Clinical Case Report

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Primary small bowel mesentery de-differentiated liposarcoma causing torsion with no recurrence for 5 years

A case report and review of the literature

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Abstract

Rationale: Liposarcoma (LPS) is a rare malignant soft-tissue tumor. Management of LPS is relatively difficult, because there are no characteristic symptoms, or biomarkers, nor any established effective treatment. Hence, the report of the accumulation of each LPS case is necessary. We experienced an extremely rare case of torsion caused by a primary small bowel mesentery LPS.

Patient's concern: A 70-year-old male consulted our hospital with the complaints of abdominal pain and sudden vomiting.

Diagnosis: No lump could be palpated, and tumor markers tested were within normal limits. However, computed tomography revealed an intestinal obstruction caused by torsion of the small bowel due to an LPS tumor.

Interventions: After decompression of the intestinal obstruction by use of an ileus tube, surgical treatment was performed with rapidity.

Outcome: The torsion was found to be caused by the tumor that originated from the small bowel mesentery. The tumor was resected along with a portion of the small bowel. The growth of adipose tissues of various sizes and containing atypical cells was detected by histopathological examination. Also, immunohistochemical examination resulted in positive immuno-reactions for MDM2, CDK4, and p16INK4, which indicated the tumor to be a de-differentiated LPS. The patient was discharged on postoperative day 14 without any complications, and no recurrence of the tumor was observed at 5 years after the operation.

Lessons: LPS should be considered in differential diagnosis of bowel torsion, and careful management is required because of the high possibility of recurrence. Patients should be followed carefully for at least 5 years, and further accumulation of data will be required in order to establish the appropriate management of LPS.

Abbreviations: BUN = blood urea nitrogen, CA19-9 = carbohydrate antigen19, CDK4 = cyclin-dependent kinase 4, CEA = carcinoembryonic antigen, CT = computed tomography DDLPS = de-differentiated LPS, LPS = liposarcoma, MDM2 = mouse double minute2, WDLPS = well-differentiated LPS.

Keywords: de-differentiated, liposarcoma, small bowel mesentery, torsion

1. Introduction

Liposarcoma (LPS) is a malignant soft-tissue tumor usually found in the retroperitoneum, extremities or cervical area.[1] It is often discovered in a considerably advanced state, because it has no characteristic symptoms or effective serum biomarkers. This is one of the clinical problems regarding the management of LPS. In addition, the establishment of appropriate treatment including adjuvant chemotherapy and a follow-up system are needed in order to improve the prognosis of patients with LPS. Herein, we report an extremely rare case of torsion due to a primary small bowel mesenteric de-differentiated LPS (DDLPS) that required urgent surgical treatment. The patient had no recurrence for 5 years without adjuvant chemotherapy. Also, we present a review of LPS in the literature.

2. Case report

A-70-year-old male consulted our hospital with the complaints of abdominal pain and vomiting that occurred suddenly. On physical examination, abdominal pain was detected around his epigastric region, but no lump could be palpated. Routine

