Case Report

T-cell Rich Large B-cell Lymphoma: A Rare Variant of Diffuse Large B Cell Lymphoma

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Keywords: *Non Hodgkin's Lymphoma, Diffuse large B cell Lymphoma, T-cell histiocyte rich B cell lymphoma*

ABSTRACT

T-Cell Rich-B cell Lymphoma (TCRBCL) is separately defined by World Health Organisation(WHO), it is a subtype of Diffuse Large B cell Lymphoma (DLBCL). It is an aggressive disorder treated same as that of DLBCL. But some times it may not respond to therapy and it has poor prognosis. It requires careful histopathological examination and immunohistochemistry (IHC) for confirmation. We reported a rare case of subcutaneous T cell rich B cell lymphoma, which is confirmed by IHC which shows CD20 and CD3 marker positive.

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Introduction

It is an entity, accounting for approximately 1-2% of all non-Hodgkin's lymphoma and generally believed to be a subtype of diffuse large B-cell lymphoma in the Revised European-American Lymphoma classification and upcoming WHO classification.[1] In 2008 according to WHO classification, THRBCL is included as specific subtype of diffuse large B cell lymphoma.[2] TCRBCL are recently described ,unusual non-Hodgkin lymphoma that has diffuse morphology, a predominance of reactive T-cell and minority of neoplastic cell.[3] The neoplastic large B cell constitute less than 20% to 25% of the total cell population scattered among predominant component of reactive T lymphocytes and variable number of histiocytes including epitheloid cells and occasional eosinophil in background.[4] The diagnosis of TCRBCL can be confirmed with immunohistochemical analysis because neoplastic B cell are immunoreactive for CD20 , LCA and reactive T cell are positive for CD3,CD43 and CD4.[3]

Case Report(S)

A Fifty five years old male presented with back pain since fifteen days which aggravate on sitting position and during walking .Pain radiates to para spinal region. On examination the mass of size of 8x3 cm located on left subscapular region, fixed to subcutaneous tissue. Covering skin was free, no signs of inflammation. Clinicaly diagnosed as sarcomatous lesion. MRI study of dorsolumber spine (plain and contrast) revealed, partial collapse of D2 and L1 vertebral bodies with enhancing lesion involving multiple vertebrae (i.e C3, D1, D2, D3, D6, D10, D12 , L1, L2 and L5) causing significant obliteration of subarachnoid spaces and compression of spinal cord at thelevel D2, D12, L1 and L2 (figure1,2) Lesion was diagnosed as metastatic in nature. Excision biopsy performed from subscapular mass. Biopsy mass measured 4x3x1 cm. Cut section showed greyish white appearance.(Figure 3) On Haematoxylin and Eosin stain (figure-4,5) biopsy examination showed tumour along with areas of necrosis. Tumour cells are arranged in sheets and cords. Cells are separated by thick fibrous septa. Individual tumour cells are round, large with large pleomorphic hyperchomatic nuclei, irregular nuclear margin, prominent 1-2 nucleoli and scanty cytoplasm. Tumour cells are infiltrating skeletal muscle. Vascular proliferation along with lymphocytic infiltration between tumour cells noted. Diagnosis of Diffuse Large B cell Lymphoma was given. Advised immunohistochemistry for confirmation. It was done (By polymer detection kit) and positive for B cell marker CD20 (Figure 6) and T cell marker CD3(figure7). Final diagnosis of T-cell rich Diffuse Large B cell lymphoma was reported.

