

Imaging Characteristics of Uncommon Case of Jejunal Leiomyosarcoma in Middle Aged Women

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Abstract: ***Introduction:** Primary Malignant small bowel neoplasms are uncommon. Of this Leiomyosarcoma of jejunum is rare with late onset of clinical manifestations and poor prognosis. **Case report:** A 40 years middle-aged woman presented to our hospital with mass per abdomen, loss of appetite, vomiting and pain abdomen since 1 year. Ultrasonography showed a large intraperitoneal solid irregular mass measuring approximately 23x20 cms, which is extending from epigastric to hypogastric region, laterally to bilateral lumbar and iliac fossa displacing adjacent bowel loops with no bowel obstruction. Solid organs were unremarkable. CECT showed mild to moderately enhanced large exophytic mass inseparable from small intestinal bowel loops, however exact site of origin couldn't be elicited. Provisional diagnosis was small intestinal mesenchymal tumors. Patient underwent explorative laprotomy showing 27x25cms irregular solid mass arising from jejunum with lymph nodes was resected and Histopathological analysis revealed grade 3 leiomyosarcoma. **Conclusion:** A leiomyosarcoma was diagnosed in a hypertensive and diabetic middle-aged woman with late onset of clinical manifestations. Radical surgical resection was done with chemo radiation, however it showed poor prognosis with early intraperitoneal metastasis with in 3 months of follow up scan. Advanced multidisciplinary approach of radiological investigations with immunohistochemistry helps in early diagnosis and management.*

Keywords: middle-aged women, asymptomatic, small intestine mesenchymal tumor, Immunohistochemistry

1. Background

Primary small bowel tumors constitute about 0.5% of all cancers and 3% of all gastrointestinal tumors (1). Most common occurrence of leiomyosarcoma is in 5th-6th decade of life with, 2-9% higher incidence in males with predominance of blacks than whites (2). Small bowel leiomyosarcomas are usually asymptomatic in the initial stages and difficult to visualize by esophagogastrosocopy and colonoscopy thus presenting with late onset of clinical manifestations resulting in poor prognosis. A better approach with high-resolution images provided by Multi detector Computed Tomography (MDCT) with contrast administration in combination with CT enteroclysis is used to know about tumor characteristics and enhancement of growth. Thus helping in arriving at best possible provisional and differential diagnosis and finally histopathological analysis with advanced immunohistochemistry is ultimate for confirmation (3).

2. Case Presentation

A 40 years old female, housewife by occupation presented to our hospital with mass per abdomen, loss of appetite, dull aching pain abdomen and non-projectile vomiting's since 1 year.

Past History—Known case of hypertension and type-2 diabetes on treatment since 7 years.

On USG Examination: A Large ill-defined lobulated, intraperitoneal heterogeneous hypo echoic solid mass (figure -1) extending superiorly from epigastric region to pelvis downwards and laterally to bilateral lumbar and iliac fossae which was measuring approximately 23x20cms causing displacement of adjacent bowel loops with no bowel obstruction and solid organs were unremarkable.

On Colour Flow Mapping

Mild-moderate color uptake of mass noted (figure-1).