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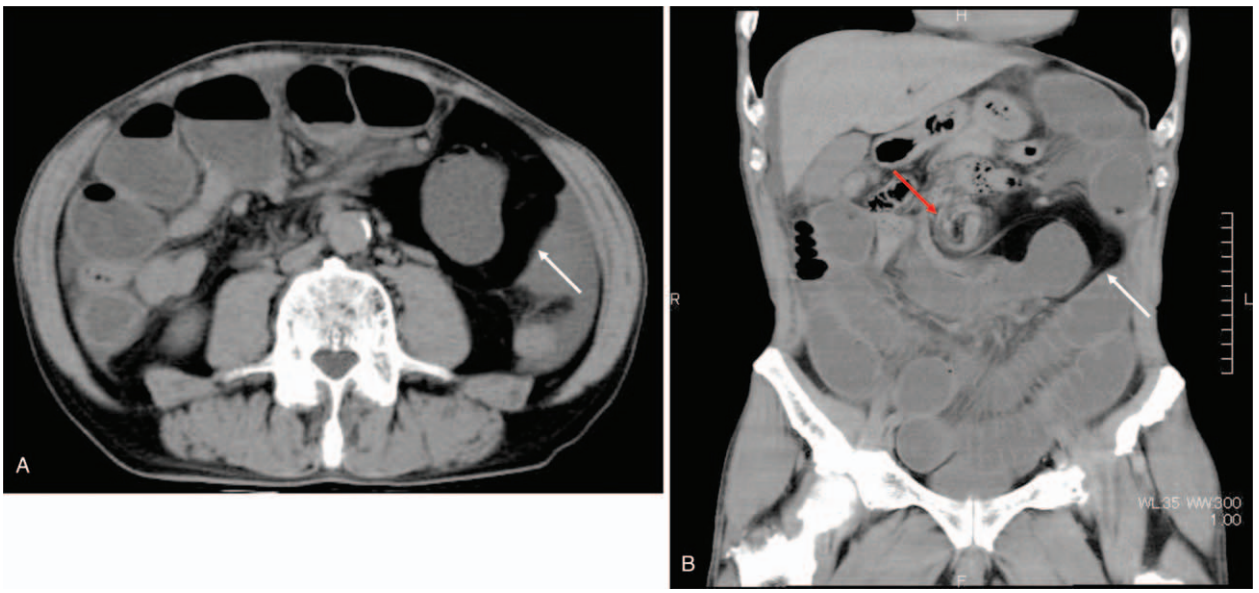

Figure 1. Highlights of preoperative images. (A) Abdominal CT image showing a low- density mass covered with a part of the small bowel (white arrow) and intestinal obstructions. (B) Coronal CT images. CT image shows a low-density mass, same as seen in "A" (white arrow). This image displays a whirl sign, indicative of torsion of the bowel mesentery (red arrow). CT = computed tomography.

laboratory investigation revealed an increased blood urea nitrogen (BUN) level (BUN: 36.3 mg/dL), suggesting dehydration. Tumor markers were normal (CEA, 1.8 ng/mL; CA-19–9, 9.6 U/mL). Computed tomography (CT) revealed an intestinal obstruction and a low-density mass covered with a part of the small bowel (Fig. 1A and B).

An ileus tube was inserted, and surgery was promptly performed after decompression of the small bowel. During surgery, torsion of the small bowel was found (Fig. 2A). After unleashing of the torsion, a tumor was found in the mesentery of the small bowel (Fig. 2B). Therefore, resection of the portion of the small bowel associated with this tumor was performed.

Macroscopically, a diverticulum was detected, and the tumor was found to have originated from the small bowel mesentery covering this diverticulum (Fig. 2C and D). Histopathological examination revealed evidence of the growth of adipose tissue of various sizes and containing atypical cells (Fig. 3A and B). Also, immunohistochemical examination showed positive immunoreactions for mouse double minute2 (MDM2; Fig. 4A), cyclin-dependent kinase 4 (CDK4; Fig. 4B), and p16INK4 (Fig. 4C). Based on these findings, we concluded that this tumor was a de-differentiated LPS (DDLPS) of the small bowel mesentery. The patient was discharged on postoperative day 14 without any complications and was followed up by both CT and ultrasound sonography every 3 months. At 5 years after the operation, no recurrence of the tumor was observed without adjuvant chemotherapy.

3. Discussion

LPS has 4 subtypes: atypical lipomatous tumor/well-differentiated LPS (WDLPS); de-differentiated LPS (DDLPS); myxoid LPS; and pleomorphic LPS according to the current World Health Organization Classification for soft-tissue and bone tumors.[1] DDLPS is an undifferentiated or pleomorphic sarcoma mixed in with a well-differentiated LPS.[1] Generally, DDLPS is detected more often as a recurrent or metastatic WDLPS rather than as a primary DDLPS.[1] DDLPS occurs in approximately 10% of WDLPS cases.[1] Primary DDLPS is relatively rare as in the present case. Several immunohistochemical examinations are helpful for diagnosing DDLPS. It was reported that the sensitivity and specificity of MDM2 and CDK4 were 97% and 92%, 83% and 95%, respectively.[2] Also, another report indicated that p16 INK4 is the most sensitive and specific marker for detecting WDLPS/DDLPS (sensitivity: 93% specificity: 92%).[3] In our case, histopathological and immunohistochemical examinations relatively clearly indicated the typical features of DDLPS (Figs. 3 and 4). Hence, we concluded that the tumor in the present case was a DDLPS that arose from the primary small bowel mesentery.

