Surgical complications of hemolytic uremic syndrome: Single center experiences

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ABSTRACT

Purpose: To determine the prevalence, outcome and prognostic factors in children with hemolytic uremic syndrome (HUS) who underwent laparotomy. Materials and Methods: The medical records of 104 patients with HUS who presented to our center between 1986 and 2003 were reviewed retrospectively. Data were analyzed using Student’s t test for comparing means, Fisher’s exact test for frequencies and Pearson’s correlation for finding the correlations. Results: 78% of cases presented with vomiting and diarrhea. Seven out of 104 needed surgical exploration. The indications of surgery were acute abdomen, severe abdominal distention and the sign of peritonitis. The findings at laparotomy were intussusceptions, perforation (colon, ileum), gangrene of entire colon, rectosigmoidal tearing, duodenal obstruction and toxic megacolon. Pathological findings were transmural infarction in two cases in which staged surgical management was performed (cecostomy, resection, later anastomosis). Four out of seven patients died because of pulmonary failure, coma and multiple organ failure (P < 0.05) compared to those who did not need laparotomy. The patients requiring surgery were young (<3 years), had high leukocyte count (>20000 mm³) and low albumin level (<3 g/dl) (P < 0.05). Conclusion: Surgical complications of HUS are rare but are assorted with high mortality due to respiratory failure and multiple organ failure. Early decision of laparotomy associated with intensive care, including mechanical ventilation, adequate dialysis and ultrafiltration, are recommended.

KEY WORDS: Bowel obstruction, bowel perforation, hemolytic uremic syndrome, ischemic colitis

INTRODUCTION

Hemolytic uremic syndrome (HUS) consists of acute renal failure, hemolytic anemia and thrombocytopenia. Gastrointestinal symptoms may be preceded or manifested during the course of HUS.[1-4] Surgical complications of HUS are rare and have been reported individually as colonic necrosis with or without perforation,[5-10] intussusceptions,[11] toxic megacolon,[12] colonic stenosis and esophageal stricture.[13] Severe colitis is associated with higher extraintestinal complication.[13,14] The aim of this study is to review the prevalence of surgical complications, prognostic factors and their outcomes in our center.

MATERIALS AND METHODS

The records of all patients with HUS admitted to our hospital from 1986 to 2003 were reviewed retrospectively. Inclusion criteria were Coombs negative hemolytic anemia, bur cells and helmet cells on peripheral blood smear, platelet count less than 150000/mm³ and increased serum creatinine in comparison to the normal ranges for age in two consecutive days. All cases had stool, urine and blood cultures, but peritoneal fluid cultures were only performed in patients on dialysis. No specific test for E. coli 0157:H7 was available. Dialysis catheter was inserted percutaneously at bedside by physicians. Two groups were formed: the first one was the patients who required surgical intervention, and the second one was those who did not require surgery. The primary clinical findings, laboratory data, complications and final outcomes were compared between these two groups. Differences were tested using Student’s t test for the mean values, Fisher’s exact test for frequencies and Pearson’s correlation for determining the correlations. The P values less than 0.05 were considered as significant.
RESULTS

The records of all 104 children (70 boys, 34 girls) with diagnosis of hemolytic uremic syndrome were collected. Around 78% of cases presented with vomiting and diarrhea. Seven out of 104 patients developed complications requiring surgical intervention [Table 1]. The indications of surgery were acute abdomen, severe abdominal distention and the sign of peritonitis. The findings at laparotomy were intussusceptions, perforation, gangrene of intestine, duodenal obstruction and toxic megacolon. Pathological findings were transmural infarction in two cases in which staged surgical management was performed (cecostomy, resection, later anastomosis). All the factors that were believed to influence the severity of gastrointestinal involvement were analyzed. The patients who required surgery were younger, had high leukocyte count, high polymorphonuclear count and low albumin levels [Tables 2, 3]. There were no statistically significant differences between the two groups for sex, prodromal duration and other laboratory results on admission. We found no correlation between the severity of gastrointestinal disease and severity of renal or central nervous system (CNS) involvement (P > 0.05). The early death rate was high in patients with surgical complications who underwent surgery (P < 0.05). Four out of seven patients died because of pulmonary failure, coma and multiple organ failure (P < 0.05) compared to those who did not need laparotomy. There was no significant difference between two groups regarding the CNS involvement. Three survived cases exhibited no renal insufficiency, proteinuria or hypertension after a mean follow-up of 2.66 years.

DISCUSSION

HUS is defined by hemolytic anemia, thrombocytopenia and acute renal failure. Gastroenteritis usually precedes typical HUS. Sometimes patients may present with significant peritoneal irritation that mimic acute abdomen leading to unnecessary surgery.[1,11,13,15] The surgical complications of HUS are rarely reported. The incidence was 6.7% in our series; this is similar to other studies.[13-16] It predominantly involves the colon. Ischemic colitis due to small-vessel thrombosis is self-

