IP Indian Journal of Neurosciences 2022;8(3):191–193

Content available at: https://www.ipinnovative.com/open-access-journals

IP Indian Journal of Neurosciences

Journal homepage: https://www.ijnonline.org/

Case Report

T cell rich B cell lymphoma of CNS in a young female- An unusual presentation

Swati Mishra[1,*], Swati Tyagi[1], Anshul Singh[1], Vatsala Misra[1]

[1] *Dept. of Pathology, Moti Lal Nehru Medical College, Prayagraj, Uttar Pradesh, India*

ARTICLE INFO

Article history:
Received 02-07-2022
Accepted 30-07-2022
Available online 03-09-2022

Keywords:
Primary CNS lymphoma
Round cell tumors of CNS
DLBCL of brain
NonHodgkin lymphoma of CNS

ABSTRACT

T cell rich B cell lymphoma (THRBCL) is described as a rare subtype of Diffuse large B cell lymphoma (DLBCL) accounting for < 10% of all DLBCL and 1-3% of all B cell lymphoma. There is a paucity of neoplastic B cells constituting < 10% of tumor and an abundance of population of non-neoplastic T cells and histiocytes. Most of the patients presents with mainly lymphadenopathy with frequent bone marrow and reticuloendothelial organ infiltration. It affects mainly middle-aged men. This is the first case report of Primary THRBCL in a young female.

1. Introduction

Primary central nervous system lymphoma (PCNSL) is belligerent extra nodal Non-Hodgkin lymphoma with most cases classified as diffuse large B-cell lymphoma (DLBCL) on histology. T cell/histiocyte rich B-cell lymphoma (THRBCL) embody an uncommon morphologic variant of DLBCL, representing 1-3% of all DLBCL and is characterized by the presence of scattered large neoplastic B-cells in a background of abundant T cells and histiocytes. The usual site of involvement is lymphoreticular system and the usual age group affected is middle age.[1] Only one case of Primary THRBCL has been reported in the Central nervous system (CNS) till date in a middle-aged male.[2] making ours the second case report of primary THRBCL involving CNS and the first case report documenting it in a young female.

2. Case History

A 17year old girl presented with complaints of altered sensorium, diplopia, vertigo and right sided weakness since one month. She was admitted to our hospital in emergency under semi-conscious condition. On examination her higher mental function were reduced with upper motor neuron type of hemiparesis noted on right side along with exaggerated reflexes. All haematological investigations were unremarkable nor did abdominal ultrasound reveal any evidence of organomegaly or lymphadenopathy. In MRI of brain, an impression of right frontal ICSOL (intracranial space occupying lesion) with differential diagnosis of high-grade glioma and lymphoma was given. Tumour excision was done and the excised tissue was sent to our department for histopathological examination.

Multiple, irregular, grey white pieces of tissue were received ranging from 0.5 x 0.5 cm to 1.5 x 1.5 cm in size. Sections examined showed a highly cellular tumor with proliferation of atypical lymphoid cells with prominent perivascular cuffing as well as infiltrating the glial tissue in sheets. They were composed of predominantly intermediate sized lymphoid cells with convoluted nuclei and high N:C ratio and occasionally conspicuous nucleoli (Centrocytes) admixed with scattered large cells with high Nuclear : Cytoplasmic ratio, vesicular chromatin and single to multiple prominent nucleoli (Centroblasts and

* *Corresponding author.*
E-mail address: drswatimishra456@gmail.com (S. Mishra).

https://doi.org/10.18231/j.ijn.2022.038

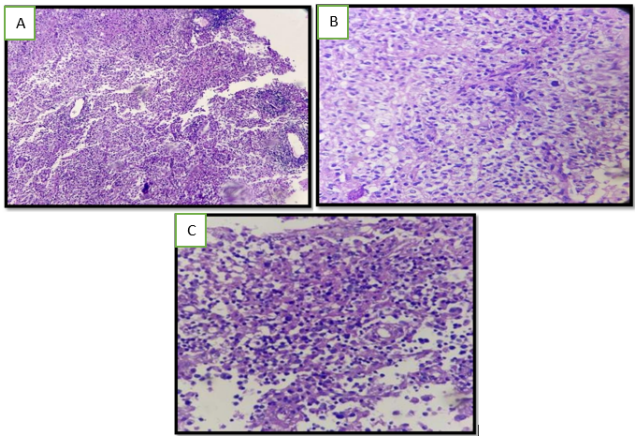

Fig. 1: A: Highly cellular tumor with proliferation of atypical large lymphoid cells and prominent perivascular cuffing (H and E 10X); **B:** Sheets of large lymphoid cells diffusely infiltrating the glial tissue. Frequent mitoses (arrow) (H & E 40X); **C:** Section showing marked vascular proliferation along with myxofibrillary degeneration (Tumor necrosis) and numerous mature lymphocytes (H & E 40X).

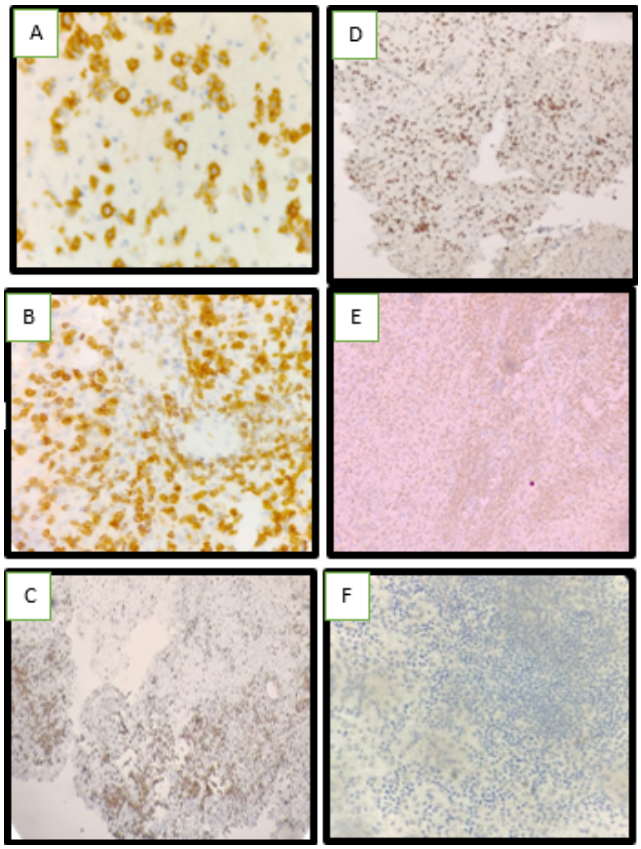

Fig. 2: A: CD 20 positivity (IHC x 4); **B:** CD 3 positivity (IHC x4); **C:** Ki67 positivity (IHC x4); **D:** BCL 2 positivity (IHC x4); **E:** CD 68 positivity (IHC x4); **F:** CD 10 negativity (IHC X4).

Immunoblasts). Many mature lymphocytes were also seen amongst the tumor cells. Frequent mitoses with areas of marked vascular proliferation along with myxofibrillary degeneration (tumor necrosis) were noted. Surrounding brain tissue showed features of reactive gliosis.

Based on the above findings a provisional diagnosis of a diffuse Large cell Lymphoma was propounded and Immunohistochemistry was advised.

The tumor cells showed positivity for LCA (CD 45), BCL-2, BCL-6, CD 20, MUM-1 and c-MYC confirming it to be a B cell lymphoma. CD 10 was negative confirming it to be of primary origin. Ki67 was high. Background showed abundant CD 3 and CD 5 positive T cells and CD 68 positive histiocytes (approximately 80% of tumor area). Thus a final diagnosis of a Primary T-cell rich B-cell lymphoma was gleaned.

Post operatively patient was treated with steroids with slight improvement but soon her condition started deteriorating again and she expired within a fortnight.

3. Discussion

Primary CNS lymphoma (PCNSL) was first expounded by Bailey in 1929 as “Perithelial sarcoma”. A rise in incidence of PCNSL from 1.5% to 6.6% has been observed largely due to AIDS epidemic. PCNSL affect all age groups with a peak seen in immunocompetent subjects during 6^{th} and 7^{th} decade of life whereas in immunocompromised patients they manifest about a decade or two earlier. 60% of PCNSL are espied in supratentorial space followed by posterior fossa (13%) and less commonly spinal cord (1%). On MRI bilateral symmetrical subependymal high signal foci are suggestive of PCNSL. The other radiological techniques used are FDG-PET scan and Thallium 201 SPECT scan needed for differential diagnosis of ring enhancing mass lesions.[1] Symptomatology includes focal neurologic deficit, neuropsychiatric symptoms, increased intracranial pressure and seizures.

Diffuse large B cell lymphomas (DLBCLs) account for more than 95% of B cell PCNSLs. THRBCL is an uncommon variant of DLBCL, representing about less than 10% of all DLBCLs.[1] It was first described by Ramsay et al in 1988[3] and later by Delabie et al in 1992 who coined the term “histiocyte rich B cell lymphoma”.[4] The median age of presentation is fourth decade of life with male predilection. It primarily affects lymph node but bone marrow, liver and spleen involvement is also frequently found.[5] On histology neoplastic B cells are seen in a characteristic angio-infiltrative pattern along with large areas of coagulative necrosis. CD 68 positive macrophages with predominant population of CD4 negative and CD8 positive T-cells are common.

In addition to pan B markers (CD19, CD20 and CD79a), they also express BCL-6, MUM-1 (90-100%) and BCL-2. CD68 and CD163 (histiocytes) positivity along with CD3

and CD5 positivity (T cells) is also noted.[1] They show PAX 5/ IgH gene rearrangement which is detected by FISH.

The other entities that share overlapping features with THRBCL are Classical Hodgkin lymphoma, Nodular lymphocyte predominant Hodgkin lymphoma, [NLPHL] and indolent Non- Hodgkin lymphoma.

