Co-occurrence of Hairy Cell Leukemia and T –Cell Large Granular Lymphocyte Leukemia : Report of 2 cases

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Introduction

Hairy cell leukemia (HCL) is an uncommon chronic B cell lymphoproliferative disorder characterized by accumulation of small mature B cell lymphoid cells with abundant cytoplasm and "hairy" projections within the peripheral blood, bone marrow, and splenic red pulp. The median age at onset is 50 to 55 years(1). It has indolent course and responds well to chemotherapy**.** Some cases have continued cytopenia(s) despite achieving remission after chemotherapy. The association of HCL with large granular lymphocyte leukemia (LGLL) has been reported previously(3,4,5). It is possible that when HCL and LGLL are co-existing, associated LGL leukemia adds to the cytopenic effect of HCL and remains an etiologic factor of cytopenia(s) after remission of HCL. We are reporting 2 patients in their 8th decades of life with HCL associated with LGLL and persistence of abnormal LGLs after their HCL went into remission.

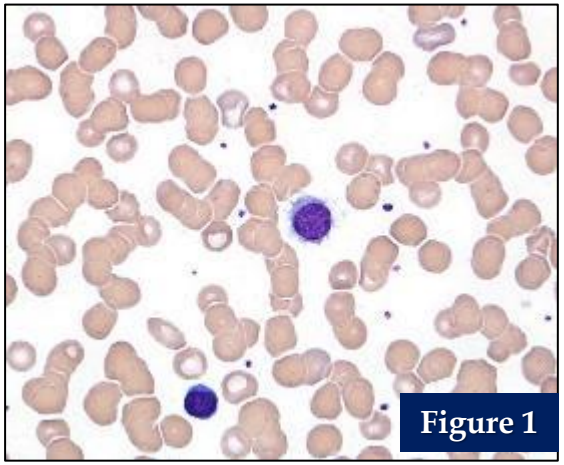

Figure 1

Case Reports

Case 1: A 75-year-old female presented with mild anemia (Hgb: 11.4 g/dl) and thrombocytopenia (PLT: 120K/ml, and low-normal WBCs (4.3 K/ml). We found 17% hairy cells and a few LGLs (Figure 1) in her peripheral blood (PB). Bone marrow (BM) biopsy showed 40% cellularity. About 95% hairy- cell component was identified (Figure 2). Immunohistochemical stains showed CD20+ (Figure 3)/CD79a+/ AnnexinA1+/ DBA.44+/BRAF V600E+ immmunophenotype. Reticulin stain showed increase in reticulin fibers. Flow cytometric (FCM) study of BM (dry tap) showed 0.1% monoclonal hairy cells. and 10% immunophenotypically abnormal (CD2+/CD3+/CD5+/ CD7+/CD8+/CD57+/CD56+ (Figures 4 and 5)/gamma-delta large granular lymphocytes. PCR for *BRAF V600* mutation was positive. Follow-up CBCs have been normal.

Case 2: A 75-year-old male presents with pneumonia and absolute lymphocytosis (4.15K/ul) and about 44% hairy cells in PB. FCM analysis showed about 38% hairy cells and about 4-5% cells with immunophenotypical abnormalalities i.e. dimCD2+/CD3+/dimCD5+/dimCD7+/CD8+/CD57+/ TCRαβ large granular lymphocytes. PCR for T-cell gene rearrangement was positive. Eighteen months follow-up CBC was normal.

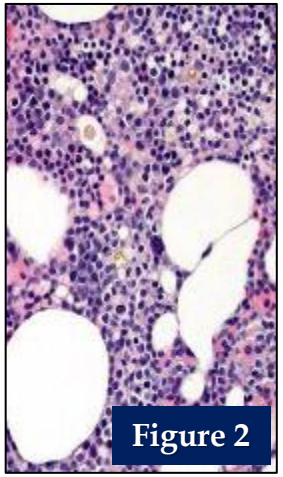

Figure 2

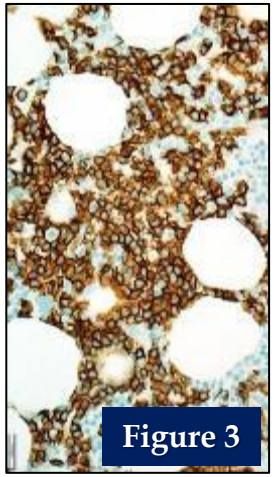

Figure 3

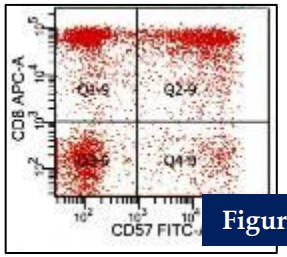

Figure 4 &5

Discussion

Co-occurrence of HCL and LGLL has been reported previously. The age of one of two patients in the previous reports was 53 years and no information is available about second patient's age and about persistent cytopenia(s) following remission in these two cases. The median age at HCL onset is 50 to 55 years.

In our retrospective review of 12 extensively immunophenotyped HCL patients in last 22 years, we found two HCL patients with co-existing LGLL. Both cases are 75 years old and went into remission with cladribine therapy. They have persistent LGLL in their blood. It appears from our small study that LGLL is more likely to co-exist with HCL when the patient is in 8th decade of life. Although CBCs normalized following chemotherapy in our two patients, persistent LGLL could be the reason for failure to normalize the PB counts after HCL remission in some patients. Association of HCL and LGLL in older patients should increase awareness of LGLL when the cytopenias persist after HCL remission.

Conclusion

Our small study shows that LGLL is more likely to co-exist with HCL in elderly population. When these two leukemias co-exist, LGLL could be a contributing factor to the cytopenia(s) and its persistence could also be the reason for persistent cytopenia(s) after HCL remission. We recommend performing extensive flow cytometric analysis in HCL patients to look for associated LGLL so that the patients with LGLL can be treated for it if the cytopenia(s) (are) do not go away following HCL remission

References

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