DDLPS from the small bowel mesentery and causing torsion is an extremely rare entity. To the best of our knowledge, only 15 mesenteric LPS cases had been reported (from 2000 until 2018 in PubMed); and 10 cases originated from the small bowel mesentery, and 3 of these cases were DDLPS. Only in our patient was torsion caused by it[4–16] (Table 1). Late diagnosis of LPS tends to be the case, and thus tumor growth easily progresses. Also, LPS is not fixed in the abdominal space. Thus, it is important to consider the possibility of LPS as a causation of torsion.

Another serious problem of DDLPS is that disease-related mortality of it is very poor (28%).[17] In the present case, the patient has had no recurrence for 5 years. However, in the literature, other patients had recurrence after 5 and 14 postoperative years.[14] Therefore, further careful observation is needed for our case. Complete surgical resection is the first choice of treatment for LPS. The effectiveness of adjuvant chemotherapy has remained unclear.[18] In 2 cases of DDLPS, adjuvant chemotherapy (such as doxorubicin, dacarbazine, and ifosfamide) was performed, and no recurrence occurred. On the other hand, no-recurrence cases including ours were experienced without adjuvant chemotherapy.[12,16] Radiotherapy was associated with improved overall survival compared with surgery alone for a patient with retroperitoneal sarcoma.[19] However, there is

