Clinical Case Report

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A rare case of primary small bowel de-differentiated liposarcoma causing intussusception

A case report

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Abstract

Rationale: Liposarcoma (LPS) is a relatively rare malignant soft tissue tumor. Management of LPS including diagnosis is difficult, because it has no characteristic symptoms and no established effective treatment. Herein we reported an extremely rare case of intussusception induced by primary small bowel LPS.

Patient's concern: A-84-year-old male was a consult to our Emergency Department with symptoms of a terrible general fatigue, abdominal pain, and vomiting.

Diagnosis: Abdominal ultrasonography and computed tomography (CT) revealed probable intussusception.

Interventions: After decompression by insertion of an ileus tube, surgery was performed.

Outcomes: The ileum and mesentery of the small intestine had invaginated into the colon. There was no evidence of metastases in the intraabdominal space. The Hutchinson maneuver could not release the invagination, and so ileocecal resection with lymph node dissection was performed. Histopathological examination showed evidence of the growth of spindle-shaped cells. Also, immunohistochemical examination indicated the tumor to be a de-differentiated LPS. The patient was discharged on postoperative day 19 without any complications; and no recurrence of the tumor was observed at 16 months post operation.

Lessons: LPS should be considered in the differential diagnosis of adult intussusception, and careful management should be required, including observation, after surgery.

Abbreviations: alpha-SMA = α-smooth muscle actin, CDK4 = cyclin-dependent kinase 4, CEA = carcinoembryonic antigen, Cre = creatinine, DOG1 = discovered on gist 1, ER = estrogen receptor, GISTs = gastrointestinal stromal tumors, HMB45 = human melanin black 45, LPS = liposarcoma, MDM2 = mouse double minute2, T-bil = total bilirubin, CA19-9 = carbohydrate antigen19.

Keywords: intussusception, liposarcoma, primary, small bowel

1. Introduction

Liposarcoma (LPS) is a malignant soft tissue tumor, usually found in the retroperitoneum, extremities, or cervical area.[1] Diagnosis of LPS is relatively difficult, because it has no characteristic symptoms.[2] Hence, it is often found by the appearance of delayed symptoms, that is, pain or a palpable lump. In adults, small bowel intussusception is caused by malignant tumors (approximately 30%).[3] Indeed, some reports indicated that metastases of LPS-induced intussusception.[4,5] However, intussusception due to primary small bowel LPS is a very rare entity. Herein, we present an extremely rare case of intussusception caused by a primary small bowel LPS that was removed by surgical treatment.

2. Case report

A-84-year-old male was a consult to our Emergency Department with symptoms of terrible general fatigue, abdominal pain, and vomiting. On physical examination, abdominal pain was detected around his epigastric region, but no lump could be palpated. Routine laboratory investigation yielded increased serum-creatinine (Cre) and total bilirubin (T-bil) levels (Cre, 1.47 mg/dL; T-bil, 2.20 mg/dL). Also, tumor markers were slightly increased (CEA, 4.1 ng/mL; CA-19-9, 46.2 U/mL). Abdominal ultrasonography showed a hypoechoic mass in the colon and hyperechoic area in the ileum (Fig. 1A and B). Computed tomography revealed an intestinal obstruction caused by invagination of the ileum into the colon with the tumor (Fig. 1C).

An ileus tube was inserted approximately 2 m, and laparotomy was performed after decompression of the intestinal tract. During

