testing confirms the diagnosis at an early stage and reduces morbidity and mortality due to the disease. WES is helpful to detect such cases.

Conclusion: Various genes such as DKC1, CTC1, RTEL1, TERF1, TINF2, TERC have been found to be responsible for DKC. RTEL1 is a DNA helicase necessary for telomere replication and stability. With the understanding of the molecular basis of the disease, patients with hematological findings at the time of diagnosis and those without skin findings were also identified. In our case, signs of bone marrow failure were observed primarily and no changes in nail dystrophy, leukoplakia and skin pigmentation and neurological findings were detected. In cases where the disease does not follow classical presentation, the use of genetic testing confirms the diagnosis at an early stage and reduces morbidity and mortality due to the disease. WES is helpful to detect such cases.

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PP 60

Acquired aplastic anemia in childhood: single-center experience

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Objective: Acquired aplastic anemia is a rare disease characterized by the irreversible loss of bone marrow function and threatens life when not treated. Bone marrow transplantation (BMT) or immunosuppressive agents (IST) are used in its treatment. In this article, we aimed to evaluate our patients with acquired aplastic anemia in epidemiological, etiological, and treatment outcomes.

Case report: Nine patients who were diagnosed with acquired aplastic anemia over ten years were evaluated.

Methodology: The patients admitted to the Istanbul Medical Faculty Pediatric Hematology-Oncology Outpatient Clinic between 2000 and 2010 were diagnosed with acquired aplastic anemia (those who underwent BMT, IST, or both) were evaluated retrospectively on patient files and computer records.

Results: Nine patients were diagnosed with acquired aplastic anemia over ten years. 4 of them were girls, and 5 were boys. The average age was 10 (1–17 years). There was a history of hepatitis in 3 cases and a history of metamizole use in 1 case. As a treatment, six patients were treated with IST, and five patients were treated with BMT. ATG 40 mg/kg/day 4 days, cyclosporin 10 mg/kg/day (6 months), methylprednisolone 2 mg/kg/day (2 months) and G-CSF 5 μg/kg (2 months) as immunosuppressive therapy. Response to immunosuppressive therapy was received at an average of 3 months. Two of them were fully responsive. One patient was lost due to septic shock before the IST response was evaluated. BMT was performed in 5 cases, three of them were unresponsive to IST. In the follow-up, two cases are in remission, and three are lost due to sepsis. When evaluating our 5 cases with dead, two of them were very severe aplastic anemia, the symptoms of sepsis were present in their first admission, and they died before the treatment started. Two of them died due to the complication of BMT in the very early period. One case was admitted with perforated appendicitis while in remission after BMT and died due to septic shock.

Conclusion: Two primary treatment modalities are used to treat patients with severe aplastic anemia; IST and BMT. The first option is BMT with the matched sibling donor. If there is no suitable sibling, IST is started first, and a fully compatible donor is searched immediately. If there is no response to IST, an allogenic BMT must be applied in the presence of a suitable donor. Our mortality rate is high compared to the literature because of severe disease presentation; most of them were late admission to the hospital due to low socioeconomic level.

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HEMOSTASIS, THROMBOSIS, AND VASCULAR BIOLOGY

PP 61

The course of intracranial bleeding in the patient with immune thrombocytopenia

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Objective: The approach to the treatment of immune thrombocytopenia always remains relevant, despite the fact that the etiology and pathogenesis of the disease is quite clear, and it is clear that the development of the disease is based on the conflict between their own platelets and autoantibodies directed at them. The goal of treatment is to resist the creation of autoantibodies, protect your own platelets and lengthen their life. The proposed standards of treatment with steroids, reticuloendothelial system blockers, anti-lymphocytic antibodies, thrombopoietin, etc.did not find a clear place for a radical change in the course of the disease.

Case report: The article presents a case of a child suffering from chronic ITP who received various medical treatments with periodic remissions for 6 years. At the age of 10, the child had convulsions and neurological disorders due to acute respiratory infection and high temperature. In blood tests: PLT-10×10^9/L. CT scan of the brain showed the presence of intracranial bleeding. The prescribed “pulse therapy” with dexamethasone and platelet transfusions allowed for intracranial surgery (PLT – 234×10^9/L).). However, a few days later, due to the ineffectiveness of “pulse therapy”, and the risk of renewed bleeding, the patient was again transfused platelet mass and prescribed high-dose intravenous immunoglobulin (IVIG), which raised the platelet level to 210×10^9/L. Soon, this therapy was ineffective, and we had to re-transfuse the platelet mass and simultaneously prescribe thrombopoietin (Revoleyd). Against the background of this therapy, the platelet level was stabilized, and the resulting effect was long-lasting.

At the moment, the child's hematological and neurological status is quite satisfactory.

