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A CASE REPORT ON ILEAL GASTROINTESTINAL STROMAL TUMOR

General Surgery

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract, though they account for only 1-2% of all GI malignancies. They arise from interstitial cells of Cajal. Clinical presentations are diverse and depend on tumor size and location. This case report highlights the rarity of ileal GIST and diagnostic confusion between adnexal and intestinal pathology.

KEYWORDS

GIST, mesenchymal tumor, ileum

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract, yet they account for only about 1-2% of all primary gastrointestinal malignancies.[1] Originating from interstitial cells of Cajal, which regulate gut peristalsis, they are primarily found in the stomach and small intestine. The clinical presentation is variable and depends on tumor size and anatomic site. GISTs originate from the muscularis mucosa or muscularis propria layers and typically exhibit an endophytic growth pattern, growing within the bowel lumen. While the overlying mucosa is usually preserved, larger and more aggressive tumors often ulcerate through it.

GISTs are distinguished from other soft tissue sarcomas by the presence of CD117, CD34, and DOG1 expression, along with the absence of smooth muscle staining markers such as actin and desmin. The primary treatment approach is a full surgical resection.[2,3]

CASE REPORT:

A 55-year-old female who is a smoker and alcoholic presented with chief complaint of diffuse abdominal pain since 2 years, which is insidious in onset, gradually progressive, dull aching type, no aggravating and relieving factors, non-radiating. Bowel habits irregular. No history of fever, nausea, vomiting, early satiety, loss of weight, malena.

No significant past medical or surgical history.

No history of similar complaints in family.

On examination, abdomen was soft, non-distended, tenderness in hypogastrium and right iliac fossa, vague mass of size approximately 6x4 cm palpable in hypogastrium, freely mobile in all directions, ill-defined margins, firm in consistency, freely falling in knee elbow position, dull on percussion, Puddle's sign negative.

Investigations:

All blood investigations and tumor marker CA 125 were within normal limit.

CECT abdomen revealed a well-defined soft tissue density lesion with heterogenous enhancement noted in right iliac fossa region measuring 5.7 x 5.4 x 5.7 cm, laterally and medially related to ileal loops with loss of fat planes and posteriorly related to right adnexa, possibility of neoplastic etiology: primary mesenteric neoplasm/ ileal gastrointestinal stromal tumor (exophytic GIST)/ ovarian fibrothecoma.

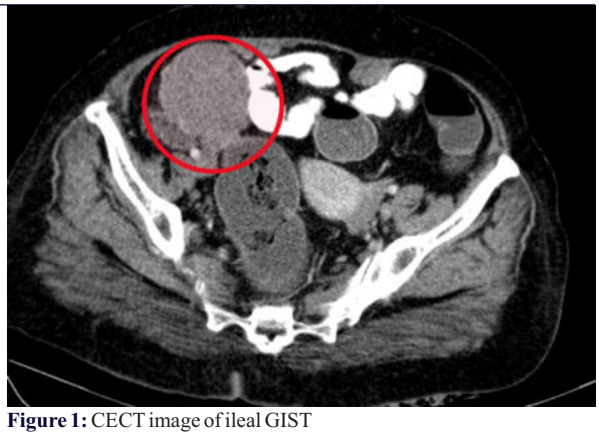

Figure 1: CECT image of ileal GIST

USG guided biopsy was suggestive of gastrointestinal stromal tumor – fibroblastic variant.

Patient was preoperatively stabilized nutritionally, thoroughly evaluated and pre-anaesthetic checkup done.

Preoperatively 1 dose of antibiotic was given before induction.

Surgical Intervention:

Exploratory laparotomy was performed and a solitary pedunculated mass of size approximately 6 x 5 cm was seen arising from anti-mesenteric border of ileum, 70 cm proximal to ileocolic junction. The ileal segment was resected taking a 5-cm margin on both sides, followed by end-to-end ileo-ileal anastomosis. Bowel was followed thoroughly and no other growth was found in gastrointestinal tract. Rest of the visualised viscera was normal.

