

Case Report

An unusual case of dedifferentiated leiomyosarcoma of the primary mesentery mimicking ovarian cancer

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Received August 16, 2019; Accepted September 27, 2019; Epub November 1, 2019; Published November 15, 2019

Abstract: Dedifferentiated leiomyosarcoma of the primary mesentery is extremely rare. We report a case of dedifferentiated leiomyosarcoma (LMS) of the primary mesentery mimicking ovarian cancer. A 62-year-old woman presented with progressive low abdominal pain. Pelvic magnetic resonance imaging (MRI) revealed a large adnexal mass with carcinomatosis peritonei. Laboratory examination revealed an elevated serum level WBC 46,520/uL (Ref. 4,000~11,000/uL), PLT 687,000/uL (Ref. 140,000~400,000/uL), CA-125 69.1 U/mL (Ref. 0~35 U/mL), and beta-hCG 43.1 mIU/mL (Ref. 0~5 mIU/mL) level. The patient underwent exploratory laparotomy under suspicion of ovarian cancer. We observed a 20-25 cm-sized huge pedunculated subserosal mass arising from the mesentery, and other masses with sizes of 15-20 cm were adherent to peritoneum and ileocecal region. There was a multiple seeding metastasis in the omentum and bowel mesentery. A frozen section revealed malignancy originating from the mesentery, and thus, total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, pelvic and para-aortic lymph node dissection, and mass excision were performed. Subsequent histopathologic examination resulted in a final diagnosis of dedifferentiated leiomyosarcoma of the mesentery. The patient was transferred to a department of hemato-oncologist for additional managements. Doxorubicin was used for adjuvant chemotherapy.

Keywords: Dedifferentiated leiomyosarcoma, mesentery, mimicking ovarian cancer, leukocytosis

Introduction

The mesentery is a site where gastrointestinal malignancy is metastasized along lymphatics and lymph nodes, and here metastatic tumors are commonly found. In the mesentery, primary tumors are relatively rare, and lymphoma is the most common primary tumor [1]. In addition, gastrointestinal stromal tumors (GISTs), leiomyosarcomas, liposarcomas, fibrosarcomas, pleomorphic undifferentiated sarcomas, and hemangiopericytomas have been reported [2]. Leiomyosarcoma (LMS) represents between 10% and 20% of all newly diagnosed soft tissue sarcomas [3]. LMS is a highly aggressive tumor with a poor prognosis. A review of the literature showed that recurrence rates range from 45% to 73% and 5-year overall survival rates between 30% and 42% [4]. Dedifferentiated LMS is mostly tumors metastasized from other primary lesions, but mesenteric or sigmoid colonic LMS is most likely derived from the

smooth muscle cells of blood vessels in the mesentery and bowel muscle [4, 5]. Among these, ileum is the most common site, but transverse and sigmoid mesocolon and gastro-hepatic ligaments are also possible [6].

Here, we report an unusual case of dedifferentiated LMS of the primary mesentery mimicking ovarian cancer, describing its imaging findings in a postmenopausal woman.

Case report

A 62-year-old postmenopausal woman presented with progressive low abdominal pain of 4 days duration. The patient had no significant gynecological history and surgical history except cesarean section. She had a medical history of hypertension and dyslipidemia. She had no associated bowel complaints. On physical examination, she had an enlarged abdominal mass accompanying tenderness without re-

