



Primary Lymphoma of the Sacrum- A Rare Entity

Amol Dilip Amonkar, Boney Perumal, Bhaskar Mallaiah*, Fayiza Musthafa

Department of Lymphoma, K S Hegde Medical Academy, Mangalore, India

Abstract

Primary lymphomas of the bone are rare neoplasm accounting for approximately 0.4% of all primary bone tumours. We report a case of 39 year old male who presented with lower backache. Computed Tomography (CT) showed a mixed sclerotic and osteolytic lesion involving predominantly the right half of sacrum with adjacent soft tissue and muscle thickening and enlarged regional lymph nodes, mimicking a sacral neoplasm. Histology of the lymph node showed features of non-Hodgkins lymphoma, immunohistochemistry confirmed the diagnosis. The patient underwent radiotherapy followed by chemotherapy (CHOP Regimen). A diagnosis of primary lymphoma of the sacrum was made as it fulfils the criteria of primary lymphoma of the bone (PBL) that is involvement of a single skeletal site with or without regional lymph node involvement. To the best of our knowledge, primary lymphoma of the sacrum is an extremely rare condition and only a handful of cases are reported till date. We report a case of primary lymphoma of the sacrum with relevant discussion on the same.

Keywords: Primary bone lymphoma; Non-Hodgkins lymphoma; Sacrum; Diffuse large B cell lymphoma; Immunohistochemistry

Abbreviations

PBL: Primary Lymphoma of Bone; FNAC: Fine Needle Aspiration Cytology; CECT: Contrast Enhanced Computed Tomography; MRI: Magnetic Resonance Imaging

Introduction

Primary Bone Lymphoma (PBL) is a rare neoplasm accounting for less than 2% of all lymphomas in adults [1] and the Diffuse Large B Cell Lymphoma (DLBCL) accounts for most of these cases [1]. PBL is defined as 1) a single skeletal site, with or without regional lymph node involvement, 2) multiple bones are involved but there is no visceral or lymph node involvement [2]. The femur and other long bones of the extremities, pelvic bone, head and neck are predominantly affected, while sacrum is a rare site of occurrence of this tumour [1]. Sacral tumours are relatively rare, accounting for 1-2% of all musculoskeletal tumours [1]. Chordoma is the most common primary sacral tumour while primary sacral lymphoma is very rare [1]. The disease occurs predominantly in elderly men and the clinical symptoms include low back pain with or without radiculopathy [1]. CT/MRI/Radiographs are helpful in the diagnosis of primary lymphomas of the bone however histopathology and immunohistochemistry are confirmatory. The treatment of choice is chemotherapy (CHOP Regimen) followed by radiotherapy [1]. The imaging findings of sacral lymphoma are similar to those of other neoplastic diseases of the sacrum, the prognosis of sacral lymphoma is relatively good and it is important to distinguish it from other neoplastic diseases [1].

Case Report

A 39 year old male presented with the history of lower backache since 1 year, which was insidious in onset, gradually progressive, no aggravating or relieving factors were noted. On physical examination an enlarged right inguinal lymph node measuring 3 cms X 3 cms, hard in consistency and mobile was noted. X-ray spine was reported as normal. FNAC of the inguinal lymph node revealed cytological features in favour of anaplastic large cell lymphoma with a differential diagnosis of poorly differentiated carcinoma, advised biopsy for confirmation. CECT Abdomen showed a mixed sclerotic and lytic lesion involving the pelvis predominating the right half of sacrum with an adjacent soft tissue and muscle thickening and multiple regional enlarged lymph nodes, (Figure 1a and b) possibility of neoplastic etiology needs consideration. A biopsy of the enlarged lymph node was suggestive of non- Hodgkin's lymphoma- anaplastic large cell lymphoma, advised immunohistochemistry for confirmation. Immunohistochemistry showed CD45-positive, CD20-positive, CD3-positive, BCL-2-positive, BCL-6-positive, KI-67 which confirmed the diagnosis

OPEN ACCESS

*Correspondence:

Bhaskar Mallaiah, Department of Lymphoma, K S Hegde Medical Academy, Mangalore, India, E-mail: bhaskar.mallaiah@gmail.com

Received Date: 20 Mar 2017

Accepted Date: 02 May 2017

Published Date: 04 May 2017

Citation:

Amonkar AD, Perumal B, Mallaiah B, Musthafa F. Primary Lymphoma of the Sacrum- A Rare Entity. *Clin Oncol.* 2017; 2: 1273.

Copyright © 2017 Bhaskar Mallaiah.

