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Original Research Paper

General surgery

A RARE CASE OF PRIMARY MESENTERIC LEIOMYOSARCOMA

Dr. B. Santhi	MBBS, MS(General Surgery),DGO, Professor and Head of the Department, Department of General Surgery, Government Kilpauk Medical College, Chennai -600010
Dr. M. Annapoorani	MBBS, MS(General Surgery),Assistant professor, Department of General Surgery, Government Kilpauk Medical College, Chennai -600010
Dr. Deepak Bharathwaj P. P.	MBBS, Postgraduate in General Surgery, Department of General Surgery, Government Kilpauk Medical College, Chennai -600010

ABSTRACT A rare case of mesenteric leiomyosarcoma has been described in a brief review of the literature. A 25 yr old female came with chief complaints of a mass over the abdomen for five months. MRI shows a large abdominopelvic cystic lesion with multiple loculation and septation. After evaluation patient was taken for exploratory laparotomy, which revealed a large abdominopelvic mass of size26x24x20 cm arising from mesentery abutting to the loop of the ileum; hence proceed with resection of mass along with ileum and ileo-ileal anastomosis .later on, histopathological examination revealed it to be the myxoid type of leiomyosarcoma arising from the mesentery. Primary tumour originating from mesentery is rare. Mesentery Leiomyosarcoma is one of the extremely rare tumours arising from the mesentery.

KEYWORDS : mesentery, leiomyosarcoma, gastro-intestinal

INTRODUCTION

Leiomyosarcoma is one of the very rare malignant neoplasms. It is the type of soft tissue sarcoma which arises from smooth muscle spindle cells. These tumors have a female predisposition. The most common genes involved are RB(loss of ch13q), PTEN(deletion of ch10q), loss of p53, MED 12 (especially in uterine lesion)

It leads to the activation of the PI3K/AKT pathway. It also has an association with prior radiation, immunocompromised, and EBV infection. Leiomyosarcoma most commonly occurs in the retroperitoneum, intraperitoneal, out of which the uterus is a more common site.

It can also arise from smooth muscle cells of major vessels like IVC. The mesenteric origin of leiomyosarcoma is extremely rare, with an incidence of 1:3,50,000, which is likely to develop from vessels of the mesentery.

Case Study

A 25-year-old female came with chief complaints of abdominal mass with occasional abdominal pain for five months duration. On examination, mass extending from xiphisternum to below pubic tubercle. Ultrasonography revealed a large loculated cystic lesion with normal bowel peristalsis and solid organs that looks normal. MRI shows a large abdominopelvic cystic lesion with multiple loculation and septation with the septal thickness of 7.5 mm, uterus and bilateral ovaries normal. No lymph node enlargement, invasion into other organs, and ascites. On exploratory laparotomy, an encapsulated and lobulated mass, 26×26×20 cm in size, arising from the mesentery of the ileum, and it is adherent to the ileal wall. The mass was removed along with 30-cm of distal ileum which is five centimeters proximal to ileo-cecal junction and anastomosis of the ileum, and the specimen was sent for histopathology. On gross examination, the tumor measured 25x25x20 cm and weighed 10 kg. The cut surface of the mass shows greyish white cystic areas along with areas of hemorrhage with the periphery showing myxoid changes. On histopathological examination, well-encapsulated circumscribed neoplasm arranged in sheets and fascicles consisting of elongated spindle-shaped cells with vesicular nucleus with atypia, extensive areas of myxoid degeneration, and stellate cells are seen with no capsule infiltration and necrosis. Ileal loops are free from the tumor with Mitotic activity was low, with 1-2 mitoses/10 high power fields seen. Immunohistochemistry shows desmin positive, SMA - positive, Ki 67-1% positive, EMA, c-KIT, and S-100 negative suggestive of the myxoid type of mesenteric leiomyosarcoma with FNCLCC Histological grade of Grade I. Patient started on AIM Regimen Inj.Adriamycin, Inj. Ifosfamide and Inj.MESNA. On follow up patient completed four cycles of AIM Regimen

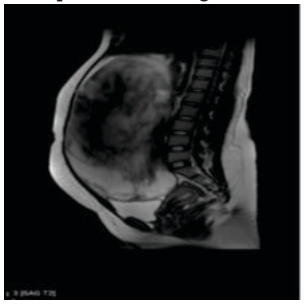


Figure 1: Mass separate from uterus Uterus appears normal

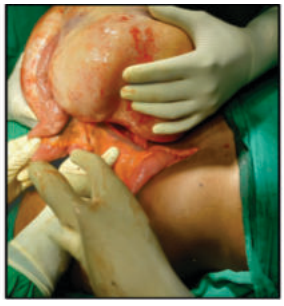


Figure 2: Mass attached to mesentery and ileum

DISCUSSION:

