

Hairy cell leukemia-variant

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ABSTRACT

Hairy cell leukaemia variant is a very rare chronic lymphoproliferative disorder and is closely related to hairy cell leukemia. We hereby describe a case of hairy cell leukaemia variant for the first time in Saudi Arabia. An elderly Saudi man presented with pallor, massive splenomegaly, and moderate hepatomegaly. Hemoglobin was 7.7 g/dl, Platelets were 134 x10⁹/l and white blood count was 140x10⁹/l with 97% being abnormal lymphoid cells with cytoplasmic projections. The morphology, cytochemistry, and immunophenotype of the lymphoid cells were classical of hairy cell leukaemia variant. The bone marrow was easily aspirated and findings were consistent with hairy cell leukaemia variant.

Keywords: Hairy cell leukaemia-variant, chronic lymphocytic leukaemia, splenic lymphoma with villous lymphocytes.

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Prolymphocytic variant of Hairy Cell Leukaemia (HCL) or Hairy Cell Leukaemia variant (HCL-V) is a very rare chronic lymphoproliferative disorder.¹⁻⁴ It is closely related to HCL. In contrast to HCL, these cases have high white cell count and are characteristically CD25 negative. Compared to Chronic Lymphocytic Leukaemia (CLL), these are CD5 negative.²⁻⁵ Keeping in view the rarity of this disorder, we hereby report a case of HCL-V.

Case Report. We received EDTA blood and bone marrow samples of a 70-year old Saudi male, admitted in King Fahd Hospital Al-Hafof with complaints of easy fatigability, pallor and abdominal distension for hematological analysis. He had pallor, huge splenomegaly reaching up to right iliac fossa, 6cm enlargement of liver below right costal margin and very small lymph nodes bilaterally in axilla and groin. The CBC findings were Hb 7.7 gm/dl, Platelets 134 x10⁹/l, and WBC 140 x10⁹/l. Blood film showed that 97% white cells were large lymphoid cells with pale blue cytoplasm. These cells

had cytoplasmic projections/villi/hairs (Figure 1a and b). Compared to classical HCL, these cells had high nuclear to-cytoplasmic ratio. Many binucleated cells were also present and nucleus was bilobed in few. The cells formed loose clumps at places (Figure 1b). The other cells were neutrophils (2%), eosinophils (1%) and rare monocytes. The bone marrow aspiration slides showed hypercellular marrow. The dominant cell in the marrow was again atypical lymphoid cell. These constituted 86% of all marrow cells. Their morphology was similar to those in blood film except villi/projections were slightly less marked. Prominent nucleoli were seen in majority of lymphocytes. Other cells were myeloid cells 11%, monocyte macrophages 1.5% and normoblasts 1.5%. Occasional megakaryocytes were also present. Cytochemistry- PAS negative; Tartrate resistant acid phosphatase (TRAP)- negative. The results of FAC SCAN immunophenotypic analysis on blood sample showed: CD3 5%, CD5 8%, CD14 12 %, CD19 93%, CD20 94%, CD22 94%, CD23 22%, CD25 0%, Kappa 77%, Lambda 2%. This phenotype is typical

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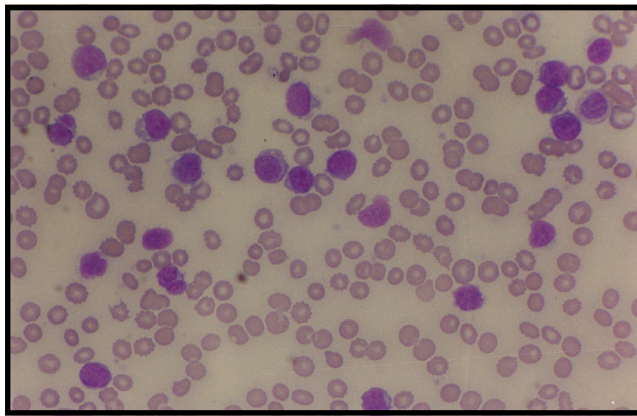


Figure 1a - Blood film showing lymphocytosis. Majority have cytoplasmic projections. Two binucleated lymphocytes are also seen (MGG x 400).

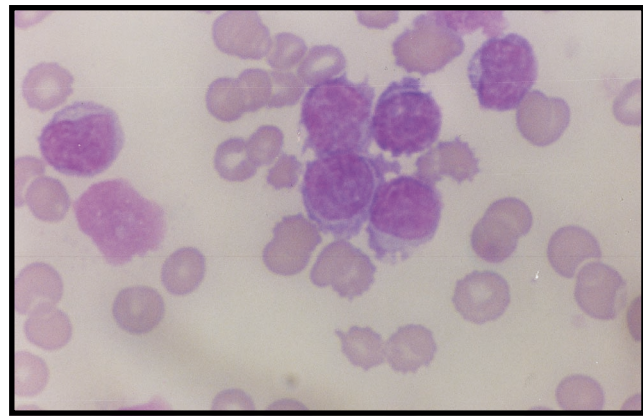


Figure 1b - Lymphocytes under higher magnification show clear cut cytoplasmic projections, high nuclear to cytoplasmic ratio, prominent nucleoli and clumping of cells (MGG x 1000).

of HCL-V. We concluded that this elderly male with huge splenomegaly, moderate hepatomegaly, insignificant lymphadenopathy, marked lymphocytosis, typical morphology of lymphoid cells, TRAP negative lymphoid cells, and characteristic immunophenotype of lymphoid cells appeared to be a typical case of variant form of Hairy Cell Leukaemia (HCL-V).

Discussion. Hairy cell leukemia variant (HCL-V) first described in 1980, is a rare disorder.¹ Until July 1990, a total of 25 cases were described in world literatures.^{2,3} These patients have huge splenomegaly, lymphocytosis, no significant cytopenias and easily aspirable bone marrow. The morphology of lymphocytes closely resembles hairy cells showing numerous surface projections but in contrast these cells have high nuclear to cytoplasmic ratio. In HCL-V the nucleus is more rounded and nucleolus is prominent. Binucleated and hyperchromatic cells are frequent (Figure 1a and b). Further, in contrast to hairy cells the lymphocytes in HCL-V are usually TRAP-ve. The immunophenotype is also characteristic, in contrast to HCL the lymphocytes in HCL-V are CD25 and HC2 negative and in contrast to CLL these are CD5 negative. Using scoring system based on positivity or negativity with CD11c, CD25, HC2 and B-Ly-7 (proposed by Matutes et al 1994), a score of 1-2 was obtained in HCL-V whereas a score of 3-4 was recorded in HCL.⁵ The other closely related entity is splenic lymphoma with villous lymphocytes (SLVL). The cells in SLVL are smaller and nucleoli are rarely prominent.^{3,6} Also immunophenotype pattern in SLVL and HCL-V differs.^{3,6} However it has been suggested that HCL-V and SLVL may belong to the same family as immunophenotype may change from HCL-V to SLVL on follow up.⁷ Our patient showed classical clinical, hematological and immunological features of HCL-V. The patient was referred for

treatment to King Faisal University Hospital in Al-Khobar. 2'-deoxycoformycin and splenic radiation are usually associated with complete or partial response^{3,8-10} a-IFN and combination chemotherapy are ineffective.^{3,9}

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