

2nd International Conference on **Hematology & Blood Disorders**

September 29-October 01, 2014 DoubleTree by Hilton Baltimore-BWI Airport, USA

Anaplastic large cell lymphoma, ALK-positive, small cell variant, with leukemic presentation and rare CD8-positive phenotype

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Anaplastic large cell lymphoma (ALCL), anaplastic lymphoma kinase (ALK)-positive is a T-cell lymphoma consisting of usually large and pleomorphic lymphoid cells. The characteristic genetic abnormality is a translocation involving the ALK gene. The disease frequently involves both lymph nodes and extranodal sites but a leukemic presentation is quite rare. Of the broad morphologic spectrum of ALCL, one particular subtype, the small cell variant (5-10% of cases), may have a leukemic presentation with peripheral blood involvement. It is described a 16-year-old female patient who presented with fever, leukocytosis and lymphadenopathy. Initially, the peripheral blood smear examination was suggestive of leukemia. Subsequent flow cytometric analysis of the peripheral blood demonstrated an aberrant T-cell population but did not show an immunophenotype characteristic for a specific type of T-cell leukemia like T-cell prolymphocytic leukemia, adult T-cell leukemia, Sezary syndrome, etc. A bone marrow study was also performed and a smaller population of similar atypical cells was identified. Since the peripheral blood and bone marrow findings were not definitively diagnostic of a specific entity, a decision was made to obtain a lymph node biopsy. The normal lymphonodular architecture was almost completely effaced and, even though the majority of neoplastic cells were small, larger cells including "hallmark" and wreath-like cells were easily identifiable, suggesting a diagnosis of ALCL which was immediately confirmed by ALK immunohistochemistry and FISH analysis. Additionally, a rare CD8-positive phenotype was present by flow cytometry.

Biography

Dragos C Luca is the Director of Hematopathology and Flow Cytometry at Children's National Medical Center and an Assistant Professor of Pathology and Pediatrics at George Washington University, both in Washington, DC. He is certified by the American Board of Pathology in Hematopathology, Pediatric Pathology and Anatomic/Clinical Pathology after completing his Pathology residency at the University of Illinois at Chicago with subsequent fellowships in Hematopathology at Loyola University and Pediatric Pathology at Northwestern University, both in Chicago. He has published in the fields of hematologic malignancies and pediatric solid tumors.

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