Case Report

Gastrointestinal Stromal Tumor of Jejunum Presenting as a Pelvic Mass: A Rare Case Report and Review of Literature

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Abstract

Gastrointestinal stromal tumors (GISTs) are mesenchymal in origin and quite rare in the gastrointestinal system. Jejunal GISTs are extremely rare, approximately 0.1%–3% of all gastrointestinal (GI) tumors. The stomach is the most common site while small intestine (usually duodenum) is the second most common site in the GI tract. The clinical manifestations of GISTs range from asymptomatic to mild abdominal pain, mass, mechanical obstruction, and intestinal hemorrhage as well as perforation. Final diagnosis is made by combined histopathological examination (HPE) and immunohistochemistry (IHC) examination of resected specimen. Here, we present the rare case of jejunal GIST as pelvis mass in 48-year-old gentleman. The patient presented with pain abdomen and heaviness in lower abdomen. On contrast-enhanced computed tomography abdomen, diagnosis of pelvic mass was made. Exploratory laparotomy was done. A solid mass measuring 6 cm × 8 cm arising from antimesenteric border of proximal jejunum found into pelvic cavity. Jejunal segment with tumor resected and anastomosis done. HPE and IHC confirmed intermediate grade GIST of the jejunum. Imatinib mesylate was initiated due to probability of disease recurrence (24%). Carefully complete surgical excision and adjuvant therapy with imatinib is the cornerstone of intermediate to high degree GISTs.

Keywords: Gastrointestinal stromal tumor, jejunum, pelvic mass

Introduction

Gastrointestinal stromal tumors (GISTs) are rare malignant tumor of gastrointestinal (GI) tract. However, GISTs are most common mesenchymal tumor of GI tract. GIST originates from the interstitial cells of Cajal in the autonomic nervous system of the intestine. The stomach is the most common site of GIST but rare in small intestine, particularly jejunum.[1] It occurs mainly in the connective tissue of GI tract with possibility of 20%–30% for malignancy. It has male preponderance with a median age of 50–70 years.[2] Clinical presentation of GIST is highly variable. They can present as simple abdominal pain to abdominal emergencies such as GI bleeding, bowel perforation, obstruction, abscess formation, and tumor rupture. Our patient had unusual presentation of jejunal GIST as a pelvis mass, which is the rarest form of its presentation.

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Case Report

We present the case of a 48-year-old gentleman, who presented with pain and heaviness in the lower abdomen for the last 3 months. Pain relieved after taking medication. No history of melena, hematemesis, and altered bowel habits. Family history and psychosocial history is not significant. General physical and systemic examination was normal. On per abdominal examination, tenderness in lower abdomen after deep palpation. Routine blood investigations of the patient were within the normal limits.

Ultrasonography of the abdomen shows a 6 cm × 8 cm hypoechoic mass in the pelvis. Contrast-enhanced computed

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tomography (CECT) of the abdomen and pelvis showed a heterogeneously enhancing solid mass measuring about 5.3 cm × 7.5 cm in the pelvis behind the urinary bladder [Figure 1a and b]. CEA and CA19-9 were within the normal limits. Colonoscopy was also normal. Provisional diagnosis of pelvic mass was made.

After proper counseling, an exploratory laparotomy was planned. On exploration through midline incision, there was a 6 cm × 8 cm jejunal mass approximately 20 cm from duodenojejunal flexure in the pelvis [Figure 2]. 15 cm of jejunum containing tumor was resected and end-to-end bowel anastomosis was done [Figure 3]. No lymph node involvement was noted and other intra-abdominal organs were normal. On histopathological examination (HPE), macroscopically, cut surface was grayish white with areas of cystic degeneration. Microscopy: Section showing mesenchymal neoplasm in the submucosa of jejunum and fibroblastic tumor cells arranged in whorls and fascicles with mitotic figures were <5/50 HPF [Figure 4]. Resection margins of bowel were free from tumor. CD117/c-kit showed positive expression on immunohistochemistry (IHC) examination. On the basis of HPE and IHC, the tumor was classified as intermediate-grade GIST.

