PRIMARY MALIGNANT NON-HODGKINS LYMPHOMA OF THE KNEE
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ABSTRACT:
Primary Bone Lymphoma (PBL) is a relatively uncommon entity. It represents approximately 5% of all Non Hodgkin Lymphomas (NHLs) and 3% of all bone malignancies. The femur, tibia, and pelvis are the most common skeletal sites involved. It can occur at any age, with a peak incidence in the fourth and fifth decades. It can mimic other disease processes, especially infection and at times arthritis if it is in the periarticular area. So, thorough and prompt investigatory workup is essential for adequate treatment. We discuss the clinical findings, diagnosis, and treatment in a case of PBL involving the distal femur.

KEYWORDS: Non hodgkin lymphoma, primary bone lymphoma, chemotherapy

INTRODUCTION:
Primary bone lymphoma (PBL) is a relatively uncommon entity. It represents approximately 5% of all Non Hodgkin lymphomas (NHLs) and 3% of all bone malignancies. The femur, tibia, and pelvis are the most common skeletal sites involved. Appropriate imaging evaluation and histopathologic examination allows early diagnosis and treatment. We present a case of NHL of the distal femur. Herein, its primary location in the knee, the presenting symptoms, and the management of the disease are discussed.

CASE REPORT:
A 45 year old immune competent female presented with a one year history of pain, swelling in right knee, and inability to bear weight on the right leg. The symptoms aggravated on walking. There was no history of associated trauma which led to the symptoms. The patient was being managed symptomatically for pain and swelling before she presented to our institution in December 2014 when the symptoms worsened. The patient was initially evaluated in the Department of Orthopedics. Patient had constitutional symptoms like loss of weight, loss of appetite, night cries and occasional on and off fever. On examination, the patient’s gait was antalgic, with inability to bear weight on her right leg. The skin over the knee was stretched, with engorged veins and mild warmth on palpation.[Fig 1]
Figure 2: X-ray right knee AP and Lateral views showing sclerotic changes in the distal femur with cortical breach and minimal periosteal reaction. Routine blood investigations were in normal limits except ALP and LDH which were raised insignificantly, 131U/L and 247 U/L respectively. X-ray of the right knee [anteroposterior (AP) and lateral views] revealed diffuse sclerotic lesion of the right knee with minimal periosteal reaction and surrounding massive soft tissue shadow. [Figure 2]. Magnetic resonance imaging (MRI) of the right knee revealed a large intraosseous lesion involving distal femur with transcortical extraosseous extension into the surrounding soft tissues. [Figure 3]

Figure 3: MRI Right femur and knee showing intraosseous lesion with mixed signal intensities involving distal femur bone with transcortical extraosseous extension into surrounding soft tissues. On computed tomography (CT) scans of the chest, abdomen, and pelvis, no lymphadenopathy or organomegaly was present. Biopsy from the infiltrative lesion was done for assessment and histopathologic examination revealed tissue infiltrated with large atypical cells with nuclei showing prominent nucleoli. Based on clinical, radiological, and histopathologic evaluation, a diagnosis of stage IV primary lymphoma of the right distal femur was established.

The patient received 6 cycles of combination chemotherapy with inj. rituximab 375 mg/m2 i.v. on day 1 (d1), inj. cyclophosphamide 750 mg/m2 i.v. d1, inj. doxorubicin 50 mg/m2 i.v. d1, inj. vincristine 1.4 mg/m2 i.v. d1, and tablet prednisone 100 mg d1–5 (RCHOX) every 3 weeks. CHOP chemotherapy is made up of C = Cyclophosphamide H = Doxorubicin hydrochloride O = Vincristine (which used to be called Oncovin) P = Prednisolone (a steroid) MRI done after completion of chemotherapy demonstrated resolution of the extraosseous component with a significant change in the intraosseous distal femur lesion. [Fig 4] On follow up, patient showed remarkable symptomatic improvement.

DISCUSSION:
Primary bony involvement at initial presentation is uncommon with NHL. An accepted definition of PBL is that of a single bony lesion that persists for longer than 6 months without evidence of systemic involvement.[3,4] The presence of regional lymph node involvement does not exclude a diagnosis of PBL, but a histological examination of the lymph node is necessary. In the case reported here, the patient presented with a single distal femur lesion without evidence of any systemic involvement, which was evaluated by imaging and pathological analysis. There was no source of dissemination at 6 months after initial diagnosis. PBL is often characterized by osteolytic defects. Evaluation with CT scan or MRI is essential to know the extent of local disease and to rule out systemic involvement. MRI has been demonstrated to be a helpful tool in the evaluation of PBL. Its great sensitivity in detecting bone marrow abnormalities can be used to plan biopsy sites or detect satellite lesions. Bone marrow involvement is confirmed by using bone marrow biopsy.

Tissue diagnosis with biopsy of an adjacent lymph node or directly from the involved bone forms the foundation of the diagnosis. High grade tumors are rare, and the most common grade identified is intermediate, followed by low grade lesions. Majority of patients with PBL have diffuse large B cell lymphoma (DLBCL). Immunohistochemical staining shows CD20, CD79a, and Bcl 2 positivity and CD3, CD5, CD10, and CD23, cyclinD1, and terminal deoxynucleotidyltransterase negativity. In the case reported here, patient presented with a long history of pain in right knee. On imaging, sclerotic lesion was seen in the right knee with soft tissue component. On histopathologic and Immunohistiochemistry examination, a diagnosis of DLBCL was made. For the management of PBLs, no universally accepted therapeutic guidelines have been developed. Combined modality therapy has been shown to yield better prognosis and results are superior to those of radiation therapy alone. Patient in the present study was treated with 6 cycles of chemotherapy. At 1 year follow up, the patient is disease free with adequate functionality and local pain control. Prognosis of patients with PBL truly limited to bone is more favorable than for patients with spread to regional lymph nodes or disseminated NHL.[6,7]

REFERENCES: