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RESEARCH ARTICLE

PRIMARY BONE LYMPHOMA OF FEMUR: A CASE REPORT

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Abstract

Primary Bone Lymphoma (PBL) is a subtype of lymphoma that exclusively affects the skeletal tissue. Primary Bone Diffuse Large B Cell Lymphoma (DLBCL) is the most common pathological type, comprising about 80% of PBL. In this case report, we discuss a patient, a 50-year-old lady who presented with solitary bone lesion, confirmed by biopsy and IHC as DLBCL. She was diagnosed as stage IAE as per WHO staging system. Chemotherapy with R-CHOP for a total of 6 cycles were given followed by IFRT. The patient is under follow up with complete response at present. Being rare case, no randomised study can be taken up for standard treatment protocol. Case to case individualised treatment is done based on stage, histopathological subtype and predictive biomarkers to response. PBL is more common in paediatric population more than the adults. The prevalence of PBL is estimated to be 3-7% amongst all lymphomas in adults.¹ WHO classification of soft tissues and bone tumors defined PBL as either single osseous lesions without regional Lymph node involvement, or tumor involvement of multiple osseous sites without associated visceral or Lymph Node disease.²

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Introduction:-

Case History:

A 55 Year old female was referred to our department for pathological fracture of left subtrochanteric region of femur with high suspicion for malignancy. The patient has other comorbid conditions, on medications for hypertension and type 2 Diabetes Mellitus. Antibody titres for Hepatitis C Virus was positive. The patient gave history of pain over left hip joint for 3 months duration, gradual in onset and progressive in nature. Then she developed sudden onset weakness for 1 week. She underwent Proximal Femoral Nailing (PFN) of Left Femur with excision biopsy on April 20, 2021. HPE confirmed the diagnosis for NHL- DLBCL. On IHC study, the tumor cells were positive for CD20, CD45/LCA, MUM but negative for CD3 and CK. Post operative, the patient was put on adjuvant chemotherapy with R-CHOP, for a total of 6 cycles. After that, IFRT was given to a total of 46Gy with Telecobalt -60 by opposing fields using conventional fractionation schedule.

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Fig 1:- X- Ray Pelvis showing pathological fracture of left subtrochanteric region.

