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Case Report

Case of Multifocal Non-Hodgkins Lymphoma of Bones of Foot and Ankle

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Abstract

We present a unique case of a 65 year old woman with complaints of multiple swellings around the left ankle. we performed preoperative work up with radiography, CT and MRI and because of diagnostic dilemma, we planned for excision biopsy and sent for histopathological examination which suggested large B cell lymphoma and was confirmed by Immunohistochemistry. The patient subsequently underwent chemotherapy and radiotherapy to the affected area.

Keywords: Large B Cell Lymphoma; Chemotherapy; Radiotherapy

Introduction

Primary intraosseous lymphoma of bone, known in the past as reticulum cell sarcoma, is an uncommon malignancy that accounts for less than 5% of primary malignant bone tumors. Over 20% of patients with lymphoma have secondary bone involvement. Most intraosseous lesions are Non-Hodgkin's Lymphoma (NHL). Nonhodgkins lymphoma of bone is found in the femur and pelvis in patients twenty years of age older. Primary Hodgkin's lymphoma of bone is very rare and might occur anywhere in the human skeleton.

Symptoms and signs

NHL present as pain in the part involved or swelling. Patients usually are in good health otherwise. Lymphoma of bone has a different picture on plain x ray, this lesion might appear as vague with mottled lucency This lesion within in the bone usually has lysis which is permeative pattern but may resemble blastic or sclerotic type followed by Periosteal reaction and cortical destruction. CT scan is useful for disease staging and delineating spinal lesions. MRI is a definitive tool in displaying bone marrow and soft tissue involvement. Bone scan shows increased uptake in lymphomas.

Differential diagnosis

The radiologic differentials include osteosarcoma, Ewing's sarcoma and osteomyelitis. the need to eliminate the possibility of metastatic disease is a must at this juncture.

Histological findings

Primary NHL of bone is a gray, white lesion when examined grossly and diffusely infiltrates bone. Pathological diagnosis requires clinical knowledge of lymphoma for handling of tissue, it is a must to obtain tissue to maintain the cell morphology without crush artifact or decalcification. Needle biopsy is not good enough. large cells with irregular cleaved nuclei and prominent nucleoli surrounded by reticulin fibers should give a suspicious picture of NHL because it is commonly presented in similar fashion. The most commonly presented subtype is diffuse histiocytic lymphoma. Hodgkins lymphoma has a mix of varied cell population like plasma cells, lymphocytes, histiocytes, and eosinophils. To make a diagnosis of hodgkins lymphoma presence of large Reed-sternberg cells, sharply delineated cells with abundant cytoplasm and double nucleus is a must. The pathologic differentials include ewings sarcoma, chronic osteomyelitis, and eosinophilic granuloma.

Treatment options

Clinically staging is done with the following serum chemistries, Chest x ray, bone scan, CBC, bone marrow aspirate and biopsy.

Radiation and chemotherapy are the prescribed treatment options to deal with NHL.

Surgery is only indication for pathological fractures.

Outcomes of treatment and prognosis

Lymphoma of bone has the best prognosis of all primary malignant tumors.

Literature survey

Primary bone lympomas are very rare. The diagnosis is based on imaging studies and histopathological examination, where other disease foci should be excluded. The lymphomas are most often found in the axial skeleton, pelvis, femur and humerus. The tumour causes extensive lytic bone destruction with soft tissue infiltration. Surgical treatment is currently not used routinely.

The aim of this study was to present rare clinical case of NHL around ankle and its most characteristic imaging features.

Case Report

65 years old woman came to orthopaedic OPD in our hospital with complaint of pain and multiple swellings around left ankle since 3 months. On clinical examination 3to 4 globular swellings on lateral side of the left ankle (Figure 1). No local rise of temperature. Skin over the swelling normal with firm consistency. Swellings are non-tender and found to be adherent to underlying soft tissue structures around ankle. Ankle movements are normal.

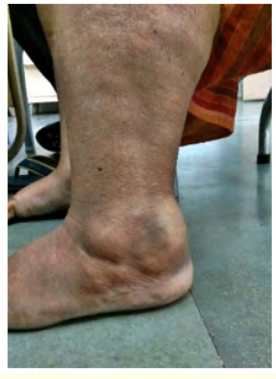

Figure 1

We subjected the patient for radiographs (Figure 2a, b) of left ankle.

