Hairy Cell Leukemia-varaint (HCL-v) with typical HCL immunophenotype

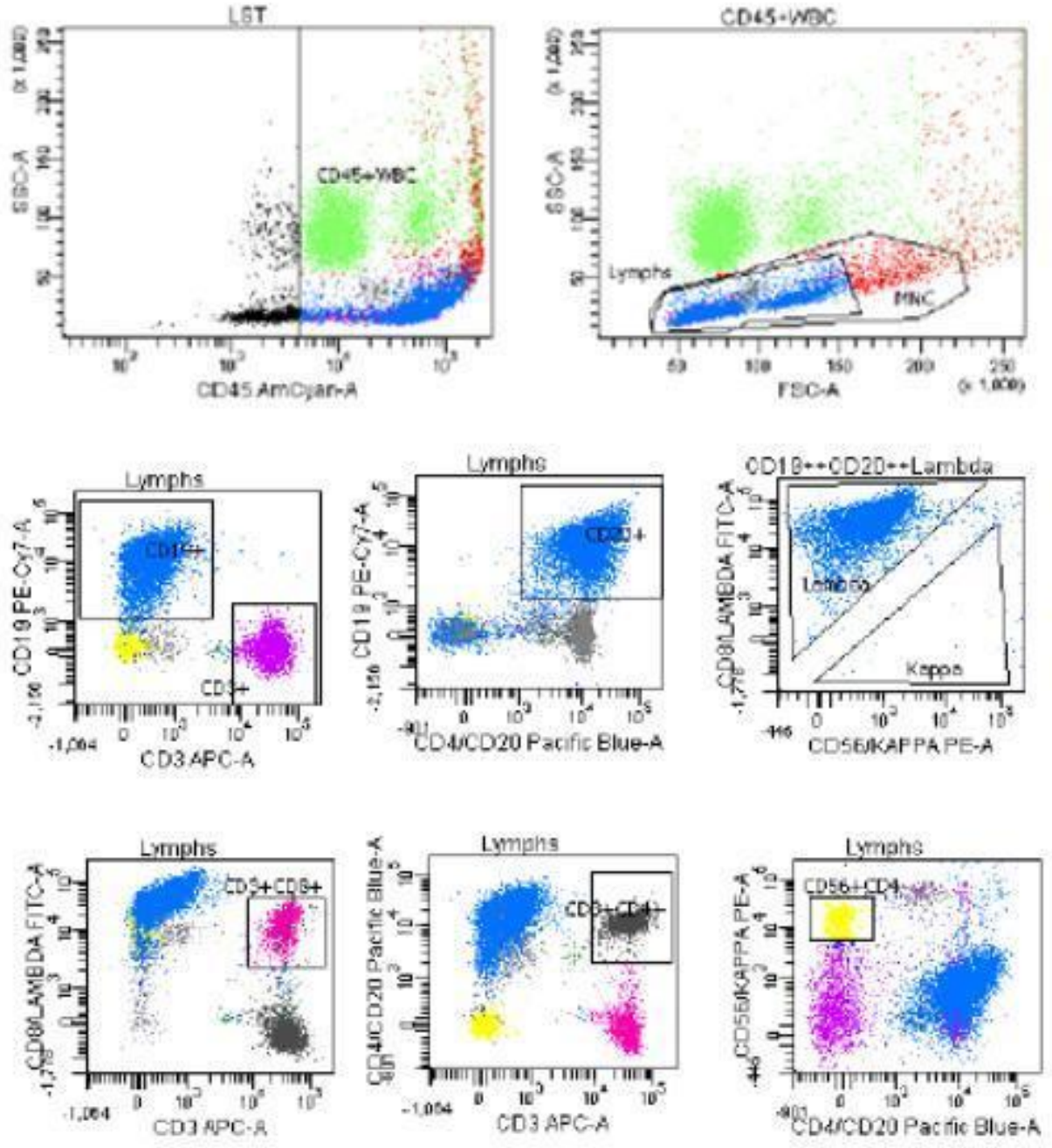

Figure 1:

A 74 year old female was referred with lymphocyte leucocytosis. Peripheral blood film revealed the presence of many abnormal medium-sized lymphoid cells with moderate N/C ratio, conspicuous nucleoli and clumped chromatin of oval, round and bean-shaped nuclei, few bilobed nuclei, slightly basophilic cytoplasm with many cells showing hairy projections (Image 1). Monocyte count was preserved.

Flow cytometry analysis demonstrated that these were clonal B-lymphoid cells with strong expression of Lambda light chain restriction and positive for CD19, CD20, CD22, CD79b and FMC-7 (Image 2). HCL panel score was 3 out of 4 (negative for CD11c and positive for CD25, CD103 and CD123).

Thus, although the peripheral blood film was suggestive of HCL-v, the immunophenotype was typical of HCL.

Following these contradictory results, a BRAF V600E molecular testing was carried out and revealed absence of this mutation.

The BCSH guidelines (Jones et al, 2011) recommend that when the HCL panel score is 3 or 4 out of 4 then it allows for differentiation of HCL from other villous B-cell disorders (recommendation grade 1C). Although the strength of this recommendation is “1 or strong” but the quality of evidence is “C or low” which means that further research is likely to have an important impact on confidence in the estimate of effect.

Accordingly, we suggest that in patients with peripheral blood morphology showing lymphoid cells with villous projections but with atypical features of HCL as leukocytosis, presence of conspicuous nucleoli and absence of monocytopenia especially if combined with flow cytometry finding of HCL panel score of less than 4 out of 4 then testing for BRAF V600E mutation is desirable.

Also in this patient the peripheral blood morphology revealed the presence of LGLs (Images 1, arrows) and concordantly the flow cytometry plots showed the incidental presence of significant proportion of remnant normal lymphocytes populations including T-helper (positive for CD3 and CD4), T-cytotoxic (positive for CD3 and CD8) and NK-cells (negative for CD3 and positive for CD8 and CD56) (Image 2). The identification of these reactive LGLs is important in practice so that not to misinterpret it as villous lymphocytes especially when their granules are sparse.