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

($n=3$), CD64 ($n=59$), CD14 ($n=20$), CD5 ($n=51$), CD3 ($n=55$), CD56 ($n=52$), CD10 ($n=67$).

Results: Among them presence of CD33, CD19 and CD56 increased the probability of remission achievement ($p=0.005$; 0.025 and 0.049 respectively) while CD14 ($p=0.028$) had a negative effect on it. It is important to note that none of these markers had a significant effect on the overall survival.

Conclusion: In conclusion, search for new prognostic factors for AML in children continues, and aberrantly expressed immunophenotypic markers may become important for clinicians.

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PP 64

The course of toxic hepatitis at the stage of treatment consolidation acute leukemia in children

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Objective: Toxic hepatitis occupies a special place among the complications of chemotherapy in the treatment of patients with acute leukemia. The research work we have presented is devoted to studying the frequency of toxic hepatitis and the choice of treatment tactics for children who are at the stage of consolidating acute leukemia.

Methodology: The study group included 110 children from both sexes who reached complete remission after a course of induction. Patients were 9 months old up to 15 years. The treatment was carried out according to the Moscow-Berlin-2015 program, where the consolidation phase was composed of 3 courses of 8 weeks. The severity of toxic hepatitis was predetermined by its criteria.

Results: According to the data obtained, 81 patients had toxic hepatitis (73.6%). In mild form it was noted in 46 children (56.7%), in moderate severe in 31 (38.4%), and in severe in 4 children (4.9%).

Conclusion: In the mild form of hepatitis from the intravenous use of Essentiale forte and Riboxin against the background of ongoing chemotherapy, a positive effect was obtained. With moderate severity, intravenous administration of Adeomethionine preparations (Heptral/Legend) in combination with Aevit per os turned out to be more effective. In 4 patients, upon transition to a severe form in the last course of consolidation, along with these drugs, ursodeoxycholic acid (Ursobil) + enhanced detoxification therapy was prescribed, which led to a complete recovery. After the treatment of toxic hepatitis, all patients with moderate and severe form, for the purpose of prevention, was prescribed combination therapy with Ursobil + Aevit + Lipoic acid, which gave a long-term positive effect.

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PP 65

Klippel–Trenaunay syndrome associated with chronic myeloid leukemia

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Objective: Klippel–Trenaunay syndrome (KTS) has been associated with capillary, venous, lymphatic and soft tissue malformations, whether it predisposes to malignancy is not clear. We report a case of chronic myeloid leukemia (CML) with KTS. We report this case because of its rarity and need for long term follow-up.

Case report: A 14-year-old boy presented with a painless mass on his left groin which was extending to his knee. Physical examination revealed splenomegaly, limb length discrepancy, left lower extremity hypertrophy and capillary hemangiomas over the posterolateral skin of the left thigh. KTS was suspected and confirmed with heterozygous mutation (c1634A>C/p.Glu545Ala) at the PIK3CA gene. The patient consulted to the hematology due to hemorrhage complication of the surgery. Complete blood counts showed a hemoglobin level of 7.3 g/dL, white blood cell as 164×10^9/L, neutrophil 76.4×10^9/L and thrombocytes 104×10^9/L). The differential was metamyelocytes 20%, bands 4%, neutrophils 70%, eosinophils 4%, lymphocytes 2%, normoblast 4%, except circulating blasts. Bone marrow aspiration showed normocellular myeloid/erythroid ratio of 23:1, granulopoiesis with left shift, increased megakaryocytes seen with normal maturation and blasts were lower than 5%. RT-PCR from peripheral blood was positive for the BCR-ABL p210 transcript. Conventional karyotyping revealed a typical 46 XY, t(9,22)(q34;q11.2) without any additional cytogenetic abnormalities in all (20/20, 100%). Chronic phase CML (CML-CP) was diagnosed, and imatinib was initiated with a 300 mg/m^2 dose daily.

Results: To our knowledge there has been no description of an association between KTS syndrome and CML in the literature. We report this case because of its rarity.

Conclusion: Klippel–Trenaunay syndrome is a rare congenital malformation involving blood and lymph vessels and abnormal growth of soft and bone tissue. The exact cause of KTS is not clear, several genes and pathways have been identified in its pathogenesis. Remarkably, PIK3CA gene mutations have been detected in some cases of KTS. PIK3CA encodes for a subunit of the phosphoinositide 3-kinase enzyme, which is involved in cell proliferation and migration. The angiogenic gene VG5Q has also been implicated in KTS. We report the case of a 14 year-old boy with diagnosed KTS, who presented with a bleeding from the surgical region that was found to be a chronic myeloid leukemia. To our knowledge there has been no description of an association between KTS syndrome and CML. In the literature, there are cases where KTS is associated with Wilms tumor, neurofibromatosis and osteoblastoma, but no hematologic malignancy has been so far.

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