ORIGINAL RESEARCH PAPER

Surgery

KEY WORDS:
leiomyosarcoma, soft tissue neoplasms. Local recurrence

SMALL BOWEL LEIOMYOSARCOMA:A RARE CASE REPORT

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ABSTRACT

leiomyosarcoma is a rare type of malignant tumor that arises from smooth muscle cells.it is classfied as a soft tissue sarcoma.tumors of small bowel amount for less than 5 % of all gastrointestinal malignancies with sarcomas ranking 5 th (~1.2%). small bowel tumors are usually asymptomatic at the early stages & difficult to visualise by upper & lower endoscopy LMS belongs to a heterogeneous group of soft tissue neoplasms, and with its aggressive nature, it often presents local recurrence and visceral metastases. As in the presented case, clinical manifestation of this disease, as in other small bowel tumours, is unspecific.here we discuss a rare case presentation of the same.

INTRODUCTION :

Primary small bowel sarcoma are extremely rare accounting to 5% of the total gastrointestinal malignancies. Most common small intestinal tumors are Adenocarcinoma, carcinoid,lyphoma and sarcoma in decreasing order of frequency.[1]

Histologically, most common variant being leiomyosarcoma which commonly occurs in 5th to 6th decade of life totaling to 2-9%.[2]Few cases were reported of primary small bowel leiomyosarcoma following vigorous immune histological techniques.[3]

Small bowel sarcomas are usually asymptomatic at earlier stages and difficult to visualize by colonoscopy or upper gastroscopy.[4]The jejunum (32 %), ileum (25,2 %), and duodenum (12,6 %) represent the most common sites. LMSs arise in the submucosa and bulge out the mucosa and submucosa. [5] In later stages they may present as intestinal obstruction, bleeding and perforation with poor chances of survival and palliative therapy being the only mode of treatment.[6].

CASE STUDY

45 year old female, homemaker by profession presented with chief complaints of lower abdominal fullness and pain since 15 days. Patient also had a history of constipation.

Computed tomography study of abdomen was suggestive of mass arising from left adnexa with mesenteric and omental fat stranding suggestive of CA Ovary with metastasis and ascites .

Patient had similar complaints three years back and was diagnosed with left adnexal mass for which myomectomy was performed in an outside hospital.On histopathological examination it suggested of leiomyoma.

Patient was admitted at our hospital and evaluated further. Tumor markers like CA-125 were sent which was found to be elevated (401.5). Ascitic fluid was tapped and sent for further investigation which were negative for malignant cells. Decision was hence taken to perform a diagnostic scopy for the patient.

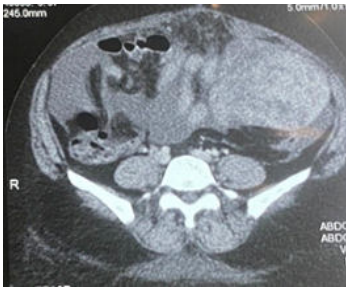
Intraoperatively, on scopy to our surprise, bilateral ovaries and uterus were normal post which decision was made to perform an exploratory laparotomy for the same.

A 14x12x7cm mass was located at the mid jejunal loop around 10 cms distal to the ligament of trietz adhered to the mesentery and omentum suspected to be GIST .

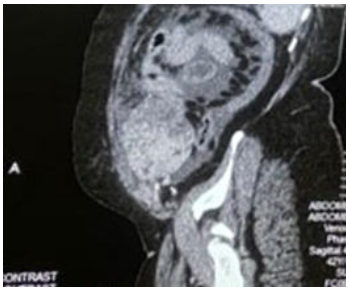
The mass was excised after thorough adhesiolysis with 5cm jejunal marigns and an end to end double layer jejunojejunostomy was performed.

The specimen was further sent for histopathological examination and immunohistochemistry studies.

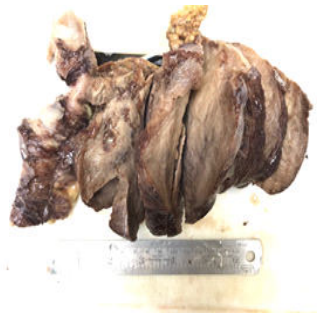
On histopathological examination the diagnosis was suggestive of spindle cell neoplasm (3-5 mitotic figure/10hpf) Immunohistochemistry was done for the specimen which suggested of low grade leiomyosarcoma.



