Case report of primary mesenteric leiomyosarcoma

Muhammad Areeb Ashfaq[1*], Summaya Saeed[2], Ammara Salam[3], Pirhay Fatma[4], Danish Faheem[5], Aun Ali Khawaja[3]

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ABSTRACT

Background: Mesenteric Leiomyosarcoma (LMS) is a rare soft tissue tumor that takes its origin from smooth muscle cells, usually mesenteric blood vessels. Immunohistochemical (IHC) staining is deemed necessary for the diagnosis. Surgical resection with negative microscopic margins is the best treatment available, with limited role of chemo and radiotherapy.

Case Presentation: We present a case of 46-year-old male who presented with large abdominal mass and obstructive symptoms. Surgical excision was performed and diagnosis of LMS was established on the basis of IHC stains. Currently, the patient is under follow up and has not developed any recurrence.

Conclusion: Due to limited cases recorded globally, definitive guidelines for the management of this tumor are lacking. Work needs to be done to explore further about Primary Mesenteric LMS.

Keywords: Leiomyosarcoma, biopsy, large core-needle, mesenchymal tumor, case report.

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Correspondence to: Muhammad Areeb Ashfaq
*Aga Khan University Hospital, Karachi, Pakistan.
Email: areeb249@gmail.com
Full list of author information is available at the end of the article.

Background

Leiomyosarcoma (LMS) is an aggressive soft tissue tumor that arises from smooth muscle cells. It is a rare tumor with an incidence of 1 in 350,000 cases [1,2]. The common sites include retroperitoneum, uterus, and abdominal vessels, although it can also arise from the mesentery [3]. Among the mesenteric origin tumors, around two thirds arise from small intestinal mesentery usually the ileum [4]. Immunohistochemical (IHC) staining is regarded necessary for the diagnosis of LMS as it helps to differentiate it from gastrointestinal stromal tumors [5,6]. As like other soft tissue sarcomas, LMS are usually painless as they grow along the tissue planes and present at a late stage as large abdominal masses.

Case Presentation

Clinical presentation

A 46-year-old male patient presented at the outpatient clinic with complaints of intermittent non-projectile vomiting and constipation associated with generalized weakness and malaise, all of which had developed and worsened in the past one and a half month. His appetite and sleep had also deteriorated. He had no known co-morbidities but had a family history of first degree relatives with diabetes mellitus type 2. Patient was not using any medications previously. He had no prior surgeries or hospitalizations. He had a long term addiction of beetle nut chewing.

On examination, general physical examination was unremarkable other than the abdomen where a large, non-tender mass was palpable, extending from right iliac fossa up to left hypochondrium, around 15 × 20 cm in size, hard in consistency with irregular margins and mobile in vertical axis.

Workup

Abdominal magnetic resonance imaging

Multiple heterogeneous masses of varying sizes seen in abdomen with one measuring 14.4 × 10.2 cm, adjacent to the aorta. Liver echotexture is altered. Rest is unremarkable.

Abdominal CT scan

Large heterogeneously enhancing multi-lobulated soft tissue density mass seen in the abdomen, predominantly on left side of midline (Figures 1 and 2). 19 × 12 × 20 cm in dimensions. The lesion is infiltrating walls of distal jejunum/proximal ileal loops resulting in effacement. Anteriorly abutting the anterior abdominal wall and posteriorly reaching up to the prevertebral space and causing effacement of inferior vena cava, aorta, and its branches. Superiorly abutting the tail of the pancreas. Laterally

