Diagnostic Challenges of $CD4^+/CD56^+/CD123^+$ hematological neoplasms*

Desafio diagnóstico de neoplasias hematológicas $CD4^+/CD56^+/CD123^+$

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In the January/February 2013 edition, Maio *et al*[1] describe a patient with $CD4^+/CD56^+/CD123^+$ ascribed as blastic plasmacytoid dendritic cell neoplasia (BDCN). However, since $CD4^+/CD56^+/CD123^+$ neoplasms are highly heterogeneous, the precise diagnosis requires an extensive immunophenotypic panel.[2,3] Although highly suggestive, the cytochemical positivity for CD4, CD56 together with CD123 in the absence of myeloperoxidase, CD3, CD2, CD5, and CD7, is not sufficient to determine the BDCN malignant nature. Despite the expression of CD123, the aforementioned phenotype could also correspond to acute myeloid dendritic cell leukemia or acute myeloid leukemia (myeloid leukemia cutis), especially with monocytic differentiation. The diagnostic work-up of these entities relies on a comprehensive antibody panel that should also include CD13, CD33, CD15, CD14, CD64, CD16, CD34, CD117, BDCA-2 (CD303), BDCA-4 (CD304), BDCA-3 (CD141) and TCL1. BDCN are phenotypically recognized by expression of specific plasmacytoid dendritic cell proteins (CD303 and CD304) in the absence or dim expression of myeloid markers. Conversely, acute myeloid dendritic cell leukemias specifically express CD141 along with some myeloid markers. By exclusion, the absence of CD303, CD304 and CD141, along with the presence of myeloid and monocytic markers, supports the diagnosis of myelomonocytic leukemia. Additional molecular tests targeting well-characterized abnormalities could serve as ancillary diagnostic tools.

Although the panel herein proposed could not be entirely performed on skin biopsies, it could be easily applied by flow cytometry on circulating cells during the disseminated phase.

It is clear that strong collaborative efforts are required to improve diagnosis and management of these rare diseases.

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