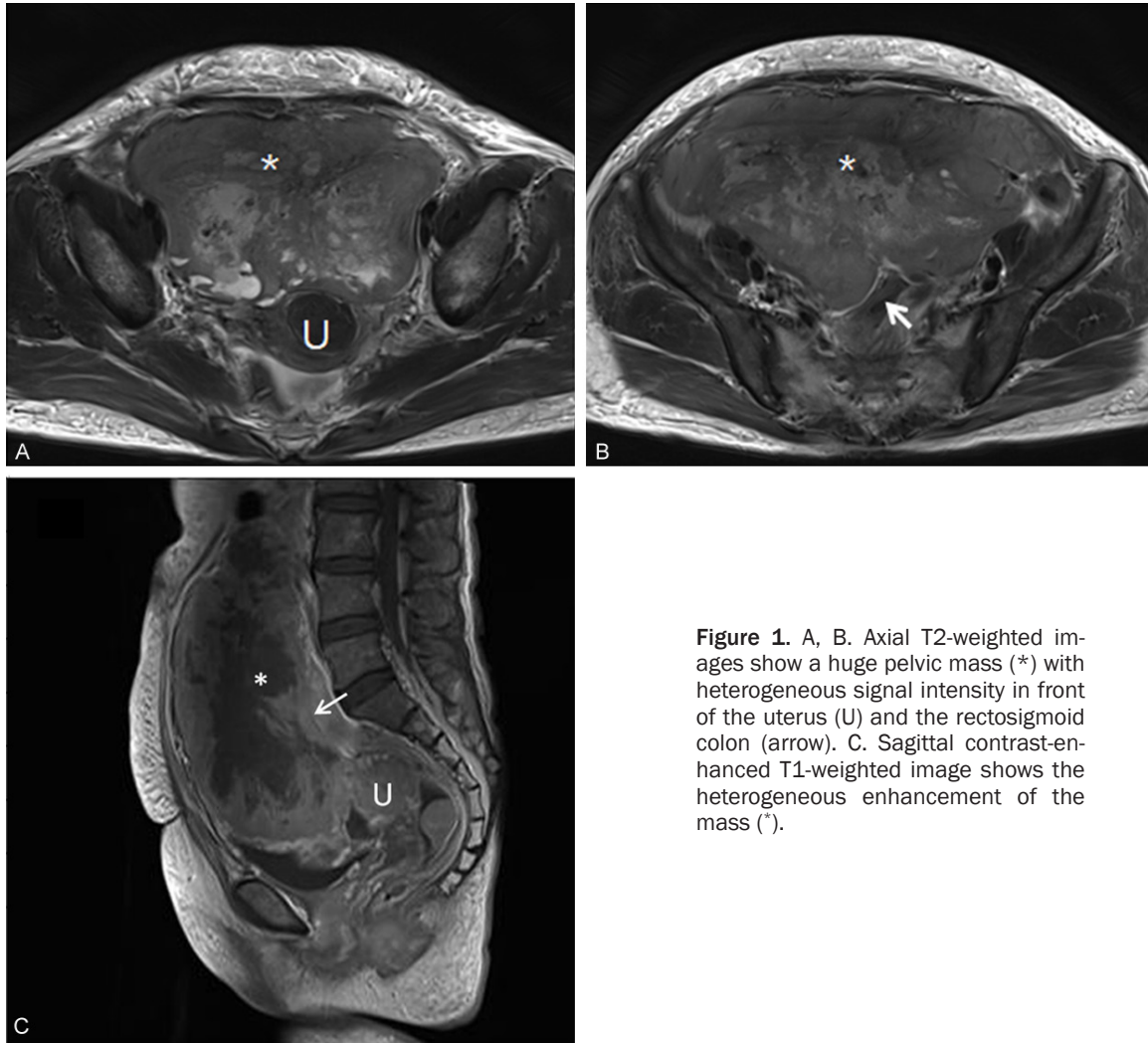


Figure 1. A, B. Axial T2-weighted images show a huge pelvic mass (*) with heterogeneous signal intensity in front of the uterus (U) and the rectosigmoid colon (arrow). C. Sagittal contrast-enhanced T1-weighted image shows the heterogeneous enhancement of the mass (*).

bound tenderness. A large and complex adnexal mass was found by subsequent pelvic ultrasonography and magnetic resonance imaging (MRI). In MRI, axial T2-weighted imaging showed a 21 cm sized solid, cystic mass with heterogeneous signal intensity in the pelvic cavity. On sagittal and axial T2-weighted images, the mass abutted to the uterus (**Figure 1**). Heterogeneous enhancing lesion in front of the uterus and the recto sigmoid colon. There was no enlarged lymph node.

