Hematologic malignancies in pediatrics inborn errors of immunity

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Context

Inherited errors of immunity (IEI) or primary immunodeficiencies are a group of genetic rare diseases predisposing to severe infections, autoimmunity, and malignancies. The overall risk for malignancies in pediatric IEI-patients is estimated to range from 4–25%, of which 60% of cases are of hematological origin.

Objective

to evaluate the hematological malignancies in pediatrics IEI.

Materials & Methods

Were included 13 patients 3 of which developed hematological malignancies and were evaluated for demographics, clinical features, and prognosis.

Results

The prevalence of hematological malignancies among our patients was 23% (3/13). The male-to-female ratio was 2:1. The mean age at diagnosis of malignancy was 8.6 years (range 5-14). Non-Hodgkin lymphoma was the malignancy type diagnosed in all the pediatric patients (n=3, of which Wiskott-Aldrich-1, ataxia-telangiectasia-1, CVID (common variable immunodeficiency)-1). However, 2 patients (52.5%) died (1-during first line-chemotherapy, 1-NHL was established postmortem). 1 patient underwent bone marrow transplantation and is currently under supervision.



Figure 1. Cervical lymphadenopathy in a child with IEI, later diagnosed with NHL

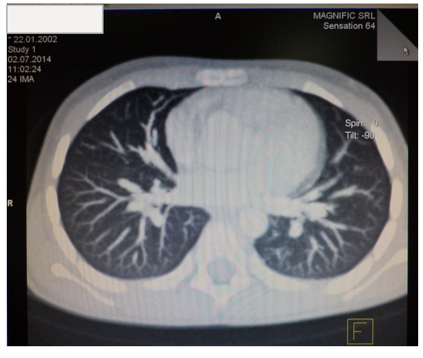


Figure 2. Pulmonary CT showing para-aortic lymph nodes on the left up to 0.9x1.4 cm
Intrathoracic lymph node biopsy by open thoracotomy with histopathological investigations confirmed the diagnosis of non-Hodgkin's lymphoma, microlymphoblastic variant

Conclusion

In this study, we related an association between hematological malignancies and IEI. Lymphoid malignancies were the most common malignancy, this warns of the need for multidisciplinary monitoring of these patients.