Case report

Surgery of intraabdominal giant dedifferentiated liposarcoma of ascending colon mesentery: A rare case report

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ABSTRACT

Introduction: Liposarcoma, a lipogenic tumor of large deep-seated connective tissue space, presents the most common type of soft tissue sarcoma arising in the retroperitoneum. Liposarcoma that arises from colonic mesentery is especially a very rare disease. The present case describes a surgery of giant dedifferentiated liposarcoma at ascending colon mesentery.

Presentation of case: A 47-year-old South Korean man was admitted and presented with palpable abdominal mass. Abdominal pelvic computed tomography scan revealed a huge mass at his right sided abdomen (about 25 × 19 cm sized mass at right abdomen with encapsulation). After the surgery, the entire mass was completely excised en bloc with the ascending colon. The specimen consisted of multinodular, pinkish tanned, focally myxoid tissue, which measured up to 25.5 × 19 × 12.5 cm. Final pathological analysis reported dedifferentiated liposarcoma (high grade sarcoma) with MDM2 and CDK2 (+) in immunohistochemistry.

Conclusion: The present case report concerns a 47-year-old male with giant dedifferentiated liposarcoma arising from colonic mesentery and achieved en-bloc resection of liposarcoma with right hemicolectomy.

1. Introduction

Intra-abdominal soft tissue sarcoma is a rare kind of tumor, which accounts for less 1 % of all malignant tumors. Retroperitoneum is the primary site in about 15 % of soft tissue sarcomas [1,2]. Of all the kinds, liposarcoma presents the most common type of arising soft tissue sarcomas, taking up about 45 % [3] and about 41 % at lower extremities, and about 11 % chiefly in thighs [4]. Liposarcoma is a large lipogenic tumor deeply seated in connective tissue space. Retroperitoneal liposarcoma alone comprises 0.07–0.2 % of all neoplasms [5]. Liposarcomas arising from the mesentery are extremely rare and also scarce to report. Liposarcomas have five histologic subtypes according to embryogenic mesodermal origin: 1) well-differentiated liposarcoma, 2) dedifferentiated liposarcoma, 3) myxoid, 4) round cell and 5) pleomorphic liposarcoma [3,6]. Dedifferentiated liposarcoma, round cell and pleomorphic liposarcoma are aggressive high-grade, tumors with metastatic potential, while well-differentiated liposarcoma and myxoid liposarcoma are low-grade tumors that follow a more indolent clinical course [7]. Dedifferentiated liposarcoma is an uncommon subtype of liposarcoma, with poor prognosis. The present case reports a surgery of giant dedifferentiated liposarcoma that arose from the mesentery of ascending colon. This case report has been reported in line with the SCARE criteria [8].

2. Case report

A 47-year-old South Korean man was admitted and presented with palpable abdominal mass. The patient had no specific medical and surgical history. He had light diffuse abdominal pain with huge palpable mass and had abdominal distension with no other symptom. The physical examination revealed neither tenderness nor rebound tenderness, nontender intra-abdominal mass measuring around 18 × 18 cm in size with well ill-defined margins at right sided abdomen. The vital sign of patient was stable (blood pressure 140/90 mmHg, heart rate 89 bpm), body temperature was 36.2 °C, and BMI was 21.3 kg/m². The patient's bowel function and defecation was normal. Laboratory testing revealed a white blood cell count of 8070 cells/mm³, and hemoglobin count of 12.5 g/dL. The other lab finding was not reported specific finding. Carcinoembryonic antigen was normal (3.53 ng/mL) and alpha fetoprotein was also normal (4.6 ng/mL). Abdominal pelvic computed...
tomography (CT) scan revealed a huge, heterogeneous mass at retroperitoneum (about 18 × 18 cm sized mass at right abdomen with encapsulation) (Fig. 1). The mass was displaced at lower pole of right kidney in right mid/lower quadrant area. Most of the intestine was weighted to the left-side of abdomen (Fig. 1). The surgery was decided because abdominal discomfort persisted due to the huge mass and intestinal obstruction was expected in the future. And malignant mass was also not excluded. The surgery for excision of the mass was performed with diagnostic and therapeutic tools. During the surgery, the huge mass was verified to be conjugated with ascending colon mesentery, and the mass looked like it was closed by the intestine wall. Thus, hemicolectomy was performed to remove the entire mass with adequate margin. The specimen was a well-demarcated ovoid solid mass adhesive to the serosal surface of colon, measuring up to 25.5 × 19.0 × 12.5 cm in size (Fig. 2). The cut surface was multinodular, pinkish, soft and fleshy, including focally myxoid and hemorrhagic portion on one part, and focally firm and fibrotic portion on the other. Resection margin was not involved microscopically. Final diagnosis was dedifferentiated liposarcoma involving serosa and outer layer of proper colonic muscle with features of high-grade sarcoma and variable cellularity. The result of immunohistochemistry was positive to MDM2, CDK4 and c-kit and negative to DOG1, desmin, actin, S100 and CD34. We also conducted the MDM2 gene amplification test for diagnosis and it had positive finding with 6.0 of MDM2/CEP1 ratio and average 19 copies of MDM2 gene (Fig. 3). After surgery, the patient recovered well and discharged on postoperative day 10. In multidisciplinary discussion, the adjuvant chemotherapy was decided as docetaxel, ifosfamide, and mesna (AIM) regimen for 6 cycles every 3 weeks. During 21 months of follow-up period, no recurrence has occurred at abdominal pelvis according to the trimonthly computed tomography examination.