The postoperative period was uneventful and patient discharged on the 7th postoperative day from hospital. On the basis of oncologist's opinion, imatinib mesylate 400 mg once daily was given as adjuvant chemotherapy. He has been on regular follow-up for 6 months without any symptoms of recurrence.

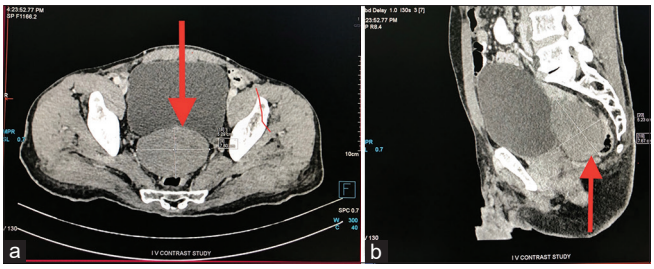

Figure 1: Contrast-enhanced computed tomography of the abdomen showed heterogeneously enhanced solid mass in the pelvis (a) axial and (b) sagittal view. Red arrow indicates tumor

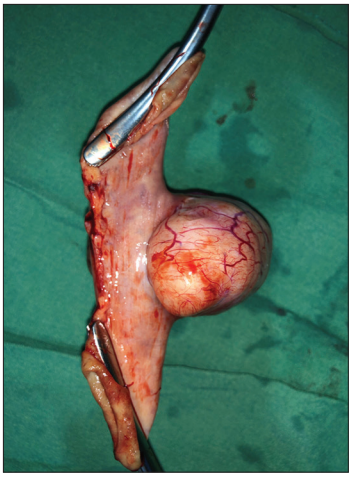

Figure 3: Resected specimen of segment of jejunum with tumor mass

Discussion

Small intestine tumors are rare. Only 1% of GI malignancies arise from the small bowel. GIST are smooth muscle tumors of GI tract and can be classified as leiomyomas, leiomyosarcomas, and high grade leiomyosarcomas.[3] The incidence of GIST is 2 in 100,000, considered as rare tumor. Jejunal GISTs are extremely rare and accounting for 0.1%–3% of all GI tumors.[4] Two-thirds of GIST present in the stomach while about one-fourth develop in the small intestine but rare in jejunum.

The clinical presentation of GIST depends on size, site of tumor, and tumor's relationship with surrounding tissue. Abdominal pain, abdominal mass, and bleeding are main presenting features of GIST. Other clinical features include abdominal distension and fullness, palpable abdominal mass, nausea, and vomiting. Approximately 20% case remain asymptomatic and detected incidentally.[5]

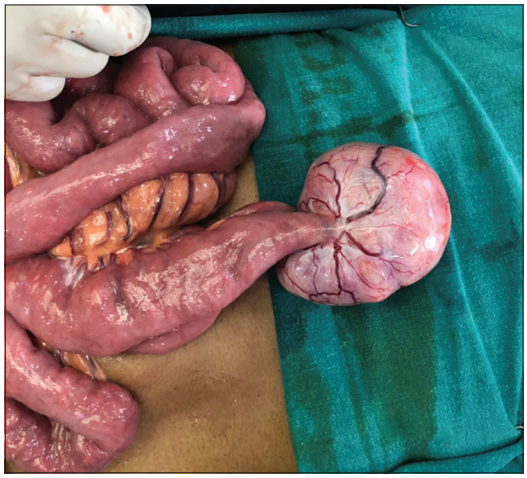

Figure 2: Intraoperative photograph showing an encapsulated tumor mass in the jejunum

