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Case Report

A primary leiomyosarcoma of small intestine: A rare case report

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ABSTRACT

Primary leiomyosarcoma of small intestine originating from smooth muscle cell is extremely rare entity. It mimics gastrointestinal stromal tumor (GIST) because of their common morphological appearance. Microscopic and immunochemistry findings are necessary to differentiate Leiomyosarcoma from gastrointestinal tumor (GIST). We report a case of 58-year-old male patient presented to tertiary care hospital with complain of generalized abdominal pain, vomiting, early satiety and constipation. Physical examination revealed no significant findings. Imaging findings showed Ileo-Ileal intussusceptions. Neither CT scan nor MRI reveal any evidence of tumor. The patient underwent exploratory laparotomy which revealed polypoidal mass in ileum. Resection and anastomosis of distal Ileum done & specimen received in histopathology department. On microscopic examination, Diagnosis of GIST & Leiomyosarcoma was considered. Immunohistochemistry was positive for Desmin marker and negative for CD 117, DOG 1, S-100 & ALK marker. The diagnosis of High grade Leiomyosarcoma was confirmed.

1. Introduction

Malignant tumors of small bowels are rare and accounts less than 5% of all gastrointestinal cancers.[1] Primary Leiomyosarcoma of the small intestine is an extremely rare form of gastrointestinal malignant tumor. The incidence of Primary Leiomyosarcoma of small intestine has been estimated to be less than 0.1 per 1, 00,000 people per year. Amongst the variety of soft tissue sarcomas, leiomyosarcomas represent 10–20% of these malignancies. LMS most commonly originates in the uterus, GI tract, and retroperitoneum. Within the GI tract, the stomach is the most common site followed by the small intestine, colon, and rectum.[2] Leiomyosarcoma of small intestine is most often observed in the jejunum, ileum and duodenum.[3] Patients aff[4] ected by small bowel leiomyosarcoma are usually admitted to the emergency department due to abdominal pain, bowel obstruction or occult gastrointestinal bleeding.[5,6] It is difficult to diagnose tumors of small intestines at early stages because of indolent nature, therefore, diagnosis is delayed. Usually, patients are treated in advance stage of disease when their general conditions are compromised. Prognosis is poor and local recurrence is frequent. Surgery plays an important role in treatment.

2. Case Report

A 58-year-old male presented with a complain of generalized abdominal pain which was associated with non-projectile vomiting, early satiety and constipation. Physical examination as unremarkable. All hematological investigations were within normal limits. Patient was nonreactive for HIV, HBsAg and HCV. Ultrasonography (USG) and Computed tomography (CT) scan report revealed ileo ileal intussusception and changes of proximal small bowel obstruction. Investigations like Magnetic

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Resonance Enterography (MRE), CT colonography (CTC) and Wireless Capsule Endoscopy (WCE) was not done. Exploratory laparotomy revealed polypoidal mass in ileum so resection and anastomosis of distal ileum performed and specimen received in histopathological department.

2.1. Gross

The specimen received in formalin labelled as ileum resected with polypoidal mass in it, total measuring 24 cm in length. Proximal part of ileum is dilated and in central part of already cut open ileum revealed growth. Circumference of proximal surgical margin was 7 cm and that of distal surgical margin was 5 cm. On cut section of ileum, rugosity is partially lost near growth. Polypoidal growth was identified in central part of resected ileum measuring 4.5 x 4.0 x 1.47 cm in the lumen and obstructing 90% of lumen. On cut section of growth, it is solid, white and reached upto serosa.

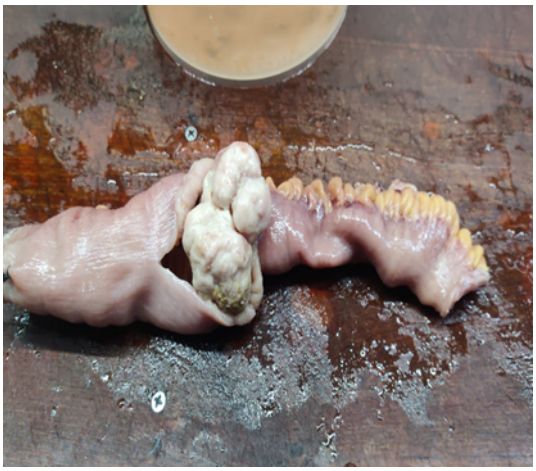

Figure 1: Left): Shows gross image of received specimen of ileum (Central portion was already cut open)

2.2. Microscopic examination

Sections reveal tumor mass is composed of proliferation of spindles shaped neoplastic cells showing cellular atypia, pleomorphism and nuclear hyperchromatism arranged in fascicles and sheets. Brisk atypical mitosis 20-25/10 high power field are seen. No evidence of necrosis is seen. A differential diagnosis of primary leiomyosarcoma of small intestine and Gastrointestinal tumor (GIST) were made. Immunohistochemistry was advised to confirm the diagnosis.

