

CASE REPORT

AGGRESSIVE PERIOSTEAL NONHODGKIN'S LYMPHOMA IN A 3 YEARS OLD BOY

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ABSTRACT: Non-Hodgkin's disease is a type of lymphoma and accounts for approximately 1% of all cancers.^[1] It spreads contiguously and along lymphatic pathways. Non-Hodgkin's disease is usually confined to the lymph nodes, although uncommon may be found in any organ system, either as a primary manifestation or as dissemination of systemic disease.^[2] Extra nodal disease is commoner in children and in the immunocompromised host.^[3] Primary Non-Hodgkin's disease of bone (Reticulum cell sarcoma) is a rare, extra nodal lymphoma histologically identical with those arising in lymphoid tissue but presenting initially as a localized solitary bone lesion accounting for approximately 3% of malignant bone tumours. The metadiaphyses of long bones are most commonly involved, particularly the femur, tibia and humerus.^[2] Flat bones are also involved, especially the ilium and scapula. We report a rare case of non-Hodgkin's lymphoma with bony metastasis showing aggressive periosteal reaction.

KEYWORDS: Non-Hodgkin's, aggressive.

CASE REPORT: 3 year old boy presented with complaints of swelling in the right lower limb for 6 months. The swelling gradually progressed to involve the thigh and the right inguinal region associated with intermittent fever. On clinical examination diffuse swelling of the right lower limb with multiple bilateral enlarged inguinal nodes and mild restricted movement around right knee joint noted. Distal pulse and neurological examination were normal.

Plain radiograph of the right tibia with knee joint revealed ill-defined periosteal infiltrative lesion involving subperiosteal and cortical region of the diaphysis extending through the entire length of tibia with an aggressive looking periosteal manifestation.

To characterize the lesion and define the extent of involvement, plain computed tomography (CT) was performed on 128-slice somatom definition MDCT scanner which revealed ill-defined lesion with wide zone of transition seen predominantly in the surface of the diaphysis extending through the entire length of right tibia.

Periosteum showed features of aggressive reaction and appeared multilayered with spiculated and interrupted margins. There was evidence of soft tissue swelling with displacement of adjacent fat planes, however no evidence of obvious soft tissue infiltration seen. The lesion was seen to involve the right distal femoral epiphysis, proximal and distal tibial epiphysis and both foot bones also showed small lesions.

Plain and contrast CT scan of abdomen revealed multiple, discrete, enhancing, enlarged lymph nodes in the retroperitoneum displacing the abdominal aorta, celiac trunk, renal arteries and veins with the largest measuring 33 x 37 x 31 mm in the region of coeliac trunk. The lymph nodal mass lesions were seen compressing the ureters causing bilateral hydronephrosis.

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Similar nodal mass lesions were seen in the pelvic cavity along the right iliac vessels and right inguinal region.

Based on the above findings a diagnosis of lymphoma with bony metastasis arrived at.

DISCUSSION:

Types of Lymphoma: Lymphomas are currently classified according to the 2008 WHO classification of tumours of haematopoietic and lymphoid tissues. The main division is into:

- 1) Hodgkin lymphoma (40%).
- 2) Non-Hodgkin lymphoma (60%).
 - Mature B-cell lymphoma.
 - Mature T-cell and NK-cell lymphoma.
 - Post-transplant lymphoproliferative/lymphoproliferation disorders.

The radiographic appearance varies widely and is often not characteristic; it can mimic osteosarcoma, Ewing's sarcoma, and other bone tumors, as well as osteomyelitis.

In Non-Hodgkin's lymphoma, mediastinal lymph node involvement is the most common thoracic abnormality. Abdominal involvement includes preaortic, paraaortic and inguinal lymph nodes.

