Hemolytic uremic syndrome (HUS) – case report

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ABSTRACT

Introduction Hemolytic uremic syndrome (HUS) is characterized by hemolytic anemia, thrombocytopenia and acute renal failure. In 90% of children, HUS most often develops after an intestinal infection caused by verotoxin-producing E. coli. The remaining 10%, without any preceding diarrhea, are diagnosed with atypical HUS.

Aim The objective of the study was the presentation the case of two patient with hemolytic uremic syndrome.

Results The case report concerns two patients diagnosed with haemolytic uremic syndrome. Both cases shared many symptoms. Initially gastroenteritis, dehydration followed by thrombocytopenia, renal failure and anemia. These are examples of a typical hemolytic uremic syndrome. Many additional tests were carried out, including morphology, urinalysis, blood chemistry, but the final diagnosis was made on the basis of the clinical picture and the triad of symptoms typical of HUS.

Conclusions Hemolytic uremic syndrome is a disease that threatens the life of a child. It is important to recognize symptoms as soon as possible and begin treatment to prevent dangerous complications.

Key words: hemolytic uremic syndrome, kidney

INTRODUCTION

Hemolytic uremic syndrome (HUS) is a condition that can occur when the small blood vessels in your kidneys become damaged and inflamed. This damage can cause clots to form in the vessels. The clots clog the filtering system in the kidneys and lead to kidney failure, which could be life-threatening. By definition, it is a thrombotic microangiopathy with a characteristic triad of symptoms: haemolytic anemia, thrombocytopenia and acute renal failure [1]. HUS most often develops in 90% of children after an intestinal infection caused by verotoxin-producing E. coli. The remaining 10%, without any preceding diarrhea, are diagnosed with atypical HUS [2]. The hemolytic uremic syndrome has a different clinical manifestation, it is divided into typical and atypical. Typical (D +) - preceded by diarrhea, atypical (D -) is not preceded by an intestinal catarrh. Is also a secondary form of HUS, which may be e.g. drug-induced, may occur in the course of neoplastic disease, glomerulonephritis, HIV infection, after bone marrow transplantation [3]. The seasonality of occurrence is characteristic of the typical form - most cases are recorded in the spring and summer months.
The typical form appears in infants and younger children, practically never in newborns or older children - which may be associated with a greater maturity of the immune system [4]. Atypical HUS is not preceded by diarrhea, is not characterized by a seasonal nature of its occurrence, and may occur in families. It is a heterogeneous group of disorders of the immune system and coagulation related to the abnormal functioning of the complement system, consisting in its permanent activation [5].

**CASE REPORT**

A 13-month-old patient admitted to the Pediatric Department due to progressive dehydration in the course of gastroenteritis. For 3 days there has been watery diarrhea, currently about 10 stools per day. In also, vomiting occurs after oral hydration, which continues from the day before admission. No fever. He was taking Nifuroxazide. According to the mother's report, there was a partial improvement in the child's health. The patient is not under the care of a specialist clinic. He has a delayed vaccination schedule. Negated allergies. Boy at the admission in general average condition. The child is apathetic and dehydrated in a moderate state. Single petechiae are visible on the skin in the neck area and on the forearms. Regular heart rate - 130 beats per minute. Auscultatorically normal alveolar respiratory murmur over the lung fields. Abdomen palpable soft, peristalsis intensified. Liver and spleen not enlarged. Crotch area unchanged. In lab studies leucocytosis is 14,56 10⁹/l. There are anemization features: hemoglobin 9,7 gr/dl, and thrombocytopenia platelets 56,0 10⁹/l. There are elevated markers of inflammation: PCT 4,94ng/ml, CRP 1,27 mg/dl. Clotting times normal. Gasometric disturbances increase over time: pH 7,14, BE -24,2 mmol/l, hyperkalemia, hypoglycemia. Test reveals features of liver and kidney damage: creatinine 2,11 mg/dl, hyperurycemia, urea retention up to 157 mg/dl, ALT 122U/l, hyperbilirubinemia. Stool test for rotavirus, noravirus, adenovirus - negative. During the morning round, the general condition deteriorated. The patient was asleep, with no appetite. Anuria was diagnosed - last micturition 8 hours earlier. Edema appeared on the feet and hands. Blood was found in numerous diarrheal stools. Continuous monitoring of vital signs was carried out. Blood pressure 166/67 mmHg, heart rate 130-150 / min SpO2 96%. The patient was catheterized - urine was present in the drain. The child is irrigated intravenously under glycemic control. Due to developing renal failure, haemolytic uremic syndrome was suspected.