2.3. Immunohistochemistry report

Desmin: Positive
CD117/C-kit: Negative
DOG 1: Negative

Figure 2: (Right): Shows gross image of resected (cut open) ileum

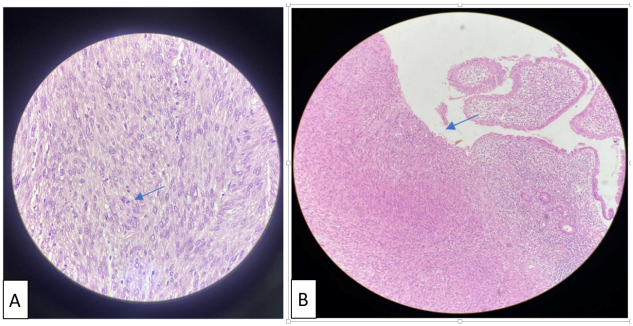

Figure 3: A): Top left: Proliferation of spindled shaped neoplastic cells showing cellular atypia, pleomorphism, and nuclear hyperchromatism arranged in fascicle. (Blue arrows show mitotic figure); **B):** Top Right: Section shows tumor cells bulge out into mucosa and surface erosion is evident. (Blue arrow shows surface erosion)

S-100: Negative
ALK/ CD 246: Negative
KI 67: 30%

Overall findings were reported as high grade leiomyosracoma.

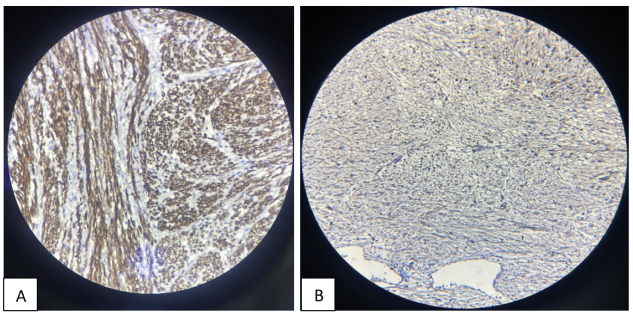

Figure 4: A): Left image: IHC: Desmin-positive; **B):** Right image: Ki 67 – 30%

3. Discussion

We report a rare case of high grade ileal Leiomyosarcoma. Leiomyosarcoma of small intestine is are extremely rare entities, particularly following the advance in immunohistological diagnostic methods allowing differentiations from gastrointestinal stromal tumor (GIST).[7] There are only few cases of leiomyosarcoma of small intestine have been reported. Majority of cases were reported in middle age and there was male preponderance is seen in the previously reported cases.[3] Several etiological factors have been determined but most of leiomyosarcoma are sporadic in nature. LMS more commonly occurs in patient having history of HIV, Retinoblastoma, immunocompromised patients after transplantations and children having congenital immunodeficiency. Tumors also have been associated with Epstein Barr virus (EBV) as a nuclear antigen-2 protein and specific EBV receptor in smooth muscle cells are expressed in immunocompromised individuals.[7]

The most common clinical presentation of leiomyosarcoma of small intestine are hemorrhage and chronic abdominal pain followed by chronic anemia and recurrent melena.[8] Leiomyosarcoma most frequently arises from retroperitoneal space, uterus, vascular wall and soft tissue. Ileal leiomyosarcoma originates from smooth muscle cells within the muscularis mucosa or muscularis propria.[3] Leiomyosarcomas are distinctive from other malignant tumor of small intestine by their greater tendency to bleed and tendency to attain large size without obstruction. Approximately 50 percent of patient with leiomyosarcoma survive 5 year or more than 5 years.

Histologically, leiomyosarcoma mimics gastrointestinal stromal tumor (GIST) due to its common morphological appearance. Leiomyosarcoma presets as a smooth muscle cell malignant neoplasm with high mitotic count, pleomorphism, necrosis and cytological atypia.[9]

It is important to distinguish the diagnosis of LMS from other mesenchymal tumor of gastrointestinal tract, particularly gastrointestinal stromal tumor (GIST) due to different treatment and prognosis. On histomorphological features, Differential diagnosis of Leiomyosarcoma are gastrointestinal tumor (GIST), Desmoid fibromatosis, Leiomyoma, Schwannoma, Intra-abdominal Fibromatosis etc.[10]

Histological diagnosis of LMS is confirmed by immunohistochemistry which is positive for SMA, Desmin and Caldesmon and negative for C-KIT, CD 34, DOG.[8,11] The hallmark histochemical stain that differentiates LMS from GIST is C-KIT, which is uniformly positive in gastrointestinal stromal tumor (GIST) but usually negative in LMS.

The long-term outcome of patients with small intestinal leiomyosarcoma is unknown.

4. Conclusion

We report this case of primary leiomyosarcoma of small intestine for its rarity. Immunohistochemistry is necessary to differentiate LMS from gastrointestinal stromal tumor (GIST). Confirmation of diagnosis is utmost important due to different treatment and prognosis. Although ileal leiomyosarcoma is rare entity, it should be considered in differential diagnosis when patient presents with intestinal mass with morphology of spindle cell neoplasm.

5. Source of Funding

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6. Conflict of Interest

No competing interest.

7. Confirmation

The authors confirms that the content of the manuscript has not been published or submitted for publication elsewhere.

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