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Title: An unusual presentation of Hairy cell Leukemia

Short running title: Hairy cell leukemia.

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Title: An unusual presentation of Hairy Cell Leukemia

Dear Editor,

The aberrant expression of CD5 in both hairy cell leukemia (HCL) and HCL-variant (HCL-v) is very rare, only 26 cases of have been reported in the literature (1). Simultaneous absence of splenomegaly and cytopenia(s) is even rarer which may pose a diagnostic dilemma. We described a case of CD5 positive HCL with absence of splenomegaly and cytopenia. To the best of our knowledge, only one case of HCL without cytopenia and splenomegaly has been reported in literature till date (2), but without CD5 positivity. Our patient was a 59 year-

male, presented with intermittent cough with expectoration for last 3 to 4 years with no history of fever.

Radiological investigations including X ray and computed tomography scan were normal. Complete blood counts showed hemoglobin of 15.1 g/dL, white blood cell count of 7.73×10^9 /L (neutrophils 52%, lymphocytes 45%, Monocytes 2%) and platelet counts of 153×10^9 /L. Peripheral blood smear (PBS) and bone marrow aspirate (BMA) showed 10% and 24% abnormal lymphoid cells, respectively (fig. 1 A). These cells were small to medium in size, with abundant pale blue cytoplasm and circumferential hairy projections. BM biopsy showed interstitial aggregates of abnormal lymphoid cells (fig. 1 B) which were CD20 and Annexin 1 positive. Flowcytometric immunophenotyping (fig. 1 C) revealed these cells positive for CD19, CD20, CD22, CD103, CD11c, CD123, CD25, CD5 (heterogenous), CD200, CD23 (dim), kappa and negative for CD10, FMC7. The patient was found to be positive for BRAF V600E mutation. Diagnosis of hairy cell leukemia with aberrant CD5 was given.

HCL is an indolent small mature B lymphoid malignancy accounting for 2% of lymphoid leukaemias (3). Three most important findings for diagnosis are: splenomegaly, cytopenia(s) and BM dry tap resulting from marrow fibrosis (4). In unsuspected cases with unusual presentation, the best approach for the diagnosis is by careful examination of morphological details on PBS and BM to identify the morphologic features of hairy cells which further confirmed on characteristic immunophenotypic profile, as in our case. Differential diagnoses of HCL include chronic lymphocytic leukemia, prolymphocytic leukemia, splenic marginal zone lymphoma(SMZL), HCL-v, and the mantle cell lymphoma, which can be excluded based on characteristic morphologic and immunophenotypic features. Hairy cells are 10–15 μ m in diameter, with central or eccentric round, oval or indented nuclei, reticular or netlike chromatin pattern, indistinct or absent nucleoli, pale blue cytoplasm with fine, hair-like projections or ruffled borders, stain positively for tartrate resistant acid phosphatase (TRAP) (5). Typical combination of immunophenotypic markers expressed by hairy cells as CD19, CD22, and CD79b, with brighter expression of CD20, along with co-expression of CD103, CD123, CD25 and CD11c, confirms the diagnosis (6).

To conclude, this case posed a diagnostic challenge as patient had no cytopenias or splenomegaly along with CD5 positivity. The importance of this case is to create awareness of this uncommon presentation of HCL and to emphasize that the best approach of diagnosing HCL is to give careful attention to morphological detail while interpreting peripheral blood, as in our case, which prompted detailed evaluation on bone marrow with immunophenotyping in such cases for early diagnosis and management of the patient.

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Figure Legends:

Fig 1: Bone marrow aspirate showing hairy cells (A: arrow), Bone marrow biopsy (B) showing abnormal lymphoid cell infiltration which were positive for CD20, Annexin A1. C, immunophenotyping showing CD45 bright positive further gated CD19 positive abnormal lymphoid cells which were positive for CD20, CD22, CD103, CD11c, CD25, CD5, CD123, CD200, kappa and negative for CD10.