Laboratory findings revealed a raised serum level WBC 46,520/uL (Ref. 4000~11000/uL), PLT 687,000/uL (Ref. 140,000~400,000/uL), LDH 392 IU/L (Ref. 135~225 IU/L), CRP 9.72 mg/dL (Ref. 0~0.5 mg/dL), CA-125 69.1 U/mL (Ref. 0~35 U/mL), and Beta-hCG 43.1 mIU/mL (Ref. 0~5 mIU/mL) level. Urine analysis results were normal. Based on radiologic imaging findings and the elevated serum tumor marker level, an ovarian malignancy was suspected.

So, the patient underwent exploratory laparotomy under suspicion of ovarian malignancy. A 20-25 cm-sized huge pedunculated subserosal mass arising from the mesentery as observed, and other masses with sizes of 15-20 cm were adherent to peritoneum and ileocecal region. There was a multiple seeding metastasis in the omentum and bowel mesentery. The right ovary showed an 8 cm-sized solid mass and the left ovary was grossly normal in size and contour. A mass with segmental descending colon resection was sent for frozen section, which revealed malignancy originating from the mesentery (**Figure 2**). Based on such findings, malignant bowel GIST was suspected. Resection of descending colon with ileocectomy and total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, appendectomy, pelvic and para-aortic lymph node dissection and excision of a multiple metastatic mass on peritoneum, and bladder wall and mesentery were performed. The omentum and mesentery

