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LYMPHOBLASTIC LYMPHOMA PRESENTING AS ISOLATED BONE LESION IN A CHILD - A RARE ENTITY

Pediatrics

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ABSTRACT

Non Hodgkin lymphoma especially lymphobastic lymphoma (B- Cell) having skeletal presentation is a rare entity. Among extranodal sites, skin is most common followed by bone & GI tract. It is difficult to suspect this diagnosis without clinically obvious lymph nodal mass or bone marrow involvement. We are presenting here a case report of 11 year old girl having pain and swelling in right limb just above knee joint in absence of any organomegaly and lymphadenopathy. MRI was suggestive of lytic lesion whereas tissue biopsy revealed it to be acute lymphoblastic lymphoma (B- cell). Hence, primary lymphoma of bone should be kept as one of the differential diagnosis of tumorous lesion of bone even in pediatric patients. Early and accurate diagnosis of this entity is required for better management of this condition due to its high cure rates.

KEYWORDS

bone tumor, lytic lesions, hematological

Introduction

Primary bone lymphoma (PBL) is an uncommon clinical entity and a rare presentation of non-Hodgkin's lymphoma. Among NHL, Lymphoblastic lymphoma (LL) of the B cell type is uncommon, and extranodal presentation is even rarer. Common extranodal sites include the skin, GI and, less often, bone which account for approximate 25-40% of total extranodal NHL[1].

Primary bone lymphoma constitutes 3% of all malignant bone tumors and 5% of all Non-Hodgkin lymphomas in all age groups[2,3]. Incidence in pediatric population is even less. Clinically and histologically it may be confused with other round cell tumors. Radiologically also, lymphomas do not have characteristic features and can mimic other tumorous conditions. To increase awareness amongst paediatricians about this rare entity, we report a case of B-cell Lymphoblastic lymphoma in a 11 year old girl who presented with extensive bony lesion of lower end of femur and without clinically obvious lymph nodal mass or bone marrow involvement.

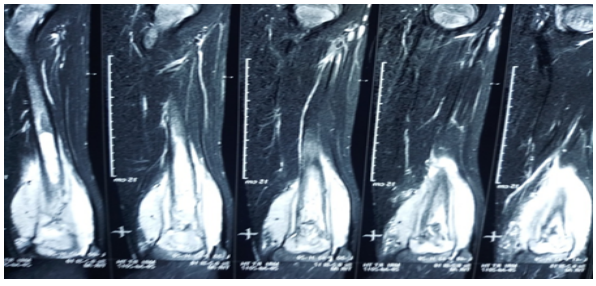
Case Report

A 11-year-old girl presented to the out-patient department with one-month history of continuous, dull aching pain and swelling in right limb just above knee joint (Image 1). This was also accompanied by loss of appetite and loss of weight. There was no history of fever or any bleeding manifestations. Initial examination revealed an approx 20 cm by 15 cm tender swelling on right thigh with indistinct margins. There was increase in temperature and erythema and it was not associated with localized or generalized lymph node enlargement. General examination was unremarkable except for pallor. Hemogram, renal function test and liver function test were within normal limits. ESR and CRP was raised for this child. MR imaging of right thigh revealed lytic destruction of cortex of lower end of femur involving metaphyseal region with periosteal reaction with mild myofascial and subcutaneous edema and patchy involvement of epiphysis with invasion of growth plate with minimal joint effusion(Image 2). Tissue biopsy showed it to be acute lymphoblastic lymphoma (B- cell) and immunohistochemistry showed CD 99, CD 79a, Tdt, HLADR positive. CT Scan of chest and abdomen did not revealed any metastasis in any organ or internal lymph node involvement. There was no evidence of lymphoma infiltration in Bone Marrow aspiration and biopsy. CSF for malignant cells was also negative.

Image 1 : Clinical photograph of patient showing swelling on right thigh



Image 2 :MR image showing lytic destruction of cortex of right femur



Discussion

Primary lymphoma of the bone (PLB) is a rare extranodal presentation of non-Hodgkin's lymphoma (NHL). An osseous involvement of a lymphoma is generally caused by 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 extra-nodal tissue involvement [4,5]. PLB can involve any part of the skeleton, but a trend exists in favour of the bones with persistent bone marrow. 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 pelvis, humerus, head and neck, and the tibia [6]. Some series have found that the long bones and the flat bones are equally affected [7]. 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' [8]. Pathological fractures may be present in approximately one quarter of the cases, as were seen in few case reports.[9] The diagnosis is established by biopsy and immunohistochemistry.

LLs are mainly of the T-cell type, with only 10-20% being of the B-cell type.[10] Mostly B- cell LL have been reported in literature as case reports or very small series.[11] Unlike T-LL, pre-B LL predominantly presents with extranodal masses, especially those involving the skin with low propensity for leukemic involvement.[11] Bone is the second most common site of extranodal involvement. There are only few individual case reports and small series of pre B-LL presenting as primary bone lesions. Skeletal radiographic changes associated with and/or suggestive of lymphoma include diffuse osteopenia, metaphyseal bands, periosteal new bone formation, geographic lytic lesions, sclerosis, mixed sclerosis/lysis and permeative destruction.[12] Bony lesions in pre-B LL are usually osteolytic as seen in our case report, although few cases with osteoblastic lesions have been reported.[13]

The treatment of PLB typically involves a combination of radiation and chemotherapy and is usually associated with good outcome .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.

To conclude, PLB is a rare malignant bone tumour with a variable radiographic manifestation. The curability of LL in contrast to other metastatic malignancies makes early and accurate diagnosis of paramount importance. Hence, LL must be considered in the differential diagnosis of extensive bony lesions, and adequate immunohistochemical markers must be used to differentiate this entity. A high index of suspicion should be maintained for this disease in the patients who present with solitary bone lesions.

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