- **CECT ABDOMEN:** A Large, lobulated, exophytic intraperitoneal mild-moderately enhancing soft tissue density mass lesion measuring approximately 26x25x21cms with few areas of non-enhancing necrosis and no calcifications. CECT shows better delineation of mass arising from the small intestine; however it was inseparable from small bowel loops resulting in displacement of small bowel loops adjacent to mass with evidence of minimal ascites and no bowel obstruction (figures 2-4). Evidence of few bilateral retroperitoneal and iliac fossae lymph nodes, largest measuring approximately 1.8x1.4 cm in right Para-aortic region was noted. Patient underwent explorative laparotomy by midline incision. A 27x25cms irregular large lobulated solid mass noted arising 10cms distal to ligament of treitz from the jejunum (Figure -5) with high vascularity and surrounding omental adhesions. Mass was excised with 5cms margin of jejunum. A single layer of end-to-end jejunojejunostomy was done. **Nature of specimen:** Small bowel (Jejunum about 10cms) with tumor and 2 lymph nodes (figure -6) excised for Histopathological analysis. Microscopic examination revealed that tumor cells were spindle shaped to epithelioid in appearance and fibrosarcomatous growth pattern (Figure -7). Polar vacuoles mimicking GIST. Furthermore Immunohistochemistry markers were done which revealed KIT 117 negative, SMA, CALDESMIN, CALPONIN, and DESMIN positive (Figure -8). **Mitotic rate:** Score 2 (10-19/10hpf)
- **Federation Nationale De Centres De Lutte Contre Le Cancer (FNCLCC) Grading:** Grade 3 with Scores 6, 7 or 8 Smooth muscle cell tumor-malignant leiomyosarcoma of jejunum.

Differential Diagnosis

- 1) Gastrointestinal stromal tumor.
- 2) Primary gastrointestinal lymphoma.

ON 3 MONTHS FOLLOW UP SCAN: There was recurrence of multiple variable sized, lobulated solid intraperitoneal masses (figure-9).

3. Discussion

Neoplasms of the small bowel are rare lesions less than 3% of all gastrointestinal tumors (1). Sarcomas rank fifth (1.2%) among all malignant small bowel tumors, after carcinoids (44.3%), adenocarcinomas (32.6%), lymphomas (14.7%), and gastrointestinal stromal tumors (GISTs) (7.2%). Leiomyosarcoma originates from smooth muscle cells between the muscularis mucosa and muscularis propria, Incidence of 22.7 per million per year and less than 30 case reports worldwide have been found in literature (3) with higher incidence in males to-female ratio ranges from 1:1 to 2:1 with predominance of blacks than whites (2).

Several theories have been proposed but proven studies showed many predisposing conditions and risk factors involved were inherited syndromes such as HNPC (Hereditary non-polyposis colorectal cancer), FAP (Familial adenomatous polyposis), Peutz jeghers syndrome (4) and MEN (Multiple endocrine neoplasia type-1). Chronic inflammatory conditions like Crohn's and celiac disease (5) and HIV infection. Habits like Tobacco, alcohol, red meat, smoked food; refined sugar and salty or fatty food (6) are also on record.

Most commonly Leiomyosarcomas arise from the subserosal side of the bowel without causing any intraluminal obstruction. Therefore, patients remain asymptomatic in the early stages but in advanced stages they present with abdominal pain, abdominal lump or intussusception (in ileal leiomyosarcoma), iron deficiency anemia, melena. (7). It is difficult to visualize the small bowel by esophagogastroscope and colonoscopy in early stage of disease.

Differentiation between benign and malignant tumors of small intestine remains very difficult so, nowadays multidetector CT performed with non ionic contrast or CT enteroclysis / enterography (7) has advantage of defining the real extension of wall lesions, possible transmural extension, the degree of mesenteric involvement and remote metastasis in a single investigation. MDCT has advantage over MRI because of fast imaging, low cost and high sensitive for detection. Other modalities in limited use like capsule endoscopy and enteroscopy delineate mucosal changes with lower accuracy of submucosal or bowel wall extensions. MRI is more sensitive for better soft tissue delineation differentiating between different tumors based on the T1 and T2 characteristics without ionizing radiation. Positron emission tomography can contribute in evaluating metastases however it is not in widespread usage.

The most important criterion for metastasis and recurrence is presence of mitosis Leiomyosarcoma with 1 mitosis per 2

high power fields or epitheloid leiomyosarcoma with 1 mitosis per 5 fields behaves aggressively. Other factors include presence of cellular pleomorphism, necrosis, gross size of tumor, regional lymph nodes and solid organs involvement.