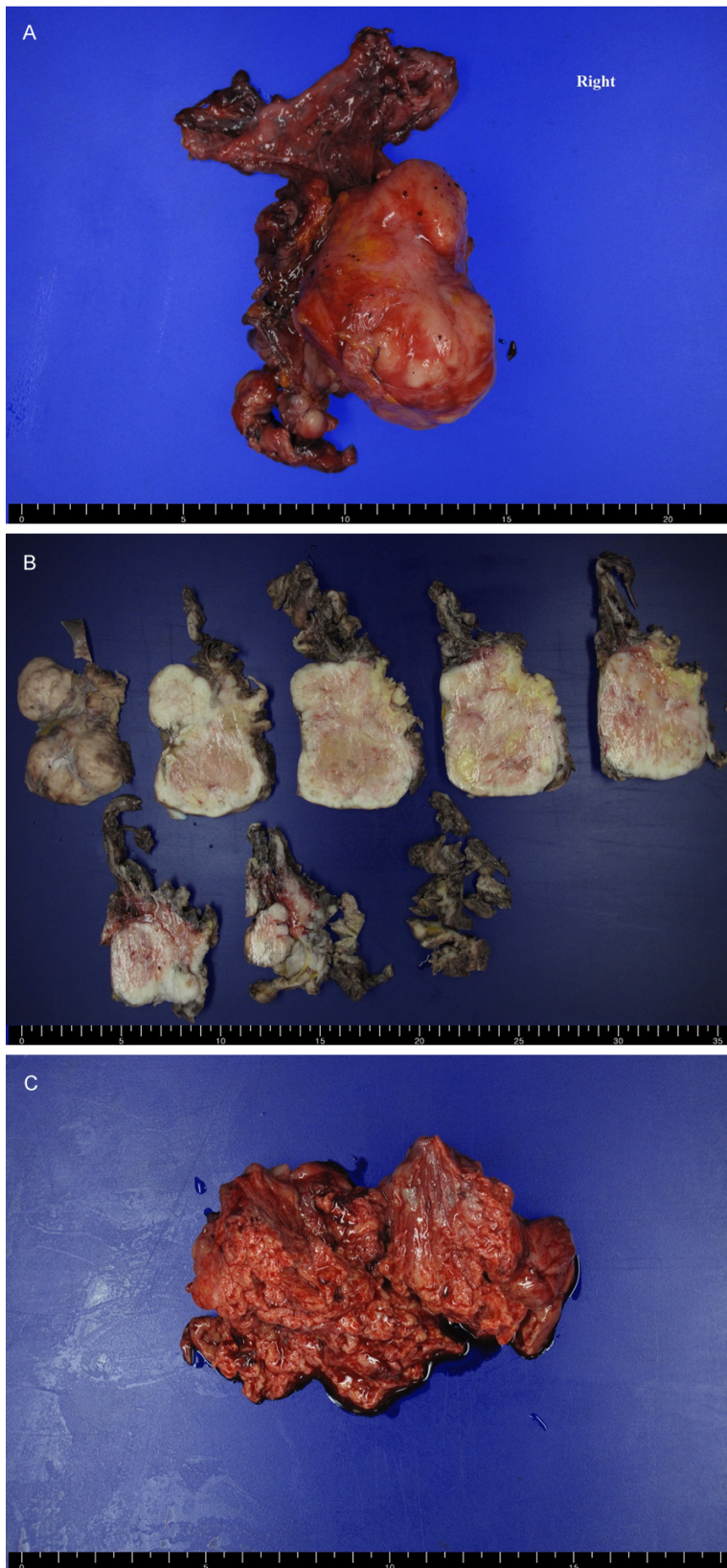


Figure 2. A. Gross appearance of patient T1-weighted image show 8 cm sized solid and cystic mass was observed. B. Cut section of right ovary and mass. C. Appearance of resected descending colon with mass removed.

around the rectosigmoid showed metastatic disease, confirming stage III disease. In our case, carcinomatosis peritonei inhibited performing R0 resection, and was only obtained for main mass at mesentery and metastatic lesion on ileocecal lesion. Pelvic and para-aortic lymph nodes were free from tumor cells, and peritoneal washings were negative for malignant cells. Excised masses showed fascicular area and pleomorphic area. The fascicular area showed positive immunoreactivity for SMA and desmin, and the pleomorphic dedifferentiated area showed reduced expression of these markers (**Figure 3**). It was inferred as dedifferentiated pleomorphic leiomyosarcoma probably originating the mesentery based on the clinical finding, although dedifferentiated leiomyosarcoma was extremely rare in this site. Afterwards, the patient was transferred to a department of hemato-oncologist for additional managements. Doxorubicin was used for adjuvant chemotherapy and CA125, WBC and serum Beta-hCG decreased to normal level at a week after surgery.

Discussion

Leiomyosarcoma of the mesentery is an extremely rare, but highly aggressive tumor and is associated with poor prognosis. The overall 5 year survival rate has been reported as only 20%~30% [4]. Yannopoulos et. al. reported the nature and behavior of the primary solid tumors of the great omentum and mesenteric leiomyosarcoma for the first time in 1963 [2]. It is an aggressive disease, and appr-