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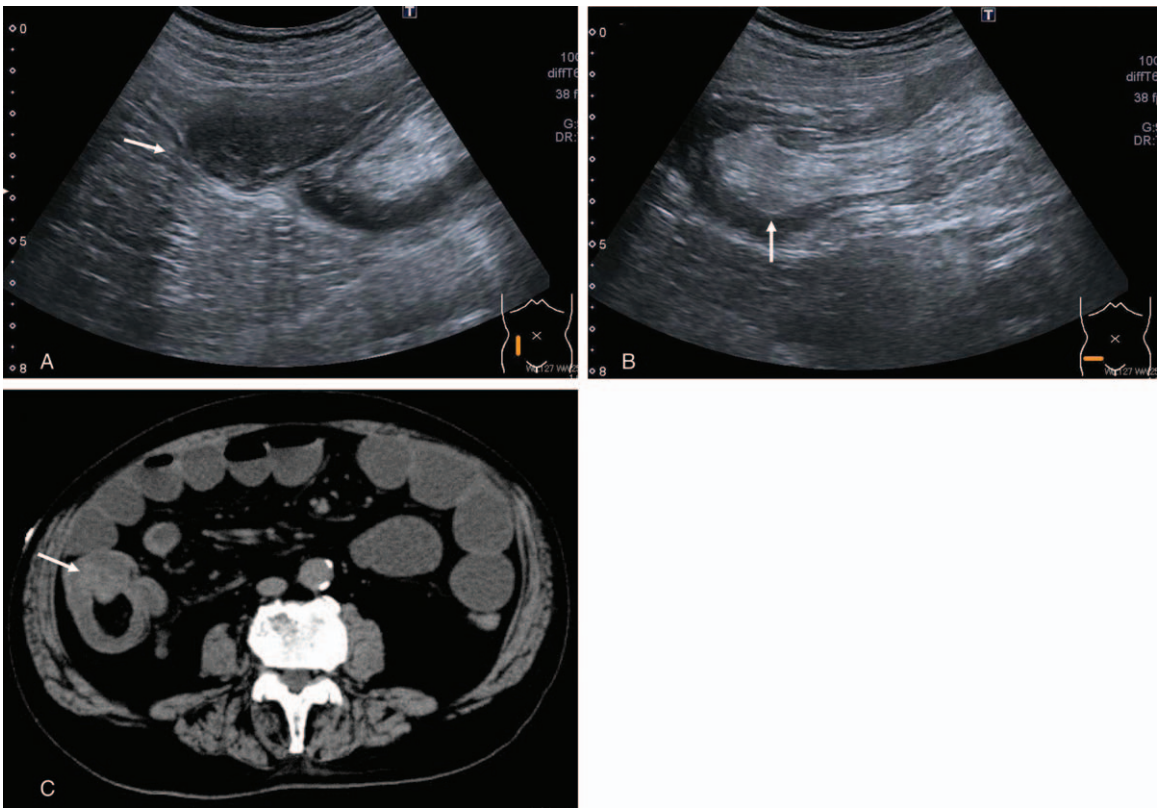

Figure 1. Highlights of preoperative images. (A) Abdominal ultrasonography image showing a 36.4 mm × 12.7 mm hypoechoic mass in the colon (white arrow). (B) That of the intussusceptional compartment (white arrow). (C) Abdominal CT image showing a continuous low-density tumorous lesion in the lumen of the colon. The white arrow indicates the tumor with ileum.

surgery, the ileum and mesentery of the small intestine were found to have invaginated into the colon (Fig. 2A). There was no evidence of metastases in the intraabdominal space. The Hutchinson maneuver could not release this invagination, and, so ileocecal resection with lymph node dissection was performed. Macroscopically, a 3-cm polypoid mass with a smooth surface was found in the ileum (Fig. 2B). Histopathological examination showed evidence of the growth of spindle-shaped cells, the size of which was shorter than that of gastrointestinal stromal tumors (GISTs) cells (Fig. 3A and B). Also, immunohistochemical examination showed a positive immunoreaction for mouse double minute2 (MDM2; Fig. 3C), but the results were negative for cyclin-dependent kinase 4 (CDK4; Fig. 3D) and for KIT, CD34, and discovered on gist 1 (DOG1), which are diagnostic GISTs markers (Supplementary Figures S1A, S1B, and S1C, http://links.lww.com/MD/C295, respectively). Also, the tumor tested negative for S100, alpha-smooth muscle actin (α-SMA), and desmin (Supplementary Figures S2A, S2B, and S2C, http://links.lww.com/MD/C295, respectively) as well as for human melanin black45 (HMB45), MelanA, estrogen receptor (ER), and

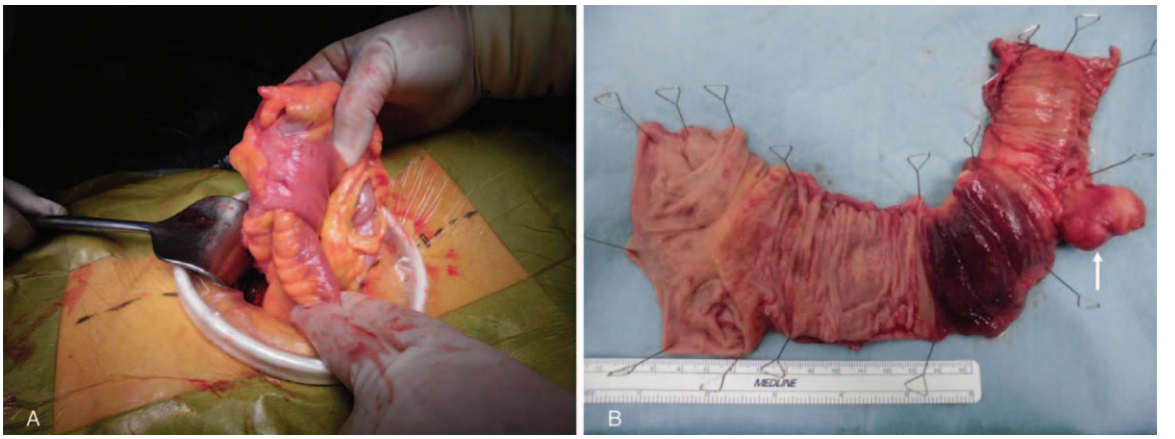

Figure 2. Highlights of intra and postoperative images. (A) The ileum and its mesentery had invaginated into the colon. (B) The polypoid tumor measured approximately 3 cm in diameter, having arisen from the ileum (white arrow).

