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Extranodal non-hodgkin's lymphoma in skeletal muscles – rare presentation: A case report

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Abstract
Approximately 40% of cases of diffude large B-cell lymphoma (DLBCL) is extranodal and can arise in any organ system. DLBCL comprises less than 1% cases of extranodal non-hodkin lymphoma. It involves soft tissue and skeletal muscles rarely. A 76 years old man presented with a soft tissue swelling, which came out to be a case of DLBCL of the left arm.

Keywords: Non Hodgkin's lymphoma, skeletal NHL, ExtraNodular NHL, Muscles NHL, upper limb NHL.

Introduction
Diffuse large B cell lymphoma (DLBCL) is a malignancy of the B cells with a significant clinical, morphological, and molecular heterogeneity. It is the most common lymphoid malignancy in adults, accounting for 30% to 40% of all non-Hodgkin lymphomas (NHLs) in western countries.[1] The median age of occurrence is in the seventh decade, and symptoms at presentation, behavior, and prognosis are dependent on the type and the primary site of the lymphoma. The malignancy typically occurs in the lymph nodes, with extranodal primary involvement being estimated at 30% to 40% of cases.[2, 3] The gastrointestinal tract is mostly affected, while extranodal sites of involvement described include the testes, skin, lung, bone, central nervous system, and the respiratory system.[2, 3] Primary skeletal muscle presentation of DLBCL is extremely rare, with an estimated incidence of about 0.5% of extranodal lymphomas,[3] presenting mostly in the lower extremities.[2] We are reporting a 76 year old male a case of primary DLBCL of the upper extremity involving skeletal muscles of arm.

Case Report
A 76 years old male came to our OPD with complain of pain and swelling over left arm since 2 months. There was no history of trauma/fever. He had undergone cardiac valve replacement surgery 2 years back and was on antiplatelet drug (Acitrom). Since 2 months the swelling is progressive and pain over the arm is gradually increasing. His Shoulder and elbow joint range of motion was complete. There was no history of any other system involvement or weight loss / loss of appetite.

On examination swelling was all around the left arm mostly on middle and distal 3rd arm. The skin over the swelling was tense, glossy with venous prominence and margins were indistinguishable (Fig 1). It was non-mobile, non pulsatile, tender with firm to hard consistency, non-fluctuant, non compressible. There was no local rise of temperature.

Axillary lymph nodes were non palpable. On General examination there was no lymph node enlargement any where in body.His blood investigation were as follows TLC-7.47 Thou/cumm, Platelet count-125 Thou/cumm, ESR-13, INR-1.16, CRP-0.75. X-ray findings suggestive of soft tissue swelling but underlying bone and joint are normal (Fig 2). MRI report suggestive of infiltrative intramedullary enhancing pathology with marked periosseous muscle altered signal intensity and enhancement? Infiltrative neoplastic process? Remote possibility of infective process (Fig 3).

With these inconclusive reports patient was planned for open biopsy from left arm after taking clearance from cardiology and anaesthesia side. Per operative the muscles of the left arm were fish flesh appearance, non bleeding non contractile. Drill hole was made in the left humerus and intramedullary sample along with muscle sample collected and send for Histopathological analysis (Fig 4).

Histopathology suggestive of round cell malignant tumour. Further Immuno-Histo chemistry analysis was suggestive of diffuse large B cell Non-Hodgin's Lymphoma (Positive for CD-45, CD-10, CD-20, CD-79a, CD-3, Ki-67 80%) Fig 5.

Fig 1: Clinical Picture showing diffuse swelling with venous prominence on the left arm

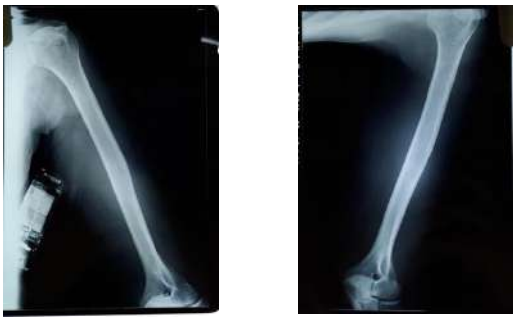

Fig 2: X-ray Left arm suggestive of soft tissue swelling but underlying bone and joint are normal