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PP 62

The cause of very severe trombocytosis: iron deficiency anemia

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Objective: Platelet count above 450,000 mm^3 is defined as thrombocytosis. It is called mild thrombocytosis if the platelet count is between 700,000–900,000 mm^3, and severe thrombocytosis between 900,000–1,000,000 mm^3. If the platelet count is over 1,000,000 mm^3, it is considered as very severe thrombocytosis. In this case report; we have showed that iron deficiency can also lead to very severe thrombocytosis by presenting the case of very severe thrombocytosis developing in an adolescent female patient.

Case report: The 12-year-old girl was referred to our hospital for anemia (Hgb: 5.8 g/dL) by an external clinic she applied due to her headache in the morning for the past month. The patient's history and family history were unremarkable. Her physical examination revealed that her general condition was moderate-poor, skin was pale, conjunctiva was extremely pale, peak heart rate: 130–140/min, TA: 90/50 mm/Hg. Lymphadenopathy and hepatosplenomegaly were not detected. In the laboratory tests of the patient, the following findings were detected; the leukocytes count was: 14,900/mm^3, neutrophil count: 11.9/mm^3, Hgb: 4.8 g/dL, Hct: 20%, MCV: 53 fl, RBC: 3.7 milyon/uL, MCH: 12.9 pg (27–31), platelet count 2,629,000/mm^3. Peripheral smear of the patient was analyzed. In eritrocytes, a high degree of hypochromic microcytes were detected and 80% neutrophils, 2% monocytes, 18% lymphocytes, abundant platelets were seen. Serum iron: 6.7 uL/dL (50–120); iron binding capacity: 525 uL/dL (155–355); ferritin: 0 ng/mL; folate: 10.6 ng/mL (0.3–24) and vitamin B12: 437 ng/mL. There was no abnormality in other biochemical examinations. Iron replacement was started at a dose of 6 mg/kg/day considering iron deficiency anemia and related thrombocytosis. Abdominal ultrasonography was evaluated within normal limits according to age. Since the patient had tachycardia, appropriate cross erythrocyte transfusion was performed. Viral serologies and autoantibodies of the patient were evaluated as normal. The control hgb level was 7.9 g/dL and thrombocyte count was 1,875,000/mm^3 after transfusion. In the bone marrow aspiration assessment, the myeloid and erythroid series in the normocellular bone marrow were seen as normal, blasts were not seen, megakaryocytes were increased. The patient had hgb: 10.4 g/dL, platelet: 732,000/mm^3 in the clinical examination performed in the second week. She is under the oral +2 valence iron treatment and had no clinical problem in her follow-up examinations.

Methodology: Information was obtained from the patient file.

Results: In childhood, thrombocytosis usually occurs due to secondary causes and thrombocytosis regresses by controlling the causing disease. Thrombocytosis due to iron deficiency is mostly seen in infancy period.

Conclusion: The cause of thrombocytosis in iron deficiency is not fully understood. The fact that the increase in EPO stimulates TPO receptors (c-mpl) in iron deficiency is known to result in thrombocytosis. However, it is very important that children should be evaluated immediately for infection and iron deficiency before performing further examinations. Keywords: Thrombocytosis; iron deficiency; child.

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LEUKEMIA/LYMPHOMA/HISTIOCYTE DISORDERS

PP 63

Immune markers are closely related to the remission achievement in childhood acute myeloid leukemia

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Objective: Immunophenotyping of the blast population at the diagnosis of acute myeloid leukemia is a routine study that supplements the data obtained by morphological, cytochemical and cytogenetic studies of tumor cells. Currently, risk-stratification of children with acute myeloid leukemia (AML) is based on initial leukocytosis and genetic abnormalities. However, those genetic aberrations which effect the prognosis of childhood AML are found only in about 35% of cases. The search for reliable factors to clarify the stratification of patients into risk groups continues, and along with chromosomal and gene abnormalities, aberrations of the immunophenotype of tumor blasts are of interest. There are conflicting data on the effect of immunological factors on the prognosis of AML. Most of them were obtained by the analysis of AML in adults. It is of interest to analyze the effect of the immunophenotypic "portrait" of blast cells on the course of the disease. The achievement of complete remission (CR) is the main prognostic factor for AML in children.

Methodology: In our study, CR was achieved in 84 of 105 children with AML (80.0%) and achieving complete remission was very significant ($p = 0.000$) prognostic factor in assessing overall survival. We analyzed the influence of gender, age, FAB-variants and immunological markers on the probability of remission achievement. The effects of age, FAB-variants and gender were not significant, though boys achieved complete remission more rarely than girls ($p = 0.11$). We analyzed effect of the following immunological markers: CD7 ($n = 69$), CD117 ($n = 37$), CD34 ($n = 93$), CD13 ($n = 97$), CD33 ($n = 96$), CD20 ($n = 47$), CD19 ($n = 84$), CD9 ($n = 9$), CD38 ($n = 50$), HLA-DR ($n = 83$), CD11b