CASE REPORT

Primary Bone Marrow B-Cell Lymphoma: An Unusual Presentation of Non Hodgkin's Lymphoma

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ABSTRACT

Non-Hodgkin's lymphoma (NHL) presenting as primary bone marrow Lymphoma is very rare. Herewe have described a 40 year old HIV positive patient presenting with right sided parietal boneswelling.MRI brain was done which revealed bony tumor. FNAC was suspicious of lymphoproliferative disorder for which true cut biopsy was done. Biopsy was suspicious for NHL. Immunohistochemistry results were consistent with Diffuse large B Cell lymphoma, post germinal type.

Keywords: Non-Hodgkin's lymphoma, NHL, FNAC

INTRODUCTION

Non-Hodgkin Lymphoma (NHL) is more common in Human immunodeficiency virus (HIV)-infected patients than the general population.[1,2]B-cell NHL contributes to90% of all NHL cases.[3]Secondary involvement of bone marrow is relatively common in NHL in contrast primary bone marrow lymphoma is a rare presenting feature of the same.[4,5] Early diagnosis and treatment needs to be advocated as prognosis is usually poor.[6]

We describe a patient with Primary bone marrow lymphoma (PBML) who was retro positive on ART presented with scalp swelling. Due to unusual manifestation , it was initially suspected to be primary malignant bone tumor on MRI brain which was later confirmed as Diffuse Large B Cell Lymphoma (DLBCL) with Immuno-histochemistry (IHC) and biopsy.

CASE REPORT

A 40-year-old male who was Retro positive on first line ART since 2014 presented with scalp swelling in the right parietal region since 4 months. Swelling was gradually increasing in size and was associated with pain. There was no specific past history and family history. On general and systemic examination, pallor was present but there was no lymphadenopathy and hepatosplenomegaly. On local examination there was a globular swelling of size 10*10*6cm which was non tender, with uniform consistency, which was fixed to the overlying scalp and underlying skull. Swelling was non pulsatile, cough reflex was absent.

Complete blood count revealed a hemoglobin level of 6.2 g%, total leucocyte count $3 \times 10^3/\mu L$, platelet count of $106 \times 10^3/\mu L$ and differential leucocyte count with neutrophils 0%, lymphocytes 0% . MRI Brain revealed Largeheterogenous enhancing lobulated lesion in extradural right parieto-occipital region and scalp with irregular cortical outline of adjacent bone features and mass effect likely to be Malignant primary bone tumourthickness of extradural component was 1.9 cm and scalp component was 7.8*4.1 cm. Routine tests including blood coagulation profile, kidney functions, liver function tests were normal. Chest and abdominal computed tomography (CT) did not demonstrate any lymphadenopathy or hepatosplenomegaly.

True cut biopsy revealed poorly differentiated (G3) malignant tumor suspicious of Non Hodgkin's lymphoma which was confirmed by IHC which was positive for CD20 , MUM-1 and Ki-67and negative for CD3 and CD10.Detection system used for IHC was polymer HRP. Morphological and IHC features were consistent with Diffuse Large B-cell Lymphoma, post germinal type.

After confirming the diagnosis, the patient was given GM-CSF and blood transfusion to raise Hemoglobin and leukocyte count after which he was administered first cycle of R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone). As

patient was having a CD4 count of 120 and a high HIV RNA viral load 69088/mL ,patient was changed to second line ART. Patient was also given Cotrimoxazole and Fluconazole as chemoprophylaxis During follow up patient the size of the swelling has decreased 6*6*2cm. Patient was administered second cycle of chemotherapy after 21 days.