Histologically the main antibodies to detect GISTs are CD117 (c-KIT), DOG1 and CD34 and leiomyosarcomas antibodies against smooth muscle are act in (SMA), desmin, caldesmin and calponin are required⁽⁸⁾. Leiomyosarcoma and gastrointestinal stromal tumor (GIST) shows a similar morphologic appearance so immune-histochemical methods should be applied to differentiate these tumors. The tumor can be graded using the Trojani or French systems (FNCLCC) for soft tissue sarcomas. (9) High-grade tumors have 10 or more mitoses per 50 high-power fields (HPF).

Main modality of treatment for all small intestinal leiomyosarcoma is radical surgical resection with adequate margins. whereas, In case of unresectable tumors, adjuvant chemotherapy agents like docetaxel, gemcitabine and trabectedin were used. However the success rate of their usage is not proven till day.

Metastases (10) of the small bowel tumors can be by intraperitoneal seeding's, direct extension from adjacent tumors or by hematogenous spread to the liver (65%), other GI locations (15%), lungs (4%). In contrast to other sarcomas, it also metastases by spread lymphatics (13%) and peritoneal route (18%).

The prognosis of small intestine leiomyosarcoma is very poor. In low-grade disease, the five-year survival rate was 55%, in contrast to 5-20% in high-grade disease (10).

4. Conclusion

Over all leiomyosarcoma of small bowel neoplasms is rare entity with 3%. In our present case report, a leiomyosarcoma was diagnosed in a hypertensive and diabetic middle-aged woman with late onset of clinical symptoms and early intraperitoneal metastasis resulting in poor prognosis, which necessitates a multidisciplinary approach of radiological investigations with advanced immunohistological techniques for diagnosis and prognosis of leiomyosarcoma to differentiate from other mesenchymal tumors of small intestine. MDCT in combination with technique of CT enteroclysis or enterography have renewed interest in bowel tumor detection and their characteristics. Radical surgical resection is the treatment of choice and role of chemo radiation has not yet been proved.

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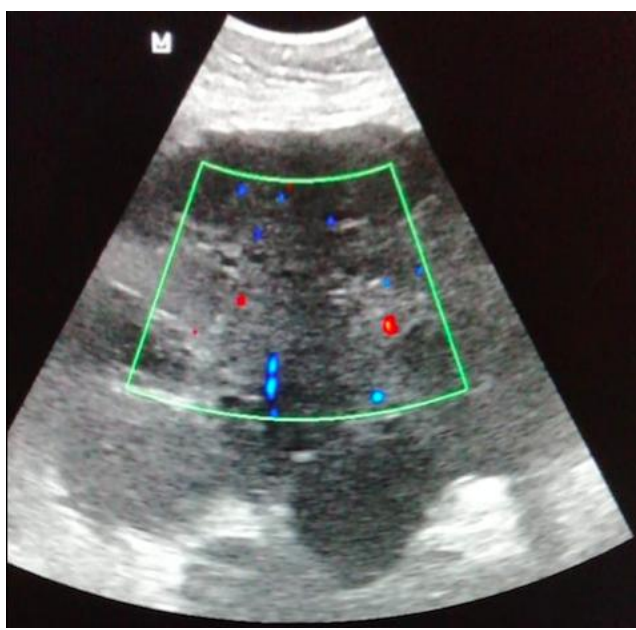


Figure 1

- ON USG :IRREGULAR HETEROGENOUS SOLID MASS WITH FEW AREAS OF NECROSIS
- ON COLOUR FLOW MAPPING: MILD TO MODERATE FLOW

Figure 2: CECT Axial Section (Pre and Post Contrast Images)

