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Case Report

Primary Skeletal Muscle Non-Hodgkin's Lymphoma in the Thigh: A Case Report

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Abstract: Primary skeletal muscle non-Hodgkin's lymphoma (PSMNHL) is a rare tumor accounting for only 0.1% of all lymphoma. We managed a case of extra-nodal primary lymphoma in right thigh exclusively with chemotherapy. A 46 year old male was referred to our center with progressively increasing mass in the anterior part of right thigh since 2 months. After proper evaluation biopsy was obtained from the mass and sent for histopathological analysis. It demonstrated a diffuse proliferation of large lymphoid cells. Immunohistochemistry examination confirmed diagnosis of NHL of skeletal muscle. We managed case with chemotherapy therapeutic approach Selective chemotherapy was administered with RCHOP (Rituximab, Cyclophosphamide, hydroxydaunomyocin, oncovin, prednisone) 6 cycle regimen. Complete radiological response was achieved after 3 cycles. Now 4 years after completing treatment he is in a state of excellent general condition without evidence of local or distant recurrence. We reported a case of PSMNHL in thigh in middle aged man with purpose of emphasizing role of intelligent diagnostic work up with radiological imaging, histopathology and immunohistochemistry techniques. Chemotherapy with RCHOP regimen with regular follow up to exclude local and distant recurrence has been resulted in good general health of the patient.
Keywords: Non-Hodgkin's Lymphoma, Primary Skeletal Muscle, thigh, chemotherapy

INTRODUCTION

Lymphomas represent clonal malignancies of lympho-reticular tissue. Extra-nodal lymphoma that is outside lymph node and spleen is a well recognized clinical entity. But primary skeletal muscle non-Hodgkin's lymphoma (PSMNHL) is a rare tumor accounting for only 0.1% of all lymphoma. [1]Owing to the rarity of tumor there is scarcity of literature about clinical profile and standard management guidelines for PSMNHL. So herein we report a case of malignant primary NHL in skeletal muscle in thigh.

CASE REPORT

A 46 year old male was referred to our center with a mass in the anterior part of right thigh. He complained of slowly growing mass since past 2 months. On acquisition of detail history, he had no previous history of trauma, infection or any other major illness in the past. He was afebrile with all normal vital parameters. The local physical examination revealed a painless single mass of size 20x15x15 cm with ill defined margins. It was immobile, firm and embedded deep in the muscle structure. Skin overlying the lesion was normal. The remainder of examination did not reveal any vascular or nervous abnormality. Peripheral lymph nodes, liver and spleen were not palpable. On chest X ray there was no evidence of mediastinal lymph nodes. Hematological and biochemical parameters were within normal range. MRI scan (Figure 1.) showed a large, soft tissue mass lesion in the vastus intermedius, bicep femoris, adductor magnus and adductor longus muscles. It appeared as hyper intense to muscle on T1W and T2W images with measurement of 18x10x10 cm. The mass was seen in the deep fibers adjacent to femur. Rest of the bones, neurovascular bundles and visualized bones appeared normal in signal intensity.

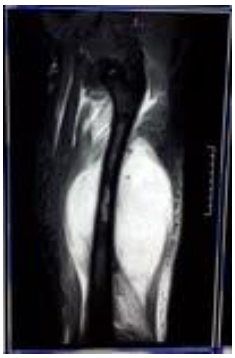

Fig. 1: Radiological characteristic of soft tissue mass in right thigh

Differential diagnoses thought at this stage were liposarcoma, synovial sarcoma and rhabdomyosarcoma. We ruled out distant spread by CT scan of chest, abdomen and pelvic region. After proper evaluation biopsy was obtained from the mass and sent for histopathological analysis. It demonstrated a diffuse

proliferation of large lymphoid cells with large vesicular hyperchromatic nuclei affecting normal architecture of striated muscle fibers. On immunohistochemistry medium to large sized neoplastic cells expressed pan B cell markers like positive CD 20 and negative CD 30, CD 15, BCL 2, Desmine and myogenine. This confirmed diagnosis of malignant stage II non-Hodgkin's diffuse large B- type lymphoma. After bone marrow biopsy we confirmed no malignant involvement of bone marrow.