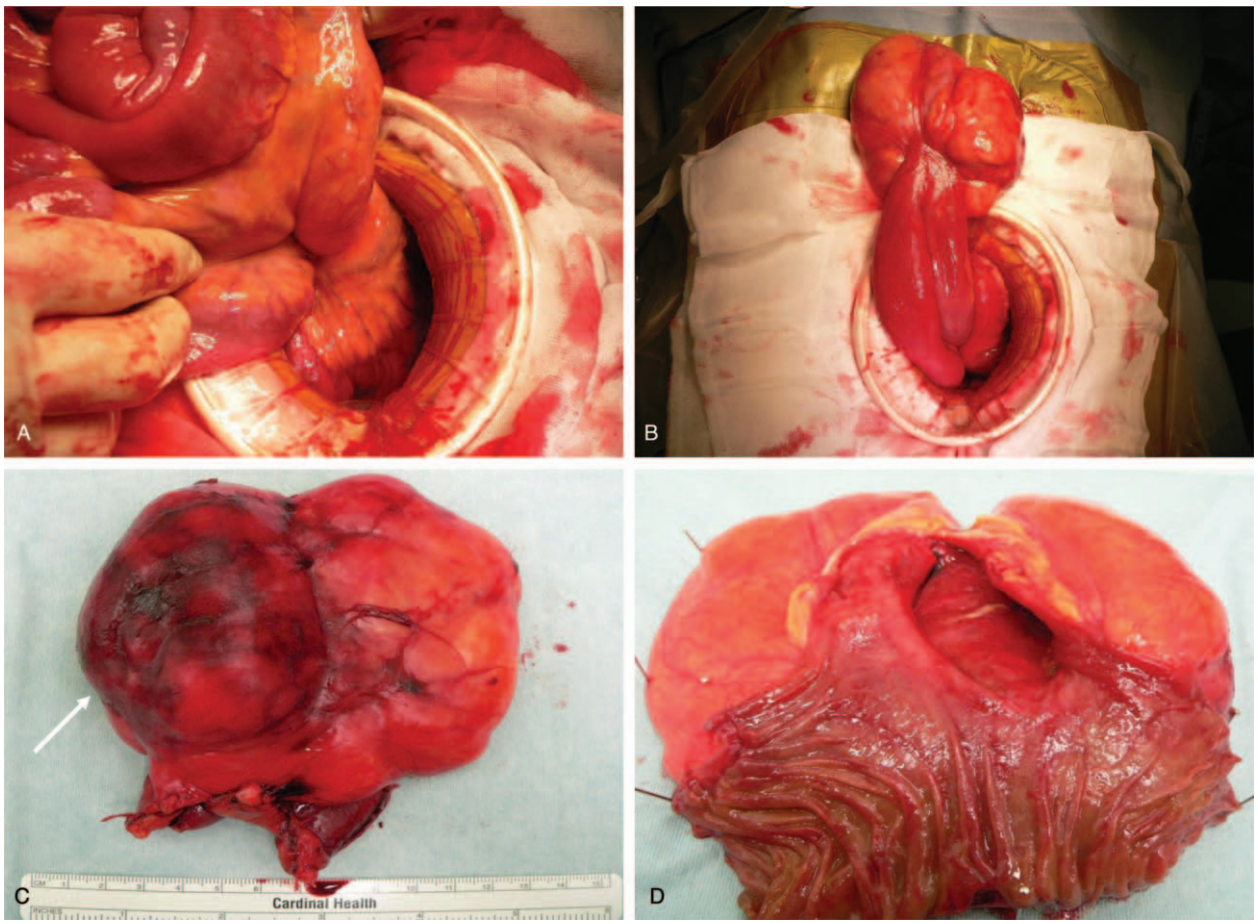

Figure 2. Highlights of intra and postoperative images. (A) Torsion of the small bowel mesentery. (B) The tumor covered with small bowel. (C) The resected tumor that originated from the small bowel mesentery (white arrow). (D) A diverticulum is seen in the lumen of the small bowel.

hesitation for performing radiotherapy for LPS originating from the bowel mesentery because of the possibility of ileitis.[20] Indeed, there is only a single reference about radiotherapy as far as we investigated.[7] These postoperative therapies should be considered depending on the patient's background such as performance status and age.

In conclusion, LPS should be considered in differential diagnosis of torsion due to a tumor, and careful management should be considered according to each patient's condition. Also, careful observation is needed at least 5 years after the operation. It is essential that further data be accumulated for the establishment of appropriate management of LPS.

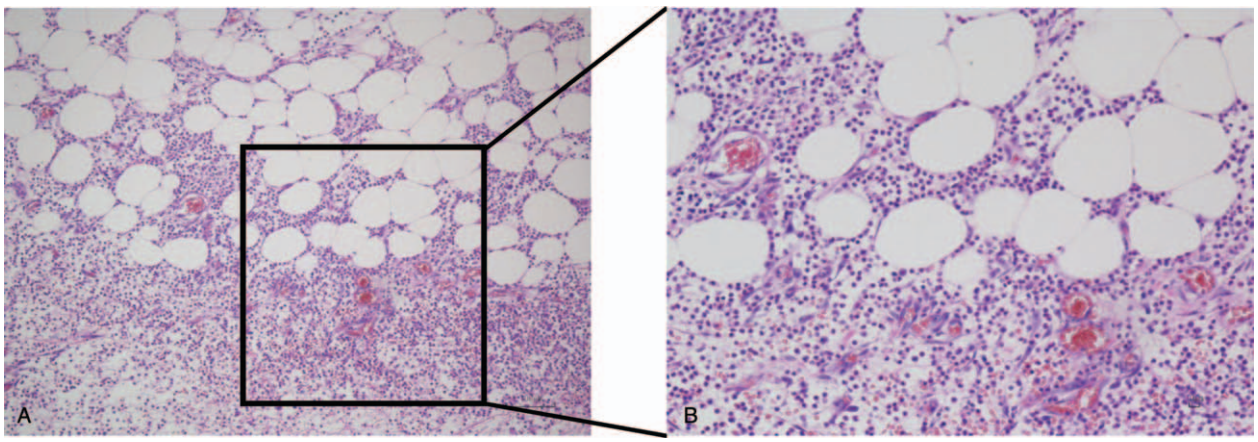

Figure 3. Highlights of the microscopic images. (A) Hematoxylin/eosin-stained section showing evidence of the growth of several adipose tissues and atypical cells in them (scale bar: 100 μm). (B) Enlarged view of boxed area in "A" (scale bar: 10 μm).