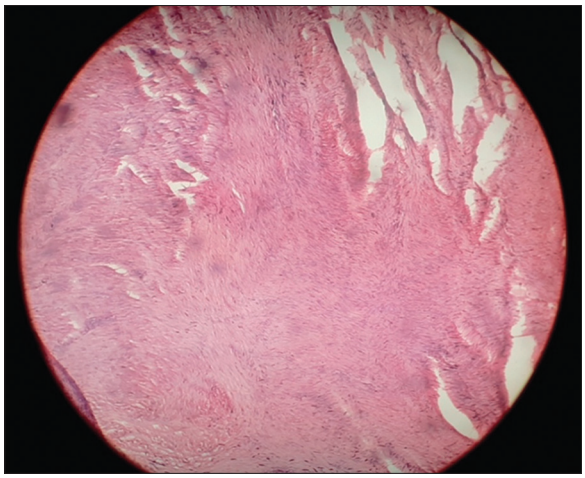

Figure 4: Microphotograph of tumor showing fibroblastic cells arranged in fascicles and whorls

The diagnosis of small bowel GIST is challenging for clinicians. Clinical history, endoscopic and radiological examinations can greatly helpful in the diagnosis. CECT is the prioritized tool to diagnose GISTs. CECT identifies the tumors and assess their range and metastasis to other organs.[5] CECT is considered better than magnetic resonance imaging (MRI) in showing, small bowel thickness and in assessment of deep loops of ileum and mesentery. CECT characteristics GISTs on the basis of tumor size, as follows: Small (<5 cm): Demarcated, homogenous masses with mainly intraluminal growth patterns. Intermediate (5–10 cm): Heterogeneous masses, irregular shape with intraluminal and extraluminal growth pattern. Large (>10 cm): Heterogeneous densities, irregular margins with local infiltration, distant and peritoneal metastases. In our case, CECT revealed a heterogeneous, solid mass measuring 5.3 cm × 7.7 cm in pelvis. MRI, endoscopic ultrasonography, F-Fluorodeoxyglucose positron emission tomography, interventional digital subtraction angiography are other diagnostic entities for GIST.[6]

According to histopathology, GISTs can be divided into spindle cells (70%), epithelioid cells (20%), and mixed subtypes (10%). IHC is always required for diagnosis confirmation. CD117, CD34, DOG-1, and S-100 markers are used in IHC. CD117 is the most common marker being used in the diagnosis of GIST. Histopathology and IHC, combination of both required to confirm the definite diagnosis.[7]

The mainstay of GIST treatment is complete surgical excision of tumor with negative margins. R0 resection with 1–2 cm clear margin is sufficient. Lymph node metastasis is rare, so lymphadenectomy is not routinely performed. Intra-operative injury of pseudocapsule of tumor can cause dissemination of tumor cells, so handling should be gentle.[8] Small bowel GISTs are more destructive and have malignant behavior as compared to gastric GISTs. Metastasis is common to the liver in most patients, while few patients have isolated local recurrence. Tumors >5 cm size and having mitotic index >10/50 HPFs, have high risk for malignancy (risk level IV category) and recurrence.[9]

Imatinib mesylate, a tyrosine kinase inhibitor targeting KIT, is being used in adjuvant therapy of GIST. It is used to prevent recurrence, in unresectable and metastatic disease cases. Sunitinib, a tyrosine kinase inhibitor is used in imatinib refractory GIST cases and in patients not tolerating imatinib. Sorafenib, dasatinib, nilotinib, and regorafenib are other newer drugs in the development for GIST treatment.[10]

Tumor size, anatomical site, histopathology, IHC, and molecular genetics are the main prognostic factors of GISTs. In follow-up, high risk patients require abdominal CECT or MRI evaluation every 3–6 months during adjuvant therapy for 5 years and low risk patients require scanning with computed tomography or MRI every 6–12 months for 5 years.[11]

Conclusion

Abdominal pain is the most common symptom of any abdominal disease. It can have varied etiologies, but GIST is a very rare cause of pain abdomen and pelvic mass. It is challenging for surgeons to diagnose preoperatively. Our case emphasizes on jejunal GIST as pelvic mass, a rare entity and cause of pain abdomen. HPE and IHC both are required to confirm the diagnosis of GISTs. CD117, a defining feature of GIST and almost all GISTs show strong positivity for it. Complete surgical excision (R0 resection) and adjuvant therapy with imatinib is the cornerstone of management for successful outcomes. Regular follow-up is necessary due to reoccurrence of tumor.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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