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Letter to the Editor

Not all hairy cells are the same: a rare case of splenic B-cell lymphoma/leukaemia with prominent nucleoli

Sir,

A peripheral blood sample from an 81-year-old male was received at our stand-alone reference laboratory for evaluation in the setting of fever, progressive fatigue over 3 months, and early satiety. On clinical examination, he had significant pallor and a firm, non-tender splenomegaly extending 6 cm below the costal margin. There was no hepatomegaly or lymphadenopathy.

Complete blood count analysis revealed anaemia (haemoglobin: 67 g/l), marked leucocytosis (total leucocyte count: 44.6×109/l), and thrombocytopenia (platelet count: 28.3×109/l). Other laboratory findings showed normal liver and renal function tests. Contrast-enhanced abdominal CT scan revealed splenomegaly (17.2 cm) without focal lesions. No significant lymphadenopathy or hepatic lesions were noted.

Peripheral smear examination showed presence of atypical lymphoid cells ranging from medium to large in size. These cells exhibited abundant basophilic to pale cytoplasm with irregular borders and variably defined cytoplasmic projections. Nuclei were round to oval with single large, prominent nucleoli (Figure 1).

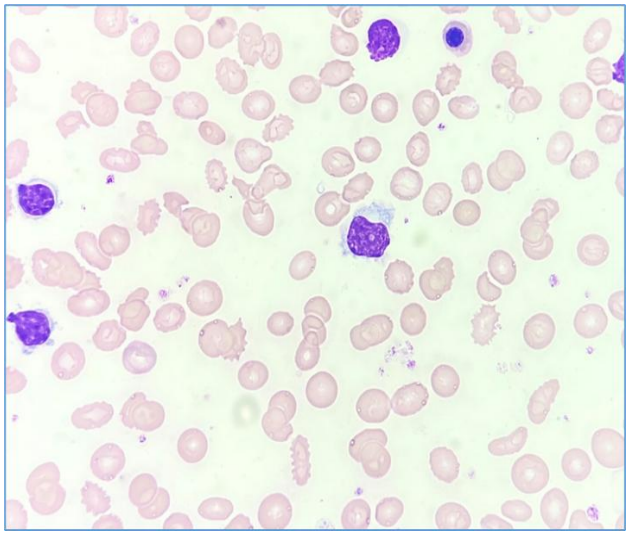

Figure 1: Giemsa stained smear shows atypical lymphoid cells ranging from medium to large in size with abundant basophilic to pale cytoplasm, irregular borders and variably defined cytoplasmic projections. Nuclei were round to oval with single large, prominent nucleoli (x1000).

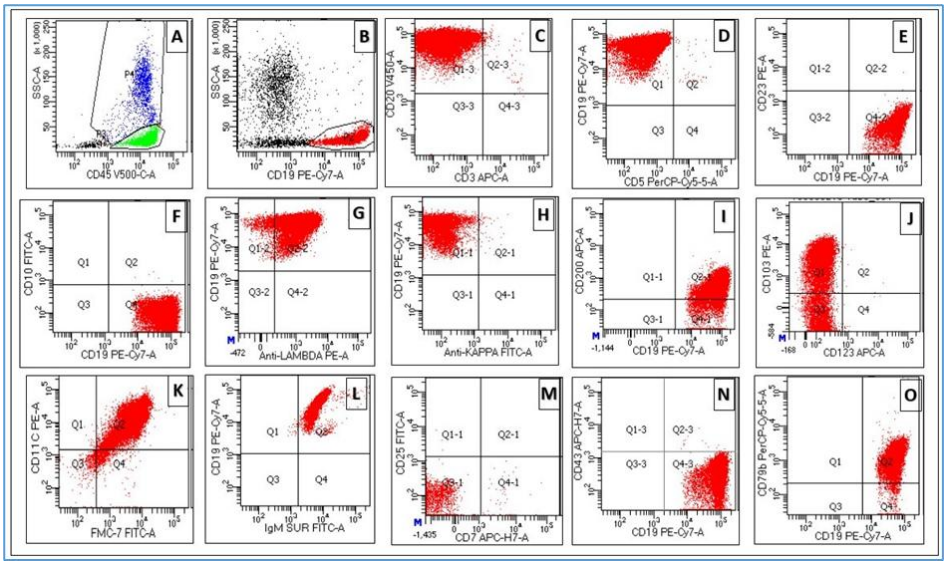

Figure 2: On flow cytometry, (A & B) bright CD45 and CD19 atypical cells were gated. These atypical cells were (C) Bright CD20 positive, (D) CD 5 negative, (E) CD 23 negative, (F) CD10 negative, (G) Lambda restricted, (H) Kappa negative, (I) dim to moderate CD200 positive, (J) CD103 Dim positive and CD123 negative, (K) Dim to moderate CD11c and bright FMC7 positive, (L) IgM positive, (M) CD25 and CD7 negative, (N) CD43 negative and (O) Dim to moderate CD79b positive.