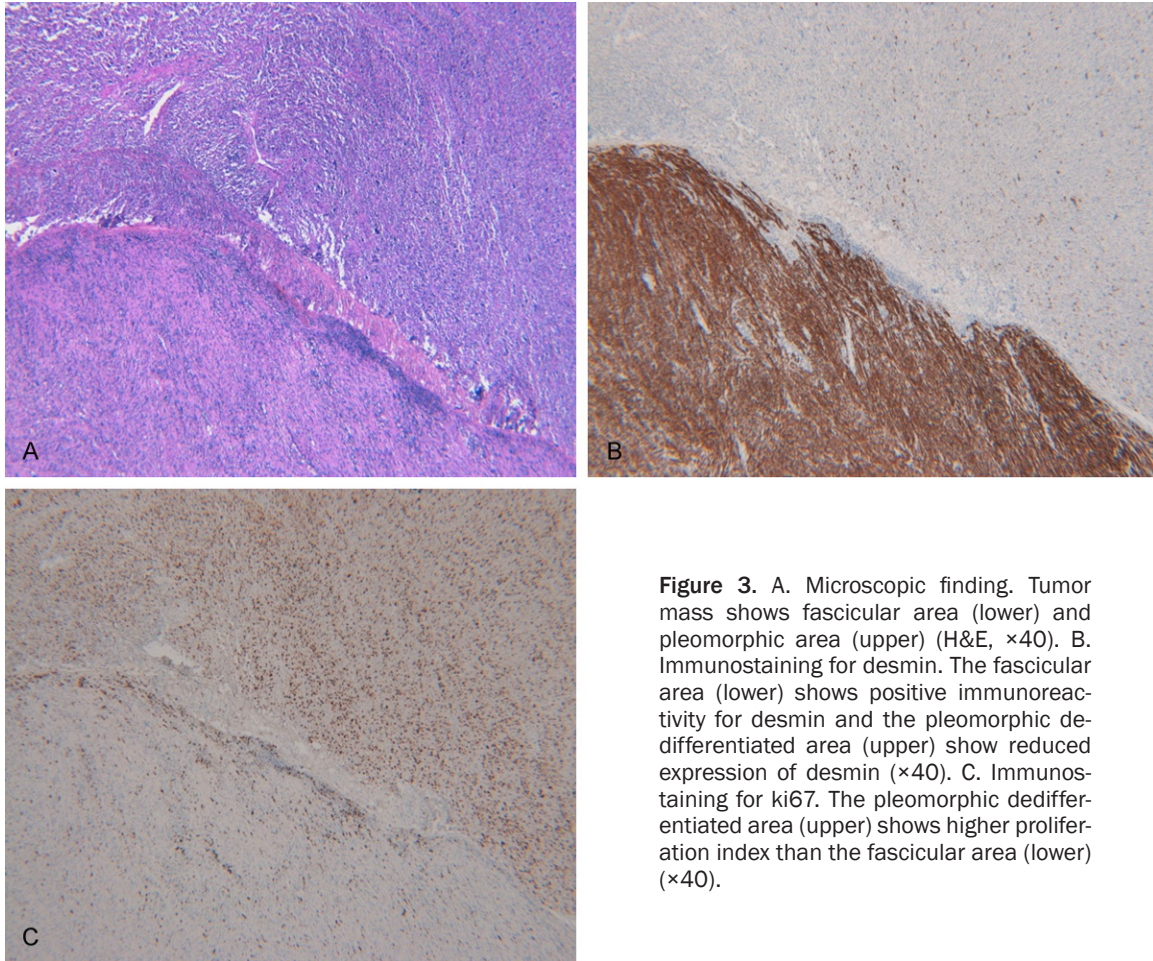


Figure 3. A. Microscopic finding. Tumor mass shows fascicular area (lower) and pleomorphic area (upper) (H&E, ×40). B. Immunostaining for desmin. The fascicular area (lower) shows positive immunoreactivity for desmin and the pleomorphic dedifferentiated area (upper) show reduced expression of desmin (×40). C. Immunostaining for ki67. The pleomorphic dedifferentiated area (upper) shows higher proliferation index than the fascicular area (lower) (×40).

oximately half of all patients are known to develop distant metastasis despite adequate local control and detection, partly because mesenteric LMS often remains undetected until late in the course of disease development [7]. Abdominal distension and discomfort are common presenting symptoms of leiomyosarcoma [3]. Serrano et. al. have reported that abdominal leiomyosarcoma might show altered bowel movements, weight loss, and non-specific blood tests, except anemia; but, the patient of our case showed leukocytosis (46,520/uL), thrombocytosis (687,000/uL) and elevated LDH, serum beta-hCG and CA-125. Leukocytosis and thrombocytosis were normalized after chemotherapy and low-molecular weight heparin (LMWH) administration. As far as we know, this is the first dedifferentiated leiomyosarcoma with leukocytosis. G-CSF is known to function as a hematopoietic growth factor to be responsible for leukocytosis. Normally, the serum G-CSF level is very low. Production of G-CSF by

tumor cells was first identified in lung carcinoma in 1977, and since then few reports of liposarcoma has been described the expression of leukocytosis and granulocyte-colony stimulating factor (G-CSF) [8]. Yet, dedifferentiated liposarcoma with leukocytosis has rarely been reported. LMS and dedifferentiated liposarcoma with leukocytosis reported is described in **Table 1**. It would have been better to measure the serum G-CSF level to clarify whether the malignant mass secreted G-CSF. If G-CSF secreting cancer was present, serum G-CSF measurement could have been used as a marker of cancer recurrence in such patients.

