain and an additional lump over his right thigh.

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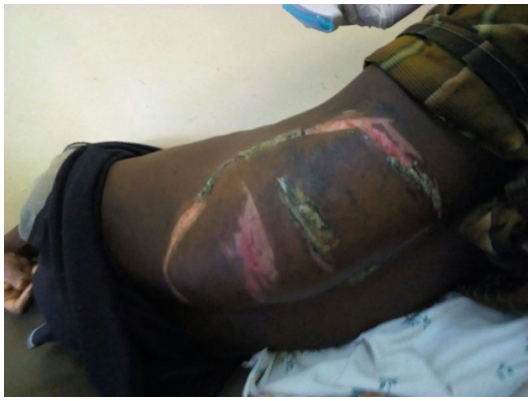
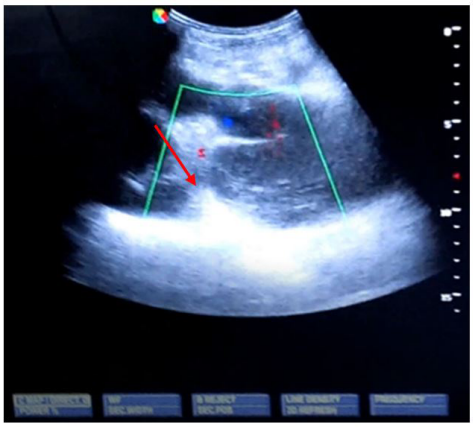
On admission, he was chronically sick-looking, and his vital signs were stable. There were palpable tender masses over the superficial anterior abdominal wall and about a 15×12 cm raised flat lesion over his right posterior flank area with a circumferential healing wound from traditional cautery. There is also a palpable 3×2 cm tender, ill-defined mass in the deep right anterior thigh mass, which was mobile sideways and able to reach below the mass (Figure 1). There was no significant palpable lymphadenopathy. Laboratory tests showed mild anemia with hemoglobin levels of 10.1 g/dL and a raised creatinine level of 1.83 mL/dL. Other complete blood count (CBC) profiles, liver function tests, and random blood sugars were within normal limits. The chest X-ray was normal.

Figure 1. *The Right Thigh Anterior Compartment was Swollen, and the Skin was Ulcerated due to the Traditional Cautery*



US of the lumps showed diffuse enlargement of bilateral psoas, anterior muscle compartment of right thigh, bilateral lower abdominal rectus muscle, left calf muscles, and left brachialis muscles with hypoechoic regions of irregularity infiltrated muscle tissue, with a texture resembling muscle fibers retaining continuity with the surrounding muscles. The architecture of the adjacent muscle appeared to be reserved (Figure 2).

Figure 2. *Ultrasound Examination of Right Psoas and Iliopsoas Muscle is Diffusely Enlarged, with Areas of Poorly Defined Hypoechoic Regions (arrows) within the Proximal Right Psoas Muscle. The Hypoechoic Regions Irregularly Infiltrated Normal Muscle Tissue, with Texture Resembling Muscle Fibers Retaining Continuity with the Surrounding Muscles (arrow)*



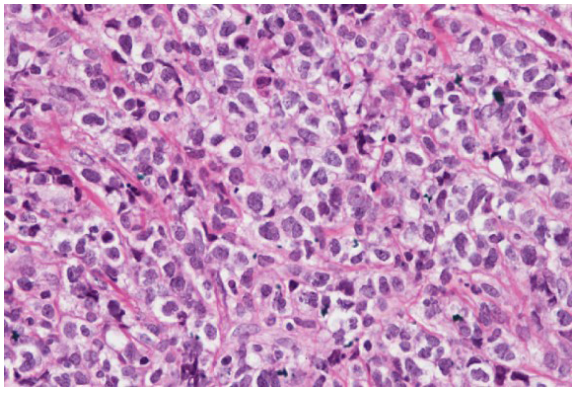
The architecture of the adjacent muscles appeared to be preserved. Color and power Doppler ultrasound showed hypervascularity of the iliopsoas muscle compared to the other normal muscles.

On color Doppler, there is hypervascularity compared to the contralateral and other normal skeletal muscles. The adjacent subcutaneous tissue is thickened. Otherwise, no organomegally or lymphadenopathy (LAP) is seen in all accessible areas. The X-ray of the lower and upper limb bones is normal, with no cortical erosion or lytic lesion.