Flow cytometric immunophenotyping was performed using BD FACSCanto II and analysed by FACSDiva software. Based on CD19 and side scatter gating, a dominant population comprising 81% of total events was identified. These cells demonstrated bright expression of CD19, CD20, CD45, and FMC-7, with dim to moderate expression of surface IgM, CD79b, and CD200. Lambda light chain restriction was observed. The neoplastic cells also expressed CD103 and CD11c, while lacking expression of CD5, CD10, CD23, CD25, CD38, CD43, ROR1, and CD123 (Figure 2).

The immunophenotypic findings were consistent with a CD5-negative and CD10-negative mature B-cell lymphoproliferative disorder. The co-expression of CD103 and CD11c, in the absence of CD25 and CD123, is characteristic of Splenic B-cell lymphoma/leukaemia with prominent nucleoli (SBLPN)—a rare entity previously referred to as the variant form of hairy cell leukaemia (HCLv). These findings fulfill the essential diagnostic criteria as outlined by the World Health Organization (WHO) classification for SBLPN.[1] BRAF V600E studies were negative by PCR.

At 4-month follow-up, the patient remains under observation without active intervention, in line with the family's informed decision for conservative management. Hematologic parameters have remained stable, and there has been no clinical or radiological progression.

SBLPN is a rare splenic B-cell neoplasm that shares some cytomorphological and immunophenotypic features with hairy cell leukaemia (HCL), but differs significantly in its clinical behaviour and response to treatment. SBLPN is characterized by splenomegaly, high lymphocytosis, and cytopenia, with the notable absence of monocytopenia, which is a hallmark of classic HCL. This malignancy predominantly affects elderly patients and accounts for approximately 0.4% of all chronic lymphoid malignancies. While autoimmune phenomena are rare, patients often experience chronic disease progression, with B-symptoms and features of frank disease progression being uncommon.[1]

The clinical course of SBLPN is relatively indolent, with a long lymphocyte-doubling time. In many cases, the white blood cell count may remain stable for several years without significant change. The primary clinical complications arise from splenomegaly and hypersplenism, which can lead to significant cytopenias. Despite the chronic nature of the disease, the median survival of patients is relatively favorable, with a reported 9-year median survival in some cohorts.[2]

However, the rarity of SBLPN and its distinct clinical features pose challenges in treatment, as conventional therapies for HCL tend to be ineffective. In contrast to typical HCL, patients with SBLPN are generally resistant to interferon-alpha (IFN-α) therapy, and responses to purine analogues such as pentostatin and cladribine are limited and often transient.[3] Combination chemoimmunotherapies, such as cladribine and rituximab, have yielded the best results to date in HCL-V.[4] This highlights the need for alternative therapeutic strategies. Given the limited therapeutic options and disappointing responses to standard treatments, further research is necessary to elucidate more effective approaches for managing this rare and challenging malignancy.

In summary, this case highlights the diagnostic features of SBLPN—a rare mature B-cell neoplasm presenting with splenomegaly and circulating atypical lymphoid cells in elderly patients.

Accurate morphological assessment combined with comprehensive immunophenotyping is essential to differentiate SBLPN from other B-cell lymphoproliferative disorders and to guide appropriate clinical decision-making, especially given its distinct treatment approach and prognosis.

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REFERENCES

1. Traverse-Glehen A. Splenic B-cell lymphoma/ leukaemia with prominent nucleoli. In: WHO Classification of Tumours Editorial Board. Haematolymphoid tumours. Lyon (France): International Agency for Research on Cancer; 2024. (WHO classification of tumours series, 5th ed.; vol. 11). Available at: https://publications.iarc.who.int/637. Accessed on 15th August 2025.
2. Matutes E, Wotherspoon A, Brito-Babapulle V, Catovsky D. The natural history and clinico-pathological features of the variant form of hairy cell leukemia. Leukemia. 2001;15(1):184-6.
3. Paillassa J, Maitre E, Troussard X. Hairy Cell Leukemia (HCL) and HCL Variant: Updates and Spotlights on Therapeutic Advances. Curr Oncol Rep. 2022;24(9):1133-43.
4. Tran J, Gaulin C, Tallman MS. Advances in the Treatment of Hairy Cell Leukemia Variant. Curr Treat Options Oncol. 2022;23(1):99-116.

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