An unusual case of primary colonic dedifferentiated liposarcoma

Mehmet Akit Türkoğlu a,*, Gülsum Özlem Elpek b, Volkan Doğru a, Hasan Çalış c, Ashlı Uçar b, Cumhur Arıcı a

a Department of General Surgery, Akdeniz University School of Medicine, Antalya, Turkey
b Department of Pathology, Akdeniz University School of Medicine, Antalya, Turkey
c Department of General Surgery, Antalya Training and Research Hospital, Antalya, Turkey

ARTICLE INFO

Article history:
Received 20 August 2013
Received in revised form 25 October 2013
Accepted 28 October 2013
Available online 14 November 2013

Keywords:
Colonic dedifferentiated liposarcoma
Intraabdominal mass
Dedifferentiation

ABSTRACT

INTRODUCTION: In this paper, we present a rare case of primary dedifferentiated liposarcoma (DDLS) of the colon, management of which is unclear and difficult to cope with.

PRESENTATION OF CASE: 71 year old female patient with complaints of abdominal pain and swelling was referred to our clinic with the diagnosis of intraabdominal mass. 23 cm × 19 cm × 18 cm tumor starting from the neighborhood of left liver lobe and extending toward pelvic floor was detected on computed tomography. At laparotomy, a multilobulated, soft and yellowish mass was arising from transvers colon and invading greater curvature of stomach. En-bloc removal of the tumor including segmental colon and gastric wedge resection was performed. Postoperative histopathological diagnosis was consistent with dedifferentiated liposarcoma.

DISCUSSION: Liposarcomas are rarely encountered in the gastrointestinal tract. Previously, only ten cases of primary liposarcoma of the colon have been reported worldwide and to our knowledge DDLS of transverse colon is the first case reported in the literature. DDLS is a high-grade aggressive tumor carrying the ability to metastasize. Despite complete removal of tumor recurrence is common in DDLS.

CONCLUSION: The constellation of findings in our patient demonstrates that liposarcomas which histologically exhibit dedifferentiation are associated with a poor clinical prognosis and advocating surgery alone is not recommended.

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1. Introduction

Liposarcoma is one of the most common types of soft tissue tumors in adults. The incidence of liposarcoma peaks between 40 and 60 years.1 Liposarcomas are usually reported in the extremities and retroperitoneum whereas it has been rarely seen in the gastrointestinal tract, and colon is extremely uncommon site.2,3 Here, a case of DDLS which arising from transverse colon is presented, and the literature is reviewed.

2. Presentation of case

71 year old female patient was admitted to outside hospital with complaints of abdominal pain and swelling lasting for three months was referred to our clinic with the diagnosis of intraabdominal mass. There was no change in bowel movements, no sign for bowel obstruction and no weight loss. Triphasic computed tomography (CT) revealed a large macrolobulated peripherally enhancing mass compatible with gastrointestinal stromal tumor, approximately 23 cm × 19 cm × 18 cm in size starting from the neighborhood of left liver lobe and extending toward pelvic floor (Fig. 1). While not invading main vascular structures it was occupying most of the area adjacent to bowel loops and the tumor was invading gastric wall in a small portion at the greater curvature. There was no radiologic hint of colonic invasion or obstruction reported. Patient had a notably distended abdomen on physical examination. Tumor markers were normal. These findings were accepted as sufficient for preoperative diagnosis and no additional investigation was performed in order not to delay surgery. Since neoadjuvant therapy has been found to be inefficacious in such cases, patient was booked for operation. At laparotomy, a multilobulated, soft and yellowish mass approximately 25 cm × 18 cm × 18 cm in size was encountered (Fig. 2). It was arising from transvers colon and invading greater curvature of stomach. After a thorough exploration of abdominal cavity neither peritoneal implants nor ascites was noticed. The mass underwent en-bloc resection including segmental colon and gastric wedge resection. Patient was discharged on day 7 without any complications.

The cut section showed a large grayish yellow nodular mass with few mucoid areas, along with a variegated appearance. At places, it appeared to be fleshy with focal areas of hemorrhage and necrosis (Fig. 3). The tumor center was located in the colon

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* Corresponding author. Tel.: +90 5068864003; fax: +90 2422278837.
E-mail addresses: makturko@gmail.com, makturko@hotmail.com

M.A. Türkoğlu.
wall and invasion to stomach was minimal. Thus macroscopic image had a significant role in identifying the primary origin of the tumor which was accepted as colon. On microscopic examination there were areas with fibrosarcomatous, myxofibrosarcomatous, hemangioperistomatous, gastrointestinal stromal tumor like appearance adjacent to a lipogenic component that was composed of fat cells of varying sizes, which were separated by slender connective tissue septae. In these areas the neoplastic cells were uni or multivacuolated (lipoblasts) with moderately pleomorphic hyperchromatic nuclei, with a multivacuolated, clear cytoplasm (Fig. 4). Immunohistochemical staining revealed that tumor cells were positive with vimentin, CDK4 and S-100. EMA, C-kit, CD34, CD31, CD1A, SMA, S-100, NF, GFAP, HMB-45, NSE, ALK, CD99, BCL-2, Caldesmon, β-catenin, myosin, desmin and FXIII was negative. On basis of these findings, the histopathological diagnosis was consistent with dedifferentiated liposarcoma.
Postoperative adjuvant radiotherapy was planned but patient voluntarily refused the treatment. Two months later, follow up CT studies revealed multiple peritoneal implants; 6 cm × 5 cm at most. The tumor followed an aggressive course with widespread metastatic disease and patient died within about 3 months of operation.