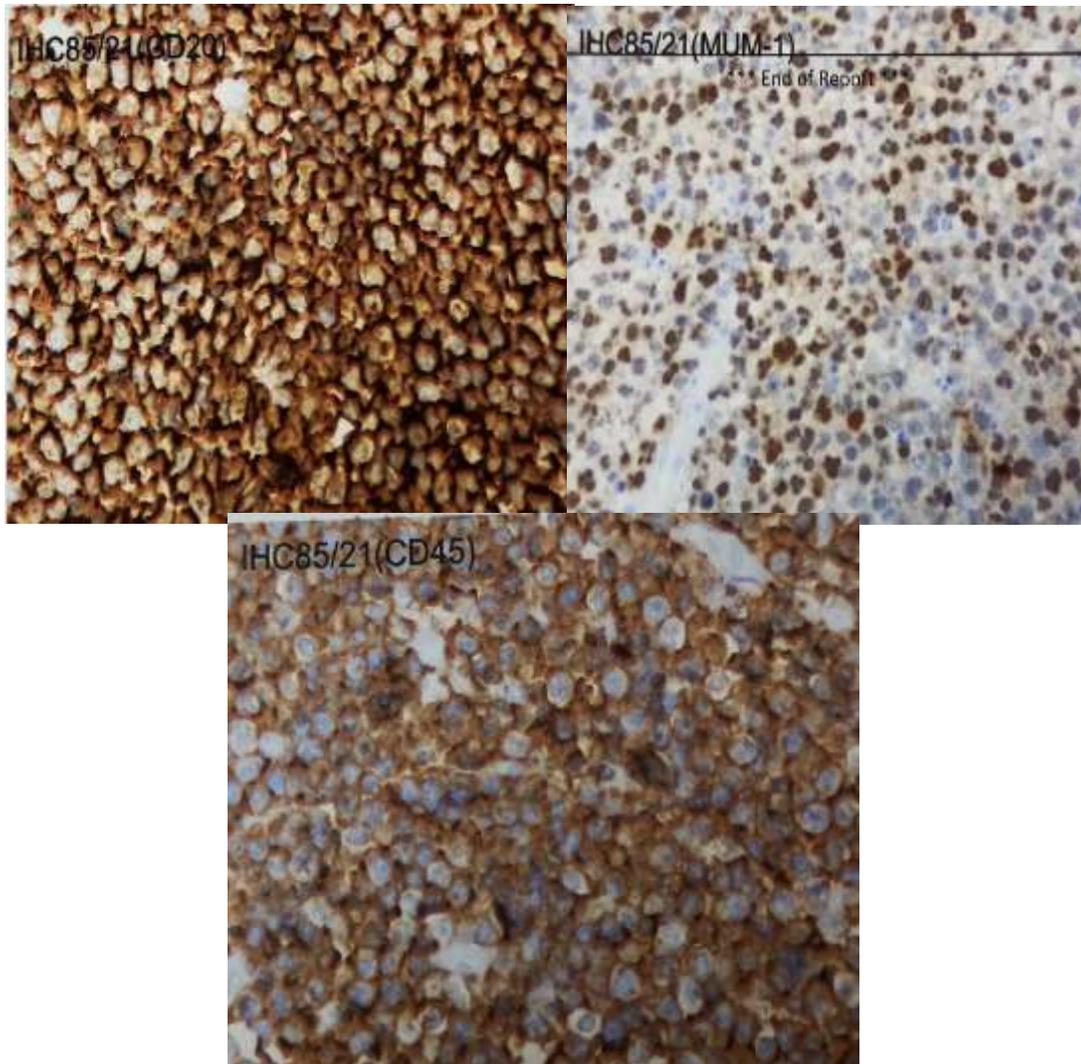


Fig 3:- IHC Marker positive for CD20, MUM1 and CD45.



Fig 4:- PET/CT Scan showing complete response.

Discussion:-

The most common presentation of PBL consists of bone pain and appearance of soft tissue mass. Constitutional symptoms (B symptoms) such as weight loss, fever and night sweats are present in fewer than 10% of patients with PBL. Pain and swelling of the involved site are two of the most common clinical manifestations of this disease.³

Based on radiographic examination, and taking into consideration the lesion's exact radiographic appearance and on the age of the patient, the differential diagnosis includes Metastatic carcinoma, Ewing's Sarcoma, Fibrosarcoma of bone, Multiple Myeloma and Paget's Disease. Lab studies include CBC and basic metabolic panel including Calcium levels. In plain X Ray, the potential presentations include essentially normal appearing bone with only a subtle shadow from an associated soft tissue extension or focal well circumscribed lucent lesion or permeative destructive

lesion with an aggressive periosteal reaction.⁴CT scan can be used to further delineate these lesions and remains the primary modality for staging, restaging and follow up of PBL. On histology, it is characterized by large cells that are centroblastic, immunoblastic or anaplastic. The centroblastic variant is most common and has the appearance of large lymphocytes with scant cytoplasm. PET-CT is recommended as a standard tool for the initial evaluation, staging, and response assessment of FDG-avid lymphomas by the recent Lugano Classification System.⁵The most common used staging criteria for PBL has been proposed by the Lugano Classification System. Fluorine-18 Fluorodeoxyglucose (F-FDG) Positron Emission Tomography PET/CT has been employed in staging and restaging of PBL. A solitary bone lesion with PBL is classified as stage IE according to the Ann Arbor System. This represents one extranodal site of involvement. If regional lymph nodes are involved, it is stage IIE. Disseminated disease is deemed stage IV. Currently there exist multiple treatment modalities for PBL including chemotherapy, localized radiation therapy and surgical intervention. Most of PBLs have characteristics consistent with DLBCL; hence, chemotherapeutic regimens usually consist of anthracycline-based combination therapies such as cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) with or without rituximab (R-CHOP). A prospective study conducted by Christie et al. involving cases of PBL treated with three cycles of CHOP followed by 45 Gy of involved field radiation listed 5-Year local control rate of 72% and overall survival rates of 90%.⁶Therefore, it is currently recommended that patients with diagnosed PBL receive treatment with combined chemoradiation in order to achieve best response.

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