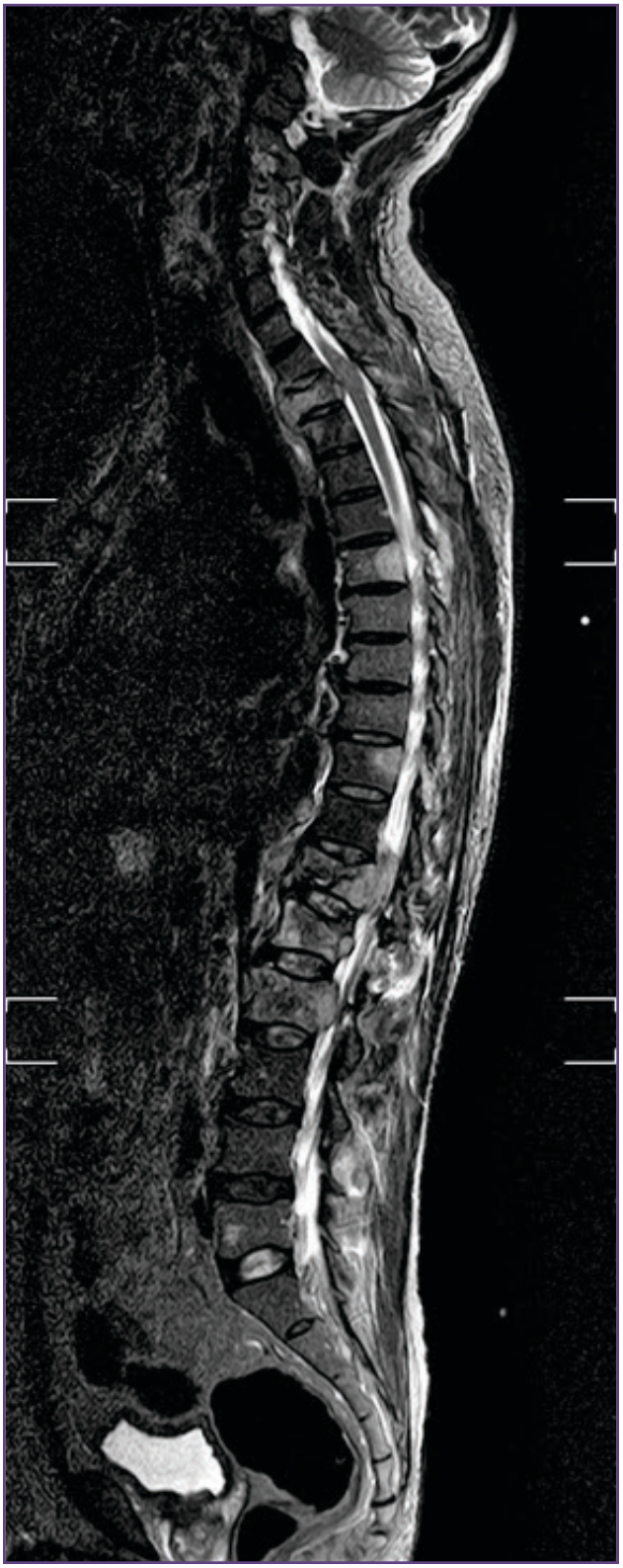

Fig. 1: STIR SAG showing fracture of D2, D12 and L2 BMP (560X1484)