3. Discussion

Liposarcoma is the most common type of soft tissue sarcomas. About 24% of liposarcomas originate from limbs and 45% originate from retroperitoneal area.2 However liposarcomas are rarely encountered in the gastrointestinal tract.3–12

Liposarcomas have five histological subtypes; well-differentiated liposarcoma (WDL), dedifferentiated liposarcoma (DDLS), pleomorphic, myxoid and round cell liposarcoma.13,14 Liposarcomas originating from the colon are very rare.2,15 Previously, only ten cases of primary liposarcoma of the colon have been reported worldwide but these reports were focusing on other locations of the colon and most histological subtypes were different from our case (Table 1).3–12 Also to our knowledge DDLS of transverse colon is the first case reported in literature.

Since DDLS is a high-grade sarcoma that originates in a background of a preexisting well-differentiated liposarcoma, it is considered a variant of well-differentiated liposarcoma.15 Histological subtype of the tumor is very important during the course of the disease. While dedifferentiated, round-cell and pleomorphic liposarcomas are high-grade aggressive tumors carrying the ability to metastasize, well-differentiated and myxoid liposarcomas are low-grade tumors and progressing more slowly.2,16

DDLS is often observed at sixth decade of life. It is more common in males and often exhibit abdominal cavity involvement.15 The most common abdominal involvement is retroperitoneum. However, intraperitoneal origin is extremely rare.2,12,17

DDLS may occur when well-differentiated liposarcoma transforms into non-lipogenic sarcoma areas. Dedifferentiation develops in 20% of the first local recurrence and 44% of the second local recurrence, and this has been shown to be related to poor progression and metastasis.16

When DDLS develops inside the abdominal cavity it presents with a space occupying mass lesion. The pathognomonic finding in computed tomography (CT) and magnetic resonance imaging is a heterogeneous and nonlipogenic encapsulated mass.18 These findings are sufficient for the diagnosis and needle biopsy is not necessary.

Surgery is the best method of treatment for dedifferentiated liposarcomas. DDLS sometimes invade adjacent structures. In such cases, when safe dissection plan cannot be obtained, en bloc resection can be done. It is important to remove tumor entirely.13–12

In cases where total resection cannot be achieved, debulking surgery has not been demonstrated to contribute to the overall disease-free survival rates. The targeted chemotherapeutic agents and radiation therapy are being investigated for these unresectable cases.19 In our case, en-bloc resection including colon segmental and gastric wedge resection was performed to the mass arising from transverse colon invading gastric greater curvature.

Even though tumor size, histologic subtype, dissemination are important prognostic factors, it is still difficult to predict prognosis of colonic liposarcoma.12 As in our case, liposarcomas which histologically demonstrate dedifferentiation are associated with a poor clinical prognosis.20

Recurrence is common in liposarcomas. There are studies recommending chemotherapy as the initial course of action in DDLS exhibiting local recurrence where tumor growing faster than 1 cm/month in CT imaging.15 Neoadjuvant therapy has not demonstrated clinical benefit.21,22 Therefore, neoadjuvant therapy was abandoned in our case considering that it does not provide an additive effect on overall disease-free survival rates. There are studies favoring adjuvant radiotherapy in maintenance of local control.23

Table 1

| Age-gender | Size (cm)/location | Histological subtype |
|------------|--------------------|----------------------|
| Wood and Morgenstern | 62-F | 12/ileocecal valve | Myxoid |
| Parks et al. | 54-E | 6/ascending colon | Pleomorphic |
| Magro et al. | 65-F | 5/carcen | Well-differentiated |
| Chen | 52-F | 7.5/descending colon | Well-differentiated |
| Gutsu et al. | 46-M | 12/ascending colon | Myxoid |
| Shahidzadeh et al. | 56-F | 3.5/heptic flexure | Well-differentiated |
| Chaudhary et al. | 66-F | 4.5/descending colon | Well-differentiated |
| Jarbou et al. | 69-M | 7/splenic flexure | Dedifferentiated |
| D’Annnabile et al. | 79-F | 5.2/heptic flexure | Pleomorphic |
| Choi et al. | 41-E | 20/ascending colon | Myxoid |
| Our case | 71-K | 23/transverse colon | Dedifferentiated |
4. Conclusion

In DDLS, local recurrence, distant metastasis, and disease-related mortality rates are 52%, 15% and 30%, respectively. In general, it should be kept in mind that DDLS follows a dreadful course in terms of treatment and prognosis as the tumor exhibits rapid progression characteristics despite chemoradiotherapy. Surgery is still the most effective treatment option, but relying on this approach alone is not sufficient for disease-free survival.

Conflict of interest statement

None.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient’s relatives for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Mehmet Akif Türkoğlu contributed to study design, data collections, data analysis, writing. Gülşüm Özlem Elpek contributed to writing. Volkan Doğru contributed to writing. Hasan Çalış contributed to data collections, data analysis, writing. Aslı Uçar contributed to data analysis. Cumhur Arıcı contributed to data analysis.

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