After careful evaluation, we opted medical therapeutic approach for our patient. Selective chemotherapy was decided with RCHOP (Rituximab, Cyclophosphamide, hydroxydaunomyocin, oncovin, prednisone) 6 cycle regimen. Complete radiological response (Figure 2) was achieved after 3 cycles.

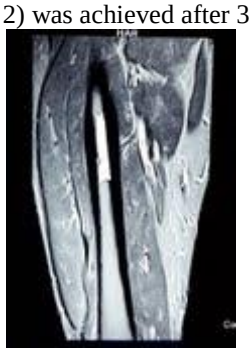

Fig. 2: Regression of lymphoma after 3 cycles of chemotherapy

Patient tolerated all cycles of chemotherapy without any adverse hematological, cardiovascular or infectious incidence. He was further treated with adjuvant radiotherapy. During follow up with MRI scan there was complete regression of the mass. Patient was on regular clinical and radiographic follow up every 3 months in first year and semiannually since second year. Now 4 years after completing treatment he is in a state of excellent general condition without evidence of local or distant recurrence.

DISCUSSION

Soft tissue tumors are commonly seen as benign (lipoma, hematoma, myoscitis etc) or malignant (fibro sarcoma, liposarcoma) lesion [1]. Prevalence of malignant lymphoma is rising due to AIDS pandemic. Although NHL affects the organs with lymphoid tissues, it may originate from almost every organ of the body. Neoplastic proliferation of lymphocytes at sites other than native lymph nodes and lymphoid tissue is described as primary extra-nodal or extra lymphatic lymphoma [2]. Primary extra-nodal NHL involving skeletal muscles is rare type of lymphoma. Commonly it affects skeletal muscles of lower extremity, pelvic region and upper arm [3].

In our present case we first thought probability of rhabdomyosarcoma from clinical and radiological findings. So we were going to opt for surgical resection of the tumor. But thanking to histopathology and immunohistochemistry, it diagnosed as NHL of skeletal muscle and we immediately changed over to exclusive chemotherapy management. Mass regressed completely after 3 cycles of chemotherapy.

E. Bölke reported extra-nodal diffuse B cell lymphoma in right thigh which was managed with chemotherapy followed by radiotherapy [4]. Ueno A et al reported two cases of PSMNHL, one originated in upper arm and another in right thigh. But age of presentation in both cases was 82 and 87 years respectively [5]. This type of malignant lesions commonly occurs in old age. But our patient is 46 year old man without any significant risk factor. Belaabidia B and colleagues described PSMNHL in right thigh in 70 year old woman. They managed it with wide surgical resection followed by chemotherapy [6].

Jean-Michel Laffosse and colleagues reported an authentic case of malignant NHL in a skeletal muscle and its management which was purely medical and conservative using selective chemotherapy [1]. In the similar way we also managed our case exclusively by chemotherapy and avoiding mutilating surgical procedure. In another report 25 year female was presented with atypical manifestation with leg edema which later turned to malignant lymphoma of skeletal muscle [7]. Yu-Huan Gao recommended management of skeletal muscle NHL without surgical excision as it is both, chemo sensitive and radiosensitive [8].

Prior histopathological diagnosis from biopsy tissue plays important diagnostic role so as to avoid amputation of limb. Primary extra-nodal NHL commonly arises from gastrointestinal tract, bone marrow and rarely in skeletal muscle. In case of skeletal muscle involvement, common site is upper arm and glutei muscles [9]. Although numerous cases have been reported regarding NHLs from soft tissues, but management of primary skeletal muscle NHL with exclusive chemotherapy without surgical resection will add to the literature [10- 12].

CONCLUSION

We reported a case of PSMNHL in thigh in middle aged man with purpose of emphasizing role of intelligent diagnostic work up with radiological imaging, histopathology and immunohistochemistry techniques. Chemotherapy with RCHOP regimen with regular follow up to exclude local and distant recurrence has been resulted in good general health of the patient.

Conflicts of interests: There is no financial or any other conflict of interest.

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