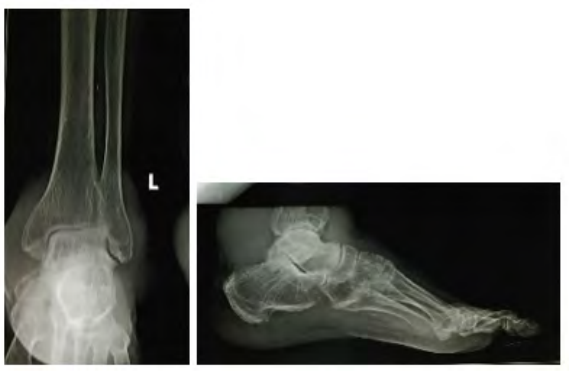

Figure 2a, b

As there is no bony involvement on radiographs, we did high resolution ultrasound left ankle which reported as bursitis/ synovitis left ankle. Ultrasound guided FNAC shows features suggestive of granulomatous inflammation. MRI of left ankle was done which shows well defined lobulated soft tissue lesion around ankle joint with altered intensities and cortical irregularities in talus and calcaneum (Figure 3) probably chronic infective etiology. For confirmation of diagnosis we did excision biopsy and sent for histopathological examination (HPE). HPE reported as malignant tumor composed of round cells that exhibit little pleomorphism, scant cytoplasm, hyper chromatic nuclei with a little intervening stroma which is hyalinized showing areas of necrosis with infiltration into surrounding fat and skeletal muscle. As histopathology is inconclusive, immunohistochemistry was done for CD3, CD20, KI-67 and final interpretation was b-cell non hodgkins lymphoma, large cell type.

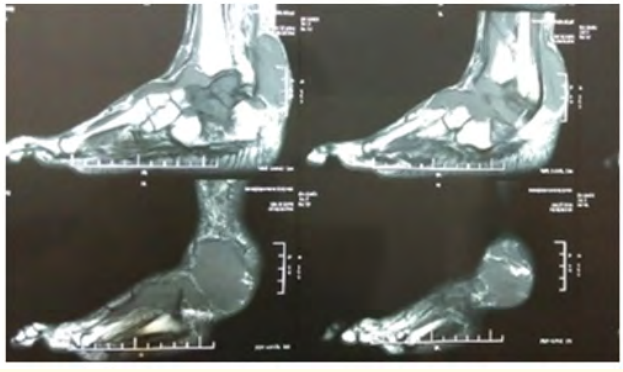

Figure 3

To rule out other any lesions else were in body we ordered CT scan from vertex to foot which shows small nodule of about 5mm in right middle lobe there is no mediastinal or hilar lymphadenopathy.

Small cyst involving right hemi pelvis size 3. 1 *2. 6 cm appears as simple cyst.

After that patient received chemotherapy and radio therapy under the supervision of medical oncologist.

Presently she is not having any complaints and swellings are almost disappeared in leg (Figure 4).

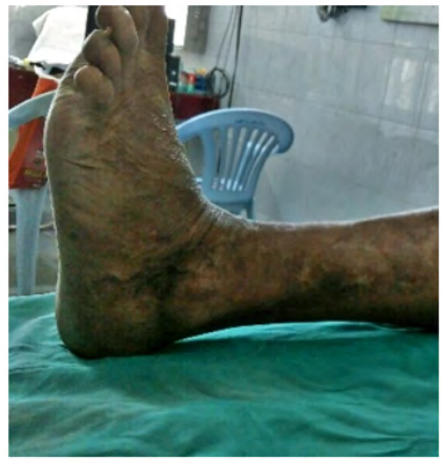

Figure 4

Discussion

Initially Primary bony involvement is uncommon with NHL, an accepted definition of Primary Bone Lymphoma (PBL) is that of a solitary bony lesion which is persistent for more than 24 weeks without the evidence of systemic involvement. The presence of regional lymph nodal enlargement can't exclude a diagnosis of PBL, but it is necessary to have a histological examination.

In this case patient presented with ankle swelling without evidence of any systemic involvement, which was evaluated by imaging and pathological analysis. there was no source of dissemination even after an year of initial diagnosis. To our clinical knowledge this case is an unique presentation.

PBL most commonly occur in fourth and fifth decades, can present at any age and the incidence is less than 15% in cases below 20 years of age. pain at the involved region and type b symptoms like generalized weakness, night sweats, and weight reduction, may be present. These are often nonspecific findings but should prompt further diagnostic work up. type b symptoms were absent in our case presented in orthopedic opd. the most common sites to be involved are the femur, pelvis, tibia, humerus, spine, mandible, skull, scapula, radius and ulna according to Beal., *et al.* the clinical characteristics of 82 patients and reported.

PBL is often characterized by osteolytic defects. to know the extent of local disease and to rule out systemic involvement ct scan or mri options are used. due to maximum sensitivity in detecting abnormalities of bone marrow, MRI is very helpful tool in the evaluation of PBL and can be used to plan biopsy sites or detect lesions elsewhere. bone marrow biopsy confirms the diagnosis of involvement of bone marrow. diagnosis is based on the tissue biopsy taken from the involved bone or from an adjacent lymphnode. the most common grade seen is intermediate, followed by low grade and coming to High grade tumors, they are rare, and Maximum number of cases present with diffuse large b cell lymphoma. in PBL. CD20, CD79a, and bcl-2 positivity is seen in Immunohistochemical stains and negative CD3, CD5, CD10., andCD23, along with cyclinD1, and terminal deoxy nucleotide transferase [1-7].

Conclusion

Primary Bone Lymphoma (PBL) is case which was very difficult to have a diagnosis clinically or radiologically was confirmed with combination of histopathology, immunohistochemistry and PET scan. Lymphoma presenting as soft tissue swelling is relatively rare entity and can get confused with a variety of inflammatory conditions, as well as infectious diseases like tuberculosis.

Future Scope

In the future during evaluation of any swelling around ankle joint with inconclusive diagnosis we can consider even primary bone lymphoma as one of the differential diagnosis.

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