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Splenic diffuse red pulp small B-cell lymphoma with overlapping clinical and immunophenotypic features with hairy cell leukaemia

Background

Splenic diffuse red pulp small B-cell lymphoma accounts for less than 1% of all lymphoid malignancies with peripheral blood involvement. The main diagnostic challenge is to differentiate splenic diffuse red pulp small B-cell lymphoma from other splenic B-cell lymphomas and leukaemias including hairy cell leukemia, splenic marginal zone lymphoma, and splenic B-cell lymphoma/leukaemia with prominent nucleoli, due to high overlapping clinical, morphologic and phenotypic features in the absence of splenectomies for diagnostic purposes.

Case presentation

We describe a case of a 78-year-old woman who presented with weight loss, pancytopenia, lymphocytosis, and splenomegaly. Peripheral blood smear showed a population of small to medium-sized lymphocytes having oval nuclei, condensed chromatin and villous cytoplasmic projections. Bone marrow showed B-cell infiltrate predominantly in intrasinusoidal pattern. Immunophenotype was similar to that of a hairy cell leukemia, except for the lack of expression of cyclin D1 and BRAF. This was further supported by the lack of BRAF p.V600E mutation by polymerase chain reaction. A diagnosis of splenic diffuse red pulp small B-cell lymphoma was thus favored.

Conclusion

Awareness of this infrequent clinical and immunophenotype presentation of splenic diffuse red pulp small B-cell lymphoma is essential for diagnosis and treatment.

Keywords

Splenic B-cell lymphomas and leukaemias; Splenic diffuse red pulp small B-cell lymphoma; Hairy cell leukaemia; Splenic B-cell lymphoma/leukaemia with prominent nucleoli; Splenic marginal zone lymphoma.

Biography

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Hematopathologist and adjunct assistant professor at Department of Pathology and Laboratory Medicine, Royal Victoria Regional Health Center and Department of Laboratory Medicine and Pathobiology, University of Toronto, Canada, Ontario, Canada.

Dr. Hanna completed a residency training in hematologic pathology at the University of Ottawa in Canada. In addition, she earned a PhD in experimental medicine at Université Laval in Quebec, Canada