Primary lymphoma of the bone involving epiphyseal humerus: report of a case

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ABSTRACT
Lymphoma is considered to be primary in bone when it arises in the medullary cavity without concurrent regional lymph node or visceral involvement. Some of these tumors present with a large extrasosseous soft tissue mass with little cortical destruction and periostial reaction on plain radiographs. This may result in a delay in the diagnosis and improper therapy.

Keywords: Malignant lymphoma, Bone lymphoma, non-Hodgkin's lymphoma.

Case Report
A 41 year-old previously asymptomatic patient presented with 2 year history of pain in right shoulder followed by a gradually increasing swelling of the right shoulder and upper arm over the previous 10 months. There was no history of trauma or systemic symptoms. After preliminary investigations elsewhere, he has received multiple courses of antibiotics without relief. Examination here, showed a well nourished patient, afibrile and there was no anemia, cyanosis, jaundice or lymphadenopathy. The right shoulder and arm were grossly swollen with stretching of the overlying skin (Fig. 1). The swelling was warm, firm, tender, nonfluctuant and fixed to underlying humerus. Investigations revealed a raised LDH of 1384 iu/l. The plain radiograph of the right shoulder and upper arm showed a permissive lesion of the humeral head with little periostial reaction and a large soft tissue mass. Computerized tomography (CT) scan of the right upper humerus showed cortical erosions with small cyst formation in the epiphyseal, metaphyseal and diaphyseal regions with a disproportionately large soft tissue component (Fig. 2). Abdominal ultrasonography as well as the CT scan of thorax and abdomen were normal. A fine needle aspiration from the swelling suggested non-Hodgkin's lymphoma (NHL) of large cell type, which was confirmed on needle bone biopsy (Fig. 3). Bone marrow aspiration and biopsy from iliac crests, however, were normal. A diagnosis of NHL stage 1/E A was made and he received 3 cycles of CHOP, consisting of cyclophosphamide, Adriamycin, vincristine and prednisolone with a partial remission and was lost to follow up.

Discussion
Extranodal presentation of NHL is quite frequent in this part of the world. The majority of these patients, however, present with gastrointestinal tract involvement. Primary lymphoma of bone (PLB) is a rare subset of extranodal lymphomas that accounts for 1% of all non-Hodgkin's lymphomas and 5% of all primary bone tumors. It affects all age groups, but is rare in patients 10 years or younger. Any bone can be affected, the most frequent being the femur (24%), ilium (17%), spine (14%), and the humerus (11%) as in our patient. The majority of lesions in long bones are metaphysically or diaphysically centered and the size varies from 3 to 25 cms. Pain and/or swelling with increased warmth are the most common presenting features. Neurological symptoms and signs may be the presenting feature if lymphoma involves the spine. The radiological features of PLB are by no means specific and include a permissive-destructive lesion and periostial reaction. However, the amount of cortical destruction may be minimal as compared to the large contiguous soft tissue component as seen in our patient. This is attributed to the permeation of cortex by the tumor gaining access.

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Fig. 1 Photograph showing diffuse swelling of the right shoulder with stretching of the overlying skin.

Fig. 2 Computerized tomographic scan of right humerus showing cortical erosion with small cyst formation. The large soft tissue component is seen.

Fig. 3 Photomicrograph of bone biopsy from right humerus showing sheets of neoplastic lymphoid cells infiltrating into lamellar bone (H and EX40).

seen in other conditions like aggressive primary neoplasms, hyperparathyroidism and osteomyelitis. Morphologically PLB are heterogenous and have been classified as diffuse large cell or diffuse mixed cell lymphomas. As PLB are identified to be derived from B-cell lineage, these tumors have been classified as diffuse large B-cell lymphomas in recently proposed classification of lymphomas. The therapy for PLB includes chemotherapy with or without radiation and the single most important predictor of survival is the stage of disease. The 5 year survival rate varies from 58% in patients with localized tumor to 22% in those with associated nodal or soft tissue disease. PLB may go unrecognized due to lack of specific and gross plain radiographic abnormalities. However, it should be suspected in patients with plain radiographic abnormalities in the wake of a large soft tissue mass. Proper imaging studies and biopsy usually prove diagnostic.

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