Adult multicentric burkitt lymphoma with bowel obstruction due to intussusception

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ABSTRACT

Primary malignant tumors of the small intestine are very rare, accounting for 2%-3% of all gastrointestinal malignancies. Lymphoma constitutes about 15%-20% of all small intestine neoplasms and 20%-30% of all primary gastrointestinal lymphomas. The ileum is the most common site for gastrointestinal lymphomas. Because the symptoms and physical findings are non-specific, the preoperative diagnosis is usually difficult. In this case report, we describe the highly unusual case of sporadic Burkitt lymphoma with complete intestinal obstruction due to intussusception of the proximal jejunum and discuss the treatment options.

Keywords: *Gastrointestinal lymphoma, intestinal obstruction, malignant small intestine tumors*

INTRODUCTION

Burkitt lymphoma (BL) is an uncommon and aggressive form of non-Hodgkin's lymphoma in adults, and it represents less than 5% of adult lymphoma cases (1). There are four clinical variants of BL based on the underlying mechanism of cancer formation, namely, endemic, sporadic, immunodeficiency-associated, and a type of post-transplant lymphoproliferative disorders (PTLD) (2). Sporadic BL accounts for 1%-2% of adult lymphoma cases worldwide and usually involves lymph node formation in the abdomen (3). Extranodal involvement is also very common, usually occurring in the gastrointestinal tract, spleen, liver, kidneys, pancreas, adrenal glands, and testes (4). Clinical presentation depends on the site of involvement. Here we describe a very unusual case of sporadic BL in a 37-year-old female with complete intestinal obstruction due to intussusception of the proximal jejunum.

CASE PRESENTATION

A 37-year-old female presented to the emergency department with epigastric and right lower quadrant pain, nausea, and vomiting for 1 day. She had mild abdominal and epigastric discomfort for nearly 2 months, with negative upper endoscopic examination. She had no history of previous abdominal surgery.

Physical examination

The patient was conscious and stable. Her abdomen was distended and bowel sounds were normal. She had abdominal tenderness and rigidity on palpation. She also had abdominal rebound tenderness in her left abdominal quadrants.

Laboratory findings

The patient's laboratory results were unremarkable: hematocrit was normal, white blood cell count was 6.77×10^3/μL (Normal: 4×10^3-10×10^3/μL), C-reactive protein level was 0.41 mg/dL, creatinine level was 0.59 mg/dL (Normal: 0.7-1.2 mg/dL), urea level was 21 mg/dL (Normal: 18-55 mg/dL), and aspartate aminotransferase level was 15 U/L (Normal: 5-30 U/L).

Imaging work-up

Abdominal plain X-ray imaging showed air-fluid level in her left upper quadrant. Abdominal ultrasound revealed proximal bowel dilatation with a mural thickening of up to 9.3 mm and an intraluminal echogenic appearance (target sign) in a 10-cm-long proximal intestinal segment. A plain and contrast-enhanced abdominal computed tomography (CT) demonstrated multiple polypoid solid lesions in the jejunal segment, as well as a jejunojejunal

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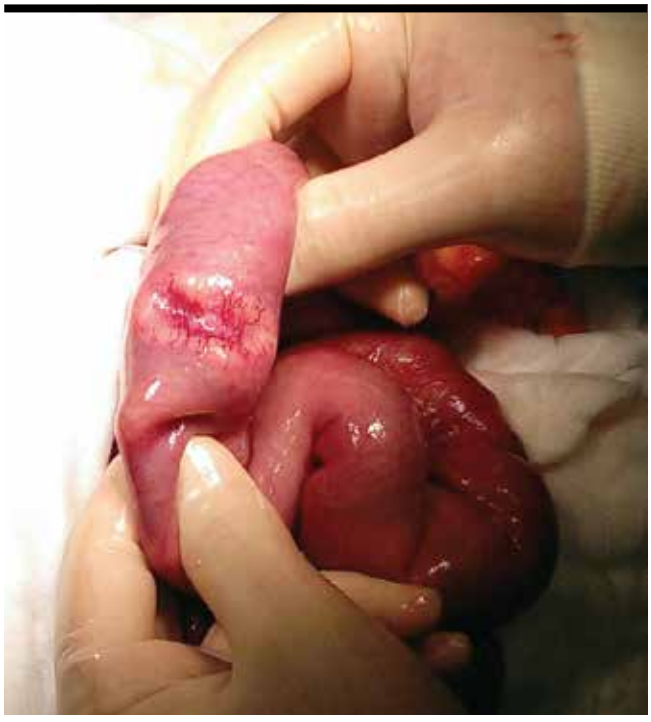