This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

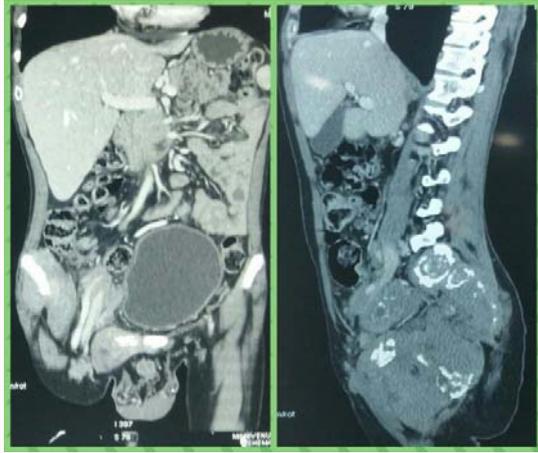


Figure 1a and b: Computed tomograph showing mixed sclerotic and lytic lesions involving the pelvis predominating the right half of sacrum with adjacent soft tissue and muscle thickening and multiple regional enlarged lymph nodes.

of a non-Hodgkin lymphoma, diffuse large B cell lymphoma germinal center type. The patient was treated with radiotherapy (30GY/10 fractions) by linear accelerator over 2 weeks followed by chemotherapy (6 cycles of CHOP regimen). Patient is now symptomatically doing well and post-treatment CECT abdomen and pelvis showed significant reduction in the size of the sacral lesion. A diagnosis of primary non-Hodgkin's lymphoma of sacrum was made as it fulfils the criteria of PBL, which is involvement of a single skeletal site with or without involvement of the regional lymph nodes.

Discussion

Primary lymphoma of the bone is rare, accounting for approximately 0.4% of all primary bone tumours [3]. Malignant lymphoma is associated with a low incidence of B symptoms such as fever and weight loss. Our patient showed no B symptoms. Laboratory findings are often non-specific including increased CRP, LDH and ALP with low frequencies of abnormal values and their diagnostic usefulness is low [1]. The most common imaging finding is an osteolytic bone destruction which is observed in approximately 70% of all patients [4]. In patients in whom osteolytic bone lesion is not appreciated, FDG-PET/CT is very useful in detecting lesions in these cases and also helps in determining the disease stage at the time of diagnosis and for assessing the treatment effects after completion of treatment [5]. Primary benign and malignant tumours of the sacrum are rare, accounting for less than 7% of all primary spinal tumours [6]. Approximately 40% of all primary sacral tumours are chordomas and primary sacral lymphoma is extremely rare [1]. Literature shows that till 2013 only 6 cases have been reported [1] so hence the incidence and prognosis have not been clearly defined.

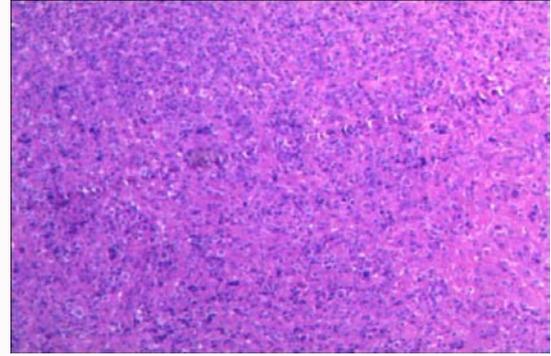


Figure 2: On microscopy, diffuse loss of architecture with replacement by highly pleomorphic tumour cells having abundant eosinophilic cytoplasm with pleomorphic nuclei having coarse chromatin and prominent nucleoli, admixed with these are lymphocytes and neutrophils suggestive of non Hodgkin's lymphoma.

Conclusion

To conclude, primary lymphoma of the sacrum is condition which exhibits features similar to other primary sacral neoplasms, definitive diagnosis can be made only with the help of histopathology and immunohistochemistry. This case of primary lymphoma of sacrum is presented because of its rarity, the incidence & the prognosis is still not yet well defined. The treatment modality is chemotherapy (CHOP Regimen) followed by radiotherapy and the recurrence rate is still unknown as only a few cases have been reported till date.

References

1. Shimada A, Sugimoto K, Wakabayashi M. Primary sacral non-germinal centre type diffuse large B-cell lymphoma with MYC translocation: a case report and a review of the literature. *Int J Clin Exp Pathol.* 2013;6(9):1919-1928.
2. Unni KK, Hogendoom PCW. World Health Organisation. Classification of tumours. Lyon, France: International Agency for Research on Cancer (IARC). Pathology and Genetics of Tumours of Soft Tissue and Bone. 2002.
3. Kelley SP, Ashford RU, Rao AS. Primary bone tumours of the spine: a 42-year survey from the Leeds Regional Bone Tumour Registry. *Eur Spine J.* 2007;16:405-409.
4. Durr HR, Muller PE, Hiller E. Malignant Lymphoma of bone. *Arch Orthop trauma Surg.* 2002;122:10-16.
5. Yamamoto Y, Taoka T. Superior clinical impact of FDG-PET compared to MRI for the follow up of a patient with sacral lymphoma. *J Clin Exp Hematop.* 2009;49:109-115.
6. Llauger J, Palmer J. Primary tumours of sacrum: diagnostic imaging. *AJR Am J Roentgenol.* 2000;174:417-424.