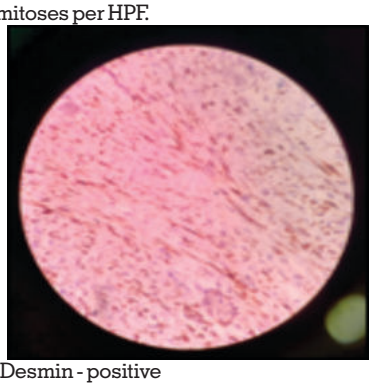
Leiomyosarcoma is a type of soft tissue sarcoma which is generally a heterogeneous, well-circumscribed tumor with an often cystic or necrotic central area. leiomyosarcoma is mainly found in the retroperitoneum, uterus, and medium to large abdominal vessels arises from smooth muscle cells of vessels, especially in IVC, it can rarely arise from the mesenteric smooth muscles. Mesenteric leiomyosarcoma most commonly involves the meso-ileum, but there are rare reported cases involving the transverse, ascending, descending, and sigmoid mesocolon. The mesentery is usually a common site for the metastasis of gastrointestinal malignancies. Primary tumors arising from the mesentery are relatively rare, lymphomas, gastrointestinal stromal tumors (GISTs), leiomyosarcomas, liposarcomas, fibrosarcoma, pleomorphic undifferentiated sarcomas, and hemangiopericytoma. The mesenteric leiomyosarcomas with a reported incidence of 1:350000.

Leiomyosarcoma presents clinically with various symptoms such as nausea, vomiting, diarrhea, abdominal mass, weight loss, abdominal distention, and nonspecific abdominal pain.

Leiomyosarcoma tends to grow along the tissue planes and thus can compress surrounding structures, leading to the formation of a pseudo-capsule with fingerlike projections that can infiltrate adjacent tissues. Approximately 50% of patients will develop distant metastasis that occurs through the hematogenous spread, most commonly to the liver, lungs and rarely to adjacent lymph nodes.

A definitive diagnosis of leiomyosarcoma can only be made by the histopathologic study, including immunohistochemical staining and genetic analysis. IHC is essential for the accurate diagnosis of leiomyosarcoma. Percutaneous biopsy is contraindicated because skin metastasis and peritoneal dissemination may be induced by biopsy.

Surgical excision with wide margin is primary mode of treatment Pathologically leiomyosarcoma usually has high mitotic activity. Factors increasing survival rate are the histological grade of mesenteric leiomyosarcomas, based on cell differentiation, cellularity of the tumor, anaplasia, and the number of mitoses per HPF.



IHC shows Desmin - positive

Historically, doxorubicin, ifosfamide, docetaxel, and gemcitabine have been used for leiomyosarcoma. Recently Olaratumab is a recombinant human monoclonal antibody that binds PDGFRα. In combination with doxorubicin, it showed an improvement in overall and progression-free survival for patients with advanced, unresectable leiomyosarcoma

Postoperative serial imaging such as a CT chest, abdomen, and pelvis is recommended in leiomyosarcoma. 63% of patients had positive metastasis in a median of 13-months, and 21% showed recurrence of primary tumors within a median of 6.5 months. Therefore, in patients who underwent complete tumour resection, strategic surveillance is recommended every three to six months for two to three years, and then annually overall 5-year survival rate for this tumour is only 20%–30% and complete primary surgical resection is critical for achieving the best outcome. Conversely, recurrence can occur within five years, so that close and long term follow-up of such patients for five years or more, with particular attention to the gastrointestinal tract, liver, and lung, is necessary

Conflict Of Interest: No conflict of interest.

CONCLUSIONS

This is the rare case of primary mesenteric leiomyosarcoma present as abdominal mass then proceeded with laparotomy surgical excision. Post operatively on histopathological examination revealed mesenteric leiomyosarcoma. Later, patient sent for chemotherapy patient started on Inj.Adriamycin, Inj. Ifosfamide and Inj.MESNA. patient completed four cycles of AIM Regimen

Mesenteric leiomyosarcoma is mainly an histopathological diagnosis and surgical excision is main line of treatment

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