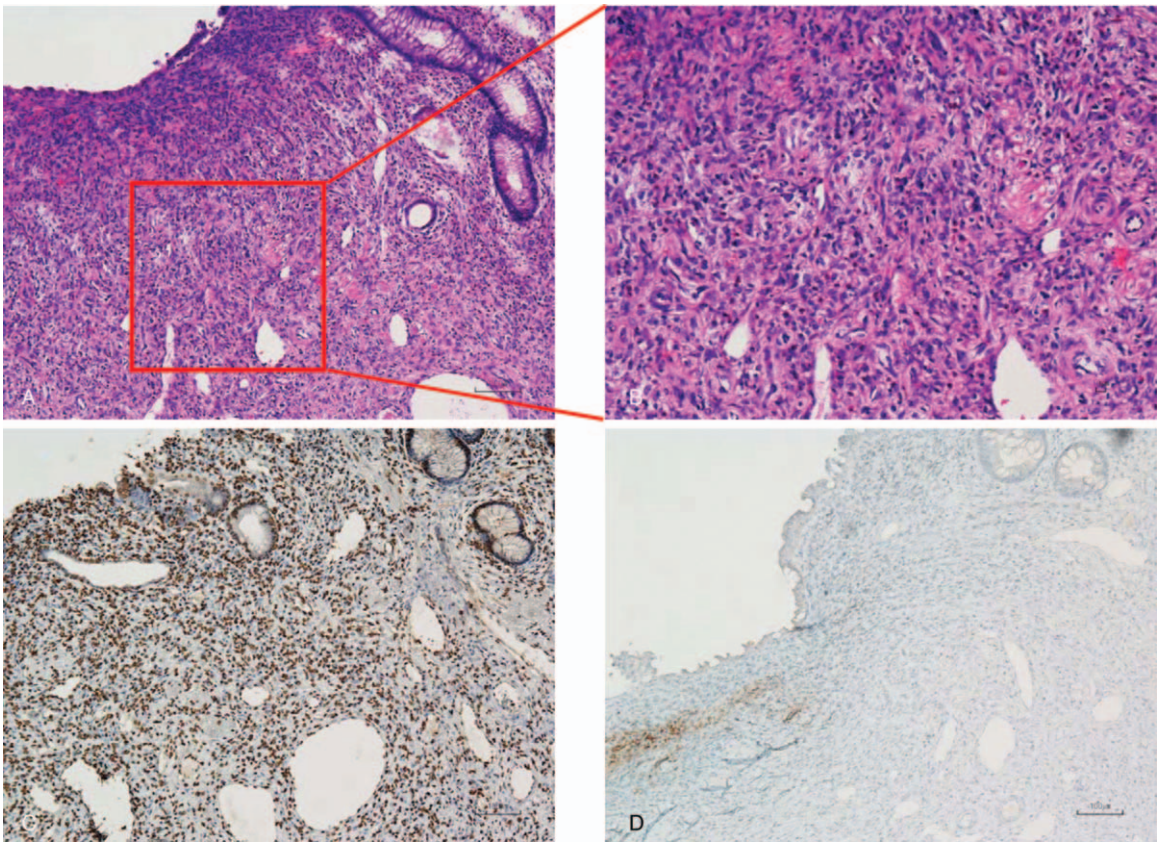

Figure 3. Highlights of the microscopic images. (A, B) Hematoxylin-eosin-stained section showing evidence of the growth of spindle-shaped tumor cells. (C, D) Immunohistochemical staining reactions for MDM2 and CDK4. MDM2 was positive; and CDK4, negative (scale bar: A, C, D 100 μm B:10 μm).

β-catenin (Supplementary Figures S3A, S3B, S3C, and S3D, http://links.lww.com/MD/C295, respectively). Based on these findings, we concluded that his tumor was a de-differentiated LPS. The patient was discharged on postoperative day 19 without any complications. Also, at 16 months after the operation, no recurrence of the tumor was observed.

