Acute intestinal obstruction requiring surgery in pediatric malignancies: Summary of 7 cases

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Article info

Article history:
Received 16 May 2015
Received in revised form 27 May 2015
Accepted 30 May 2015

Key words:
Intestinal obstruction
Children
Malignancy

Abstract

Catheter-related or perianal problems are common surgical complications in patients with pediatric malignancies. Acute intestinal obstructions requiring surgical intervention are rare, however. We herein report our experience of 7 patients with separate primary diagnoses who developed acute intestinal obstruction that mandated surgery before or during chemotherapies. The original diseases were hematologic malignancies in 5 patients (3 leukemia, 1 lymphoma, 1 Langerhans cell histiocytosis (LCH)) and solid tumors in 2 patients (1 intra-abdominal desmoplastic small round cell tumor (DSRCT), 1 rhabdomyosarcoma of vagina). The intestinal obstructions were treated by resection of the involved segment with (N = 2) or without (N = 3) enterostomy. Two cases of intestinal obstruction were treated with an enterostomy alone. Three patients died due to the progression of the original disease, and 3 patients are currently alive. One patient was lost to follow-up. Although rare, acute intestinal obstructions can occur during the treatment course of pediatric malignancies or be an initial symptom of disease. Operative intervention is mandatory and effective in the treatment of such complications. Overall patient prognosis is dependent on the original disease.

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The overall 5-year survival rate from pediatric malignancies has improved to over 75% over the past 30 years along with advances in diagnostic modalities and chemotherapeutic agents [1], and the variety of complications requiring surgical intervention has also increased [2,3]. Catheter-related or perianal problems are common surgical complications [4,5], but the development of an acute intestinal obstruction, either before or during chemotherapy, is rare and unfamiliar to most of pediatric surgeons [6]. We herein report 7 cases of pediatric malignancies with acute intestinal obstruction that mandated surgical interventions and discuss the role of surgical treatment and prognosis of each case.

1. Case summary

There were 3 male and 4 female patients, and the median age at the time of the diagnosis of intestinal obstruction was 36 months (range, 5 months to 6.2 years). Intestinal obstruction occurred during the course of chemotherapy in 4 patients and was the initial presentation in 3 patients. All patients showed typical symptoms and signs of mechanical bowel obstruction. The underlying disease included 5 hematologic malignancies and 2 solid tumors. The 5 hematological malignancies included 3 leukemias, 1 Burkitt’s lymphoma, and 1 Langerhans cell histiocytosis (LCH). The 2 solid tumors included 1 intra-abdominal desmoplastic small round cell tumor (DSRCT) and 1 rhabdomyosarcoma arising from the vagina. There were no cases of neuroblastoma or Wilms’ tumor (Table 1). The causes of intestinal obstruction in patients with hematological malignancies included a paralytic ileus due to diffuse infiltration of leukemic cells along the entire length of the small intestine (Case No. 1, Fig. 1A and B), ileo-cecal obstruction by a leukemic mass (Case No. 2) or lymphoma (Case No. 3), ileal stricture by grade 4 graft versus host disease (GVHD) in a post-bone marrow transplantation (BMT) leukemic patient (Case No. 4, Fig. 2A and B), and ileal stricture due to the histiocytic infiltration in a patient with LCH (Case No.5, Fig. 3A and B). In case 1, we had decided exploratory laparotomy for two reasons; there were no radiologically demonstrable lesions to explain the obstruction and the clinical deterioration of the patient despite of conservative managements. The causes of intestinal obstruction for solid tumors included 1 DSRCT involving the ileum (Case No. 6, Fig. 4) and 1 rhabdomyosarcoma...
compressing the rectum externally (Case No. 7). Conservative management failed to relieve the obstruction in all cases. Treatment options for the intestinal obstruction were resection and anastomosis in 3 cases (Case No. 4, 5, 6), resection with enterostomy in 2 cases (Case No. 2, 3), and enterostomy alone (Case No. 1, 7). The median follow-up period was 13.5 months (range, 1–22 months) after the operation. Three patients died during the observation period due to progression of their underlying disease (Case No. 1, 3, 7), and 3 patients are now alive receiving chemotherapy. One patient has been lost to follow-up.

2. Discussion

The diagnosis of a surgical abdomen is usually made by clinical history and typical findings on the physical examination. When it occurs in patients receiving chemotherapy for malignant disease however, the diagnosis of a surgical abdomen can be difficult or often delayed if the surgeon confuses the symptoms of the underlying disease with the abdominal symptoms. This case report aims to describe the wide variety of clinical presentations and treatment outcomes of rare acute intestinal obstructions requiring surgery in patients with pediatric malignancies.

GVHD is a well-known post-BMT complication and usually involves the skin, liver, and gastrointestinal (GI) tract [7]. Gavel et al. reported a case of obstruction of the entire small bowel with GVHD in a 10-year-old boy; the pathological examination revealed transmural necrosis with fibrosis [8]. In our case, the pathological findings were identical to the previous report, but the lesion was more localized, and an ileocecectomy cured the obstruction. The extent of GI tract GVHD is likely to vary in different cases, and localized GVHD can be successfully treated by resection. The possibility of recurrent GVHD and subsequent obstruction should be kept in mind.