Figure 1. Invaginated/obstructed small bowel segment due to Burkitt lymphoma

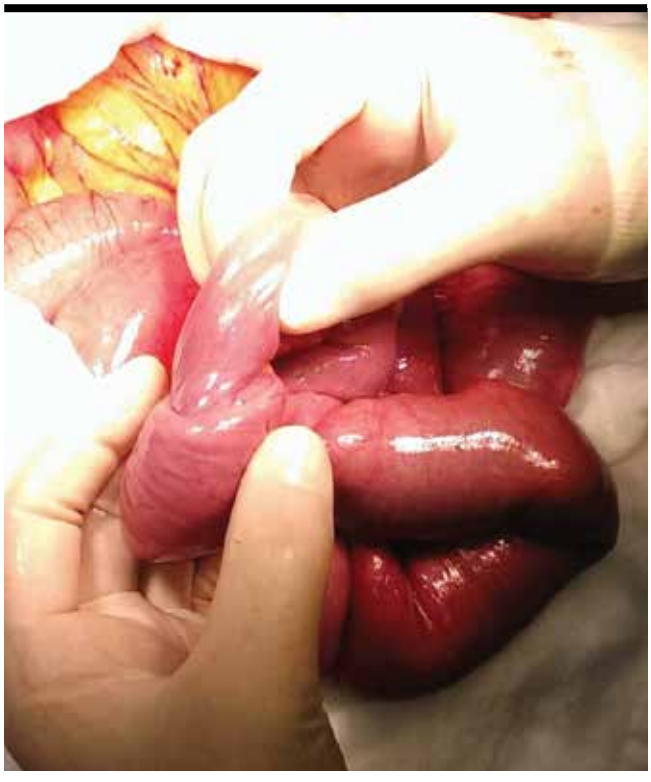

Figure 2. Envaginated small bowel segment due to Burkitt lymphoma

intussusception with a round, soft-tissue, mass appearance. The CT scan also demonstrated two other mass lesions with dimensions of 75X35 mm and 50X40 mm located at the right iliac artery and right adnexal regions, respectively. The patient also underwent a colonoscopy, which showed no pathological finding up to 50th cm of the ileum. The patient underwent an exploratory laparotomy based on the preoperative diagnosis of small bowel obstruction possibly due to intussusception secondary to small bowel tract neoplasm.

Operative findings

During exploration, multiple intramural masses (5-10 mm in diameter) were found across the small bowel surface. Proximal jejunal segments were dilated, and there was an intussusception at around the 100th cm of the jejunum from the ligament of Treitz. An intramural tumoral mass (3 cm in diameter) was found after the initial reduction of the intussusception (Figure 1,2). Segmental small bowel resection and end-to-end primary anastomosis were performed for the invaginated jejunal segment, resecting most but not all of the small bowel tumoral involvement. Since the other lesions were small, their resection was not justified.

Pathological evaluation

Biopsy specimens were carefully examined by an expert pathologist who found a diffused growth pattern of mature β-cells without nodularity. Immunohistochemical analysis revealed that β-cells expressed monotypic surface IgM, CD19, CD20, CD79a, CD10, and BCL6, with Ki-67 proliferative fraction >95%, which were diagnostic for BL.

Postoperative course

The patient's postoperative course was uneventful. After being discharged, she was referred to the oncology department and received Hyper-CVAD (cyclophosphamide, vincristine, dexamethasone, methotrexate, ARA-C) treatment protocol for six cures. Since 3 years of treatment, she has remained relapse-free and in a good condition.

The patient provided an informed consent for publication.

