PRIMARY BONE LYMPHOMA: A CASE REPORT OF A 22 YEAR YOUNG MALE AND REVIEW OF LITERATURE

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ABSTRACT

Primary bone lymphoma (PBL) is comparatively rare disease process and distinct clinical entity. Previously reported most cases showed its particular tendency to affect senior adults. Although PBL can occur at any age and site with slight male predominance but femur bone has been reported the most commonly involved single site. Majority of PBL patients have early stage disease, and the stage 1 disease is the most important prognostic factor. Majority of the case series were reported from West with a little share from Asia including Japan. We report a case of DLBCL involving the upper end of humerus in a 22 years old male with review of literature.

Keywords: Primary bone lymphoma.

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INTRODUCTION

Primary bone lymphoma (PBL) was first explained by Oberling in 1928. Before that it was known as reticulum cell sarcoma. In 1939, Jackson and Parker reported a series of 17 Cases [1]. PBL comprises of about 7% of total bone malignancies. It consists about 4-5% among all extranodal lymphomas. It is less than 1% among all malignant lymphomas. Non-Hodgkin lymphoma (NHL) is common and DLBCL is most common subtype [2]. According to WHO, PBL is that which involve single skeletal location with or without involvement of regional lymph nodes or involving multiple bones but no involvement of viscera or lymph nodes. There is slight male predominance. Majority of PBL patients had early clinical stage disease, which is the most important prognostic factor. Histopathology proved that the majority of PBL cases were DLBCL type [3].

CASE REPORT

A 22 years old young male presented with 4 months history of moderate to severe pain in right arm, fever and weight loss. X-Ray showed aggressive looking lytic osseous lesions in metadiaphyseal region of right upper humerus with soft tissue extension and gross periosteal reaction (Fig-I). MRI images revealed an enhancing abnormal signal intensity mass involving metadiaphysis of humerus with fractures seen at neck and proximal shaft. Technetium-99m (99mTC) radionuclide bone scan showed active bone pathology in region of shaft and upper end of right humerus along with two active bone lesions in left 5th & 6th ribs posteriorly.

The incisional biopsy was performed and sent to Department of Histopathology, Armed Forces Institute of Pathology (AFIP), Rawalpindi for histopathological evaluation of the case. Morphology revealed sheets of large atypical lymphoid cells with marked pleomorphism and vesicular nuclei along with prominent eosinophilic nucleoli. The atypical cells were diffusely permeating the bone (Fig-2,). Immunohistochemical evaluation of the tumor cells showed positive staining for leukocyte common antigen (LCA) and cells were negative for cytokeratin (CK), S100 protein, CD99 and osteopontin. On further sub typing, the malignant cells showed positivity for CD20 and were negative for CD3, CD79a and CD138. Ki67 index was 80%. Further workup included normal bone marrow aspiration and biopsy, and Computed Tomography (CT) scan of abdomen, chest and neck which showed right axillary lymphadenopathy. Serum lactate dehydrogenase level was within normal limits. Based on above findings, a definitive diagnosis of Primary Bone lymphoma, Non-Hodgkin Lymphoma, Diffuse Large B Cell Type of right Humerus, Ann Arbor stage IV was established.

Patient got treatment with 8 cycles of CHOP chemotherapy and post chemotherapy CT scan showed residual disease at primary site while the rest of lesions regressed. Radiotherapy of primary site

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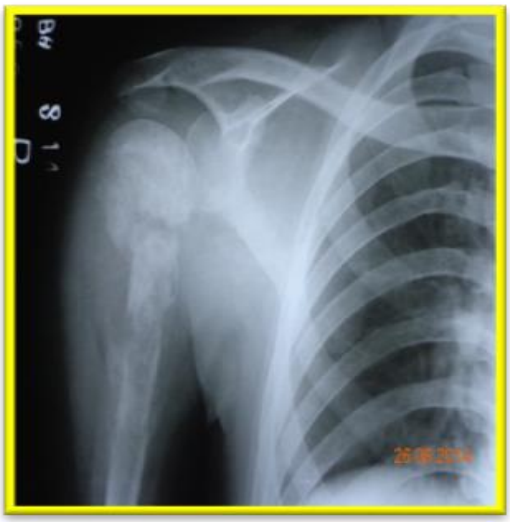
along with debulking and cementing was done and patient showed complete regression of the disease.



