

Case Report-II

Primary Bone Lymphoma - A Case Report

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ABSTRACT

Primary bone lymphoma is rare. A 33 year old male presented with pain and swelling of left upper thigh. X-ray left femur revealed pathological fracture of left femur shaft. Biopsy revealed - non Hodgkin's lymphoma, diffuse, intermediate grade, B cell type. Patient received 4 cycles of CHOP chemotherapy followed by radiotherapy. He continues to be disease-free, and functionally normal, 16 months after diagnosis. Conclusion: primary bone lymphoma have excellent prognosis following chemotherapy and radiotherapy.

CASE

A 33 year male patient presented with complaints of pain & swelling of left thigh of one year duration. Examination revealed a young male in good general condition, afebrile with no evidence of peripheral lymphadenopathy. Per abdomen- liver and spleen not enlarged. X-ray of left thigh revealed: soft tissue swelling upper thigh with periosteal reaction. Hemogram, biochemistry was within normal limits. X ray chest was normal with no evidence of mediastinal widening. Ultrasound of abdomen revealed no evidence of retroperitoneal lymph node enlargement. Open biopsy from left femur was done. Per operative findings revealed - bone surrounded by whitish gelatinous necrosed tissue, thickening of bone with cortical irregularity. Histopathology - non Hodgkin's lymphoma: diffuse large B cell, intermediate grade. The bone marrow aspiration revealed no evidence of marrow infiltration. While being

investigated, patient developed pathologic fracture at the site of the lesion for which internal fixation was done. With final diagnosis of primary non Hodgkin's lymphoma of left femur, patient received six cycles of CHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone) chemotherapy. This was followed by radiation therapy 35Gy in 15 fractions over 3 weeks followed by a boost of 500cGy with a reduced field. Patient is now on regular follow up & is disease-free 16 months after treatment.

DISCUSSION

Bone involvement by non Hodgkin's lymphoma (NHL) is uncommon. A frequency of 20% have been recorded in autopsy studies. Most of these are due to metastasis from systemic lymphoma. To be defined as primary bone lymphoma (i) there should be a primary focus in a single bone (ii) positive histological diagnosis (iii) there should be no evidence of distant soft tissue or distant lymph node involvement. Regional lymph node involvement at diagnosis is not considered as exclusion criteria.¹ Currently, it is recognized that primary bone lymphoma may involve multiple bones, as long as the other 2 criteria are met. Present case fulfills above criteria of being a primary NHL of bone.

The median age at diagnosis is 35 years, 20% of cases occur in sixth decade of life. The common sites are - femur followed by ilium. Rarely, small bones of the hands may be involved. Clinical presentation include- local pain in a patient in otherwise good general health. Pain is a constant feature often present for several months and occasionally years. Swelling, tenderness and warmth in local area

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may be present. B symptoms are not present initially. Patients with primary spine lymphoma may present with neurological symptoms.

Radiologically, bony lesions may be either extensive, destructive or infiltrative.² Extensive lesions involve 25-50% of affected bone, sometimes entire bone. Destructive lesions are radiolucent, mottled, patchy, moth eaten and sometimes the outline of the bone is completely lost. Infiltrative or permeative lesions have poorly defined interface with normal bone, 50% have a mixture with small areas of sclerosis. Nearly all variants destroy cortical bone and 25% thicken the cortex. Often large, obvious soft-tissue extension may be present along with bony lesion. In present case there was evidence of periosteal sclerosis. Such a presentation is rare.² Sclerosis may precede diagnosis by several years and in flat bones may resemble Paget's disease. When confined to marrow cavity with no cortical invasion, plain x-ray may be negative but bone scan and MRI may indicate evidence of bone marrow involvement. Osteosclerotic lesions e.g. ivory vertebra seen in Hodgkin's lymphoma is rare. About 25% of patients may present with pathologic fracture. Histopathologically most cases are of high grade, B cell type. Less frequently- it could be follicular or low grade subtype.^{3,4}

Diagnosis is established by biopsy. Other investigations- skeletal survey, bone scan, bone marrow biopsy, CAT scan of whole abdomen & chest to assess lymph node involvement and serum LDH estimation are done as part of the staging procedure. Plain radiographs often underestimate the extent of the lesion. CAT scan is useful for disease staging and delineating spinal lesions. MRI is helpful in demonstrating bone marrow and soft tissue involvement. Lymphoma has an increased uptake on bone scan. Primary bone lymphoma needs to be differentiated from metastatic carcinoma,

Ewing's sarcoma, osteosarcoma, eosinophilic granuloma (skeletal) and chronic osteomyelitis.^{5,6}

Treatment for diffuse large cell NHL includes- 4 to 6 cycles of CHOP chemotherapy followed by local radiotherapy. Surgery is only indicated for pathologic fractures. Disease free and overall survival rates have been reported to be 78% and 91% at 5 years and 73% and 87% at 10 years, respectively.⁶ The complications noted by the combined modality treatment include - pathologic fracture & avascular necrosis of the irradiated bone.^{7,8} Thus, primary lymphoma of bone should be considered in the differential diagnosis of bony tumours in young patients in 2-4th decade. Chemotherapy followed by radiotherapy is the treatment of choice and is associated with good outcome.

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