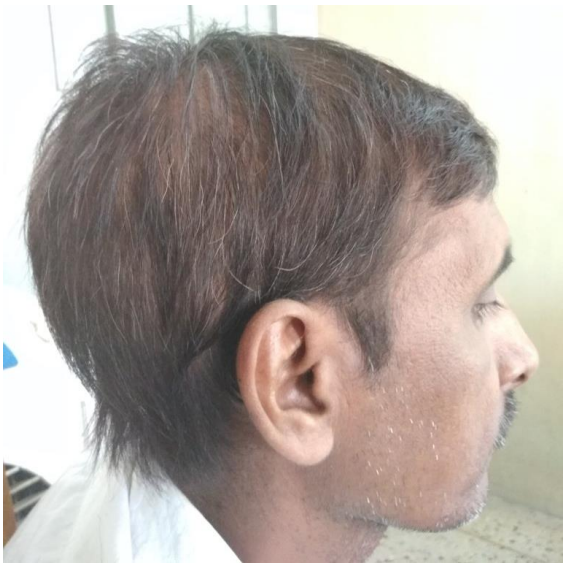

Figure 1: Case of Non Hodgkin's lymphoma

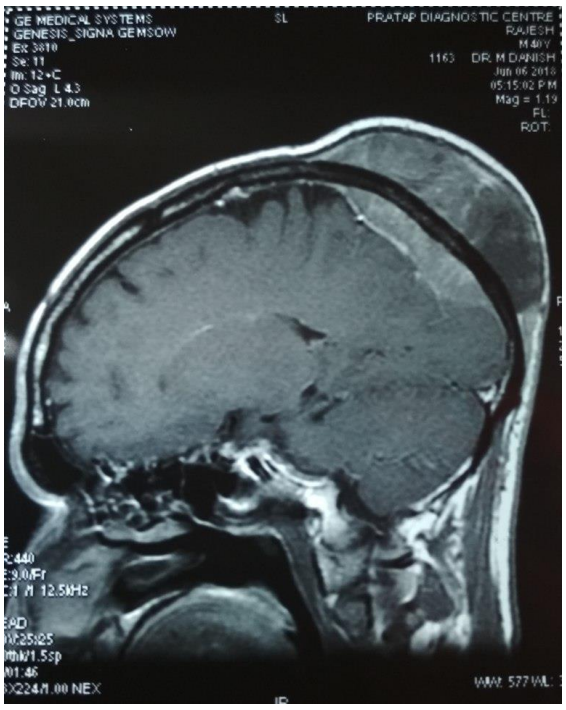

Figure 2: MRI Brain of Case

DISCUSSION

NHL is more common in the elderly than in young age.NHL is also more common in HIV infected patients than the general population. Lymphoma varies widely in its mode of presentation, variety and progression. Primary extranodal involvement occurs in approximately 40% of patients at the time of diagnosis.[3] PBML presenting with primary bone marrow involvement is uncommon.[6]Investigation like Biopsy and IHC, are useful for confirming the diagnosis of PBML.[7]

For the diagnosis of PBML, the following criteria is advocated: 1) isolated bone marrow infiltration of lymphoma cells regardless of peripheral blood involvement; 2) no evidence of lymph node, spleen, liver, or other extra bone marrow involvement on physical examination or imaging studies 3) absence of localized bone tumors; 4) no evidence of bone trabeculae destruction in the bone marrow biopsy; and 5) exclusion of leukemia/lymphoma cases.[8,9]

In our case, the patient was a 40 year old male whowas retrovirus positive on first line ART presented with a globular scalp swelling. There was no organomegaly and MRI Brain revealed malignant primary bone tumour. Complete blood count revealed bicytopenia with normal platelet count.

Truecutbiopsy was suspicious of Non Hodgkin's Lymphoma in a background of patient being retropositive. Further IHC was positive for CD20,Ki-67 and MUM-1 and negative for CD3 and CD10 which confirmatory of Diffuse B Cell lymphoma.

CONCLUSION

NHL should be taken into account while diagnosing unusual hematologic manifestation in Retropositive patients. Biopsy and IHC are more specific methods to confirm the diagnosis. Correlation of morphological findings with investigations leads to early diagnosis and treatment increasing survival for the patient.

REFERENCES

1. Dal Maso L, Franceschi S. Epidemiology of non-Hodgkin lymphomas and other haemolymphopoietic neoplasms in people with AIDS. Lancet Oncol. 2003;4:110–119.
2. Frisch M, Biggar RJ, Engels EA, Goedert JJ. Association of cancer with AIDS-related immunosuppression in adults. JAMA. 2001;285:1736–1745.
3. De Gruchy GC. Tumours of lymphoid tissues. In Firkin F, Chestermann C, Penington D (eds). De Gruchy's Clinical Haematology in Medical Practice.5th edition. Oxford University Press. 1989;257-278.
4. Kajiura D, Yamashita Y, Mori N. Diffuse large B-cell lymphoma initially manifesting in the bone marrow. Am J ClinPathol. 2007;127(5):762-769.
5. Lee Y Ja, Kim H Jae, Shin H Jeong, Kim R Hye, Lee N Jeong, Kim H Ki, Lee S Won, Joo D Young, Sohn H Chang, Kim CH. Primary isolated bone marrow diffuse large B-cell lymphoma with hemolytic anemia as the first manifestation. Korean J Hematol. 2008;43(1):48-52.

6. Nishida H, Hori M, Obara K. Primary isolated bone marrow diffuse large B-cell lymphoma with the initial presentation as severe thrombocytopenia, successfully treated with chemo-therapy: a case report and review of the literature. Journal of Cancer Therapeutics and Research. 2012;1:28.

7. Kagoya Y, Sahara N, Matsunaga T, Uekusa T, Irie S, Hat-anaka K. A Case of Primary Bone Marrow B-Cell Non Hodgkin's Lymphoma with Severe Thrombocytopenia: Case Report and A Review of the Literature. Indian J Hematol Blood Transfus. 2010;26(3):106-108.

8. Yamashita T, Ishida M, Moro H, Yumoto H, Uchibayashi S, Yoshii M, Nakanishi R, et al. Primary bone marrow diffuse large B-cell lymphoma accompanying cold agglutinin disease: A case report with review of the literature. OncolLett. 2014;7(1):79-81.

9. Martinez A, Ponzoni M, Agostinelli C, Hebeda KM, Matutes E, Peccatori J, Campidelli C, et al. Primary bone marrow lymphoma: an uncommon extranodal presentation of aggres-sive non-hodgkin lymphomas. Am J SurgPathol. 2012;36(2):296-304.