Table 1: Patients with surgical complications

<table>
<thead>
<tr>
<th>Patient number</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td>M</td>
<td>M</td>
<td>M</td>
<td>M</td>
<td>F</td>
<td>M</td>
<td>M</td>
</tr>
<tr>
<td>Age (years)</td>
<td>2</td>
<td>2.5</td>
<td>2</td>
<td>2.5</td>
<td>0.9</td>
<td>1</td>
<td>2.5</td>
</tr>
<tr>
<td>Time required for laparotomy (days)</td>
<td>10</td>
<td>8</td>
<td>6</td>
<td>44</td>
<td>17</td>
<td>13</td>
<td>15</td>
</tr>
<tr>
<td>Days of hospitalization (day)</td>
<td>10</td>
<td>13</td>
<td>41</td>
<td>31</td>
<td>4</td>
<td>3</td>
<td>20</td>
</tr>
<tr>
<td>Gut complications</td>
<td>Intussusception of sigmoid colon, descending colonic perforation</td>
<td>Gangrene and transmural necrosis of colon</td>
<td>Rectosigmoid tearing, ileal perforation, transmural necrosis of colon</td>
<td>Duodenal Obstruction, Pancreatitis</td>
<td>Intestinal Perforation, free air on KUB</td>
<td>Toxic Megacolon</td>
<td>Toxic Megacolon</td>
</tr>
<tr>
<td>Laparotomy</td>
<td>Reduction and repair of perforation</td>
<td>Cecostomy, colon resection, ileorectal anastomosis (3 weeks later)</td>
<td>Cecostomy, colon resection, ileorectal anastomosis (3 weeks later)</td>
<td>Cecostomy, Resection, Anastomosis (2.5 months later)</td>
<td>Release Ladd’s band</td>
<td>Died before surgery</td>
<td>Died before surgery</td>
</tr>
<tr>
<td>WBC (PMN)* (mm$^3$)</td>
<td>29600 (80)</td>
<td>17500 (82)</td>
<td>22600 (75)</td>
<td>77000 (68)</td>
<td>27600 (66)</td>
<td>47300 (84)</td>
<td>55000 (80)</td>
</tr>
<tr>
<td>Platelet* (mm$^3$)</td>
<td>18000</td>
<td>10000</td>
<td>88000</td>
<td>75000</td>
<td>61000</td>
<td>61000</td>
<td>30000</td>
</tr>
<tr>
<td>Hematocrit (g/dl)*</td>
<td>32.6</td>
<td>28</td>
<td>26.2</td>
<td>28</td>
<td>15</td>
<td>16</td>
<td>33</td>
</tr>
<tr>
<td>BUN (mg/dl)*</td>
<td>17</td>
<td>24</td>
<td>40</td>
<td>41</td>
<td>155</td>
<td>68</td>
<td>40</td>
</tr>
<tr>
<td>Creatinine (mg/dl)*</td>
<td>1.3</td>
<td>1.8</td>
<td>4.3</td>
<td>2.2</td>
<td>4.4</td>
<td>6.9</td>
<td>1.5</td>
</tr>
<tr>
<td>Culture*</td>
<td>Negative</td>
<td>Negative</td>
<td>Enterobacter (peritoneal fluid culture)</td>
<td>Negative</td>
<td>Enterobacter (blood)</td>
<td>Negative</td>
<td>Negative</td>
</tr>
<tr>
<td>Peritoneal dialysis</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>CNS involvement</td>
<td>coma</td>
<td>Respiratory failure</td>
<td>Respiratory failure</td>
<td>Seizure</td>
<td>Respiratory failure</td>
<td>Seizure</td>
<td>Multiorgan dysfunction</td>
</tr>
<tr>
<td>Reason of death</td>
<td>Respiratory failure</td>
<td>Respiratory failure</td>
<td>Respiratory failure</td>
<td>Seizure</td>
<td>Respiratory failure</td>
<td>Seizure</td>
<td>Multiorgan dysfunction</td>
</tr>
</tbody>
</table>

*On admission, †Included blood, stool, urine and peritoneal fluid cultures (in the case of inserting dialysis catheter), PMN = polymorphonuclear, WBC = white blood cell, CNS = central nervous system, BUN = blood urea nitrogen
limited; however, it may progress to toxic megacolon, extensive necrosis of colon and perforation. Colonic perforation tends to occur at the end of the second week; this is similar to our findings.\[4-6,8-10,12,13,17,18\] However, there was no significant difference between the two groups regarding the duration of prodrome. We found significant correlations between the surgical complications and younger age, high leukocyte and neutrophil counts and lower serum albumin. In contrast to other studies,\[13,19,20\] there were no significant correlations between sex, CNS presentation, calcium or hemoglobin levels and surgical complications. The epidemic form of HUS with a diarrheal prodrome tends to occur in younger patients. Neutrophil traffic in colonic mucosa increases the transport of toxin across the epithelial barrier; therefore, a higher peripheral blood neutrophil count predicts the severity of HUS.\[3\] The association between low serum albumin and disease severity may indicate the gastrointestinal protein loss. The incidence of early death was significantly higher in children who underwent surgery. Two patients even died before laparotomy. We found severe pulmonary and multiple organ failure as causes of early death in this study. Peritoneal signs without documented intestinal catastrophe in a patient with HUS do not indicate laparotomy. Nevertheless, whenever a child with HUS has been suspected to acute abdomen, requesting for surgical consultation several times a day is necessary to make an earlier decision with regard to laparotomy. In addition, adequate dialysis is necessary to remove toxins and achieve fluid and electrolyte balance. In the case of severe intestinal involvement, peritoneal dialysis may not allow sufficient ultrafiltration in comparison to hemodialysis or continuous hemofiltration. The risk of death is high when these techniques are not easily available.

REFERENCES

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