Granulocytic sarcoma is defined as an extramedullary leukemic cell infiltration; it frequently involves the skin, bone, soft tissue, lymph nodes, and GI tract [9]. Intestinal obstruction secondary to a granulocytic sarcoma has rarely been reported; most of the prior cases were mechanical obstruction due to a bulky mass and were successfully treated by resection and anastomosis [9,10]. Although not confirmed on intraoperative frozen section, the possibility of residual leukemic cells at the resection margin and the development of anastomotic complications led us to avoid primary anastomosis after resection in our cases. However, primary anastomosis appears to be a safe procedure after confirmation of the status of the margins and can be attempted in similar cases. We treated one

![Figure 1](image_url)

**Table 1**

Clinical presentations and treatment outcomes of an acute intestinal obstruction in pediatric malignancies.

<table>
<thead>
<tr>
<th>No.</th>
<th>Sex</th>
<th>Age (yr)</th>
<th>Original disease (primary site)</th>
<th>Location of lesion</th>
<th>Onset</th>
<th>Surgical treatment</th>
<th>Pathology of obstruction</th>
<th>Outcome (follow-up)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>6.2</td>
<td>AML</td>
<td>Jejunum + ileum</td>
<td>D</td>
<td>Ileostomy → jejunostomy*</td>
<td>Granulocytic sarcoma</td>
<td>Died (13)</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>3</td>
<td>JMML (coccum)</td>
<td>Ileum</td>
<td>D</td>
<td>Right colectomy, ileostomy</td>
<td>Granulocytic sarcoma</td>
<td>Loss (1)</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>6</td>
<td>Burkitt’s Lymphoma (coccum)</td>
<td>Cecum</td>
<td>B</td>
<td>Ileocecectomy, ileostomy</td>
<td>Burkitt’s lymphoma</td>
<td>Died (7)</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>3</td>
<td>AML</td>
<td>Ileum</td>
<td>D</td>
<td>Ileocecectomy</td>
<td>GVHD</td>
<td>Alive (20)</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>2.5</td>
<td>LCH</td>
<td>Ileum</td>
<td>D</td>
<td>Ileocecectomy</td>
<td>Histiocyte infiltration</td>
<td>Alive (14)</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>4.3</td>
<td>DSRCT (intraperitoneal)</td>
<td>Ileum</td>
<td>B</td>
<td>Ileum segmental resection</td>
<td>DSRCT</td>
<td>Alive (22)</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>0.5</td>
<td>Rhabdomyosarcoma (vagina)</td>
<td>Rectum</td>
<td>B</td>
<td>S-colostomy</td>
<td>N/A</td>
<td>Died (12)</td>
</tr>
</tbody>
</table>


* Onset of intestinal obstruction.

* Ileostomy failed to relieve the obstruction, and jejunostomy was performed 10 days after the ileostomy.

* Died of original disease progression.

* Surgical abdomen was the initial presentation.

* Month.

Fig. 1. (A) Note the yellowish, discoid leukemic patches spread along the whole intestine. (B) Diffuse leukemic cell infiltration in the mucosa, submucosa, and muscularis propria and extensive necrosis; chronic active inflammation caused functional obstruction rather than mechanical obstruction.
case of paralytic ileus without a definite mechanical obstruction due to disseminated granulocytic sarcomatous lesions along the entire length of the small bowel. This is an extremely rare condition, and only one case has previously been reported in the literature [11]. In that case, the intestinal paralysis was explained by transmural involvement of leukemia cells with extensive necrosis and fibrosis; our case showed the same pathological results. This functional obstruction appears to be similar to an intestinal pseudo-obstruction, in which an ileostomy is usually unsuccessful for intestinal decompression. We also failed to relieve the obstruction with an ileostomy and accomplished intestinal decompression with a proximal jejunostomy. An enterostomy should be considered at the most proximal intestinal region where the intestine is free of disease.

LCH is a systemic disorder that largely affects children and has an incidence of 1:200,000 to 1:350,000 [12]. Gastrointestinal manifestations of LCH are unusual and variable; they include abdominal pain, vomiting, diarrhea, and intestinal perforation [13,14], but intestinal obstruction has not yet been described. The gold standard for the diagnosis of LCH is the identification of the ultrastructural Birbeck or Langerhans cell granules. We did not observe these in our operative specimens, although extensive histiocytic infiltration and gross strictures were noted. The exact pathological diagnosis causing the obstruction could not be made. We speculate that a transmural, extensive histiocytic infiltration destroyed the intestinal musculature and led to the stricture. We recommend that, regardless of the microscopic pathology, resection should be considered to control the intestinal obstruction associated with LCH.

Intra-abdominal DSRCT is a rare malignancy mainly affecting male adolescents and young adults [15]. The usual presentation is an extensive intra-abdominal dissemination with or without visceral involvement; extra-abdominal involvement is unusual at presentation [16]. The tumor seen in this case suggested visceral involvement by serosal implantation. Long-term survival depends on the ability to remove all gross disease after chemotherapy. Our patient is currently receiving chemotherapy without evidence of extra-abdominal metastases. The prognosis and further treatment planning should be assessed after a second look operation to determine the chemotherapeutic effects on intra-abdominal dissemination.

Total resection is recommended in the presence of localized Burkitt's lymphoma. In patients with disseminated disease, chemotherapy should be the primary treatment and surgery should be limited only to tissue biopsy [17]. Acute abdominal disease due to localized ileocecal involvement has frequently been reported, and, similar to our case, complete tumor resection with simple bowel resection can offer both a diagnosis and treatment [18].

In summary, acute intestinal obstruction can present in patients with pediatric malignancies with a variety of features either as an initial presentation or during chemotherapy. Although rare, pediatric surgeons should keep such obstructive complications in mind for early diagnosis and appropriate treatment. Surgical intervention can successfully treat the obstruction, but the patient's prognosis still primarily depends on the underlying disease.
There were no conflicts of interest.

There were no sources of funding.

References