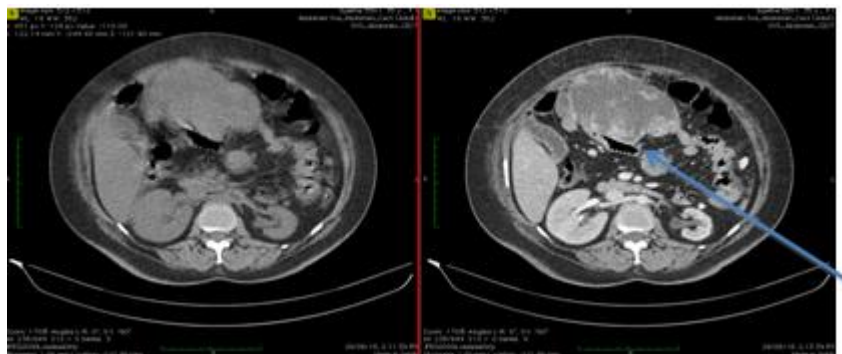


Figure 3: CECT Coronal Section (Pre and Post Contrast Images)

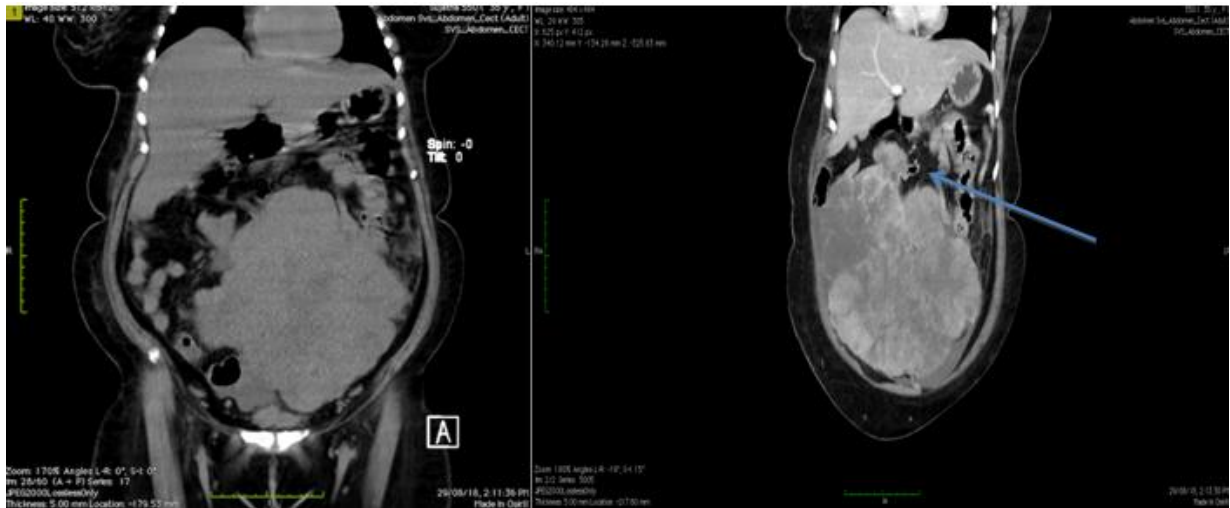


Figure 4: CECT Sagittal Section (Pre and Post Contrast Images)