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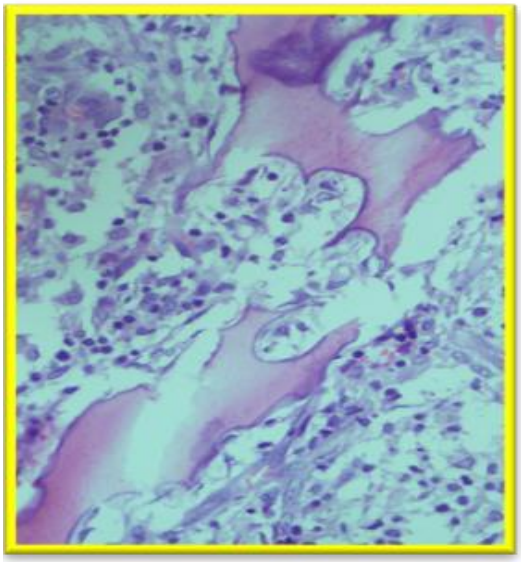

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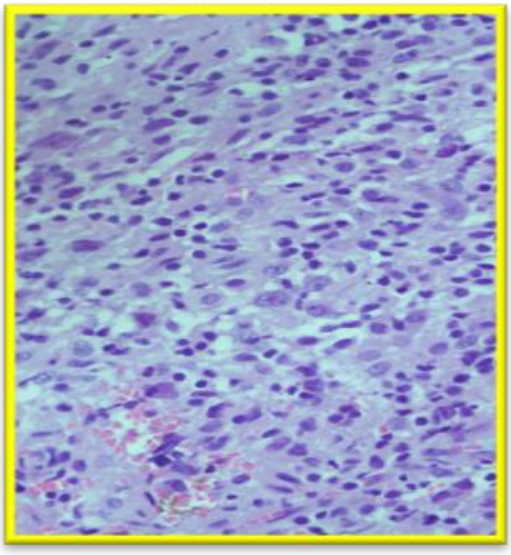

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Figures: 1. X- Ray of right humerus shaft & upper end with lytic lesions.
2. Diffuse permeative lesions of bone trabeculae.
3. Sheets of malignant lymphoid cells with vesicular nuclei & prominent eosinophilc nucleoli

DISCUSSION

Lymphomas are heterogeneous group of malignant lymphoid neoplasm formed by infiltration of malignant lymphoid cells in any tissue of the body [4,5]. Lymphomas are broadly categorized into Hodgkin lymphomas & NHLs. WHO classify NHL into B and T cell neoplasm, each is further sub divided into Immature and mature/peripheral types. Among various categories of peripheral B cell lymphomas DLBCL is most common subtype [5]. About 19.7 cases per 100,000 Population are diagnosed as NHL and almost 1/3rd die every year in USA. About 40% of all lymphomas present as extra nodal lymphomas. Less than 1% present as Primary bone lymphoma [6]. Median age is 48 years and there is a slight male predominance. The 3-year treatment failure-free survival is 72% & overall 3-year survival is 77%. Primary NHL of bone can involve any part of the skeletal system but long bones followed by the pelvic bones are the most commonly involved sites at presentation. Pain is most common presenting symptom followed by soft tissue swelling and pathological fractures. B symptoms may be present [2,7]. The variety of factors can contribute in eitiology of lymphomas including infectious causes, exposure to environmental carcinogen, immune dysfunctions, smoking, alcohol consumption, sun exposure and genetic factors [5].

Histopathology always remained the gold standard for diagnosis & IHC play vital role in exact typing of the noeplasm and exclusion of differentials. The DLBCL of bone is positive for LCA and CD20 with a high index of Ki67. The IHC panel can be modified based on morphological findings and radiological differentials [8,9]. Staging and prognostic workup includes, serum LD levels, Chest X-ray, USG abdomen, CT scan chest and abdomen, whole body bone scan and PET scan5. In PBL 6 to 8 cycles of chemotherapy are recommended. If lymphoma is CD20 positive then anti CD20 drug Rituximab is recommended. Radiotherapy is reserved for the bulky primary disease [10].

AFIP data from Jan 2010 to Dec 2014 shows only 35 cases of PBL which is comparable to regional and international studies [3,7,11].

CONCLUSION

Lymphomas involving bone are although quite uncommon but can be diagnosed and managed quite effectively with high index of suspicion and effective application of appropriate IHC panel.

AUTHORS CONTRIBUTION

Akhter Ali Bajwa; Concept and manuscript writing.
Hafeez Ud Din: Case diagnosis
Muhammad Tahir Khadim: Overall supervision and proof reading
Syed Salman Ali: Data collection and statistics
Unaiza Jamil: Photomicrography
Umair Aslam Shahzad Khan: Literature review

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