3. Discussion

Dedifferentiated liposarcoma (DDL) is one of the high-grade sarcomas and has a wide morphologic spectrum. It can usually include various components which are non-lipogenic high-grade sarcoma (such as undifferentiated pleomorphic sarcoma or spindle cell sarcoma) and lipogenic low-grade sarcoma (such as atypical lipomatous tumor or well-differentiated liposarcoma) with abrupt transition between non-lipogenic and lipogenic lesion [9]. Dedifferentiated liposarcoma is classified as primary DDL, which is discovered at the time of diagnosis alike the current case, and secondary DDL, which lost certain differentiation from previously diagnosed well-differentiated liposarcoma in the process of recurrence [10]. Primary DDL arises from the soft tissue incidentally and accounts for 57 % of liposarcoma in the retroperitoneum [11]. The ratio of primary and secondary DDL is about 9:1 [12].

DDL tends to occur at older age (peak incidence between sixties and eighties) and occurs similarly in both genders mainly within the retroperitoneum or extremities [13]. Clinical symptoms of intra-abdominal liposarcoma are mainly painless palpable masses, which are presented with inherent characteristics in relation to deep localization and expansive growth [14]. Clinically, the tumors tend to present with diffuse abdominal pain accompanied by anorexia, weight loss and an increase in abdominal girth. Most symptoms develop via displacement of nerves and vessels, or compression of adjacent organs and structures [14]. Furthermore, when the size of the tumor grows similar to that of the adjacent organs, other symptoms such as intestinal or urinary tract obstruction can occur. These tumors may grow to a large size without any symptoms, and about 20 % of the tumors are >10 cm in diameter at the time of diagnosis [15]. The tumor size of the present case was also very large (25.5 × 19 × 12.5 cm), and the patient had complained recently regarding palpable abdominal mass and abdominal discomfort.

The oncologic behavior of DDL has aggressive growth locally. About 40 % of DDL shows local recurrence, and 80 % of DDL’s local recurrence of occurs within the five years after complete resection [16]. Metastasis of DDL is within the primary site of tumor or extent to the distant organs such as liver, lung and brain or other soft tissue [17]. The rate of metastasis in DDL is 15–30 %. 5-year overall survival is 57 % for low grade DDL, and 21 % for high grade DDL. Tumor grading, subtype, complete resection with surgery, metastasis and tumor size are associated with the prognosis for liposarcoma [18]. Studies have shown the worse prognostic factor of DDL to be the location of tumor, especially when the tumor is in the retroperitoneum [10]. DDL has better survival rate than pleomorphic liposarcoma and worse survival rate than well-differentiated liposarcoma [16,19]. The complete surgical resection of

![Fig. 1. Abdominal pelvic CT scan – About 25.5 × 19 cm sized mass at right abdomen with encapsulation, A) sagittal view, arterial phase, B) sagittal view, portal phase, C) Coronal view.](image-url)
tumor is the only curative treatment for DDL. There is an issue of oncologic superiority of R0 resection for retroperitoneal DDL. Generally, for DDL, macroscopic complete excision (R0, R1) is known as the better prognostic factor, increasing the rate of overall survival than R2 resection [20]. Moreover, multivisceral excision was also studied for oncologic outcomes of DDL. A retrospective study compared between excision of primary sarcoma and surgery extent to the surrounding organs such as kidney, colon, and pancreas. Extended resection with tumor and adjacent organ had lower rates of local recurrence and had improvements in overall survival [21]. In the present case, the tumor is assumed to have originated from colon mesentery or retroperitoneum, and grossly invaded to colon wall. The patient underwent right hemicolectomy to achieve adequate margin and extended resection during the surgery of current case. On the other hand, the extended compartmental resection including other organs has a higher postoperative morbidity and mortality with poorer outcomes for liposarcoma [22].

In this case before the surgery, it was thought to be difficult to achieve complete resection of tumor because of the giant size and multivisceral invasion of the tumor at the time of diagnosis. Radiation and systemic chemotherapy could be chosen as selective modalities in neoadjuvant and adjuvant setting to reduce local recurrence rates. Neoadjuvant radiation therapy is avoided for retroperitoneal DDL due to radiation enteritis (60 % of the patients given radiation therapy) [23]. Also, systemic chemotherapy for DDL has been reported to have limited benefits with <12 % of response rate [24]. However, when the primary tumor is advanced, borderline resectable, or near the main vessel or organ, systemic therapy can be considered [25]. In the present case, although the surgery was successfully performed with grossly complete resection, residual tumor was suspicious on postoperative abdominopelvic computed tomography (APCT) on the right paracolic gutter. With the multidisciplinary discussion, 6 cycles of combination chemotherapy with doxorubicin, ifosfamide, and mesna (AIM) regimen was

Fig. 2. Surgery of Giant dedifferentiated liposarcoma of ascending colon mesentery: A) Anterior part view, B) posterior part view.

Fig. 3. Histopathologic features of dedifferentiated liposarcoma. (A) The well-defined lobulating tumor involves proper muscle layer and serosa. (B) The cut surface of tumor shows yellowish tan color and soft and fleshy texture. In low power view, the tumor presents short fascicles of spindle cells (×100). (C) In high power view, the tumor cells are mainly spindle cells with various size and multifocal bizarre nuclei are observed (×400). (D) MDM2 gene (green) amplification is confirmed by fluorescence in situ hybridization. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)
decided in patient as adjuvant therapy. It is uncertain whether the chemotherapy is favorably affected to the patient, as there has been no progression or recurrence of the tumor for 21 months so far.

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Ethical approval
This manuscript is a case report retrospectively and also is not a clinical study. The ethical approval is not necessary.

Consent
The patients have provided written informed consent for publication of the case.

Author contribution
Nahyeon Park contributed to writing the paper and data collection. Dae Ro Lim contributed to conceptualization and writing the paper. Jung Cheol Kuk contributed to data collection. Eung Jin Shin contributed to supervision and reviewing the paper.

Registration of research studies
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