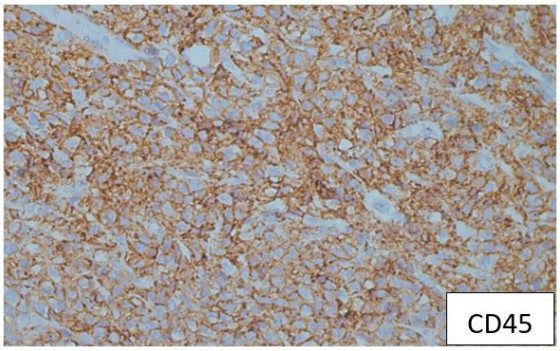
Fine needle aspiration cytology (FNAC) from flank and thigh mass showed small to medium-round cells with fine chromatin, prominent nucleoli, occasional nuclear membrane irregularity, and scant cytoplasm. With an index of round cell tumor (differential diagnosis of lymphoma and extra-skeletal atypical Ewing sarcoma), incisional biopsy and immunohistochemistry (IHC) were recommended.

An incisional biopsy was done under local anesthesia. The hematoxylin- and eosin-stained tissue sections showed sheets

Figure 3. *Round to Angulated Cells with Distinict Nucleoli with Intervening Delicate Fibrous Septa (H&E staining 40x) B. Diffuse Membranous/Cytoplasmic Staining of Lymphoma Cells (CD45)*



A



B

of cleaved cells with distinct to prominent nucleoli and scant cytoplasm with intervening fibrous strands. IHC showed diffuse CD45 reactivity, a pan-leukocyte marker including lymphoid cells (Figure 3). The tumor was diagnosed as intramuscular lymphoma.

DISCUSSION

Lymphomatous involvement of the muscle has been reported to be 0.1-1.4% of all extranodal lymphoma, and the common clinical scenario for lymphomatous involvement of the muscle is often secondary hematogenous dissemination from nodal disease elsewhere or adjacent extension from an involved site. Primary extranodal lymphomatous involvement is exceedingly rare. As seen in our patient, the most commonly involved sites are the pelvic and gluteal musculatures.[2,3]

It can present as a discrete mass or diffuse enlargement of the muscle with loss of normal fascial planes. Occasionally, it can present without any muscle enlargement and can only be diagnosed by biopsy. In our case, the average age of presentation is usually around 60-70 years of age, and a preceding trauma history is often present.[4,5]

Diagnostic imaging is pivotal in identifying skeletal muscle lymphoma. Even though ultrasonography is the most widely used medical imaging modality to evaluate masses, the features of lymphomas involving muscles are non-specific and heterogeneous. However, the predominant presentation of muscle lymphoma involves muscle enlargement characterized by extensive segmental or multi-compartmental spread, involvement of the deep fascial layers, identifiable vessels traversing the affected area, stranding of subcutaneous fat, and increased skin thickness, all of which are findings seen in our patient, but the subcutaneous thickening and stranding in our patient could be due to the traditional cautery. Nonetheless, there are no distinct radiological markers exclusively for the diagnosis of skeletal muscle lymphoma.[2,6-8]

Computed tomography (CT) findings for muscle involvement are non-specific. Muscle enlargement can be seen, with the abnormal tissue often isodense to normal muscle, and enhancement post-contrastis variable. It is useful for identifying the extent of disease and bony involvement or destruction.[9]

MRI features of muscle lymphoma show long segmental or multi-compartmental involvement, deep fascial involvement, the presence of traversing vessels within the lesion, subcutaneous fat stranding, and skin thickening. The involved muscles show enlargement without necrosis or hemorrhage and are seen as isointense on the T1-weighted images and hyper-intense on the T2-weighted images. They enhance homogeneously after the gadolinium administration. Significant diffusion restriction is seen on diffusion-weighted imaging (DWI).[2]

Although imaging can assist in the diagnosis of muscular lymphoma, histological diagnosis is still necessary to avoid inappropriate surgery, as lymphomas respond well to chemotherapy.[10]

CONCLUSION

Muscle involvement in lymphoma is uncommon, and primary intramuscular lymphoma in particular is extremely rare. Nevertheless, it should be considered a differential diagnosis in patients presenting with intramuscular tumors.

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