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Case Report:

B-cell Prolymphocytic Leukemia in a Young Male.

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Abstract: B-cell prolymphocytic leukemia [B-PLL] is a neoplasm of B prolymphocytes affecting the peripheral blood, bone marrow and spleen. The principal disease characteristics are massive splenomegaly with absent or minimal peripheral lymphadenopathy and a rapidly rising lymphocyte count. Here, we report a case of B-PLL in a 42 year old male who had come for routine health check up.

Key Words: B-PLL; Prolymphocyte; Massive splenomegaly; Immunophenotyping.

Introduction:

B-cell prolyphocytic leukemia is an extremely rare disease, comprising approximately 1% of lymphocytic leukemias.[1] It needs to be differentiated from T-cell prolymphocytic leukemia for theurapetic and prognostic purposes. Differentiation can be made by a comprehensive approach taking into account the clinical features, the cell morphology and the immunophenotype of leukemic cells.[2] B-cell usually affects elderly males over 50 years of age. The principal disease characteristics are massive splenomegaly with absent or minimal peripheral lymphadenopathy and a rapidly rising lymphocyte count. Immunological markers show a B cell phenotype such as strong expression of surface IgM+/- IgD and Bcell antigens such as CD19,CD20,CD22, CD79a and b.[1,2] Here, we present the clinico-pathologic features of a case of B-PLL with emphasis on the diagnostic features and differential diagnosis.

Case Report:

A 42 years old man had come to our centre for routine health check up. On physical examination, massive splenomegaly and minimal posterior cervical lymphadenopathy were observed. Laboratory investigation revealed hemoglobin of 12.3g/dl, total leucocyte count of 12x103/cu.mm and platelet count of 71x103/cu.mm. Blood biochemistry was normal except for mildly elevated lactate dehydrogenase level. Peripheral blood smear showed 70% atypical lymphoid cells [prolymphocytes] which were medium sized with regular round nucleus, moderately condensed nuclear chromatin, a prominent central nucleolus and a relatively small amount of faintly basophilic cytoplasm [Figure 1]. Bone marrow aspirate smears showed predominantly atypical lymphoid cells with similar morphology as peripheral blood cells [Figure 2]. There was marked reduction of normal hematopoietic cells. On flow cytometric immunophenotyping, the atypical lymphoid cells were positive for CD19, CD20, CD22, CD23, CD45 and negative for CD2, CD3, CD4, CD5 and ZAP-70. Taking into account the clinical, morphological and immunophenotypic features, a diagnosis of B-PLL was made. Patient was treated with Cytoxan, Adriamycin, Vincristine and Prednisolone regimen. A repeat count was done 6 months after chemotherapy. It showed a total leucocyte count of 9,000 cells/cu.mm with the absence of prolymphocytes. The patient is doing well, 20 months after diagnosis and therapy.

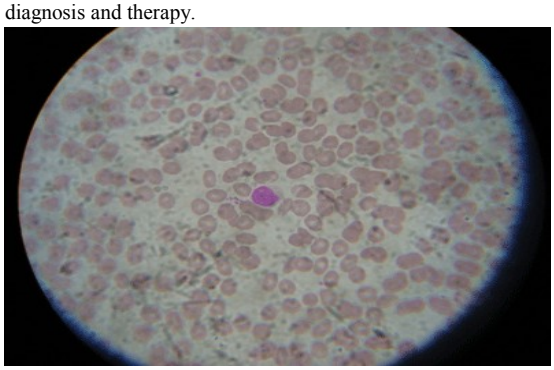

Figure 1: Peripheral blood smear showing prolymphocytes which have moderately condensed nuclear chromatin and a prominent nucleolus [Leishmanx100X]

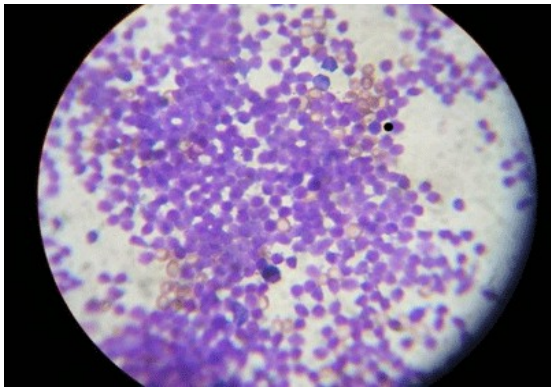

Figure 2: Bone marrow aspirate smear showing numerous prolymphocytes in diffuse sheets [Leishmanx 100X].