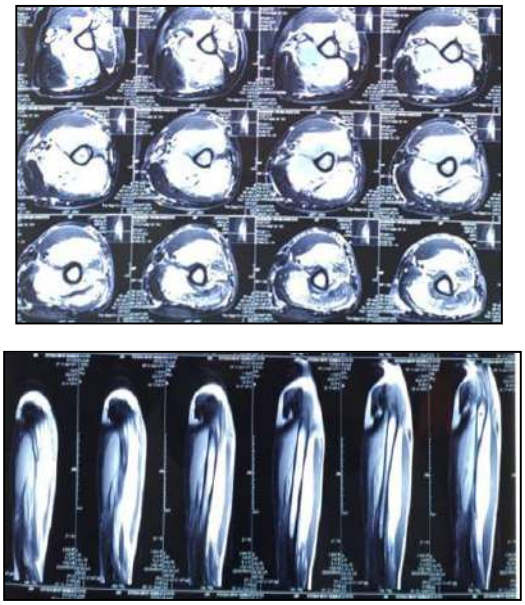

Fig 3: MRI report suggestive of infiltrative intramedullary enhancing pathology with marked periosseous muscle altered signal intensity and enhancement ? infiltrative neoplastic process ?? remote possibility of infective process

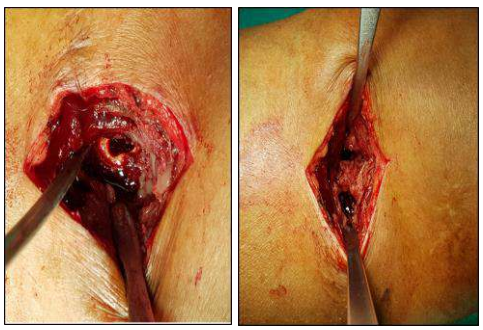

Fig 4: Per operative picture showing dusky (fish flesh like) muscles with open medullary canal

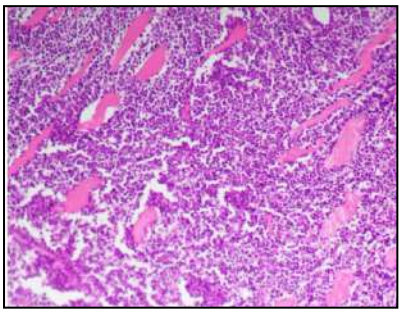

Fig 5: Histopathology Slide

Discussion

DLBCL is the most common histologic subtype of extranodal manifestation of lymphoma and is found in 40% of patients having DLBCL. These are rarely found in skeletal muscles accounting for less than 1% of all cases [4-7]. Prognosis worsen as the age advances [9].Once they infiltrate the skeletal muscles, it mimics rhabdomyoblastoma, metastatic carcinoma melanoma, osteosarcoma and rhabdomyosarcoma leading to multiple differential diagnosis with diagnostic difficulties [6]. These metastatic cells of the lymphoma penetrate deeply into more than one muscles compartment [6, 8]. These lymphomas cause lymphadenopathy characteristically not visualized in sarcomas [6]. These lymphomas primarily involve the bone followed by muscle involvement. Patients with these lymphomas complain generally of muscle pain, swelling and rapidly growing muscle mass [5, 6]. They rarely present with acute compartment syndrome [10]. It can cause changes in the soft tissue and skin, which may mimic cellulitis [11]. The most common precipitating factor is the prior injury at that site, which leads to the formation of lyphoma. Primary muscle lymphoma have been found in leg injury and needle injections and have also been found in rectum of homosexual men [12-13]. Studies have reported that the risk of Non hodkins lymphoma is increased in HIV Positive patients. Although primary muscle lymphoma are found rarely in this population [14].

Conclusion

So, hereby we conclude that all such cases which on clinical and conventional radiographic evaluation are unhelpful in reaching to a definitive diagnosis, should be further evaluated with advanced imaging technique like MRI/PET scan and biopsy should always be done to reach definite diagnosis and further management. Non skeletal condition should always be considered in such type of cases.

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