Non-Hodgkin's lymphoma of bone occurs usually as a secondary manifestation of systemic non-Hodgkin's disease. Localised pain of an intermittent nature is the most common initial symptom. Primary site of skeletal involvement involves femur, tibia and humerus. May also involve scapula, sternum and ribs.^[2] Most lesions are diaphyseal in location or affect the metaphysis adjacent to the diaphysis.

Radiologically the lesion begins in the medullary bone as a permeative lytic process. As the tumour enlarges, patchy areas of bone destruction spread to affect the adjacent cortical bone.^[1] Periosteal response is minimal and, if present, assumes a laminated pattern. Mid shaft lesions usually break the cortex at a later stage than do metaphyseal tumours.^{[4][5]} By the time cortex is disrupted, the medullary lesion becomes large. In the later stages of the destructive process, a soft tissue mass can be demonstrated, a sequel of direct extension of tumour through cortex and periosteum.^[2] Occasionally, the lesions may be sclerotic or show mixed pattern. Pathological fracture is reported in approximately 25% of cases.^[6]

In our case, enlarged retroperitoneal and inguinal lymph nodes associated with right tibial ill-defined aggressive periostitis, suggestive of non-Hodgkin's lymphoma with bony metastasis.

Histopathology findings: Tru-cut biopsy from right tibia revealed fibro collagenous tissue infiltrated by round, oval or spindle shaped crushed cells with hyper chromatic nuclei.

Wedge biopsy of right inguinal lymph node revealed greyish white tissue macroscopically. Microscopically round to oval cells with indistinct cytoplasmic margin and round to oval nucleus displaying pleomorphism, hyperchromatism and abnormal mitosis.^[7]

Histopathologically features of malignant round cell tumour favoring malignant lymphoma, large cell type.

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TREATMENT: Various types include;

- 1) Chemotherapy: Common drugs include cyclophosphamide, chlorambucil, cisplatin, ludarabine, methotrexate.
- 2) Immunotherapy: These aim in killing lymphoma cells or slow their growth. Common drugs include Rituximab, Ibinutuzumab.
- 3) Radiation and
- 4) Stem cell therapy.

Chemotherapy is the treatment of choice for this patient.

CONCLUSION: The bone metastasis secondary to non-Hodgkin's lymphoma usually presents as osteolytic lesion with no or minimal periosteal reaction. There have been several cases reported on the same. Whereas, an aggressive periosteal reaction secondary to non-Hodgkin's lymphoma, seems to be an unusual case and has not been reported.

Therefore, aggressive periosteal reaction secondary to non-Hodgkin's lymphoma should be considered in the differential diagnosis of non-Hodgkin's lymphoma with bone metastasis.

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Figure 1 & 2: Plain Anteroposterior and lateral radiograph of right tibia with knee joint showing ill-defined periosteal infiltrative lesion involving subperiosteal and cortical region of the diaphysis extending through the entire length of tibia.



Figure 1 & 2

Figure 3 & 4: CT axial and sagittal sections of right tibia showing ill defined lesion with wide zone of transition. Periosteum shows aggressive reaction and appears multilayered with spiculated and interrupted margins.

