RED BLOOD CELL DISORDERS

PP 27

ROLE OF SERUM HEPCIDIN AND RETICULOCYTE HEMOGLOBIN CONCENTRATION IN EVALUATION OF ANEMIA IN ULCERATIVE COLITIS PATIENTS

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Objective: One of the most common extra-intestinal signs of ulcerative colitis (UC) disease is anemia, which has a considerable influence on patients' quality of life. **AIM:** The aim was to evaluate the role of serum hepcidin and reticulocyte hemoglobin concentration (CHr) in the study of anemia in UC patients. **Methodology:** We recruited 80 UC patients and 30 healthy individuals of matched age and sex as controls. Subjects were subdivided into three groups – Group I: 50 anemic UC patients, Group II: 30 nonanemic UC patients, and Group III: 30 healthy controls. **Results:** CHr showed a statistically highly significant decline in Group I than Groups II and III. Serum hepcidin showed a significant difference between Groups I, II, and III. Also, a significant negative correlation between CHr, serum hepcidin and severity of UC and a significant positive correlation between CHr and hemoglobin level, MCV, serum ferritin, and transferrin S. While serum hepcidin had a significant positive correlation with hemoglobin level, MCV, serum ferritin, transferrin S., and CHr. **Conclusion:** CHr had an excellent performance in prediction of iron-restricted anemia and was the test of best performance in prediction of iron-deficiency anemia ± ACD. Serum hepcidin had an excellent performance in prediction of ACD.

https://doi.org/10.1016/j.htct.2022.09.1262

IMMUNODEFICIENCIES / NEUTROPHIL DISEASES

PP 28

GRISCELLI SYNDROME TYPE 2 –CLINICAL APPROACH AND CASE REPORT

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Objective: Griscelli syndrome type 2 is rare authosomal ressesive disorder caused by a defect in the RAB27A gene, which affects a melanosome-anchoring complex in melanocytes, affecting release of cytolytic granules from T and NK cells. Children with GS type 2 develop an uncontrolled T-lymphocyte and macrophage activation syndrome known as hemophagocytic syndrome (HS) or hemophagocytic lymphohistiocytosis (HLH). We describe a 3 years old girl patient classic features of Griscelli syndrome type 2 **Case report:** A 3-year-old girl was admitted to hematology with complaints of LAP, hepatosplenomegaly and pancytopenia (WBC- 3080/μl, Hb-7.6 g/dl, Neutr.-350/μl, PLT - 165000/μl). The patient's condition was below the percentile, her skin was bronze, her hair was silver-grey. HLH criteria were met (triglycerides 458 mg/mL, ferritin 3445 ng/mL, fibrinogen 180 mg/dl). Morphology of the bone marrow was hypocellular, signs of dyserythropoiesis (stage I) and megakaryocytes were reduced **Methodology:** According to the clinical and laboratory data (hepatosplenomegaly, increased ferritin, hypertriglyceridemia, pancytopenia, hyperthermia resistant to antimicrobial therapy, silver-gray hair, pigment balls of hair seen light microscope) and the death of another undiagnosed child in the family, suggested likely primary HLH and GS. As a result of genetic analysis (homozygous mutation c.514_ 518del-CAAGC(p.GLN172Asnfs*,rs767481076)1 in the RAB27A gene), the diagnosis of GS type 2 was confirmed. **Results:** The patient was treated according to the HLH 2004 protocol. CSA levels were measured once a week. IVIG support was given based on IgG levels. HSCT was planned from patient's healthy HLA-matched sibling, but HSCT was delayed because the brother was infant. After 45 weeks of maintenance therapy, etoposide was discontinued, dose of dexamethasone was reduced to 5 mg/kg, but CSA was continued at the same dose. Control studies are carried out once a week. As far as possible HSCT is planning **Conclusion:** The prognosis of patients with Griscelli syndrome is poor. It is usually rapidly fatal within 1-4 years without aggressive treatment and bone marrow transplantation at onset of an accelerated phase. HSCT is more successful when implemented early course of the disease. Palliative care includes treatment and prophylaxis care infections and immunosupressor therapy in accelerated phases. Some patients have died after transplantation, but others have had lasting remissions

https://doi.org/10.1016/j.htct.2022.09.1263

LEUKEMIA

PP 29

THE COURSE OF TOXIC HEPATITIS IN LEUKEMIC PATIENTS AT THE STAGE OF SUPPORT THERAPY.

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Objective: One of the main tasks in the treatment of acute leukemia is to prevent the development of complications of chemotherapy, as well as the timely choice of the correct tactics for the treatment of complications. Because forced breaks associated with complications negatively affect the end result