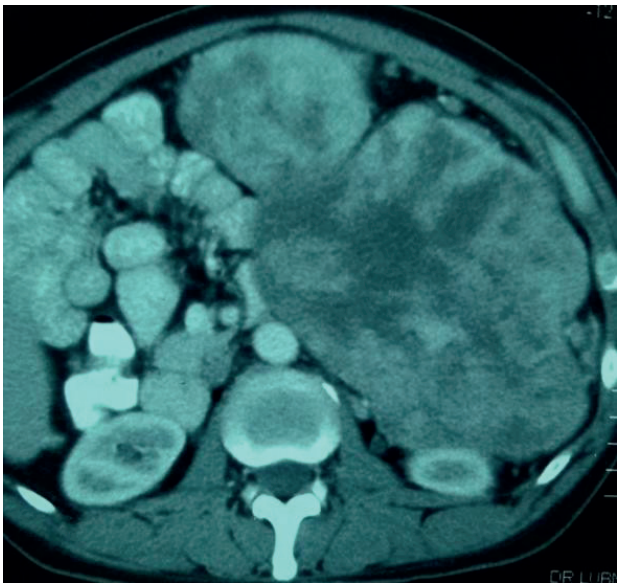

Figure 1. *Axial section of CT scan showing large tumor.*

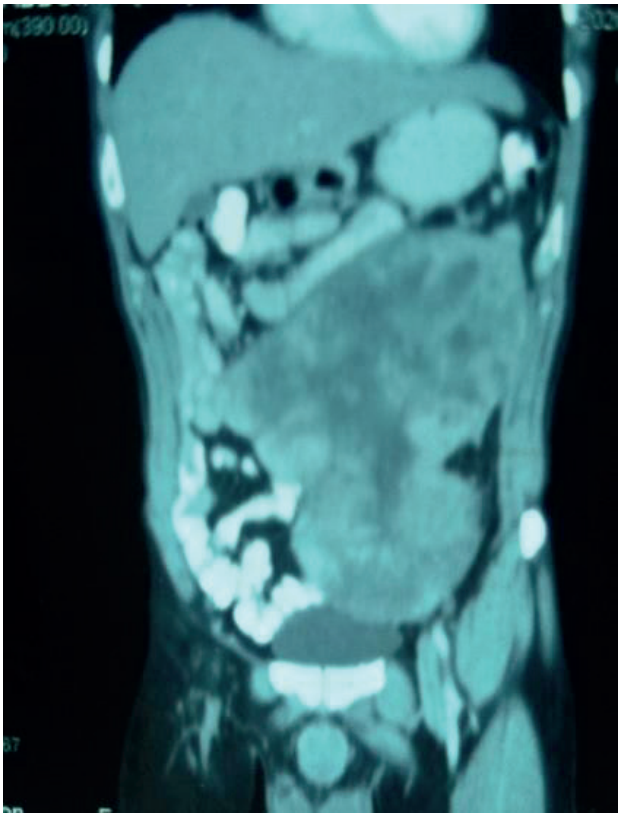

Figure 2. *Coronal View of the tumor.*

effacing left kidney causing mild hydronephrosis and inferiolaterally abutting adjacent descending colon. Multiple subcentimeteric and mildly enlarged mesenteric lymph nodes noted. Liver parenchyma is unremarkable. Rest of the scan is unremarkable.

Ultrasound guided biopsy (Trucut needle) was done and the findings were atypical smooth muscle neoplasm. Mild to moderate atypia and mitoses, low grade LMS suspected. Exploratory laparotomy was then performed on 10th April 2020. Huge lobulated mass was found arising from