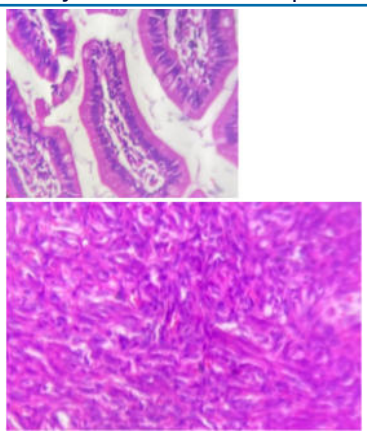
CT Axial view showing a large lobulated mass



CT ABDOMEN CORONAL VIEW



RESSECTED SPECIMEN OF THE MASS



Spindle cell malignant mesenchymal proliferation

DISCUSSION:

leiomyosarcoma originated from smooth muscle cells between musclaris mucosa and muscularis propria.it is a rare entity that typically becomes symptomatic when the tumor is highly infiltrating and spread along the abdominal viscera.early diagnosis is challenging ,mostly due to absence of specific symptoms.they most commonly grow towards the subserosal side of the bowel without causing any intraluminal obstruction hence patients are asymptomatic in initial stage.It most commonly originates in the retroperitoneal space, uterus, vascular wall, and soft tissues. Ileal LMS is a rare tumor originating from the smooth muscle cells within the muscularis mucosa or muscularis propria [7].

The highest incidence of LMS is observed in the sixth decade. There is a small preponderance of males [8].

The symptoms are not specific. Patients can present chronic abdominal pain, chronic anemia, and recurrent melena [9].

Upper endoscopy and colonoscopy are not performant in assessing the diagnosis. Thus, those tumors are discovered at advanced stages as in our case.

CT colonography (CTC) and magnetic resonance enterography (MRE) / Contrast enhanced computed tomography (CECT) represent good options to aid the diagnosis [10]. CECT is more accessible and provides a better resolution [11]. CTC acquires better soft-tissue contrast. It is also more performant to detect small mucosal lesions. It has the advantage of avoiding radiation.

Those endoscopic methods have the disadvantage of only evaluating the intestinal lumen. Thus, extraluminal growth and metastases can't be correctly evaluated.

Despite advances in imaging, determining the difference between benign and malignant tumors before surgery remains extremely challenging .Therefore, the definitive diagnosis can only be confirmed after histological examination and IHC, the case presented here being a prime example.

Histologically, LMS often has a comparable morphological appearance to GISTs. It presents as a smooth muscle cell malignant neoplasm with high mitotic counts, necrosis, and cytological atypia[12]. LMS is usually composed of elongated cells with abundant cytoplasm [13]

IHC is essential to differentiate those tumors. LMS are distinguished from GISTs by the negativity of CD 117, DOG-1, and CD 34 and the positivity of SMA and Desmin

The Tumor-Node-Metastasis classification for soft tissue sarcomas is used to stage small bowel LMS

Surgery is the only curative treatment. Those tumors have a low response rate to chemotherapy [15].

Knowing that those tumors are discovered at advanced stages, the prognosis is poor. Though it's more favorable than small bowel adenocarcinomas

Tumor size and histological grade represent independent prognostic factors for disease-specific survival. The five-year survival ranges from 10 to 48 % .We highlight the importance of performing CT and MRE in patients with chronic abdominal pain, weight loss, or recurrent melena to diagnose intestinal LMS at earlier stages.

In summary, we reported a case of jejunal LMS in a 45-year-old patient, presenting chronic abdominal pain and distension. tumor resection with jejunostomy was performed. No relapses were registered during the 4-month follow-up. Further studies with bigger sample sizes and systematic reviews can aid in proposing clear guidelines for those rare tumors.

CONCLUSIONS

Leiomyosarcoma of jejunum is a rare entity.in spite of advanced diagnostic modalities,early diagnosis of tumor is diffcult because of its asymptomatic presentation us the initial stages.radical surgical resection is the treatment of choice followed by radiotherapy.advancement of robust immunohistological diagnostic methods is allowing differentiation of leiomyosarcoma from other mesenchymal tumors of small intestine

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