Caspian Journal of Pediatrics
Babol University of Medical Sciences
e-ISSN: 2383-3106

Acute Abdomen Presentation of Hemolytic Uremic Syndrome in a Child: A Case Report

Sahar Sadr Moharrerpour [1*], Soheil Osia [2], Sajedeh Hajipour [2], Sajedeh Omidbakhsh [3]

1. Non-Communicable Pediatric Diseases Research Center, Health Research Institute, Babol University of Medical Sciences, Babol, IR Iran.
2. Clinical Research Development Unit of Amirkola Children's Hospital, Babol University of Medical Sciences, Babol, IR Iran.
3. Clinical Department, Amol Campus of Medicine, Mazandaran University of Medical Sciences, Sari, Iran

*Corresponding Author: Dr. Sahar Sadr Moharrerpour;
Address: Non-Communicable Pediatric Diseases Research Center, No 19, Amirkola Children's Hospital, Amirkola, Babol, Mazandaran Province, 47317-41151, IR Iran.
Tel-Fax: +98 1132346963 E-mail: drsaharsadr@gmail.com, s.sadr@mubabol.ac.ir

Article Info.

Article type:
Case Report

Received: 4 Oct. 2023
Revised: 11 Dec. 2023
Accepted: 8 Jan. 2024
Published: 2 March 2024

Keywords:
Acute Kidney Injury, Diarrhea, Hemolytic- Uremic Syndrome

ABSTRACT

Background and Objective: Hemolytic uremic syndrome (HUS) is the most common cause of acute kidney injury (AKI) in children, characterized by the triad of thrombocytopenia, non-immune hemolytic microangiopathic anemia, and AKI. While typically presenting with nonspecific symptoms like fatigue and pallor, severe gastrointestinal involvement can occur. This report details a rare complication of HUS, toxic megacolon, in a pediatric patient who initially presented with symptoms including diarrhea, vomiting, and abdominal pain, necessitating surgical intervention

Case Report: A 7-year-old boy presented to the emergency room with fever, vomiting, and bloody diarrhea, and was initially managed conservatively. His abdominal pain worsened with tenderness and guarding observed on examination, leading to a diagnosis of acute abdomen requiring surgical intervention. Surgery revealed toxic megacolon. Postoperatively, the patient developed pallor and oliguria. Based on laboratory findings and the presence of schistocytes on peripheral blood smear, a diagnosis of HUS was made, and he underwent hemodialysis.

Conclusion: This case reminds us that HUS can present with very rare symptoms such as toxic megacolon. Therefore, in any pediatric patient with toxic megacolon and renal impairment the diagnosis of HUS must be considered.

Cite this Article: Sadr Moharrerpour S, Osia S, Hajipour S, Omidbakhsh S. Acute Abdomen Presentation of Hemolytic Uremic Syndrome in a Child: A Case Report. *Caspian J Pediatrs* March 2024; 10: e20.

[DOI: 10.22088/CJP.BUMS.10.1.20]

Introduction

Hemolytic uremic syndrome (HUS) is one of the most common causes of acute renal failure in children. It is manifested by a triad of hemolytic anemia, thrombocytopenia and acute renal failure and is diagnosed by the presence of schistocytes in the peripheral blood smear (PBS). In addition, reticulocytes and lactate dehydrogenase are elevated and haptoglobin is decreased [1, 2].

In studies, the incidence is distributed worldwide, 1.11 cases per 100,000 children in Canada, despite 0.4 per 1,000,000 in Germany [3, 4]. The usual age of onset is 1-4 years [1]. More than 90% of cases are associated with Shiga toxin-producing E. coli (SIEC) infections [5, 6], particularly E coli O157: H7. Which is most common in Europe and the USA [7, 8].

The source of STEC is domesticated animals like cattle. Another type of HUS that occur after infection is caused by Streptococcus pneumoniae, a producer of neuraminidase, which leads to a more severe form of HUS. The genetic form of HUS is due to a deficiency in factors of the alternative complement pathway, such as factors H, I and B [2].

Symptoms of HUS following infection with STEC typically develop 3-4 days after the onset of diarrhea and vomiting. The spectrum of symptoms ranges from severe to mild and usaully includes pallor, fatigue and abdominal pain. Fever is rarely observed in these patients. Severe gastrointestinal complications such as hemorrhagic colitis, toxic megacolon, and transmural necrosis occur in 5-10% of cases [9]. The incidence of colonic perforation is reported to be 1-2 % and it is still very difficult to differentiate the abdominal symptoms associated with HUS colitis from perforation [10].

