

CASE REPORT

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Leiomyosarcoma of ileal mesentery in a middle age female: a rare case report



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Abstract

Background: Leiomyosarcoma arising from mesentery is a rare tumor associated with worse prognosis. The tumor arises from smooth muscles in vascular wall. Ileal mesentery is common site of this tumor origin.

Case presentation: A middle age female was presented with complain of abdominal distension. A mass was palpable on abdominal examination. CT scan showed large mesenteric cyst. 15 × 10 cm large mass excised with associated mesentery and small bowel segment and on histopathology was found, and it was diagnosed as mesenteric leiomyosarcoma. The patient was discharged and referred to oncology for chemotherapy.

Conclusion: Leiomyosarcoma is a rare disease with grave prognosis. There is a paucity of literature and proper guidelines for its management; however, surgical excision with healthy margins is a mainstay of management. Close follow-up is recommended post-operatively because there are high chances of recurrence and metastasis.

Keywords: Leiomyosarcoma, Ileal mesentery, Chemotherapy, Oncology, Case report

Background

Primary leiomyosarcoma of the mesentery is a rare aggressive neoplastic lesion with worse prognosis [1]. Mesenteric leiomyosarcomas have a reported incidence of 0.2% of all cancers and 15% of all soft tissue sarcomas [2]. This tumor most likely derives from smooth muscle cells of mesenteric vessels. About two-third of these tumors arise from mesentery of small bowel with ileal mesentery being the most common site, although origin from transverse and sigmoid mesocolon or other region is possible. They arise from the retroperitoneal space; therefore, diagnosis is delayed and they may therefore grow to a large size before being discovered [3].

We diagnosed a rare case of leiomyosarcoma which developed in the mesenteric cyst. Her diagnostic workup was done, surgery was planned, and histopathology of the specimen revealed definitive diagnosis.

Case presentation

A 50-year-old female was presented with complain of abdominal distension for 4 months associated with intermittent post-prandial nausea and vomiting. The patient had a past surgical history of total abdominal hysterectomy and bilateral salpingo-oophorectomy 4 years back due to fibroid uterus. General physical examination of the patient was unremarkable. On abdominal examination, a mass was palpable in central abdomen which was mobile, non-tender, and soft to firm in consistency with no overlying skin changes. All baseline investigations were within normal limits.

Ultrasound abdomen showed a complex mass in the right side of the abdomen near umbilicus, predominantly hypoechoic measuring approximately 15.2 × 9.5 cm. On color Doppler, it showed minimal vascularity. CT scan abdomen revealed large solid mass with small cystic component as well as in the lower abdomen and pelvis (Fig. 1). It was compressing and displacing the adjacent bowel loops. There was associated perilesional fat stranding. The mass measured approximately 14 × 9.2 cm. Increased vascularity was noted around the mass.

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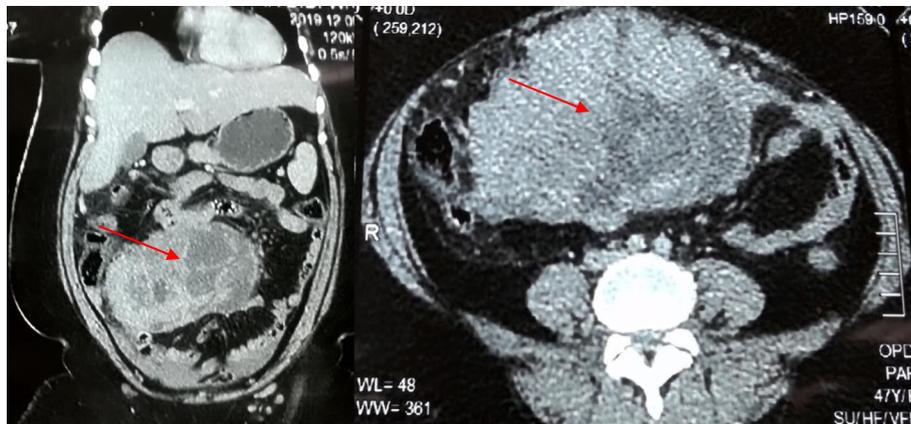


Fig. 1 CT scan (coronal and axial view) abdomen shows large mass in the abdomen (red arrows)

Mild fat stranding was noted in the right paracolic region with minimal ascites.

Chest radiograph was performed, and there was no lung metastasis. Her CA-125 level was within normal limits. A provisional diagnosis of mesenteric cyst was made.

Perioperatively large cyst of around 15 × 10 cm with enormous blood supply was seen in ileal mesentery 5 feet proximal to the ileo-cecal junction. There were multiple adhesions between cyst and anterior abdominal wall and surrounding mesentery. Excision of the cyst with wedge resection of an associated mesentery and ileal segment was done. On gross examination, it was solid in consistency (Fig. 2).

Histopathology report revealed neoplastic lesion with intersecting fascicles showing herringbone and storiform

pattern, and SMA and desmin were diffusely positive while CD117, CD-34, and SG-100 were negative and features were consistent with leiomyosarcoma. Although associated ileal segment was healthy and tumor-free.

