An Unusual Presentation of Primary Rib Non Hodgkins Lymphoma Masquerading as Male Breast Cancer

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Abbreviations

(IPI) International Prognostic Index scoring involves age > 60 years, ECOG(PS) > 1, abnormal LDH, extranodal sites > 1, stage III or IV
PBNHL Primary bone non hodgkins lymphoma

Introduction

Primary bone lymphoma accounts for less than 5 % of all primary bone tumors, 4–5 % extranodal lymphomas and less than 1 % of all non Hodgkin's lymphoma [1]. Males in their fifth and sixth decade are usually affected [2]. Long bones are most frequently involved, involvement of flat bones is uncommon and isolated rib involvement is very rare [3]. Diffuse large-B-cell lymphoma (DLBCL) accounts for the most common histology. Most common presentation is either painful swelling or pathological fracture. Due to its rarity, only a few retrospective studies have been published

describing the prognosis and treatment of primary bone lymphoma.

Case Report

A 52 year old male presented with pain and lump in right chest (inner and outer lower mammary quadrant) since 3 months. Lump had gradually progressed in size and was associated with pain. (Figure 1a). There was history of low grade intermittent fever. He was initially treated with a course of antibiotics with no relief. Chest X-Ray(PA) done was suggestive of expansile osteolytic lesion involving the anterior end of right 6th rib with minimal soft tissue component(Fig. 1b) CECT(Thorax) showed lytic lesion with homogenous mild contrast enhancement of soft tissue component. (Figure 2) Fine needle aspiration(FNAC) was suggestive of malignant lymphoma. Incisional biopsy showed small round cell tumor. Immunohistochemistry(IHC) on biopsy was positive for LCA and CD 20 and negative for CD3, CK , CD 99, desmin and cyclin D1(Fig. 3).

Other workup including complete blood count(CBC), biochemistry (LDH, alkaline phosphatase and uric acid), CT scan of abdomen , pelvis, bone marrow aspiration and biopsy were normal.

Bone scan was done to exclude other skeletal involvement. A final diagnosis of primary rib non hodgkins lymphoma (diffuse large B cell lymphoma type) was made. Patient was treated with combination chemotherapy(CHOP) along with Rituximab ,a monoclonal antibody against CD20 antigen at the dose of 375 mg/m². Four cycles of CHOP (Cyclophosphamide at 750 mg/m², vincristine at 1.4 mg/m²,

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Fig. 1 **a** Lump in right chest, lower mammary quadrant. (biopsy mark as shown by white arrow). **b** Chest X-Ray(PA) was suggestive of expansile osteolytic lesion involving the anterior end of right 6th rib with minimal soft tissue component

adriamycin at 50 mg/m², day 1 and prednisolone at 100 mg on day 1 to 4) every 3 weekly were given. Later it was consolidated with radiation to local site (30 Gy / 15#). Patient showed good response with no evidence of disease

Fig. 2 CECT Scan(Thorax) showed lytic lesion with homogenous mild contrast enhancement of soft tissue component

at the end of treatment. Patient is now on regular follow up and is disease free since last 1 year.

Discussion

Primary bone lymphoma was first described as a distinct clinicopathological entity in 1939 by Parker and Jackson [4]. Unlike other aggressive lymphomas, primary lymphomas of bone have a more favourable outcome. In 1950 Coley et al., defined the presence of 1) primary focus in a single bone, 2) a positive histological diagnosis of lymphoma and 3) no evidence of distant soft tissue or nodal involvement as essential criteria for diagnosis of primary bone lymphoma [5]. Our case report of primary non hodgkins lymphoma of rib is rare with only few reports published in the available literature. Due to its rarity and difference in the management, it needs to be differentiated from other causes of lytic bone lesions like metastatic carcinoma, Ewings sarcoma, osteosarcoma, eosinophilic granuloma (skeletal) and chronic osteomyelitis. Previous studies have used surgery (+/- RT) with chemotherapy but off lately chemotherapy with local radiation has shown equal outcomes owing to the systemic nature of disease. With the advent of rituximab in 2001, there has been a remarkable improvement in response rates, progression free survival and overall survival for patients with PBNHL compared with those treated earlier without rituximab (3-year PFS 88 % versus 52 %) [6]. In the present era, role of surgery is limited to biopsy and stabilization of pathologic fractures. Multivariate analysis of prognostic factors has identified three distinct groups, those with 1) age <60 years with IPI 1–3, 2) age ≥ 60 years with IPI 0–3 and 3) age ≥ 60 years with IPI 4–5 with markedly different 5-year OS of 90 %, 61 % and 25 %, respectively [7].

Primary bone lymphoma remains a systemic disease, so the optimal treatment is a systemic form of treatment that includes chemotherapy with or without radiotherapy

Fig. 3 **a** Diffuse sheets of tumor cells seen invading the paratrabeular region of bone (H&E stain; original magnification x40). **b** Small to medium sized tumor cells with pale pink cytoplasm and prominent nucleoli (H&E stain; original magnification x200). **c** crisp membranous LCA positivity in tumor cells (IHC; original magnification x100). **d** cytoplasmic plus membrane positivity for CD20 (IHC; original magnification x100)

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