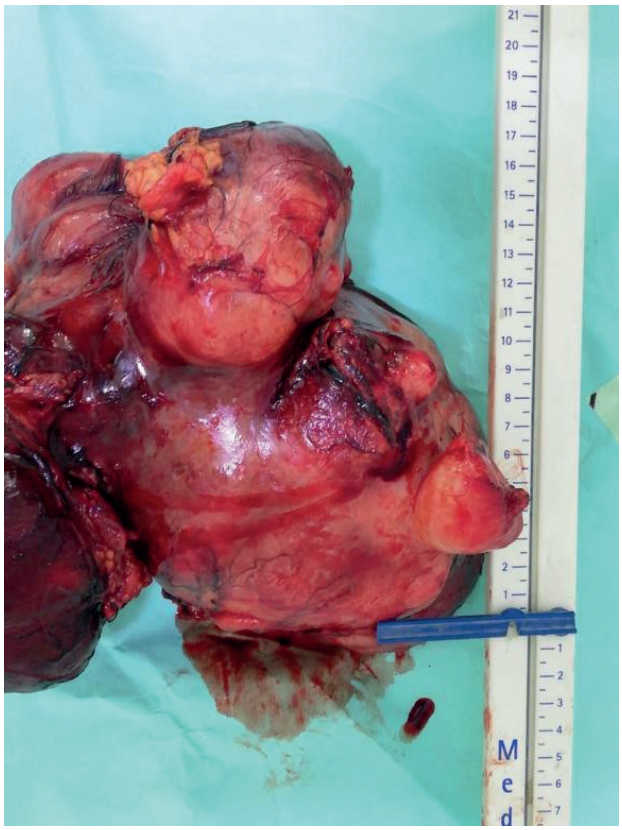

Figure 3. *Gross view of the specimen.*

the large bowel, near the splenic flexure involving the transverse mesocolon, splenic flexure and descending colon. Posteriorly, the mass was firmly adherent to the Gerota's Fascia of left kidney. The tumor was separated from the surrounding structures by ligasure, cavity washed and abdomen closed in layers. One pint of packed red blood cells was transfused intra-operatively. The excised mass was sent for biopsy and the result retrieved in 10 days. Patient spent first 24 hours post-operatively in intensive care unit for observation and one packed red blood cell volume was transfused there. The recovery was swift and on post-operative day 4, patient was discharged home.

On gross examination of the specimen (Figures 3 and 4), a brown multi nodular mass measuring 24 × 21 × 12.5 cm with nodular outer surface, thin capsule, firm and gelatinous areas and hemorrhages in certain regions. This might be the largest size of the tumor ever reported as according to a literature review done by Affas et al. [10], they have analyzed 19 case and the largest size reported was 23 cm.

Microscopic examination reveals spindle cell lesion composed of cells having elongated cigar shaped nuclei and abundant eosinophilic cytoplasm with indistinct cell borders. At places, scattered focal moderate atypia is also seen in cells. Mitoses 2/10 high power field noted. Tumor was found to be 0.2 mm at the closest margin.

IHC stains were performed and the tumor was found to be α-smooth muscle actin positive, epithelial membrane

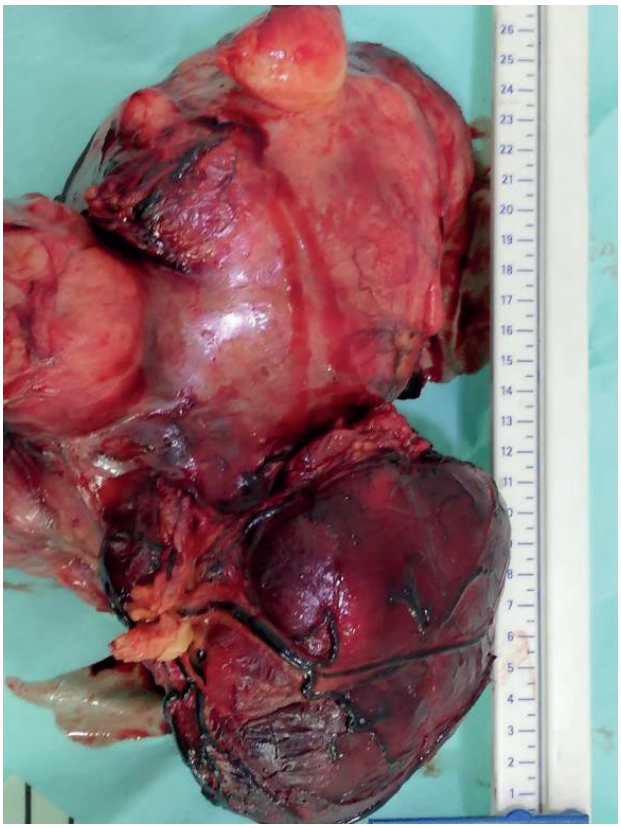

Figure 4. *Gross view of the specimen.*

antigen positive, H-Caldesmon positive, and Desmin positive. The tumor stained negative for CD117, CD34, Discovered on gastrointestinal stromal tumors protein 1, S100, and Cytokeraitv CAM 5.2.