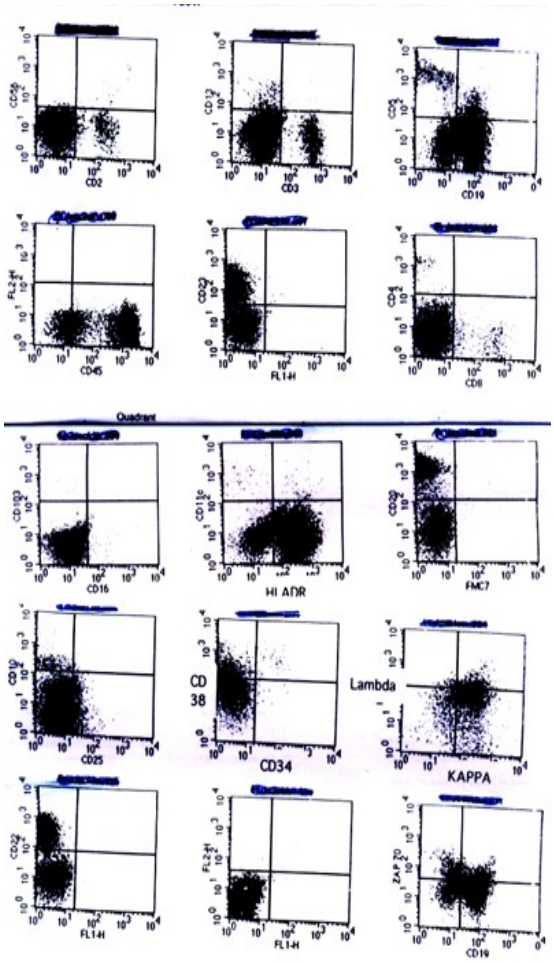

Figure 3: Dot plots showing immunoreactivity pattern of prolymphocytes in B-cell prolymphocytic leukemia

Discussion:

Prolymphocytic leukemia was first described in 1974 by Galton, as a rare variant of chronic lymphocytic leukemia.[3-6] It is a rare chronic chronic lymphoproliferative disorder that includes two subtypes, B cell and T cell, each with its own distinct clinical, laboratory and pathological features.[4] Most patients are over 60-year old, with a median age of 65-69 and similar male: female distribution.[1]

The key features of B-PLL are massive splenomegaly with absent or minimal peripheral lymphadenopathy and a rapidly rising lymphocyte count, usually over 100x109/L. Anemia and thrombocytopenia are seen in about half the number of cases.[1,3,7] T-PLL patients usually present with generalized lymphadenopathy and skin lesions.[1] Morphologically, majority (>55% and usually >90%) of the circulating cells are prolymphocytes; that is, medium sized cells (twice the size of a small lymphocyte), with a round nucleus, moderately condensed nuclear chromatin, a prominent central nucleolus and a relatively small amount of faintly basophilic cytoplasm. In contrast, the cells of T-PLL are small to medium-sized lymphoid cells with agranular basophilic cytoplasm,and have a markedly irregular nuclei.[1,2,8] Typically, B-PLL is differentiated from chronic lymphocytic leukemia [CLL] and CLL/PL with 55% prolymphocytes being a key criteria. B prolymphocytes are more uniform and have a more regular nuclear outline than those of CLL/PL. Also, CLL/PL have 10-55% prolymphocytes and a variable number of plasmacytoid lymphocytes.[4] The distinction from Hairy cell leukemia [HCL] variant is based mainly on the appearances of the cytoplasm. In HCL variant, the cytoplasm is more abundant and distinctly villous, whereas in B-PLL it is generally smooth.[1,2]

The bone marrow shows interstitial or nodular infiltrate of nucleolated cells with an intertrabecular distribution.[1,8] In the present case, the bone marrow was diffusely infiltrated by prolymphocytes with marked suppression of normal hematopoietic elements. The cells of B-PLL strongly express B-cell antigens such as CD19, CD20, CD22, CD79a and CD79b. In this case, the patient strongly expressed CD19, CD20, CD22, CD23 and CD45. The cells of T-PLL express CD2, CD3 and CD7. The reported median survival is 3 to 4 years for patients with prolymphocytic leukemia and 8 years for those with CLL. Patients with T-PLL have even poorer prognosis than those with B-PLL.[1,2,8] B-PLL responds poorly o therapies for CLL and T-PLL. Hence, typing PLL is very critical for both therapeutic and prognostic purposes.

In conclusion, a precise diagnosis by means of a comprehensive approach involving clinical, morphological and immunophenotypic features are extremely critical in this era for patient management and prognostication. Also, this case illustrates the importance of immunophenotyping as an adjunct to morphology in the diagnosis of chronic lymphoproliferative disorders.

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