Another patient, 2 years and 11 months old, was admitted to the Pediatric Department due to dehydration in the course of gastroenteritis.

In the interview, mother says that the child has had diarrhea for 3 days – several stools a day, periodically stools with the presence of fresh blood. Additionally, they report vomiting - several times a day and lack of appetite. No fever. On admission, the general condition was average. Skin without pathological eruptions. Drying, sticky mucous membranes in the mouth, pale throat. Regular heartbeat, frequency 110 / min, normal alveolar murmur above the lung fields. The abdomen is soft, peristalsis increased.In laboratory tests in morphology, leukocytosis 25 10⁹/l, no evidence of anemia, negative CRP and PCT, pH 7.45, urea 13 mg/dl, creatinine 0.44 mg/dl, hypoglycaemia. The presence of rotaviruses and noroviruses in the
stools. The gastrointestinal diseases of viral etiology was diagnosed. Intravenous hydration was applied, Hidrasec p.o. Clinical improvement was temporarily achieved. The vomiting subsided. Numerous watery stools, without pathological impurities, without fever during hospitalization. In the study control, leukocytes 20.5 \(10^9/l\), features of anemization, hemoglobin 9.8 gr/dl, PLT 33.0 \(10^9/l\) - thrombocytopenia. High CRP 8.5mg/dl, PCT 5mg/ml, OB 8mm/h. Kidney function deteriorated - creatinine level from 1.25 to 1.68 mg/dl, urea 78 mg/dl, hyponatremia, hypokalemia, INR 1.18. Deterioration in general condition was observed. Patient weakened, strenuous not taken food, fluids are reluctant to accept. Nausea and vomiting appeared. He required intravenous hydration. There was a yellowing of the skin layers. Pathological spots appeared on the forearms. Anuria. Renal failure progressed. Suspicion of haemolytic uremic syndrome.

Both patients were referred to the Department of Nephrology for further treatment.

**DISCUSSION**

In both patients, we notice typical HUS symptoms. These include: haemolytic anemia, thrombocytopenia, and renal failure. Both cases were diagnosed during the summer season. We are dealing here with a typical HUS (D+). Patients meet criteria such as the presence of diarrhea, as well as hemorrhagic diarrhea, age less than 6 months [2]. In 90% of cases, the cause is infection with a bacterium that produces verotoxin - enterohaemorrhagic Escherichia coli (EHEC, serotype O157: H7 or O104: H4) or Shigella dysenteriae (more often in children) [6]. As a result of bacterial toxin damage to endothelial cells within the kidneys, abnormal "unusually large" multimers of von Willebrand factor (UlvWF) enter the renal circulation and bind to platelets and cause local formation of platelet aggregates. Similar changes may also occur in other organs.

Additional tests are helpful in diagnosis. Blood counts include normocytic anemia, erythroblasts and schistocytes in the blood smear, increased number of reticulocytes, and thrombocytopenia. Blood chemistry tests indicate increased levels of free bilirubin and LDH activity, features of impaired renal function. Urinalysis, on the other hand, tells us about proteinuria, or hematuria. There is also an increased concentration of fibrin degradation products (FDP), sometimes also the D-dimer.

Advances in symptomatic treatment have reduced the previously reported high mortality of typical ZHM in children from 40% to less than 4%. Symptomatic treatment consists in the early initiation of dialysis therapy, meticulous monitoring of the state of hydration and arterial hypertension, correction of electrolyte disturbances and metabolic acidosis, and administration of a red blood cell concentrate (RBC) to correct anemia. Thrombocytopenia in patients with typical HUS requires the use of platelet mass in exceptional cases. Plasmapheresis and plasma infusions are not indicated in the treatment of typical HUS [7].

**CONCLUSIONS**
Hemolytic uremic syndrome is a rapidly developing disease in children. It poses a direct threat to the child's life. It is imperative to introduce treatment as soon as possible to avoid the negative effects of this disease.

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