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CLINICAL MANIFESTATIONS OF PAROXYSMAL NOCTURNAL HEMOGLOBINURIA

Introduction. Paroxysmal nocturnal hemoglobinuria (PNH) is a clonal hematopoietic stem cell disorder that manifests with hemolytic anemia, bone marrow failure, and thrombosis. Hemolysis in PNH is complement mediated and is a direct result of PNH cells acquiring a deficiency of complement regulatory proteins. The disease begins with the expansion of a hematopoietic stem cell that has a severe deficiency or absence for GPI, a glycolipid moiety that anchors >150 different proteins to the cell surface. GPI anchor deficiency in virtually all PNH cases is the result of a somatic mutation in PIGA, an X-linked gene whose product is required for the first step in GPI anchor biosynthesis. This results in the deficiency of complement inhibitory proteins CD55 and CD59 that leads to chronic complement-mediated hemolysis of the GPI-deficient erythrocytes, as well as activation of platelets, monocytes, and granulocytes. Anemia in PNH is often multifactorial and may result from a combination of hemolysis and bone marrow failure. Intravascular hemolysis with moderate to severe anemia, an elevated reticulocyte count, and up to a 10-fold increase in lactate dehydrogenase (LDH) is common in classical PNH. Thrombosis leads to severe morbidity and is the most common cause of mortality in PNH. Thrombosis in PNH may occur at any site; however, venous thrombosis is more common than arterial. Abdominal pain, esophageal spasm, dysphagia, and erectile dysfunction are common symptoms associated with classical PNH and are a direct consequence of intravascular hemolysis and the release of free hemoglobin. PNH patients have a greater than sixfold increased risk of chronic kidney disease. Renal tubular damage is caused by microvascular thrombosis and accumulation of iron deposits. Mild to moderate pulmonary hypertension is more common that previously recognized. Raised pulmonary pressures and reduced right ventricular function caused by subclinical microthrombi and hemolysis-associated NO scavenging contribute to symptoms of fatigue and dyspnea. Terminal complement

inhibition with eculizumab and allogeneic bone marrow transplantation (BMT) are the only widely effective therapies for patients with classical PNH. Improved knowledge of the molecular and cellular underpinnings of PNH over the last 2 decades has resulted in greater understanding of the biology and natural history of PNH. Recent studies with the monoclonal antibody, eculizumab, demonstrate that terminal complement inhibition controls most of the symptoms and life-threatening complications of PNH.

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THE RELATIONSHIP BETWEEN VASPIN, TNF-A LEVELS AND THE PARAMETERS OF CARBOHYDRATE METABOLISM IN PATIENTS WITH TYPE 2 DIABETES MELLITUS

Background. It is well known that the adipose tissue is an active endocrine organ. It produces a large number of substances which are involved in the regulation of metabolic and physiological processes. One of representatives is vaspin- adipokine, which is involved in some obesity-related disease states such as insulin resistance.

Aim. To determine the level of vaspin and TNF-α in blood of patients with type 2 diabetes mellitus (T2DM) depending on parameters of carbohydrate metabolism and body mass index (BMI).

Materials and methods. The study involved 31 patients (25 men) with T2DM. The following parameters have been assessed: BMI, HbA1c, immunoreactive insulin (IRI), HOMA-IR and fasting plasma glucose (FPG). The plasma level of vaspin and TNF-α were determined by ELISA. The control group included 10 healthy volunteers.

Results and Discussion. Examined patients had statistically significant deviations of the studied parameters in comparison with the control group. It was noted that the level of vaspin correlated with FPG ($r=0.62;p<0.01$), IRI($r=0.6;p<0.001$) and HOMA-IR($r=0.45$; $p<0.05$). These correlations can be estimated as a compensatory pathway in the development of insulin resistance (IR). Meanwhile the relationship between TNF-α and IRI($r=0.52$; $p<0.01$) can be a link in the development of IR. Correlation of HbA1c