DISCUSSION

Intussusception of the small bowel in an adult patient is very rare and accounts for only 5% of all intussusceptions and 1%-5% of bowel obstructions (5). Intussusceptions are more common in children, with approximately 95% of the total intussusceptions cases observed in children. In

adults, the most common cause of SBO is intraabdominal adhesions, accounting for approximately 65%-75% of cases, followed by hernias, Crohn's disease, malignancies, and volvulus (6).

Here, we describe a small bowel obstruction case in an adult caused by intussusception due to an intestinal involvement of BL. Primary and secondary malignant tumors of the small intestine are very rare, constituting 2%-3% of all gastrointestinal malignancies. Lymphomas are the third most common neoplasms, accounting for 10%-15% of cases (7). The diagnosis of small intestine obstruction is relatively easy with classical obstructive signs, such as nausea, vomiting with abdominal pain, and distension and radiological signs, such as presence of air-fluid level in plain abdominal X-rays. However, determination of the exact underlying obstructive pathology is not always easy. Abdominal tomography, with a reported accuracy of up to 100%, is the most efficient examination for the diagnosis of intestinal intussusception (7). Alternatives for imaging the small intestine are ultrasonography or contrast graphics.

The clinical presentation of the small intestine lymphoma is not specific and patients have symptoms such as fatigue, malaise, colicky abdominal pain, nausea, vomiting, weight loss and rarely, acute obstructive symptoms, intussusception, or perforation. Intussusception, as a cause of bowel obstruction in adults, is a very rare condition accounting for approximately 5% of the total number of cases (8).

BL is the most aggressive form of non-Hodgkin's lymphoma (9). There are some risk factors in the pathogenesis of gastrointestinal lymphoma including *Helicobacter pylori* infection, human immunodeficiency virus (HIV) infection, celiac disease, *Campylobacter jejuni* infection, Epstein-Barr virus, Hepatitis B virus, human T-cell lymphotropic virus-1, inflammatory bowel disease, and immunosuppression (6).

There are four epidemiological variants of BL: sporadic, endemic, HIV-related, and PTLD (2). Clinically, the disease has two forms: endemic (African) and non-endemic (American). Jaw and retroperitoneal nodal involvement is seen in the endemic form, whereas abdominal mass and extra-abdominal nodal involvement is seen in the non-endemic/sporadic form. Gastrointestinal tract involvement is more common in the sporadic form (10). Although BL is a fast-growing, high-grade, β-cell neoplasm, it is highly responsive to specifically designed intensive, rotational, multiagent chemotherapy programs, combined with the anti-CD20 monoclonal antibody rituximab. When carefully applied with appropriate supportive measures, these modern programs achieve a cure rate of approximately 90% in patients with BL (9).

The type of treatment in small bowel intussusceptions of children and adults varies. Non-operative treatment is the first choice in children, whereas operative intervention is the primary treatment option for adult patients (8). In children, the etiology of intussusceptions is generally primary and benign. However, in adults, most of the cases are associated with a pathological state and a lead point. This lead point may be a tumor, a polyp, or an adhesion, and therefore, requires a definitive treatment/resection. The resection should be performed with oncology principles, which was not possible for this case as there were multiple involvements of the small bowel and iliac regions. Total removal of the involved gastrointestinal segments would ensure short bowel syndrome. Limited segmental small bowel resection was performed, resecting only the grossly involved and invaginated small bowel segment. The patient was successfully treated with postoperative multiagent chemotherapy. The response was dramatic and complete.

Treatment of lymphomas should largely be medical. Chemotherapy response rates are very high and effective for the disease. Therefore, for these patients, surgical intervention should only be used for diagnosis or relieving emergency situations, such as obstruction, perforation, or bleeding. Extensive bowel resections should be avoided (5).

In conclusion, BL is one of the rare intussusception causes in adults. It may cause multiple and diffuse bowel involvement. In this situation, we would recommend only segmental bowel resection to the invaginated segment. Other concomitant non-obstructing intact lesions may be left intact, particularly if these are small and superficial, since these will respond to chemotherapy postoperatively.

Informed Consent: Informed consent was obtained from the patient who participated in this study.

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