3. Discussion

LPS is divided into the following 4 subtypes by the current World Health Organization Classification for soft tissue and bone tumors: atypical lipomatous tumor/well-differentiated LPS; de-differentiated LPS; myxoid LPS; and pleomorphic LPS.[1] The term "de-differentiated LPS" was first proposed by Evans in 1979 and is defined as the morphological progression from atypical lipomatous tumor/well-differentiated LPS to other subtypes of LPS.[1,6] It has been reported that de-differentiation of LPS occurs in approximately 10% of cases of well-differentiated LPS.[1,5] De-differentiated LPS is found most frequently in the retroperitoneum of adults. It also occurs in the deep soft tissues of the limbs, trunk, mediastinum, head and neck region, and spermatic cord.[1,5,6] Moreover, de-differentiated LPS is often found as a primary tumor.[1] In the present case, de-differentiated primary LPS of the ileum with intussusception was extremely unusual. To the best of our knowledge, there are only 5 case reports of LPS from small bowel [2,7–9] and only our report of intussusception caused by it (Table 1).

Various tumors of the small bowel can cause intussusception such as inflammatory fibrous polyps, lipomas, leiomyomas, metastatic lesions, leiomyosarcomas, lymphomas, carcinoid tumors, and adenocarcinomas.[3] Evidence of the growth of spindle-shaped cells allowed us to suspect GISTs (Fig. 3A and B), but our immunohistochemical examination showed that the tumor was negative for KIT, CD34, and DOG-1 (Supplementary Figures S1A, S1B, and S1C, http://links.lww.com/MD/C295, respectively) as well as for S-100, α-SMA, and Desmin denied neurinoma and myoma (Supplementary Figures S2A, S2B, and

Table 1

Case reports of LPS from small bowel.

Author, years	Age	Sex	Clinical symptom	Clinical event	Pathological classification	Treatment	Adjuvant therapy
Papadopoulos et al[9]	52	M	Abdominal discomfort, vomiting	Subtotal obstruction	Well-differentiated	Small bowel resection	None
Benaragama et al[7]	76	M	Abdominal pain	Small intestine perforation	Well-differentiated	Small bowel resection	None
Patel et al[2]	59	M	Palpable lump	A right iliac fossa mass	De-differentiated	Small bowel resection	None
Nennstiel et al[8]	60	M	Symptomatic anemia	Gastrointestinal bleeding	Pleomorphic	Small bowel resection	None
Our case (2016)	84	M	Fatigue, abdominal pain	Intussusception	De-differentiated	Ileocecal resection	None

LPS = liposarcoma

S2C, http://links.lww.com/MD/C295, respectively). Furthermore, there was no positive immunoreaction for HMB45 and MelanA denied perivascular epithelioid cell tumors (Supplementary Figures S3A and S3B, http://links.lww.com/MD/C295, respectively) or for ER and β-catenin denied mesenteric fibromatosis (Supplementary Figure S3C and S3D, http://links.lww.com/MD/C295, respectively). On the other hand, MDM2, which is a helpful diagnostic marker of atypical lipomatous tumors, well-differentiated LPS, and de-differentiated ones was positive (Fig. 3C; sensitivity, 97%; specificity, 83%).[2,10] CDK4 is also a useful marker for the diagnosis of LPS (sensitivity, 92%; specificity, 95%).[10] However, in the present case, the expression of CDK4 was negative. Nonetheless, a diagnosis of LPS was made in some cases despite the negative expression of CDK4.[10] We diagnosed this tumor as a de-differentiated LPS in light its positive expression of MDM2 and morphology, that is, unclear differentiation of adipose. Several immunohistochemical findings and the clinical context are necessary for a diagnosis of LPS.

Under existing conditions, complete surgical resection is the first choice of treatment for LPS. Effectiveness of adjuvant chemotherapy is still unclear.[11] Preoperative or postoperative radiotherapy was associated with improved overall survival compared with surgery alone for a patient with retroperitoneal sarcoma.[12] However, in this present case, the tumor arose in the ileum and exposure to radiation would be considered unwarranted to due possible ileitis. The local recurrence of de-differentiated LPS has been reported in 40%–60% of cases, with metastases in 15%–20%.[1,2] Hence, lymph node dissection should be performed as in the present case. At the present, 16 months after the operation, no recurrence of the tumor has occurred.

In conclusion, LPS should be considered in differential diagnosis of adult intussusception; and the careful management is required, including observation, after surgery.

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Author contributions

MK, KS, SY, and IM performed the patients' care including operation. KT performed pathological investigation. MK and TK designed and drafted the manuscript. LSW and UK reviewed and revised the manuscript.

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