Yannopoulos et. al. have also reported for the first time that whether benign or malignant, most of the tumors in the mesentery tend to grow to a large size enough to be palpable when first examined before it causes any symptoms [2]. Abdominal distension and related discomfort are the most common presenting symp-

Table 1. Leiomyosarcoma and dedifferentiated liposarcoma with leukocytosis reported was described as follow

Type	WBC	Report
Leiomyosarcoma	22,900/uL	Leiomyosarcoma in the humerus [18].
	19,100/uL	Recurrent uterine leiomyosarcoma [19].
Dedifferentiated liposarcoma	33,000/uL	Dedifferentiated retroperitoneal liposarcoma [20].
	103,000/uL	Dedifferentiated liposarcoma in upper arm [21].

toms in many previous reports [3, 9]. However, in our case, the patient had no complaint of abdominal distension but of low abdominal pain.

MRI is a very useful radiologic method for determining anatomic features of pelvic masses. In addition, an MRI may give diagnostic information that supports a preoperative postulation of leiomyosarcoma; however, it may not be entirely accurate. Leiomyosarcoma usually manifests as a large infiltrating myometrial mass with high signal intensity on T2W images. Final diagnosis of leiomyosarcoma is concluded by histopathological findings. In our case, an initial diagnosis of ovarian cancer was made, because ovarian cancer is more common in postmenopausal women and MRI presented a large, pelvic mass, omental smudging. Huge solid pelvic masses may have obscure origins and be misinterpreted as ovarian masses. When ovaries are visualized separately, a mass of ovarian origin can be excluded. In our case, we retrospectively analyzed MRI images and noted no bridging vessels from the mass to the bowel, mesentery, and ovary. Several case reports have been issued on considered preoperatively to be ovarian tumors, due to similar locations and imaging findings. For example, a large, solid pelvic and abdominal mass with degeneration was initially misdiagnosed as ovarian cancer due to the preoperative radiologic findings but later found out to be leiomyosarcoma of the rectum [10-12]. However, immunohistochemistry may be helpful for differentiating leiomyosarcoma from other tumors. In our case, the right ovarian mass showed fascicular area and pleomorphic area. The fascicular area showed positive immunoreactivity for SMA and desmin. The pleomorphic dedifferentiated area showed reduced expression of these markers. It is dedifferentiated pleomorphic leiomyosarcoma probably originating the mesentery based on the clinicopathological findings although dedifferentiated leiomyosarcoma is extremely rare in this site [13]. Complete surgical resection is

the cornerstone and recommended for treatment for LMS, but complete en-bloc resection is often difficult due to the large size and friable tumor and early seeding and disseminated metastasis to multiple organs and tissue [14, 15]. Cancer frequently invades the bladder wall and pelvic wall, and prognosis primarily depends on complete en-bloc resection, tumor size, histologic grade and the extent of disease at diagnosis and mitotic index. Several authors have suggested that tumor size may be an important prognostic factor [16, 17]. Although the role of adjuvant chemotherapy is unclear, for high-risk patients, after multidisciplinary assessment adjuvant treatment or clinical trials might be applied. The patient in the current study also underwent single doxorubicin treatment as an adjuvant treatment.

Conclusion

In summary, we presented an unusual case of dedifferentiated leiomyosarcoma of the mesentery that mimicked ovarian malignancy, based on anatomic location, imaging findings and serum tumor marker level in a postmenopausal woman.

This case cautions about the difficulty of diagnosing solid pelvic and abdominal mass abutting ovaries with nonspecific several elevated serum tumor markers. Such cases of leiomyosarcoma mimicking ovarian cancer might be considered for the differential diagnosis of a pelvic mass in postmenopausal women.

Acknowledgements

This work was supported by a 2-Year Research Grant of Pusan National University.

Disclosure of conflict of interest

None.

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