The diagnosis of Low Grade LMS was made on the basis of histopathology. Patient is in follow up. No recurrence of symptoms has occurred till this day. Follow up CT scan is scheduled.

Discussion

Mesenteric LMS is a rare entity and it was first recorded by Yannopoulos and Stout [7]. The tumor takes its origin from the smooth muscle of mesenteric blood vessels [5]. The clinical presentation may vary from non-specific abdominal pain, abdominal distension, nausea, vomiting and diarrhea, most of the symptoms attributing to the compression of surrounding structures by the tumor [8]. Late cases may present with a palpable abdominal mass and significant weight loss. Distant metastasis to lungs and liver is common (50%) in contrast to lymph nodes [9]. On the CT imaging, LMSs are reported as tumors showing heterogenous attenuation due to the enhancement of solid portions of the tumor along with non-enhancing areas of hemorrhage, degeneration and necrosis [10].

In a literature review done by Affas et al. [11] reports the median age at presentation is 55 years with a female predominance (68%). The tumor has a poor prognosis with 5 years survival rate of 20%-30% [10]. The 5 years survival of retroperitoneal liposarcoma is reported as 61% [12]. Surgical excision with wide tissue margin is the most effective mode of treatment since adjuvant chemo or radiotherapy has limited role [2]. Cytotoxic drugs such as Doxorubicin and Ifosamide has shown some role in advanced cases [10]. The recurrence of the tumor is the main cause of mortality among these patients and hence negative resection margins improve survival rates. Patients undergoing surgical resection need surveillance follow ups every 3-6 months for 2-3 years and then annually [13].

The role of core needle biopsy in the management of soft tissue tumors is noteworthy. As their usual presentation is of large abdominal masses, after the radiological workup is done, core needle biopsy shall be the next step. This does not only reveal the tumor pattern and histology, it is rather a fundamental step in tumor diagnosis. The diagnostic accuracy of core needle biopsy is reported to be as high as 99% [14]. Similarly in our case, it directed us towards the diagnosis with IHC staining adding up to their diagnostic yield.

Conclusion

Mesenteric LMS is an aggressive tumor, with poor prognosis. The most important prognostic indicators are the tumor size and location, as both of these factors can affect curative resection. Surgery is the mainstay of treatment. Radiotherapy can offer a better local control but does not improve 5-year survival and recurrence. For advanced stage, cytotoxic chemotherapy has shown a limited role. However, further trials need to be done on that.

We recommend that for an early diagnosis of LMS, MR, and CT imaging supplemented with core-needle biopsy shall be done. We also underline the need of Multidisciplinary Team coordination for the management of such tumors.

List of Abbreviations

IHC	Immunohistochemical
LMS	Leimyosarcoma

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

Funding

None.

Consent for publication

Written consent was obtained from the patient.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

Author details

Muhammad Areeb Ashfaq[1], Summaya Saeed[2], Ammara Salam[3], Pirhay Fatma[4], Danish Faheem[5], Aun Ali Khawaja[3]

1. Aga Khan University Hospital, Karachi, Pakistan
2. Dow University of Health Sciences, Karachi, Pakistan
3. Fazaia Ruth Pfau Medical College, Karachi, Pakistan

4. Liaquat University of Medical & Health Sciences, Karachi, Pakistan
5. Ziauddin Medical University, Karachi, Pakistan

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Summary of the case

1	**Patient (gender, age)**	Male, 46 years old
2	**Final diagnosis**	Mesenteric LMS
3	**Symptoms**	Intermittent non-projectile vomiting and constipation
4	**Medications**	None
5	**Clinical procedure**	Exploratory laparotomy
6	**Specialty**	General surgery