Patient was discharged and referred to oncology for adjuvant chemotherapy as advised by oncologist. Post-operative CT scan done after 2 months showed no recurrence or metastasis. On follow-up visit after 6 months, patient had received 6 cycles of chemotherapy and was symptoms-free with healthy healed scar and called for follow-up after 6 months in surgical outpatient department.

Discussion

Leiomyosarcoma arising from gastrointestinal mesentery is a rare but aggressive pathology. A first case report on



Fig. 2 Post-operative specimen

mesenteric leiomyosarcoma was published by Derechin et al. in 1956 [1]. Common presenting complaints in patients with leiomyosarcoma includes palpable mass, abdominal pain, and abdominal fullness, and middle age females are mostly affected. The proliferation of smooth muscle cell secondary to estrogen is a likely explanation for this increase incidence [4, 5]. Our patient was presented with complaint of abdominal distension for 4 months associated with nausea and vomiting.

In 1963, Yannopoulos et al. in their case series reported five cases of leiomyosarcoma, while three out of these five tumors were reported to be originated from mesentery [3]. Most of the primary mesenteric lesions are benign, and differential diagnosis of the malignant disease includes GIST, leiomyosarcoma, malignant myofibroblastic tumor, solitary fibrous tumor, liposarcoma, and lymphoma [3, 6, 7].

Imaging plays a crucial role in the pre-operative workup of mesenteric leiomyosarcoma. Ultrasound and CT scan both help in diagnosis, localization, identification of metastatic involvement, and characterization of disease nature. The solid lesion most likely to be malignant while cystic tends to be benign on CT scan imaging [8]. In our case, a solid area of the cyst was around 80 to 90%.

Per cutaneous biopsy is usually contraindicated as it can result in tumor seeding, thus the only means of definitive diagnosis is a histopathological examination of the specimen with immunohistochemical staining and genetic analysis [9]. CD 117 helps to differentiate GIST from leiomyosarcoma, as the former are CD 117 positive. Leiomyosarcoma universally expresses SMA (smooth muscle actin) while desmin expression is variable [4]. In our case on histopathology, SMA and desmin were diffusely positive while CD 34 and CD 117 were negative. Pathologically, Ranchod M. et al. reported in their study that high mitotic activity strongly indicates the malignant potential of leiomyosarcoma [6]. In terms of mitotic activity, in our case, up to 18 to 20 mitoses per 10 HPF including atypical mitotic figure were reported.

Surgical excision of mesenteric leiomyosarcoma with clear margins is a treatment of choice [10]. Surgeons should try to completely excise the smooth muscle cells tumor with at least 4 in. of healthy tissue and corresponding mesentery in order to improve the survival rate. The good chemotherapeutic response is reported for this condition unlike GIST [5]. Long-term follow-up is recommended as it helps in early recognition of the recurrent or metastatic disease and facilitating curative resection [5, 11]. Neoadjuvant high-dose long-infusion Ifosfamide (HLI) and external beam radiotherapy (EBRT) reduce the mass for performing a full resection. Some authors suggest a combination of preoperative

EBRT and chemotherapy with surgery and intraoperative radiotherapy (IORT) seems to improve the overall survival rate [12].

Agarwal K et al. reported that the site of tumor contributes to the grave prognosis of this disease as it prevents the detection of tumor at an earlier stage [10]. Most of the tumors are already metastatic at the time of diagnosis to lung or liver. Hashimoto et al. [7] demonstrated only 21% 5-year survival rate of 44 leiomyosarcomas arising from the retroperitoneum and mesentery. Hence, these patients need careful long-term follow-up.

Conclusion

Leiomyosarcoma of mesentery is a quite rare malignancy with dismal prognosis. It would be necessary to take a detailed history and perform a careful abdominal examination to make a proper diagnosis. In our patient, we followed her for 6 months and found her disease-free but a long-term follow-up for earlier detection of liver metastasis combined with aggressive re-excision for metastasis or recurrence is therefore recommended.

Abbreviations

CT: Computerized tomography; CA-125: Cancer antigen; CD 117: Also known as KIT, tyrosine kinase receptor; CD 34: Transmembrane phosphoglycoprotein; GIST: Gastrointestinal stromal tumor; SMA: Smooth muscle actin; HLI: High-dose long-infusion ifosfamide; EBRT: External beam radiotherapy; IORT: Intraoperative radiotherapy

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Authors' contributions

HAQK is the correspondence author, and revised the work, and submitted the case. JB and SK interpreted the data. MRA and ASM have approved the submitted version for publication. MA and MR has drafted and designed the work. No disclosure. All authors read and approved the final manuscript.

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Availability of data and materials

The data used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Ethics approval and consent to participate

This study was approved by Institutional Review Board (IRB) at Dow University of Health Sciences; reference number of approval: IRB-1799/DUHS/Approval/2020/78. The patient included in this study gave written informed consent to participate in this research.

Consent for publication

A written informed consent was taken from the patient for publication.

Competing interests

The authors declare that they have no competing interests.

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