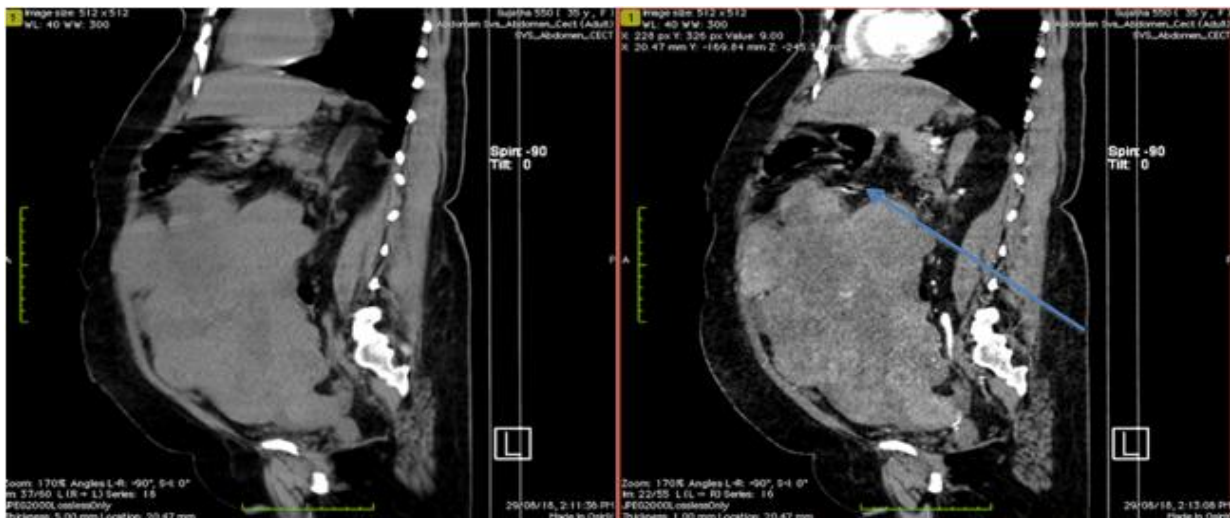


Figure 5: Intraoperative Findings

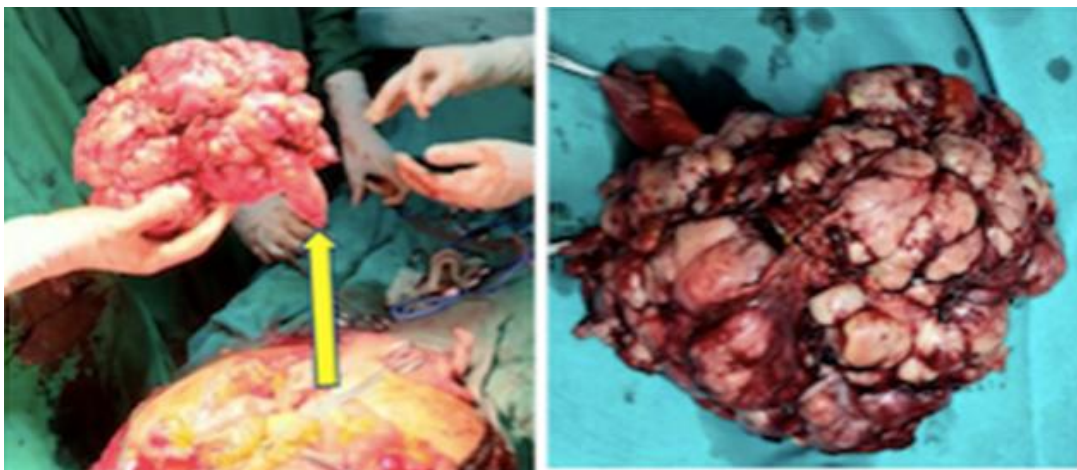


Figure 6: Nature of Specimen: Small Bowel (Jejunum About 10cms) with Tumor, 2 Lymph Nodes



Figure 7: Histopathological Analysis –Microscopy

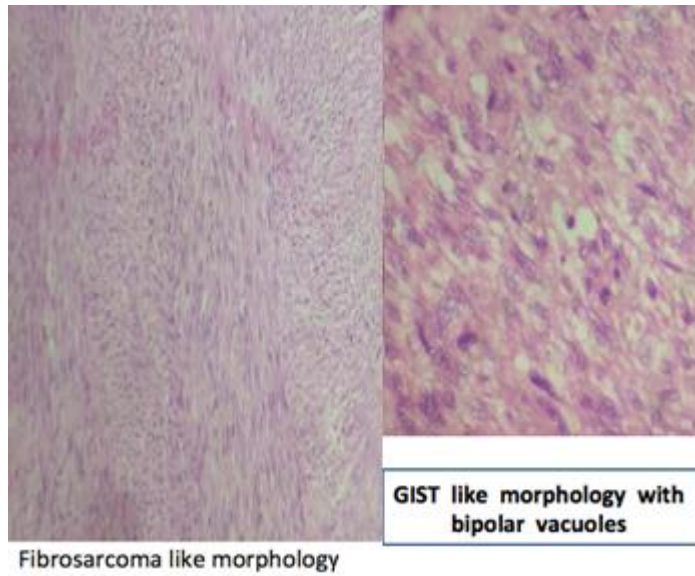
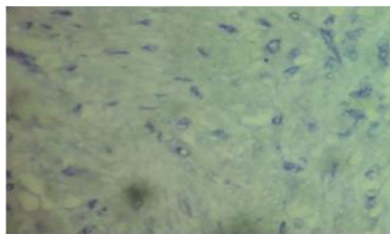
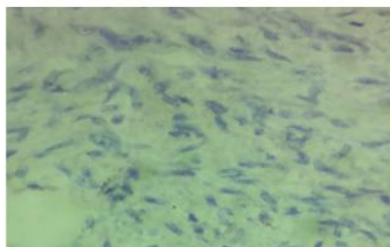


