OP 20

Risk factors and outcomes related to intensive care unit admission of children with hematological and solid organ malignancies: single-center experience

C. Kaya[1,*], Z. Guzelkucuk[2], D. Ozyoruk[2], N. Ozbek[2], N. Yarali[2]

[1] *University of Health Sciences, Ankara City Hospital, Department of Pediatric, Ankara, Turkey*
[2] *University of Health Sciences, Ankara City Hospital, Department of Pediatric Hematology and Oncology, Ankara, Turkey*

Objective: Despite the developments in the diagnosis and treatment of cancer, malignancy remains one of the important causes of mortality in children. Aggressive chemotherapy leads to severeinfections or complicationsaffecting many systems, causing admissions to intensive care units (ICU). In our study, we aimed to evaluate the demographic data, clinical findings, and prognostic factors affecting hospitalization in the intensive care unit of the patients with hematological malignancy (HM) and solid organ malignancy (SOM).

Methodology: Between June 2013 and December 2018, patients were enrolled in our study between 28 and 18 years of with HM and SOM, who were hospitalized in the Pediatric Intensive Care of the University of Health Sciences Ankara Children's Hematology Oncology Training and Research Hospital. Demographic, clinical, laboratory, and treatment characteristics and survival in ICU were recorded.

Results: During the study period, 232 admissions of 158 patients with HM and SOM who were treated in ICU were evaluated. Patients diagnosed with acute lymphoblastic leukemia (ALL) and central nervous system (CNS) tumors were the most frequently hospitalized patients in the ICU, respectively. One hundred fifty-eight patients included in our study, 89 (56.3%) died. There was no statistically significant difference between HM and SOM patients in terms of mortality rate. The overall survival rate was calculated as 51.7%. Mortality was found to be higher in patients who need ICU admission while staying in the hospital, patients between the ages of 15–18, patients needed respiratory support before ICU and underwent mechanical ventilation (MV) during the first 24 h of hospitalization, and patients needed inotropic support. Neutropenia, thrombocytopenia, hypoglycemia, hypoalbuminemia, and high levels of AST/ALT, urea, creatinine, total and direct bilirubin, LDH, and CRP values were associated with mortality. Detection of recurrenceor refractory disease and organ dysfunction is an independent risk factor on mortality.

Conclusion: One-year overall survival rate of our patients was 51.7%. Relapse/refractory disease and organ dysfunction were identified as two independent risk factors on mortality. Prospective, multicenter studies are needed to determine the increasing importance of factors in the follow-up of patients with hematological and solid organ malignancies and to determine long-term survival rates.

https://doi.org/10.1016/j.htct.2020.09.052

OP 21

A case report of RAS-associated autoimmune lymphoproliferative disorder

C. Coskun[1,*], F. Gumruk[1], M. Cemaloglu[1], M. Orhan[2], I. Tezcan[3], S. Unal[4]

[1] *Hacettepe University, Department of Pediatric Hematology, Ankara, Turkey*
[2] *Sakarya University, Department of Pediatric Hematology, Sakarya, Turkey*
[3] *Hacettepe University, Department of Pediatric Immunology, Ankara, Turkey*
[4] *Hacettepe University, Research Center for Fanconi Anemia and Other Inherited Bone Marrow Failure Syndromes, Ankara, Turkey*

Objective: Clinically, RAS-associated autoimmune lymphoproliferative disorder (RALD) is characterized by splenomegaly, peripheral lymphadenopathy and autoimmunity. The autoimmune phenotype can present in childhood or adulthood and primarily includes autoimmune hemolytic anemia, immune thrombocytopenia (ITP) and neutropenia. In this report, we present a patient with RALD. The patient showed somatic mutation for NRAS mutation.

Case report: A two-year-old boy was referred with the complaints of ecchymoses. There was no consanguinity between parents and he had a healthy sibling. It was learned that the patient presented with bruises at the age of 12 months, and was follow up with thrombocytopenia, which responded partially and transiently to intravenous immunglobin (IVIG) and steroids. Physical examination at admission revealed hepatosplenomegaly and cervical lymphadenopathies. Complete blood count showed hemoglobin (Hb) 10.6 g/dL, mean corpuscular volume (MCV) 74 fL, white blood count 11.3×10^9/L and platelet 15×10^9/L with monocytosis on blood film. Bone marrow of the patient showed megaloblastic changes and no increase in megakaryocytes. Additionaly, patient was found to have hypergamaglobulinemia. Double-negative (CD4-CD8-) T cells were 2% and a decrease in lymphocyte activation was observed with T and B cell subgroups. Mycophenolate mofetil was started. The patient was followed up with the autoimmune lymphoroliferative syndrome (ALPS) phenotype and genetic work-up revealed NRAS c.38G>A heterozygous mutation. The patient was diagnosed with ALPS type 4 (NRAS somatic mutation). Thrombocytopenia responded to mycophenolate mofetil.

Results: Genetic analysis of the RAS mutation should be performed in cases that does not meet the defined diagnostic criteria of ALPS or JMML.

Conclusion: RAS-related lymphoproliferative disease is a rare genetic disorder of the immune system and is a newly classified disease. RALD presents with autoimmunity, lymphadenopathy, and/or splenomegaly, but without a defect in FAS-dependent apoptosis or an increase in peripheral double negative T lymphocytes. The absolute or relative monocytosis in particular is an important characteristic of this disorder and help differentiate it from ALPS. JMML may be characterized with autoimmunity and may be similar to RALD as a clinical and laboratory phenotype. Approximately 15–30% of patients