LYTIC LESION OF HUMERUS PRESENTING AS NON HODGKINS LYMPHOMA

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Lytic lesion of the bone in early age group is not an unusual presentation to orthopaedic OPD. We have come across a case of lytic lesion of humerus having unusual and rare diagnosis as non-hodgkin lymphoma

Non-Hodgkin’s lymphoma of bone is a rare entity. Patients generally present with localized bone pain and, soft-tissue swelling or a palpable mass. At times, patient may present with a pathological fracture. Lymphoma presenting primarily at proximal end of humerus is unusual. A review of the literature shows that the incidence of skeletal manifestation of lymphoma of bone is less than 4%. Histopathologically, PLB usually represents diffuse large B-cell lymphoma. We report our experience with a case of Primary non-Hodgkin’s lymphoma of proximal humerus with lytic lesion & its management.

KEYWORDS: Non-Hodgkin’s lymphoma presenting as lytic lesion of bone, Chemotherapy.

INTRODUCTION: Primary lymphoma of the bone (PLB) is a rare extranodal presentation of non-Hodgkin's lymphoma (NHL). It was first described by Oberling in 1928

It accounts for approximately 3% of the malignant bone neoplasms and it comprises of less than 4% of all the extranodal non-Hodgkin’s lymphomas. An osseous involvement of a lymphoma is generally seen as a part of a multi-system dissemination. Primary lymphoma of the bone can be defined as a lymphoma which occurs in the bone without any evidence of a distal nodal or an extranodal tissue involvement.
PLB can involve any part of the skeleton, but a trend exists in favour of the bones with persistent bone marrow. The femur is the most common site and it is affected in 29% of the cases. The other sites include the pelvis, humerus, head and neck, and the tibia. The clinical presentation depends upon the rate of the tumour cell proliferation and on the initial localization. The patients generally present with localized bone pain and, less frequently, with a soft-tissue swelling or a palpable mass. On conventional radiology, PLB has a widely variable imaging manifestation which consists of either a 'lytic destructive pattern' or a 'blastic sclerotic pattern'. Pathological fractures may be present in approximately one quarter of the cases, as were seen in our patients.

**CASE:** A 17-year-old male presented with pain and swelling of the left upper arm of 4 months duration. The pain worsened over the past 1 month. The patient was having painful and restricted movements of left shoulder, especially abduction since last week. Clinically, a diffuse swelling was present around the upper arm. No neck or axillary lymph nodes were palpable. There was no palpable hepatosplenomegaly or lymphadenopathy. The chest radiograph was normal. Both the WBC and the CRP levels were within the normal range. The only medications which were taken at the time of the presentation were non-steroid anti-inflammatory drugs (NSAIDS). A screening X-ray of his left shoulder with humerus showed lytic lesion in the proximal humerus. After obtaining informed consent, patient was posted for curettage and bone grafting. Intra-operatively, in addition to lytic lesion, there was non-encapsulated soft tissue mass outside the bone below the deltoid was excised totally and sends for biopsy. During surgery, the tumor mass was having normal vasculature without excess bleeding. The tumor mass was like bunch of plenty of variable sizes of pumpkin or bean-like seeds lytic lesion was filled with cancellous bone from iliac crest. Biopsy report came as non-hodgkin lymphoma. CT abdomen and chest was done to rule out evidence of lymphoma in any form.

The patient was put on systemic chemotherapy. 6 courses of chemotherapy with the use of the CHOP regimen were completed. Patient was put on active physiotherapy. The therapeutic effects were assessed monthly for 6 months. The patient showed a clinically complete remission, and neither relapse nor metastasis has been recognized till his last follow-up at 18 months. Long followup of such cases is essential.

**DISCUSSION:** PLB occurs commonly between 20 and 50 years of age and it shows a male preponderance with a male to female ratio of 3:2. The femur (29%) is the most common site, followed by the pelvis (19%), humerus (13%), skull (11%) and the tibia (10%) Some series have found that the long bones and the flat bones are equally affected.