There is no specific treatment for typical HUS, the treatment is supportive. Supportive treatment includes controlling the body's fluid and electrolyte balance. If diarrhea and vomiting have led to dehydration, the patient's fluid and electrolyte deficits must be compensated. If the patient is oliguric and has hypervolemia on examination, fluid restriction can be performed. If the patient does not respond to fluid restriction and remains hypervolemic despite treatment, dialysis may be performed for resistant hypervolemia. Packed red blood cells (PRBCs) are administered to treat the anemia if the hemoglobin is less than 6 g/dL. Elevated potassium levels must also be treated. Thrombocytopenia is treated if the patient has acute bleeding or needs surgery, in which case platelet transfusions are administered. Vasodilators and diuretics are used to treat high blood pressure. Eculizumab is a monoclonal antibody against C5 that inhibits the complement pathway and is used in atypical HUS. Antibiotic treatment is generally contraindicated in HUS; however, if the cause is Streptococcus pneumoniae, antibiotics are indicated. Treatment for atypical HUS may include plasmapheresis [11]. The aim of this case report is to present a case of HUS with an unusual presentation such as fever, abdominal pain which was finally operated because of toxic megacolon.

Case presentation

A 7-year-old boy was admitted to the emergency department of Amirkola Children's Hospital because he had been suffering from bloody diarrhea, vomiting, and fever for 4 days. The patient had no complaints of upper respiratory tract infection, tea-colored urine, pallor, jaundice, oliguria or anuria. Clinical history revealed no evidence of travel or contact with animals. He had no edema, petechiae or hepatosplenomegaly. He had abdominal pain without tenderness or guarding. On physical examination, the child was alert and had normal consciousness on admission. His vital signs, blood pressure, and heart rate were normal, but the patient was febrile. On the second day of admission, he was evaluated for severe abdominal pain and after being diagnosed with an acute abdomen, he underwent surgery and was found to have a toxic megacolon. The day after the operation, he became pale and oliguric. There was a decrease in hemoglobin and platelets and an increase in creatinine in the blood, which led to a suspicion of HUS. The results of the laboratory tests at the time of admission were as follows: white blood cell (WBC) count: 172000/mm3, red blood cell (RBC) count: 4.15*106/mm3, hemoglobin: 11.3 gr/dl, platelet count: 88000/µl, C-reactive protein (CRP): 26.4 mg/L, blood urine nitrogen (BUN): 5 mg/dl and serum creatinine (Cr): 0.6 mg/dl. Analysis of the urine sample revealed proteinuria (trace), RBC 0-

[DOI: 10.22088/CJP.BUMS.10.1.20]

1/HPF, and WBC 0-1/HPF. Examination of the stool revealed WBC 1-2/HPF and RBC 6-7/HPF. He was treated with a third-generation cephalosporin. On Day 1, the urine sample showed a trace amount of protein and 0-1 red blood cells. By Day 3, the protein level had increased to 4+, and the red blood cell count was 8-10. On the second morning of the admission day, his abdominal pain intensified and physical examination revealed diffuse abdominal tenderness with guarding. The patient underwent ultrasonography, which revealed a significant amount of free fluid in the right lower quadrant. On the advice of the surgeon and because of the suspicion of an abscess or colitis, the patient was treated with metronidazole and amikacin. After a further consultation, an emergency operation was recommended with the diagnosis of appendicitis. On surgical exploration, he was found to have 250 CC of free fluid in the peritoneal cavity with diffuse oozing hemorrhage from the entire surface of the colon without perforation at the bleeding sites, there was multimesenteric lymphadenitis and toxic megacolon.

The day after surgery, the patient became pale and oliguric. The laboratory values were as follows: WBC count=12000/mm3, hemoglobin=7.5 mg/dl, platelet count=69000/mm3, BUN=42 mg/dl, serum creatinine=2.6 mg/dl, lactate dehydrogenase (LDH)=3320 U/ml, SGOT=302 U/L, SGPT=180 U/L, alkaline phosphatase=202 IU/I, C3=49 mg (90-180) and C4=6mg (10-40) (Table 1). The HUS panel was sent in due to the low complement levels, and factors H, I, B and Adamts13 were tested, all of which were reported as normal. The following results were found: reticulocyte count=2.3. coombs=negative, haptoglobin=decreased, urine RBC=8-10, urine protein=1gr, and schistocytes were noted in PBS suggestive of HUS.

