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Rapidly deteriorating B-CLL at presentation: clonally related Burkitt leukemia as an atypical Richter transformation?

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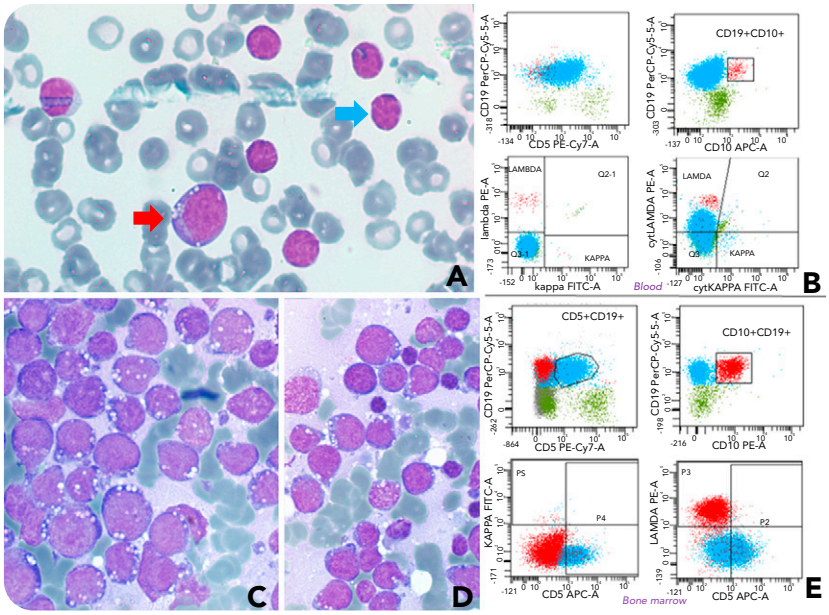

An 81-year-old man presented with acute back pain, leukocytosis-lymphocytosis (57.62 × 10^9/L), deteriorating severe thrombocytopenia (121 × 10^9/L to 29 × 10^9/L within 1 week), rapidly rising serum lactate dehydrogenase up to 13 464 units/L (×59), and ferritin levels (9896 ng/mL). The clinical examination was normal. Peripheral blood (PB) smear revealed numerous B-cell chronic lymphocytic leukemia (B-CLL)–like lymphocytes, but also very rare lymphoid cells with typical Burkitt morphology (panel A; 100× objective, original magnification ×1000, May-Grünwald-Giemsa stain, blue/red arrow, respectively), accounting for 1% of PB lymphocytes on immunophenotype (panel B; red-colored population; CLL blue-colored population). Bone marrow (BM) aspiration revealed typical Burkitt leukemic infiltration admixed with mature, small lymphocytes (panels C-D; 100× objective, original magnification ×1000, May-Grünwald-Giemsa stain). BM immunophenotype demonstrated 2 abnormal B-cell populations, both with λ-light chain restriction: 50% typical B-CLL lymphocytes CD5$^+$, CD23$^+$, CD43$^+$, CD200$^+$, CD20$^+$ (weak), CD79b$^+$ (weak), sIgλ$^-$, cIgλ$^+$ (panel E; blue-colored population) and 45% Burkitt lymphoma/leukemia lymphoid cells CD10$^+$, CD38$^+$, sIgλ$^+$ (moderately), cIgλ$^+$, CD20$^+$, CD79b$^+$, CD5$^-$, CD23$^-$ (panel E; red-colored population), and the BM biopsy findings were similar, also revealing bcl2 expression by both B-CLL and blast cells. *MYC* but not *BCL2* rearrangements and 47,XY,+7,t(8;14)(q24;q32)[18]/47,idem,+1,der(1;15)(q10;q10)[2] were shown by BM fluorescence in situ hybridization and conventional cytogenetic analysis. DNA genetic analysis demonstrated a single identical B-cell clone in blood and BM.

This is a unique case of unusual B-CLL presentation due to the complication by Burkitt transformation or Burkitt-like high-grade transformation at initial presentation under the wide definition of Richter syndrome.

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