Case Report
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Hemolytic Uremic Syndrome Still Confuses Minds: A Case With All Three Components of the Triad

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Abstract: Hemolytic uremic syndrome (HUS) is a condition that presents with a triad of microangiopathic hemolytic anemia, thrombocytopenia and acute renal failure. It is one of the leading causes of acute renal failure in the pediatric population. Its etiology includes infection, systemic diseases and various other causes. Prognosis depends on immediately starting plasmapheresis and if necessary, eculizumab. We report a patient that was admitted with bloody diarrhea and presented with all three components of the triad. The patient's renal function normalized after treatment with plasmapheresis and hemodialysis.

Keywords: Hemolytic uremic syndrome; hemolytic anemia; thrombocytopenia; acute renal failure.

Introduction

Hemolytic uremic syndrome (HUS) is a condition that presents with microangiopathic hemolytic anemia, thrombocytopenia and acute renal failure. In HUS, clinical signs begin after thrombotic microangiopathy. Pathologic lesions are artery and capillary wall thickening as well as endothelial thickening and damage. Thrombi made up of fibrin and thrombocytes can block arteries, leading to multiple organ damage including the kidneys. HUS is one of the leading causes of acute renal failure in the pediatric population. It must be considered in the differential diagnosis of children with anemia, decreased urine output and hematologic issues secondary to a recent viral infection and diarrhea. It is important to rule out thrombotic thrombocytopenic purpura (TTP) in the differential diagnosis of HUS based on ADAMTS 13 activity, which tends to be low in TTP.

Case Report

A 12-year-old female was referred to our clinic with complaints of sharp abdominal pain and bloody diarrhea for 10 days, and no urine output for 4 days. On day 3 after the complaints started, the family had applied to a different clinic and outpatient treatment was started. On day 3 of treatment, urine output began to decrease, and hemodyalysis was started 2 days before the patient was referred to our clinic. At the same time, the patient's mother and grandmother were being treated for bloody diarrhea at the first clinic at the same time.

On admission, the patient was conscious, weak and in respiratory distress with accessory muscle use. Skin and mucous membranes were pale. On auscultation, the patient had dyspnea and wet fine crackles; heart sounds were muffled. Abdominal palpation showed sharp pain, liver and spleen could not be palpated. Laboratory findings were as follows: WBC 11.700 /mm3, HGB 5.7 g/dL, PLT 33.000/mm3, creatinine 4.4 mg/dL, urea 95 mg/dl, BUN 44mg/dl, LDH 2125 U/L, total bilirubin 1.29 mg/dl, direct bilirubin 0.6mg/dl, direct Coombs negative, complement C3 1.13 g/l, complement C4 0.23g/l. Urinalysis showed 3+ blood and 3+ protein. Stool analysis showed high amounts of mucus, leukocytes and erythrocytes. Stool culture revealed no pathogenic bacteria (antibiotic treatment had been started at the previous clinic). Abdominal USG showed

intestinal wall inflammation and signs of grade 1 bilateral renal parenchymal disease; kidney size was normal. Liver and spleen echogenicity and size were normal.

The triad of hemolytic anemia, thrombocytopenia and acute renal failure pointed to the diagnosis of hemolytic uremic syndrome. Plasmapheresis and hemodialysis were started. Due to respiratory distress, the patient was placed on mechanical ventilation. After 3 days, the patient was extubated. On day 10 of treatment urine output started to improve, and on day 16 output normalized and treatment was stopped. Laboratory findings were normal: WBC 4.590/mm3, HGB 10.9 g/dl, PLT 179.000/mm3, creatinine 1.1mg/dl, urea 38.9 mg/dl, BUN 17mg/dl. The patient was discharged on day 22 and remains on outpatient follow-up with no problems.

Discussion

HUS is the leading cause of acute renal failure in children and is characterized by the triad of microangiopathic hemolytic anemia, thrombocytopenia and acute renal damage. It is often observed after upper respiratory tract infections and gastroenteritis. The two types of HUS are diarrhea-positive (typical) and non-diarrhea-associated (atypical). Typical HUS is most often caused by E.coli O157:H7 as well as streptococcus pneumaniae, HIV and various other infections. Among the genetic causes of HUS are mutations in genes of the complement system, cobalamin C metabolism deficiency and DGKE gene mutations. In cases of atypical HUS, genetic testing is recommended in addition to first line treatment of plasmapheresis and eculuzimab when necessary. If a genetic mutation is confirmed and renal failure progresses to the final stage, a renal transplant is recommended.

Diagnostically HUS is characterized by spherocytes and schistosytes in peripheral blood smear, anemia, thrombocytopenia, reticulocytosis, azotemia, elevated LDH and indirect bilirubin, and low haptoglobin.

The symptomatic approach to HUS includes red blood cell transfusion if HGB is 6-7 g/dL or hematocrit is <18%. Thrombocyte transfusions are necessary in cases of clinically significant hemorrhage or during invasive procedures. Other measures include maintaining daily fluid volume, correcting electrolyte imbalances, stopping medications that are nephrotoxic or implicated in HUS etiology, and providing normal nutrition.

Prognosis in hemolytic uremic syndrome depends on timely treatment; recovery follows in 90% of cases. Lasting complications can develop as a result of microthrombosis of various organs.

Conclsuion

It is a fact that hemolytic uremic syndrome in children and infants may not be diagnosed easily and quickly. The main safeguard against mortality and morbidity remains a high index of suspicion.

References

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