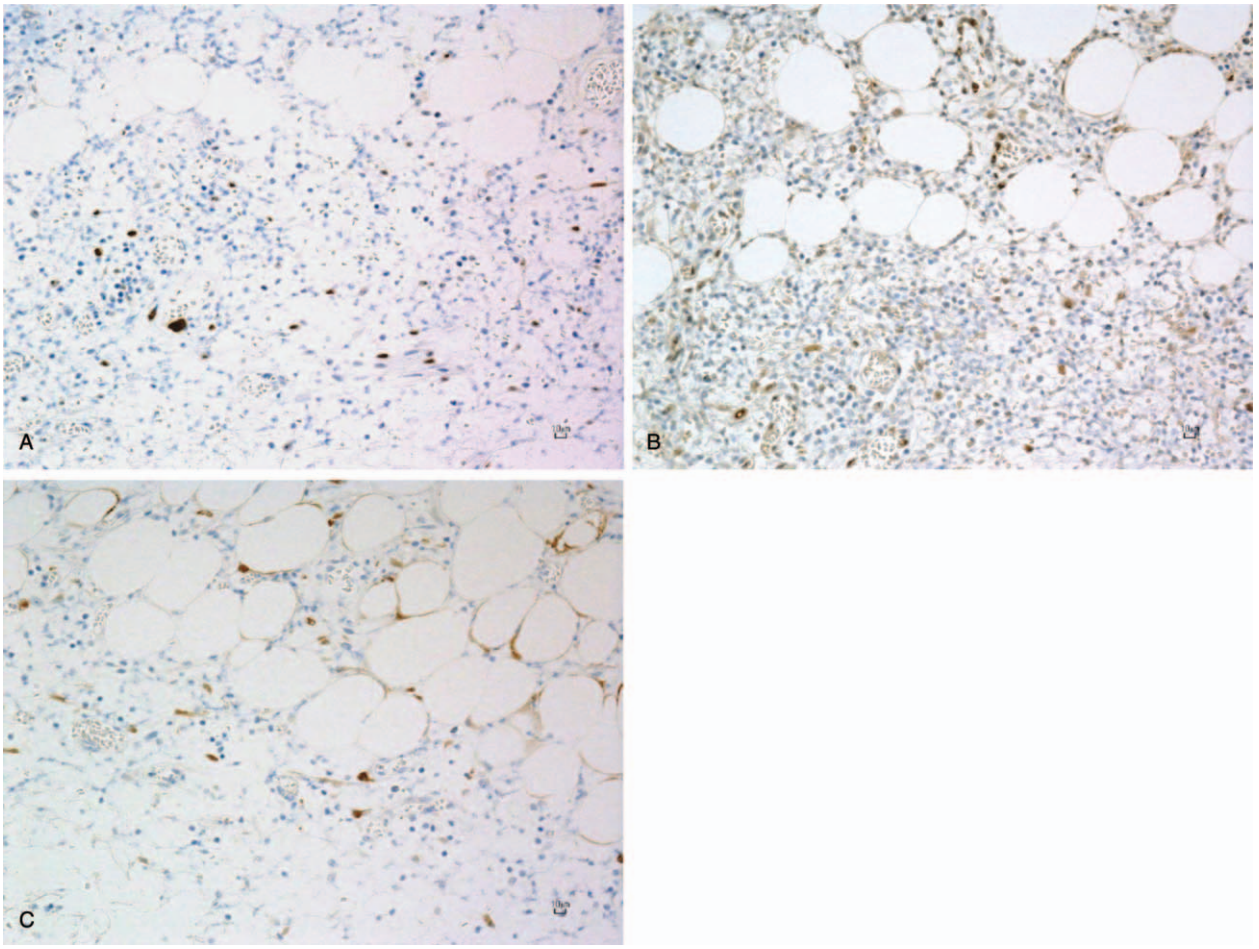

Figure 4. Highlights of the microscopic images. (A, B, C) Immunohistochemical staining reactions for MDM2 (A), CDK4 (B), and p16INK4 (C). All markers revealed positive reactions (scale bar: 10 μm). CDK4 = cyclin-dependent kinase 4, MDM2 = mouse double minute2.