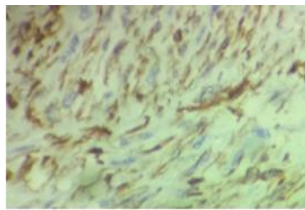
Figure 8: Immunohistochemistry



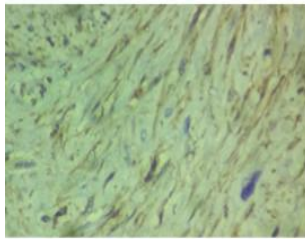
GFAP -Negative



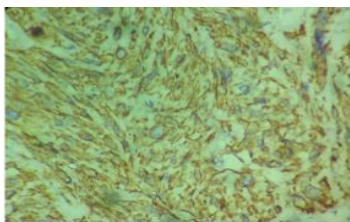
KIT 117 -Negative



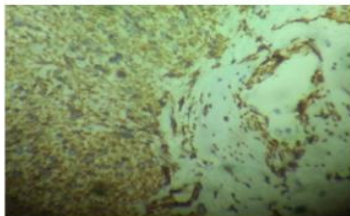
SMA - Positive



Calponin - Positive

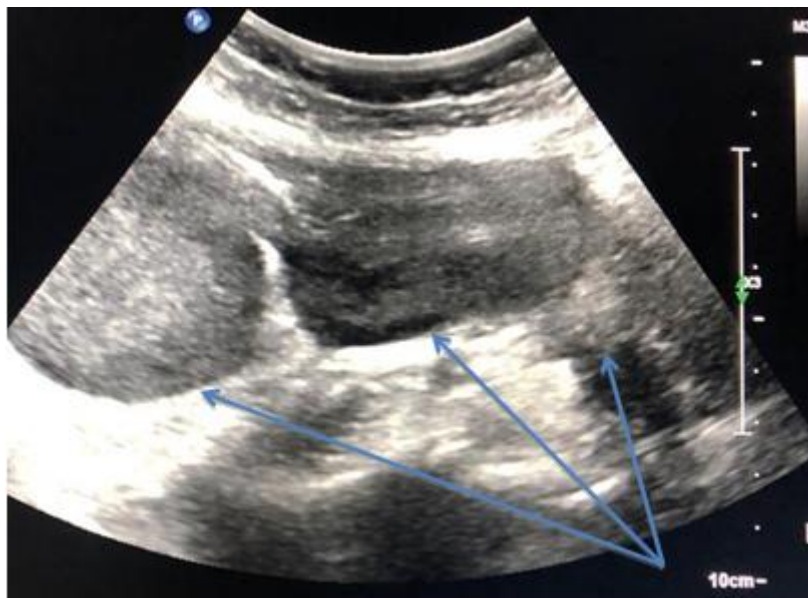


Caldesmonin - Positive



Desmin - Positive

Figure 9: Post operative status: 3 months follow up scan:



Multiple, Solid Variable Sized Intraperitoneal Masses