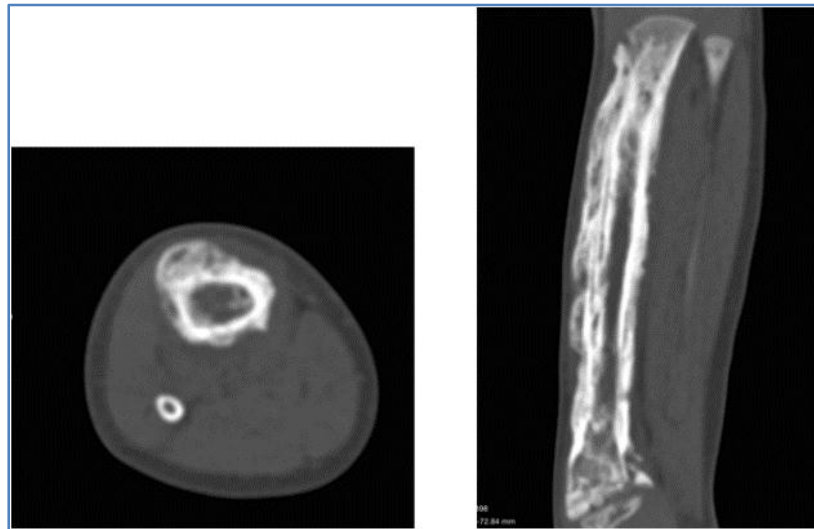


Figure 3 & 4

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Figure 5 & 6: CT axial sections of abdomen and pelvis showing enlarged lymph nodal mass in the retroperitoneum and right iliac and inguinal nodal mass lesions.

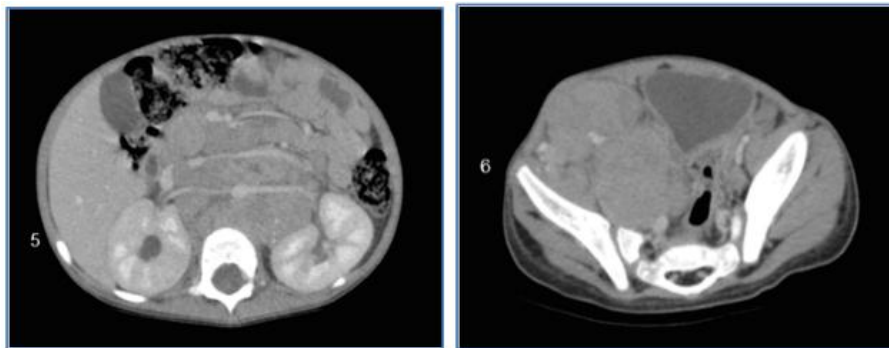


Figure 5 & 6

Figure 7: Wedge biopsy of right inguinal node: smear examination revealed round to oval cells with indistinct cytoplasmic margin and round to oval nucleus displaying pleomorphism, hyperchromatism and mitosis.



Figure 7

Figure 8: Image of bilateral leg showing diffuse swelling of the right lower limb.



Figure 8

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Figure 9: Blood picture showing reduced hemoglobin and increased ESR.

REPORT ON EXAMINATION OF BLOOD.

- TOTAL COUNT:-
W.B.C :- 9'600 /cubic mm.
- DIFFERENTIAL COUNT:-
Neutrophils:- 68 %
Lymphocyte:- 30 %
Eosinophils:- 02 %
Monocytes:- 00 %
Basophils:- 00 %
Abnormal cells:- Nil
- BLOOD FOR HB% :-
Blood HB%:- 9.3 gm% ✓
- ERYTHROCYTE SIDEAMENTATION RATE:-
1st hour :- 66 mm
- BLOOD FOR C.R.P :-
Blood C.R.P :- Positive 48 mg/dl.
Normal C.R.P :- Up to 6 mg/ dl.

Figure 9

Figure 10: Biopsy report of right inguinal node showing features of malignant round cell tumour.

SURGICAL PATHOLOGY REPORT

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Nature of Specimen - Labelled as wedge biopsy of right inguinal lymphnode.

Macroscopic Examination -
Specimen consists of two greyish white bits of tissue largest measuring 1.2x1x0.5 cms.
Cut section is greyish white

Microscopic Examination -
Multiple section studied shows complete effacement of lymphoid architecture by diffuse arrangement of round to oval cells with indistinct cytoplasmic margin and round to oval nucleus displaying pleomorphism, hyperchromatism and abnormal mitosis. Also cells are few scattered large bizarre cells.

IMPRESSION: Shows features of malignant round cell tumour.

The possibilities to be considered are

1. Ewings sarcoma, large cell / atypical type.
2. Malignant lymphoma, large cell type.

Advised Immunohistochemistry for confirmation.

Encl : 1 | 1 Slide/s. Preserve them carefully.

Figure 10

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