Small-cell variant of ALK\(^+\) anaplastic large-cell lymphoma with a leukemic phase

This peripheral blood (PB) smear from a 24-year-old man with retroperitoneal lymphadenopathy and a white blood cell count of \(77 \times 10^9\) cells/L (66% lymphocytes) showed variably-sized atypical lymphocytes with irregular nuclear contours and basophilic vacuolated cytoplasm including numerous flower cells and apoptotic cells (panel A). *Hallmark cells* with reniform nuclei, small to large in size, were a minor proportion (panels A and B). Flow cytometric analysis (FC) of the PB and bone marrow (BM) samples detected a T-cell population with low forward scatter and CD2\(^{+}\) (dim), CD3\(^{+}\) (dim), CD7\(^{+}\), CD8\(^{+}\) (dim), CD4\(^{-}\), CD5\(^{-}\), and CD25\(^{+}\); CD30 expression was dim in the BM and negative in the PB by FC. BM morphology showed scattered interstitial infiltration of small- to medium-sized atypical lymphocytes that were variably CD30\(^{+}\) and ALK\(^{+}\) (nuclear staining) by immunohistochemistry (panel B). BM fluorescence in situ hybridization and karyotype studies revealed ALK rearrangement and t(2;5)(p23;q35), respectively.

Strong uniform CD30 expression, typical of the common variant, was not observed in these samples. Furthermore, the flower cells seen raise the possibility of adult T-cell leukemia/lymphoma. This case highlights the broad morphologic spectrum of ALK\(^+\) ALCL and underscores the importance of a multimodal diagnostic approach in T-cell lymphoma that includes investigation of ALK rearrangement and/or staining, defining features of this entity.
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