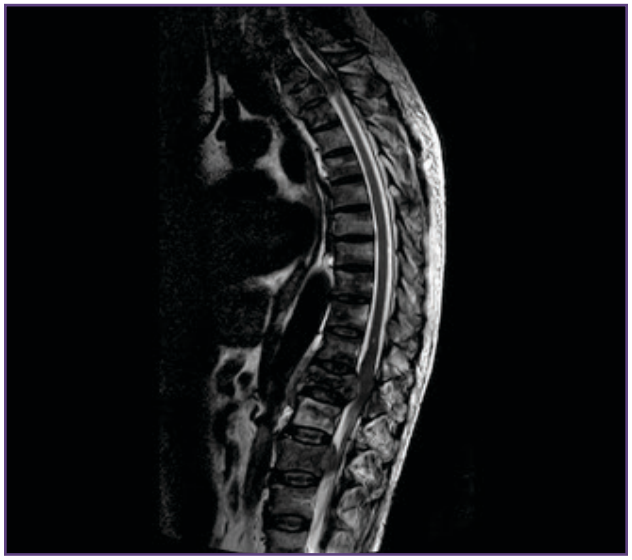

Fig. 2: T2W SAG showing multiple metastasis

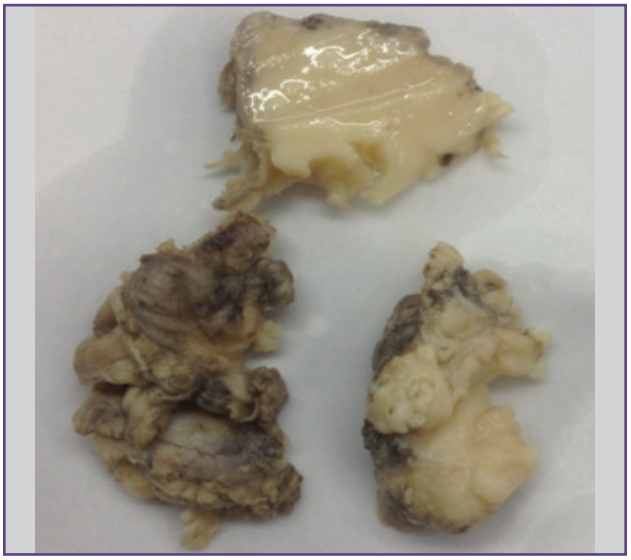

Fig. 3: Cut section shows greyish white appearance

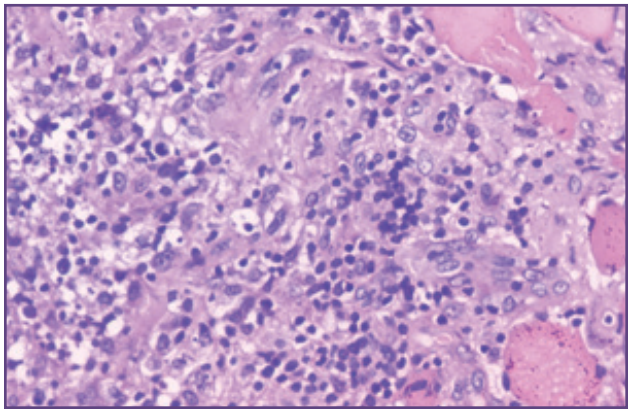

Fig. 4: H & E X 100: Shows Infiltration Large cell with infiltration of T cell on background along with subcutaneous tissue (skeletal muscle.)

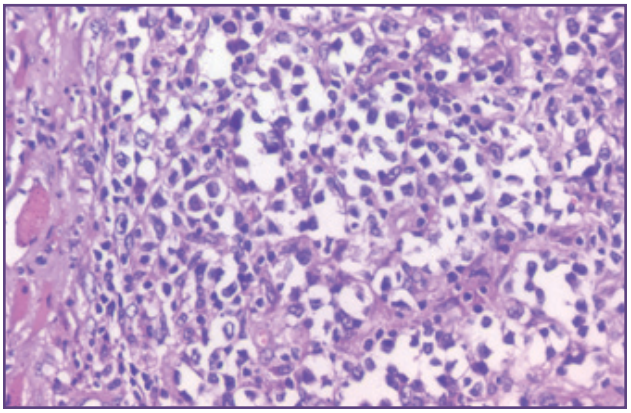

Fig. 5: Section shows Infiltration Large cell with infiltration of T cell on background with skeletal muscle (H&E, X100).

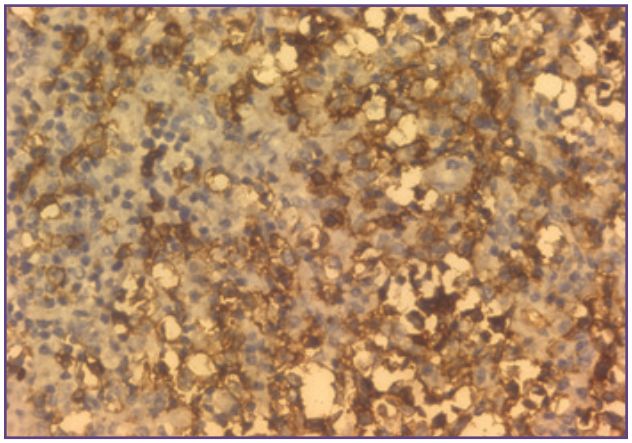

Fig. 6: CD20 staining X100: Shows membranous staining of large B cell.

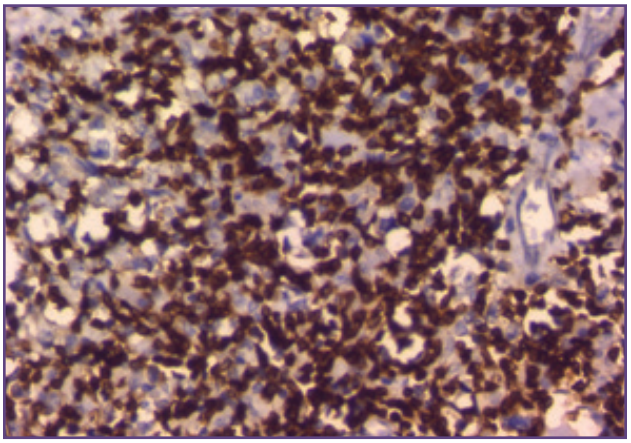

Fig. 7: CD3 staining X100: Membranous staining of reactive T cell.