The patient became anuric, blood pressure was 130/80 mm/hg>95% percentile and the patient had epistaxis once. The patient was diagnosed with HUS and conservative treatment was startesd. Hydralazine was used to control blood pressure. A Shaldon catheter was placed, and the patient was dialyzed daily Hemodialysis was initially performed for one hour and then increased to three-hour sessions on subsequent days. On postoperative day 3, his hemoglobin decreased to 5.6 mg/dl and packed cells were transfused. The thrombocytopenia resolved spontaneously on postoperative day 5. Twenty sessions of hemodialysis and four times of plasma exchange were performed. The patient was followed up for two years after dialysis, he had no problems and no kidney problems.

Table 1. Laboratory findings of HUS patient

Day	BUN	Cr	Na	K	PH	Hco_3	Co_2
1	5	0.69	140	3.95	6.36	21	37
2	10	0.91	139	3.65	6.35	20	35
3	42	2.64	129	3.6	7.23	13	31
4	78	6.1	128	4.38	7.21	10	25
5	79	10	127	5	7.27	10.1	23
6	91	6.13	129	4.35	7.29	13	28

Discussion

This study presents a rare case of Hemolytic Uremic Syndrome (HUS) complicated by severe gastrointestinal involvement, specifically toxic megacolon requiring surgical intervention. While HUS is generally characterized by the classic triad of microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury [1], its manifestations are usually non-specific, including fatigue, pallor, and decreased urine output. Although abdominal pain, especially when associated with Shiga toxin-producing E. coli, is common, fever is rarely observed. In a small percentage of cases (5-10%) of severe gastrointestinal tract involvement, severe complications such as toxic megacolon, transmural necrosis of the colon, and colonic perforation with hemorrhagic colitis occur [1]; the incidence of reported colonic necrosis and perforation in case studies varies between 1-8% [6].

Diagnosis of HUS with hemolysis (see schistocytes in PBS), increase in lactate dehydrogenase, decrease in haptoglobin, platelets below 150000 and acute renal failure (1.5-fold increase in blood creatinine) with oliguria or anuria [12].

Management of HUS primarily involves supportive care .Blood transfusions should be avoided in HUS, but some suggest doing so only when hemoglobin is below 6 gr/dl [11]. Our case received blood transfusions when hemoglobin was 5.6 gr/dl on postoperative day 3 of post-operation.

[DOI: 10.22088/CJP.BUMS.10.1.20]

The thrombocytopenia observed in HUS resolves spontaneously within a week [11]. It should be avoided to transfuse more platelet than PRBCs. The indication for palate administration should be limited to the following cases: 1- severe bleeding leading to death, 2- the patient requires surgery [11]. In our patient, the thrombocytopenia resolved spontaneously on the 5th day after surgery. Most children with HUS develop some degree of renal failure. About two-thirds of children with HUS will require dialysis [13].

Our patient needed twenty dialysis sessions and plasma exchange four times. Poor prognostic factors for HUS have been suggested. These include: Leukocytosis of more than 20,000/l, oligoanuria for more than 5-14 days, dialysis for more than 4 weeks, central nervous involvement, and hypertension [5, 14-16]. The patient in our study had white blood cells <20000/dl, but he was oliguric for 5 days and was dialyzed 20 times.

Conclusion

HUS usually manifests as abdominal pain, cramps, diarrhea, nausea and vomiting. The predominant symptom is diarrhea, which usually starts after 3-4 days. However, in 5-10% of cases, severe complications such as hemorrhagic colitis in the colon and intestinal perforation may occur, and need to be considered. A diagnosis of HUS should be considered in any child who complains of diarrhea, especially if it is bloody. Awareness of HUS is important to avoid inappropriate surgical interventions. This case reminds us that HUS can present with very rare symptoms such as toxic megacolon. Therefore, it is necessary to be familiar with all the symptoms of HUS.

Acknowledgments

The authors would like to thank the Clinical Research Development Unit of Amirkola Children's Hospital, Babol University of Medical Sciences for their contribution to this study.

Funding

This study was self-funded.

Ethical approval

This study was approved by the Ethics Committee of Babol University of Medical Sciences (IR.MUBABOL.REC.1402.009).

Conflict of interest

The authors declare that there is no conflict of interest.

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[DOI: 10.22088/CJP.BUMS.10.1.20]