Primary Segmental and Multiple Adenocarcinomas of the Small Bowel

Pei-Tu Ren and Hong Fu

Abstract

Small bowel tumors are rare; most are single and located in the duodenum. When a patient presents with unreasonable abdominal pain and distension, with normal upper gastrointestinal endoscopy and colonoscopy, it is important to consider this disease. Here, we report a case of segmental and multiple adenocarcinoma of the small bowel presenting with unreasonable abdominal pain and distension in a 76-year-old woman, and provide a brief review on this subject. Our report highlights the fact that segmental and multiple small bowel tumors are very rare and the clinical characteristics are generally vague and nonspecific.

Key words: small bowel tumors, clinical characteristics, prognosis

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Introduction

Small bowel tumor is uncommon, comprising 1-2% of the gastrointestinal malignancies (1). The overall incidence of small bowel malignancies has increased in the US (2). Small bowel tumors are usually single and most located in the duodenum. Adenocarcinoma is the most common histologic subtype of primary small bowel malignancy, accounting for about 40% of all malignancies of the small bowel (1, 3) The clinical presentation of these cancers is often vague and nonspecific, which makes early diagnosis difficult (1). We herein report a case of segmental and multiple adenocarcinoma of the small bowel presenting with unreasonable abdominal pain and distension in a 76 year-old woman.

Case Report

A 76-year-old woman was admitted because of severe abdominal pain and distension. She had many episodes of abdominal pain, accompanied with abnormal bowel habits, distension and nausea, and was diagnosed as incomplete intestinal obstruction. The symptoms were relieved after management with spasmolysis, odynolysis and anti-inflammatory drugs. She had a medical history of operation associated with bowel obstruction 50 years previously, a 20-year history of rheumatoid arthritis, duodenal diverticulum and coronary heart disease, but no tuberculosis or tumors.

A physical examination revealed conspicuous emaciation, a 10 cm scar in the left middle quadrant, abdominal distension, peristalsis and shifting dullness. High-pitched, metallic tinkling sounds along with rushes and gurgles could be detected. Laboratory tests showed an enhanced level of carcinoma antigen 125 and carcinoma antigen 19-9, and a low level of albumin. However, the laboratory tests revealed that the levels of carcinoembryonic antigen (CEA) and denylate deaminase were normal and occult blood test was negative (Table 1).

Plain abdominal radiography demonstrated an intestine with increased fluid levels in the middle-lower abdomen (Fig. 1A). An air-barium contrast enema was negative (Fig. 1B). Contrast-enhanced computed tomography revealed gas-fluid levels in the lower abdomen and segmental thickness and stenosis in intestine (Fig. 1C).

A laparotomy demonstrated multiple and regional stenosis and extension in the intestine from 80 cm jejunum followed by the Treitz ligament to 130 cm ileum, adjacent to the ileocecal junction. The most severe lesion was located in the ileocecal terminal. Mesenteric lymphadenopathy and 2,000 ml ascites were found. Radical resection was performed. Pathological examination showed that the rigid and pale lesion was multiple, and its intervals varied from 5 cm to 35
cm. Tapering of the mucosal fold was found in the severe part of the lesion, while intestinal strictures caused by the broadened mucosal fold were detected in the slight part of the lesion (Fig. 2A, B). A histological examination showed a diffused and invasive signet-ring cell carcinoma, accompanied with partial mucosa necrosis (Fig. 2C). The tumor cells infiltrated the full wall of the intestine and 7/29 lymph nodes were positive. Immunohistochemistry staining revealed positivity for cytokeratin (CK, Fig. 2D), epithelial membrane antigen (EMA, Fig. 2E) and carcinoembryonic antigen (CEA). The present case was classified as T3N3M0 stage according to the UICC staging system. Chemotherapy (combination of 5-fluorouracil and platinum) was administered after the operation.