Table 1

Case reports of mesenteric LPS from 2000 until 2018 in PubMed.

Author (Year)	Age	Sex	Clinical symptom	Clinical event	Location	Pathological classification	Treatment	Adjuvant therapy	Recurrence
Rosato L (2018)[14]	55	Male	Abdominal pain nausea, vomiting	Constipation	Right meso-colon	Myxoid	Resection of mass	—	Recurrence after 5 years and 14 years
Rosato L (2018)[14]	55	Male	Abdominal pain nausea, vomiting fever	None	Mesenterial intestine	Well-differen-tiated	Resection of mass	—	No recurrence for: 144 months
Vats M (2016)[16]	36	Female	Dull aching pain, Palpable lump	Constipation	Jejunal mesentery	De-differentiated	Complete excision of mass with jejunum and its mesentery	6 cycles of Doxorubicin; Dacarbazine; Ifosfamide	No Recurrence; after 6 cycles
Shen Z (2014)[15]	49	Female	Abdominal pain abdom-inal mass	None	Sigmoid mesocolon	Myxoid	Complete resection with part of the sigmoid colon	None	No recurrence for 17 months
Khan MI (2013)[10]	52	Male	Abdominal distension	None	Small gut mesentery	Well-differen-tiated	Complete resection	Eight full cycles of chemother-apy	No recurrence for 5 years
	77	Male		None		Myxoid		—	—

(*continued*)

Table 1

(continued).

Author (Year)	Age	Sex	Clinical symptom	Clinical event	Location	Pathological classification	Treatment	Adjuvant therapy	Recurrence
jukic Z (2012)[9] (abstract only)			Abdominal mass		Small bowel mesentery		Resection of mass		
Jain SK (2012)[8]	50	Male	Palpable lump	None	Mesentery of the jejunum	Pleomorphic	Resection of mass with jejunum and its mesentery	None	Recurrence after 26 months
Korukluoglu B (2009)[12]	61	Female	Abdominal distension	endocrine disorder	Synchronously bilateral mesenteric liposarcoma	De-differentiated	Resection of mass	chemotherapy	No recurrence for 12 months
cerullo G (2007)[5] (abstract only)	55	Male	Abdominal distension	None	Mesentery	Well-differentiated	Resection of mass	—	—
Khan N (2007)[11] (abstract only)	55	Male	—	—	—	Well-differentiated with mixed	—	—	—
calo PG (2007)[4] (abstract only)	43	Male	Abdominal pain	Constipation	Small bowel mesentery	Well-differentiated (atypical lipomatous tumor)	Surgical excision with a tumor-free margin	—	No recurrence for 33 months
Hirakoba M (2007)[6]	65	Female	Palpable lump	None	Mesentery of the jejunum	Well-differentiated	—	—	—
Ishiguro S (2006)[7]	30	Male	Abdominal distension	None	Mesentery of the terminal ileum and right-sided colon	Myxoid	Resection of mass with terminal ileum, right-sided colon and their mesentery and post-operative irradiation	pre-operative chemotherapy (doxorubicin, cisplatin and ifosfamide)	No recurrence for 26 months (in the present, recurrence)
Núñez Fernández MJ (2005)[13] (abstract only)	67	Female	Occasional discomfort	None	Mesentery of the jejunum	Myxoid	Complete resection	—	No recurrence for 12 months
our case	84	Male	General fatigue abdominal pain	Torsion	Mesentery of the jejunum	De-differentiated	Complete resection of mass with jejunum and its mesentery	None	No recurrence for 5 years

LPS = liposarcoma.

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Author contributions

Matsuo K, Kagota S, Shirai Y, and Inoue M performed the patient's care including the operation. Kataoka T performed pathological investigation. Matsuo K and Taniguchi K designed and drafted the manuscript. Lee SW and Uchiyama K reviewed and revised the manuscript.

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