Discussion

Diffuse large B cell lymphoma (DLBCL) is a most common lymphoid malignancy in adults accounting for 40% of all non-Hodgkin lymphoma.[3] T cell / histiocytic rich B cell lymphoma is uncommon morphologic variant of DLBCL and it represents 1 -3% of all DLBCLs in recent series.[3,5,6] It accounts for 1-2% of all Non Hodgkin lymphoma.[3,1] TC/HRBCL currently categorized as a variant of diffuse large B-cell lymphoma as per WHO and Revised European American classification of lymphoid neoplasm.[1,7,8] The entity was first described as "T-cell-rich B cell lymphoma" in 1988 By Ramsay et al.[5]

TCRBCL is recently described as histologic variant of B Cell Lymphoma characterized by minor population of clonal B cell distributed in background of numerically predominant polyclonal T lymphocyte.[9,10] As per WHO classification of 2001, T-cell / histiocytic-rich large B cell lymphoma (THRBCL) is defined by the presence of limited number of scattered large B cell in a background rich in T cell with or without histiocytes.[11]

Most of the patients with TCRBCL present with nodal disease involving various sites of body. Extra nodal involvement has been reported in liver, soft tissue, spleen, nesopharynx, brain, tongue, mediastinum and bone, dura, stomach.[3,4] TCHRBCL occurs at a younger age than traditional DLBCL with a median age in the fourth decade of life compared with the sixth decade for DLBCLs as a whole. A male predominance has been noted in most series.[5]

The reported mean age of occurrence of this disease is 40 years.[7]

Our patient is of older age group and presented with extra nodular subcutaneous lesion at left subscapular region. The neoplastic B lymphocyte display a heterogeneous spectrum of morphology, which includescentroblast, immunoblast, multinucleate RS like cell along with lymphocytes and histiocytes, as well as large cleaved and noncleaved cells.[4] The malignant B cell constitutes less than 10%.[2,7,8,9,12] and reactive T cell on the background constitutes more than 50%.[7,13,14]

The diagnosis of TCRBCL can be confirmed with immunohistochemical analysis because of neoplastic B cell are immunoreactive for CD20 , LCA ,CD79a[4] and reactive T cell are positive for CD3, CD43,CD4 and CD45RO positive[3, 7]

Jeremy S Abrason stated that on immunohistochemical analysis malignant B cell of TC/HRBCL shows mark positivity for CD4, CD20 and B cell transcription factor BAX/BSAP,OCT2 BOB1. The cells are uniformly negative for CD15 and rarely show weak positivity for CD30 .CD5 and CD138 are uniformly negative. BCL-6 and CD79a are expressed by majority of tumor. CD10 are positive in in only a minority of cases.[5] Our case is immunoreactive for CD20 and CD3.

First, primary cutaneous TCRBCL is so rare that only few of cases have been reported in the literature. Second, TCRBCL in general is frequently misdiagnosed as pseudolymphoma, pleomorphic peripheral T-cell lymphoma or lymphocyte-predominant Hodgkin's lymphoma.[1]

According to author, who considered this variant as an aggressive lesion and it is associated with poor prognosis by other author.[3] TCRBCL is an aggressive B cell NHL and it should be treated as high grade large cell lymphoma according to some author.[10]TCHRBCL is very aggressive disorder, which often does not respond to therapy.[11]

CHOP (Cyclophosphamide, doxorubicin, vincristine and prednisolone) chemotherapy in combination with rituximab is used in TCHRBCL treatment as it is used in all CD 20 positive nodal and extra nodal lymphoma. Response to the treatment and prognosis of cases with TCHRBCL are similar to the DLBCL cases at the same stage.[6]

Recent series have shown complete response rate to CHOP-like therapy in the range of 56- 63% with 3- year and 5- year overall survival rates, estimated at 50-64%.[5]

More than 90% of patientsdetected to have stage III or IV disease, and bone marrow involvement is common (>50%). The prognosis in such cases is poor, with 5 year survival by 20%.But this may be explainable by the high proportion of patients havingadvanced stage disease .[13]

Conclusion

We reported rare case of T cell rich large B cell lymphoma. This is an aggressive disorder, treated as high grade large cell lymphoma, which requires proper clinical evaluation, histopathologicalstudy as well as immunohistochemistry to confirm diagnosis and for proper management.

Acknowledgements

None

Funding

None

Competing Interests

None declared

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