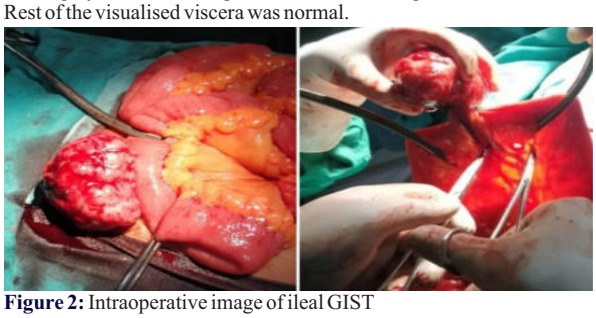

Figure 2: Intraoperative image of ileal GIST

Postoperatively patient was managed with IV broad spectrum antibiotics, antacids and analgesics. Postoperative period was uneventful and patient was discharged on postoperative day 5.

On follow up, histopathological examination showed mucosa and submucosa with underlying tumor composed of short spindle cells admist fibrillary fibro collagenous stroma, these cells appear uniform with minimal mitotic activity 2/10 HPF. Random sections from the tumor shows focal myxoid degeneration.

Impression: Gastrointestinal stromal tumor (GIST) – intermediate risk.

Patient was referred to oncology centre for further follow up as it is a case of GIST - intermediate risk. Patient is under our follow up for last 1 year.

DISCUSSION:

Gastrointestinal Stromal Tumor (GIST) is the most common mesenchymal tumor of the gastrointestinal tract. GISTs are classified morphologically as spindle (70%), epithelioid (20%), or mixed (10%) tumors that express either KIT or PDGFRA. The most common localization of GISTs is in the stomach (50-60%), followed by the small intestines (20-25%) and the rectum (5%). GIST originates from interstitial cells of Cajal (ICC) that proliferate in the myenteric plexus of the gastrointestinal tract.

The clinical presentation of GISTs varies depending on tumor size and location. Their submucosal position can lead to local obstructive symptoms, especially when they occur in the oesophagus or small intestine. Other presentations include vague upper abdominal pain, a sense of fullness, GI bleeding, and a palpable mass. Occasionally, GISTs are discovered incidentally during barium studies, endoscopy, or abdominal scans conducted for unrelated issues.[4]

Diagnosing small bowel GIST is challenging for many surgeons and physicians due to several factors. Firstly, GISTs have a low incidence and present with non-specific symptoms. Additionally, radiological assessment can be difficult because of the wide variation in radiological appearances and the potential for surrounding bowel loops to obscure the tumor.[1]

Prognosis depends on tumor size, mitotic rate, and site of origin. Affected individuals with no family history of GIST typically have only one tumor, known as sporadic GIST. However, individuals with a family history of GISTs are classified as familial GISTs.[5] A prognostic classification system proposed by Fletcher et al. (Table 1) categorizes GISTs into very low, low, intermediate, and high-risk groups for malignancy, based on tumor size and mitotic count.[6]

Table 1: Prognostic Classification System Of GIST By Fletcher et al

Class	Size (largest dimension in cm)	Mitotic count (HPF)
Very low risk	<2	<5/50
Low risk	2-5	<5/50
Intermediate risk	<5	6-10/50
	5-10	<5/50
High risk	>5	>5/50
	>10	Any mitotic rate

Surgery remains the standard of care for patients with primary resectable GIST. The goal of surgery should be R0 resection [negative microscopic margins]. Imatinib is a targeted treatment for KIT-positive unresectable or metastatic GIST. Sunitinib, a multitarget TKI, was approved as a second-line treatment for advanced GIST after imatinib failure. Regorafenib, a multitarget TKI, is a competitive inhibitor of the ATP-binding site for KIT, which has been approved as a third-line treatment of advanced GIST after progression on imatinib and sunitinib.[5]

CONCLUSION:

Small bowel GIST is an uncommon tumor. Due to nonspecific symptoms and signs, it is difficult to diagnose ileal GIST preoperatively. The mainstay of treatment is surgical complete R0 resection and close follow up.

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