The clinical presentation includes local pain, swelling and sometimes even a pathological fracture. The diagnosis is established by biopsy. The other investigations which are done to establish the diagnosis include a skeletal survey, a bone scan and a bone marrow biopsy. CT scan of the whole abdomen and the chest to assess the lymph node involvement and serum LDH estimation are done as a part of the staging procedure.
In the younger patients, the differential diagnosis of PLB mainly includes osteosarcoma, Ewing's sarcoma, and osteomyelitis and benign lytic lesions like aneurismal bone cyst, unicameral bone cyst.

A review of the literature shows that the incidence of the skeletal manifestation from NHL is less than 4% and that in all these cases, a bony involvement was reported, many years after the presentation of the primary cancer. The histological features of primary non-Hodgkin's lymphoma of the bone are identical to those of the conventional, non-osseous forms of non-Hodgkin's lymphoma.

Krishan et al. in their evaluation of 20 cases of established primary lymphoma of the bone. The most common pattern which was noted was the 'lytic destructive pattern', which was reported in around 70% of the cases, followed by the 'blastic sclerotic pattern'. The 'lytic destructive pattern' may present as permeative, moth-eaten or focal lyses with well-defined margins. A cortical erosion or destruction may occur, but there is usually a little periosteal reaction. Sequestra formation has also been demonstrated by CT in the cases of PLB. An associated soft tissue mass usually heralds a poorer prognosis. Our patient with the humeral lesion demonstrated radiographic findings which were consistent with a 'lytic destructive pattern', with a large focal lytic lesion, a permeative pattern which extended into the metaphysis. The plain radiographs often underestimate the extent of the lesion, as was observed in our patient, with absence of the soft tissue extension of the tumour mass and a periosteal reaction on the plain radiographs The other 'blastic sclerotic pattern', is more common in metastatic disease.

Hodgkin's lymphoma, the rarer type of primary bone lymphoma, may occasionally present with this pattern, but even in Hodgkin's lymphoma, the lytic pattern predominates. Our patient with the humeral lesion demonstrated radiographic findings which were consistent with a lytic pattern', and soft tissue shadow. As with conventional radiology, the findings of the other imaging modalities such as bone scan, MRI, CT and PET are not specific. CT scan is useful for the disease staging and for delineating the spinal lesions, whereas MRI is the most sensitive imaging technique which can be used for the diagnosis of PLB.

On the other hand, in the follow-up examinations, it cannot reliably differentiate between a persistent disease and a healing bone. Although the role of the positron emission tomography (PET) scan is not well defined in PLB due to the rarity of the lesion, it is preferred over MRI for assessing the remission status. Lymphoma has an increased uptake on a bone scan.

The treatment of PLB typically involves a combination of radiation and chemotherapy. Even with the use of PET, the assessment of the remission status is rather difficult in PLB; hence, the chemotherapy should involve a sufficient number of treatment cycles, i.e., six to eight cycles. The role of surgery in the treatment of PLB is restricted to biopsies and the management of pathological fractures. The role of radiation in PLB is controversial. In a study which was done by Ramadan et al. the patients with the advanced-stage disease who received chemotherapy plus irradiation, actually had a poorer outcome as compared with those who received chemotherapy alone. A major reason for avoiding radiotherapy, was the risk of the late effects from this treatment modality, especially the delayed bone growth in children and the development of a second cancer within a previously irradiated field.

To conclude, PLB is a rare malignant bone tumour with a variable radiographic manifestation. The prognosis, nonetheless, is much better than that of other malignant bone tumours. Thus, primary lymphoma of the bone may be considered in the differential diagnosis of bony lytic lesions in young patients.
patients in the second decade of life. Chemotherapy, followed by radiotherapy, is the treatment of choice and these are associated with a good outcome. Thus, a high index of suspicion should be maintained for this disease in the patients who present with solitary bone lesions.

REFERENCE: