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Review Article

The Blushing Broadcast-Splenic Diffuse Red Pulp Small B Cell Lymphoma

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Abstract

Splenic diffuse red pulp small B cell lymphoma is a mature splenic B cell lymphoma characterized by diffuse infiltration of splenic red pulp with miniature, monomorphic B lymphocytes and configures as a provisional entity of splenic B cell lymphoma/leukaemia, unclassifiable within the contemporary WHO classification of lymphomas. Neoplastic cells depict a villous morphology and appear to invade sinusoids of bone marrow and peripheral blood. Lymphoma is determined as stage IV disease with incrimination of spleen, bone marrow and peripheral blood. Segregation from disorders such as hairy cell leukaemia, hairy cell leukaemia variant and splenic marginal zone lymphoma is necessitated. Median age of disease emergence appears at 65.5 years to 77 years. A male predominance is observed with male to female proportion of 1.6 to 2.4:1. Splenic diffuse red pulp small B cell lymphoma demonstrates chromosomal del 7(q), partial trisomy 3q, trisomy 18 or del 17p. Peripheral smear exhibits miniature, villous lymphocytes with basophilic cytoplasm, clumped nuclear chromatin, smooth nuclear outline and a polar distribution of broad cytoplasmic extensions. Cytological examination delineates monomorphic, miniature to intermediate lymphocytes imbued with spherical to elliptical nuclei with compact, clumped nuclear chromatin and occasional, distinct, miniature nucleoli. Morphologically, spleen delineates diffuse incrimination of red pulp cords and sinusoids by monomorphic lymphoma cells. Bone marrow exhibits an intra-sinusoidal pattern of tumour infiltration. Minimal lymphocytosis, thrombocytopenia or leukopenia is observed. Splenic diffuse red pulp small B cell lymphoma appears immune reactive to CD20, PAX5, CD79a and DBA-44. Segregation from hairy cell leukaemia, hairy cell leukaemia variant or splenic marginal zone lymphoma is necessitated. Rituximab may be beneficially employed for cogent therapy.

Keywords: Leukaemia; World Health Organization (WHO)

Introduction

Splenic diffuse red pulp small B cell lymphoma is a mature splenic B cell lymphoma characteristically demonstrating diffuse infiltration of splenic red pulp with miniature, monomorphic B lymphocytes. The cells commonly depict a villous morphology and appear to invade sinusoids of bone marrow and peripheral blood.

Splenic diffuse red pulp small B cell lymphoma configures as a provisional entity of splenic B cell lymphoma/leukaemia, unclassifiable within the contemporary World Health Organization (WHO) classification 2017. Lymphomas depicting inadequate or non confirmatory features may be nomenclated as splenic B cell lymphoma/leukaemia, unclassifiable. The lymphoma is additionally designated as splenic lymphoma with villous lymphocytes or splenic diffuse red pulp lymphoma.

The provisional splenic diffuse red pulp small B cell lymphoma, configuring an entity within splenic B cell lymphoma/leukaemia,

unclassifiable along with hairy cell leukaemia variant (HCL-v) exhibits an indolent clinical course.

The lymphoma is frequently diagnosed as stage IV disease with incrimination of spleen, bone marrow and peripheral blood. Microscopically, the lymphoma characteristically enunciates miniature to intermediate B lymphocytes which diffusely invade red pulp cords of splenic sinuses. Peripheral blood smear delineates small lymphocytes with villous cytology. Bone marrow biopsy exhibits intra-sinusoidal dissemination of lymphoma cells.

The lymphoma necessitates distinction from disorders such as hairy cell leukaemia, hairy cell leukaemia variant and splenic marginal zone lymphoma.

Splenic diffuse red pulp small B cell lymphoma is an infrequent condition configuring < 1% of non-Hodgkin lymphomas and ~10% of splenic B cell lymphomas [1,2].

Generally, adult subjects > 40 years are incriminated with median age of disease emergence at 65.5 years to 77 years. A male predominance is observed with male to female proportion of 1.6 to 2.4:1 [1,2].

Splenic diffuse red pulp small B cell lymphoma commonly incriminates spleen, bone marrow and peripheral blood. Regional or disseminated lymph nodes are exceptionally involved [1,2].

Splenic diffuse red pulp small B cell lymphoma demonstrates chromosomal del 7(q), partial trisomy 3q, trisomy 18 or del 17p. BRAF V600E genetic mutation is absent. Genomic mutations within NOTCH1, MAPK21 or TP53 genes are associated with progressive disease. Additionally, CCND3 genetic mutations are documented.

Majority of instances exhibit massive splenomegaly. Typically, cogent clinical symptoms are represented with stage IV disease along with incrimination of peripheral blood and bone marrow. Enlargement of splenic hilar lymph nodes is frequent [1,2].

B clinical symptoms associated with non Hodgkin's lymphoma as pyrexia, night sweats and >10% loss of body weight are exceptionally encountered. An estimated 10% implicated subjects depict erythematous and pruritic cutaneous papules [2,3].

Discussion and Conclusion

Peripheral smear examination exhibits miniature, villous lymphocytes pervaded with clumped nuclear chromatin and smooth nuclear outline. Cellular cytoplasm is basophilic and demonstrates a polar distribution of broad based, cytoplasmic extensions [2,3].

Upon cytological examination, monomorphic population of miniature to intermediate lymphocytes is discerned. The lymphocytes are imbued with spherical to elliptical nuclei with compact, clumped nuclear chromatin and occasional, distinct, miniature nucleoli [3,4].

Upon gross examination, spleen is diffusely enlarged. Cut surface is homogenous and demonstrates a beefy, reddish/brown hue. Sub-capsular region exhibits wedge shaped infarcts [3,4].

Upon microscopy, the spleen is diffusely incriminated and delineates involvement of red pulp cords and sinusoids by monomorphic, neoplastic cells. Blood lakes appear to be layered with lymphoma cells. White pulp is devoid of lymphomatous involvement [3,4].

Residual lymphoid nodules are composed of T lymphoid cells. Exceptionally, residual lymphoid nodules may be confined to white pulp. Besides, secondary effacement or obliteration of white pulp architecture may ensue [3,4].

Bone marrow preponderantly exhibits an intra-sinusoidal pattern of tumour infiltration. Occasionally, an interstitial or nodular configuration of neoplastic infiltration may be discerned. Lymphoid follicles are absent. Mild fibrosis of the marrow may ensue [3,4].

Splenic diffuse red pulp small B cell lymphoma appears immune reactive to CD20, PAX5, CD79a and DBA-44. MIB1 proliferative index is minimal.

Splenic diffuse red pulp small B cell lymphoma is immune non reactive to tartrate resistance acid phosphatase (TRAP), CD3, CD5, CD10, CD11c, CD23, CD25, CD103, CD123, cyclin D1, Annexin A1 or BCL6.

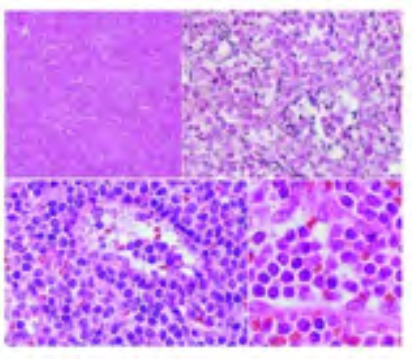

Figure 1: Splenic diffuse red cell small B cell lymphoma depicting diffuse infiltration of red pulp with monomorphic, miniature to intermediate lymphoid cells with spherical to elliptical nuclei and clumped nuclear chromatin. Blood lakes are lined by lymphoma cells [9].

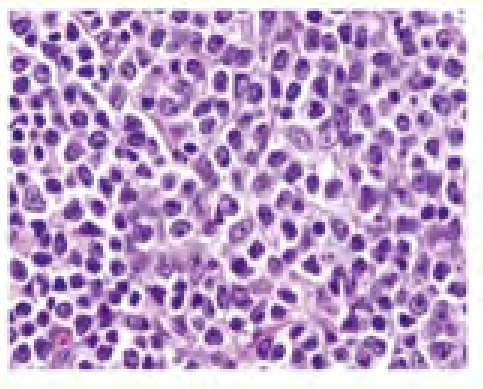

Figure 2: Splenic diffuse red pulp small B cell lymphoma delineating a monomorphic population of miniature to intermediate lymphoid cells with spherical to elliptical nuclei, clumped nuclear chromatin and occasional, distinct nucleoli [10].

	Peripheral blood	Spleen	Bone marrow	Immuno-phenotype	Chromosomal aberrations	Altered genes
Splenic diffuse red pulp lymphoma	Polar, broad based, small villous projection, condensed chromatin	Red pulp, blood lakes (possible)	Intrasinusoidal, interstitial ±, nodular ± (no follicles)	CD25-, CD103 ±, CD123-, DBA44 ±, Annexin A1-, Cyclin D3+, IgG+	Uncommon trisomy 3q, trisomy 18, del 7q	CCND3, IGHV3-4, BCOR, MAP2K1
Hairy cell leukaemia-variant	Circumferential, shorter villous projections, prominent nucleoli	Red pulp, white pulp effacement, blood lakes (uncommon)	Intrasinusoidal, interstitial, normal fibrosis	CD25-, CD103+, CD123-, DBA44+, Annexin A1-, TRAP ±	del 17p, del 7q, gain of 5	MAP2K1, IGHV4-34, U2AF1, ARID1A, TP53
Hairy cell leukaemia	Circumferential, long hairy projections, oval nucleus, inconspicuous nucleolus	Red pulp, blood lakes (common)	Interstitial, diffuse, prominent fibrosis (dry tap)	CD25+, CD103+, CD123, DBA44+, Annexin A1+, CD200+, Cyclin D1 ±	Chromosome 5 and 7 anomalies	BRAF V600E
Splenic marginal zone lymphoma	Polar shorter villi, condensed chromatin	White pulp with marginal zone expansion	Nodular ±, interstitial ±, intrasinusoidal ± (residual follicles)	CD25 ±, CD103 ±, CD123, DBA44 ±, Annexin A1-, IgD+	Gain of 3q, del 7q	NOTCH2, IGHV1-2, KLF2

Table 1: Differential diagnosis of splenic diffuse red pulp small B cell lymphoma [3,4].

Upon flow cytometry, lymphoma cells depict surface IgM+D, M alone, M+G or G alone and an equivalent distribution of immunoglobulin light chains kappa and lambda. Intense reactivity to CD20, CD22, moderate reactivity to CD11c and partial reactivity to CD103 is encountered. Besides, CD123, CD25, CD38, CD24, CD27, CD5 or CD23 appear non reactive [5,6].

Appropriate discernment of splenic diffuse red pulp small B cell lymphoma mandates assessment of constellation of clinical features, peripheral blood smear and bone marrow along with morphological, immuno-phenotypic and cytogenetic evaluation of spleen. Additionally, splenectomy may be necessitated for disease confirmation. Haematological parameters display minimal lymphocytosis, thrombocytopenia and leukopenia. Anaemia or pancytopenia is exceptionally discerned [5,6].

Upon radiographic imaging, massive splenomegaly is encountered.

The optimal, recommended therapeutic option for treating splenic diffuse red pulp small B cell lymphoma is splenectomy. Additionally, the indolent disorder may be subjected to simple observation [6,7].

Lymphoma cells appear resistant to diverse chemotherapeutic agents, in contrast to hairy cell leukaemia, hairy cell leukaemia variant and splenic marginal zone lymphoma. Nevertheless, rituximab may be beneficially employed [6,7].

Splenic diffuse red pulp small B cell lymphoma demonstrates an indolent biological course and is resistant to alleviation by diverse treatment options. 5 year survival appears at an estimated 93%. On account of resistance to conventional chemotherapeutic agents and superior prognostic outcomes, segregation from similar conditions as hairy cell leukaemia, hairy cell leukaemia variant and splenic marginal zone lymphoma is necessitated [7,8].

Neoplasms with chromosomal mutations within NOTCH1, MAP2K1 and TP53 genes document decimated progression free survival [7,8].

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9. Image 1 Courtesy: Haematologica.com

10. Image 2 Courtesy: Hindawi.com