In immunocompromised patients Hodgkin lymphoma comes across as closest differential wherein THRBCL may imitate it by occasional presence of Hodgkin Reed Sternberg (HRS) like neoplastic cells and dominant small T cell population but the polymorphous reactive cells seen in classical Hodgkin lymphoma are uncommon in THRBCL. The distinction can be confirmed by IHC wherein HRS cells show immunoreactivity for CD15 and CD30 and negativity for CD20, CD45 and CD79a.[6]

In immunocompetent patients NLPHL is a close mimicker of THRBCL. Although nodular pattern is observed in NLPHL compared to diffuse pattern perceived in THRBCL some literatures have encountered vague nodular pattern in the latter too. L and H cells of NLPHL share resemblance with malignant B cells of THRBCL. The immunohistochemical positivity is almost the same except PU.1, a B cell transcription marker which is frequently positive in NLPHL. Additionally CD57+ T lymphocytes and CD21 positive follicular dendritic cells are also detected in NLPHL which are commonly not glimpsed in THRBCL.[7–9]

Most of these patients present at an advanced Ann Arbor stage (stage III-IV) with an intermediate risk to high risk International Prognostic Index Score (IPI). The treatment of THRLBCL is primarily by chemotherapy (CHOP regimen) as for Primary CNS Lymphomas (PCNSL).[1] However in a recent analysis treatment with Rituximab has shown better outcomes.[10]

4. Conclusion

THRBCL is considered as an uncommon variant of DLBCL associated with an aggressive clinical course. Its presentation as a primary CNS lymphoma is exceedingly rare. It can be diagnosed appropriately only with the help of ancillary studies like IHC /molecular diagnostics hence the pathologists need to be aware of this entity while reporting CNS lymphomas. Our case is distinctive because of unusual age, gender as well as site of presentation of this rare variant of PCNSL. Its early and accurate detection is pivotal because of its adverse prognosis and close imitation with Nodular lymphocyte predominant Hodgkin lymphomas which carry a completely different etiopathogenesis, prognosis and treatment modality.

5. Conflict of Interest

The authors declare no conflict of interest with regards to the publication of this research review article.

6. Source of Funding

None.

References

1. Swerdlow SH. International Agency for Research on Cancer, World Health Organization. WHO classification of tumours of haematopoietic and lymphoid tissues. Lyon, France: International Agency for Research on Cancer; 2008.
2. Advani P, Starr J, Swaika A, Jiang L, Qiu Y, Li Z, et al. T cell/histiocyte rich large B cell lymphoma presenting as a primary central nervous system lymphoma. *Rare Tumors*. 2015;7(4):60–84.
3. Ramsay AD, Smith WJ, Isaacson PG. T-cell-rich B-cell lymphoma. *Am J Surg Pathol*. 1988;12(6):433–43.
4. Delabie J, Vandenberghe E, Kennes C. Histiocyte-rich B-cell lym- phoma. A distinct clinicopathologic entity possibly related to lymphocyte predominant Hodgkin's disease, paragranuloma subtype. *Am J Surg Pathol*. 1992;16(1):37–48.
5. A clinical evaluation of the International Lymphoma Study Group classification of non-Hodgkin's lymphoma: The Non-Hodgkin's Lymphoma Classification Project. *Blood*. 1997;89(11):3909–18.
6. Stein H, Delsol G, Pileri A. Classical Hodgkin lymphoma. In: Jaffe E, Harris N, Stein H, editors. Tumours of Haematopoietic and Lymphoid Tissues. Lyon: IARC Press; 2001. p. 244–53.
7. Stein H, Delsol G, Pileri A. Nodular lymphocyte predominant Hodgkin lymphoma. In: Jaffe ES, Harris N, Stein H, editors. Tumours of Hae- matopoietic and Lymphoid Tissues. Lyon: IARC Press; 2001. p. 240–3.
8. Achten R, Verhoef G, Vanuytsel L, De Wolf-Peeters C. Histiocyte-rich, T-cell-rich B- cell lymphoma: a distinct diffuse large B-cell lymphoma subtype showing characteristic morphologic and immunophenotypic features. *Histopathology*. 2002;40(1):31–45.
9. Torlakovic E, Tierens A, Dang HD, Delabie J. The transcription factor PU.1, necessary for B-cell development is expressed in lymphocyte pre- dominance, but not classical Hodgkin's disease. *Am J Pathol*. 2001;159(5):1807–14.
10. Batchelor TT, Grossman SA, Mikkelsen T, Ye X, Desideri S, Lesser GJ, et al. Rituximab monotherapy for patients with recurrent primary CNS lymphoma. *Neurology*. 2011;76(10):929–30.

Author biography

Swati Mishra, Junior Resident

Swati Tyagi, Junior Resident

Anshul Singh, Associate Professor

Vatsala Misra, HOD

Cite this article: Mishra S, Tyagi S, Singh A, Misra V. T cell rich B cell lymphoma of CNS in a young female- An unusual presentation. *IP Indian J Neurosci* 2022;8(3):191-193.