Discussion

In the present manuscript, we report a case of segmental and multiple adenocarcinoma of the small bowel, not intramural metastasis, which based on intraoperative exploration, was found to be without intraperitoneal dissemination and parenteral metastasis. Moreover, ascites probably resulted from hypoproteinemia, although cytology was not used in this patient before operation due to intestinal obstruction. Small bowel adenocarcinoma (SBA) represents the most common histologic types of small bowel tumors (4, 5). SBA often develops with vague and non-specific gastrointestinal symptoms, including obscure bleeding, abdominal pain, nausea and vomiting, weight loss, diarrhea, and intestinal obstruction. As a result of relative infrequency and vague symptoms, SBA is often diagnosed at an advanced stage, and only 50% of patients have the opportunity to undergo a curative operation (6). Therefore, it is of importance to develop a protocol for identifying the disease at an early stage.

Evidently, screening at-risk groups or patients with high-risk representations is an option for early identification of this disease (7). Chronic inflammation is one of the factors associated with the formation of several malignant tumors (8). Crohn’s disease, in particular, is considered to have an increased risk of SBA (9). The present case had a medical history of bowel obstruction. Therefore, it is possible that an incomplete intestinal obstruction may result in chronic inflammation and carcinogenesis in the small bowel. However, the causal relation remains to be further investigated. Those patients with familial adenomatous polyposis (FAP) (10), hereditary nonpolyposis colorectal cancer (HNPCC), 10 Peutz-Jeghers syndrome (11), and celiac disease (12) are also at an increased risk of developing SBA. Therefore, development of a risk-targeted strategy would be beneficial to identify SBA at the early stage.

An interesting issue from this case is how to identify the SBA from among at-risk people. SBA should be suspected in patients with persistent alarming symptoms (obscure bleeding, abdominal pain, nausea and vomiting, weight loss, diarrhea, and intestinal obstruction). Multidetector row heli-

Table 1. Laboratory Tests

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
<th>Normal value</th>
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<tbody>
<tr>
<td>Carcinoma antigen 125</td>
<td>295 U/mL</td>
<td>0-35 U/mL</td>
</tr>
<tr>
<td>Carcinoma antigen 19-9</td>
<td>44.9 U/mL</td>
<td>0-37 U/mL</td>
</tr>
<tr>
<td>Carcinoembryonic antigen</td>
<td>1.2 ng/mL</td>
<td>0-5 ng/mL</td>
</tr>
<tr>
<td>Occult blood test</td>
<td>Negative</td>
<td>Negative</td>
</tr>
<tr>
<td>Adenylate deaminase</td>
<td>12.8U/L</td>
<td>4-18U/L</td>
</tr>
<tr>
<td>Albumin</td>
<td>25.4g/L</td>
<td>30-55g/L</td>
</tr>
</tbody>
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Figure 1. (A) Intestine with fluid levels in plain abdominal radiography. (B) Negative imaging in a barium enema. (C) Thicken lesion (arrow) in the CT scan.
Juvent chemotherapy should be useful in the management of SBA patients would be improved substantially if they are diagnosed at an earlier stage and if they have a chance to undergo resection, which still offers the best chance for cure. Therefore, laparotomy is recommended for suspected patients.

Because of the rarity and variety of SBA, important questions regarding appropriate treatment recommendations and the prognostic factors remain to be resolved. Despite advances in chemotherapeutic management, the prognosis of SBA patients would be improved substantially if they are diagnosed at an earlier stage and if they have a chance to undergo resection, which still offers the best chance for cure. Laparotomy for aggressive resection and anastomosis of the involved segment with a 10 cm proximal and distal margin, including the mesentery and the lymphatics, should be performed (7). Based on the pattern of disease progression, adjuvant chemotherapy should be useful in the management of patients undergoing surgery for SBA (17). However, a detailed strategy for chemotherapy remains to be settled. Cytoreductive surgery and intraoperative, intraperitoneal chemotherapy using mitomycin C and 5-Fluorouracil could improve the prognosis of patients with locally advanced SBA, with postoperative survival of 12 to 40 months (18, 19). However, the limited data make it difficult to reach a consensus on chemotherapy for SBA. Thus, it is recommended that SBA patients be recruited to specialist centers for further management and multicenter prospective studies should be undertaken to resolve the diagnostic algorithms and develop chemotherapeutic regimens.

The authors state that they have no Conflict of Interest (COI).

References

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