

Jejunojejunal Intussusception and Massive Lower Gastrointestinal Bleeding in a Young Patient: Rare Presentation of Jejunal Gastrointestinal Stromal Tumor. Case Report and Literature Review

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Abstract

Background: Gastrointestinal stromal tumors (GISTs) - which originate from interstitial cells of Cajal - are the most common mesenchymal neoplasms of the gastrointestinal tract in adults. The incidence of GISTs is estimated to be approximately 10 - 20 per million people. Around 90% of GISTs occurs in the age group of greater than 40 years old with male dominance. GISTs were found to be present in stomach (50 - 60%) followed by small intestine (30 - 40%). Most GISTs are clinically silent till they grow large, bleed, rupture, cause mechanical obstruction or act as a lead point for intussusception. In literature review, only few cases were reported that Jejunojejunal Intussusception occurred as a result of GISTs.

Case Presentation: A 28 years old male, presented with Jejunojejunal Intussusception and massive lower gastrointestinal bleeding that was managed successfully by surgical resection.

Conclusion: Jejunal GIST may present with lower GI bleeding or intussusception. By reviewing the literature, it was found to be rare to present with both presentations at the same time especially in young patients. To the best of our knowledge, this might be the first case to be reported with this presentation under the age of 40.

Keywords: *Jejunal-Jejunal; GIST; Intussusception; Lower GI Bleeding; Young Patient*

Abbreviation

GIST: Gastro-Intestinal-Stromal Tumor

Background

Gastrointestinal stromal tumors (GISTs) - which originate from interstitial cells of Cajal - are the most common mesenchymal neoplasms of the gastrointestinal tract in adults. The incidence of GISTs is estimated to be approximately 10 - 20 per million people. Around 90% of GISTs occurs in the age group of greater than 40 years old with male dominance. GISTs were found to be present in stomach (50 - 60%) followed by small intestine (30 - 40%). Most GISTs are clinically silent till they grow large, bleed, rupture, cause mechanical obstruction or act as a lead point for intussusception. In literature review, only few cases were reported that Jejunojejunal Intussusception occurred as a result of GISTs.

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Case Presentation

A 28 years old Saudi male, not known to have any medical illness presented to ER complaining of sever diffuse abdominal pain for 2 days duration which is colicky in nature, mainly in the left upper quadrant associated with melena and one episode of hematochezia. No previous episodes of the same complain was noticed, and no History of nausea, vomiting, hematemesis or weight loss has been identified. His past medical history was unremarkable apart from open appendectomy done many years ago. On examination, the patient was apparently pale with Blood Pressure 95/60 mmHg, Temperature 36.1°C, Pulse Rate 109 beat/minute, Respiratory Rate 22 breath/minute. His abdominal examination showed diffuse tenderness with fullness in the left upper quadrant, and digital rectal examination showed perfuse fresh blood. Laboratory data showed WBC 12.0 K/UL, hemoglobin 8.2 g/dl. After initial resuscitation and fluid replacement, Upper GI endoscopy was done with no evidence of ulcer of obvious source of bleeding. Urgent Computed tomography (CT) scan of the abdomen with intravenous contrast showed jejunal loop thickening with suspicious of focal enhancing lesion associated with intussusception.

In relation to the CT scan result, the patient was shifted to the operation room immediately (On June 20th, 2015); exploratory laparotomy was done and results showed proximal jejunal intussusception with multiple mesenteric small lymph nodes. After reduction of the intussusception, the jejunal loop containing the tumor which is located about 20 cm distal to tritz ligament was resected with grossly free margin followed by primary anastomosis.

The patient had an uncomplicated postoperative course and started on Gleevec and is planned to continue chemotherapy for 36 weeks with follow-up in oncology clinic.

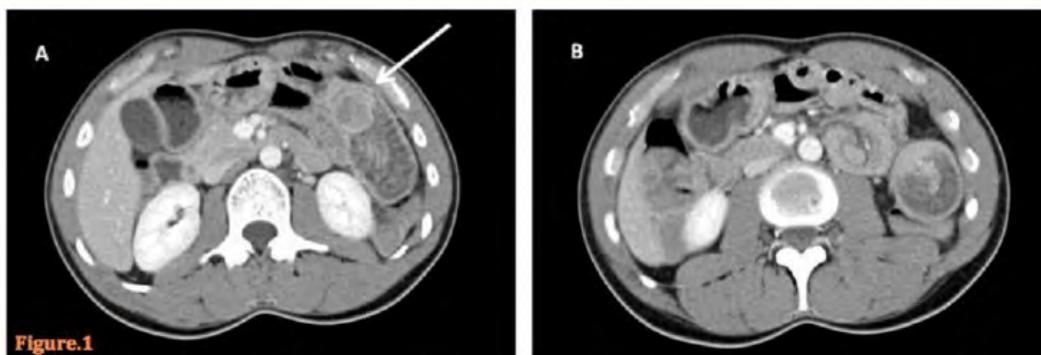


Figure 1: CT images of the mid abdomen with IV contrast shows a partially enhancing rounded mass at the apex of intussusception with a distended jejunal lumen proximal to the mass.

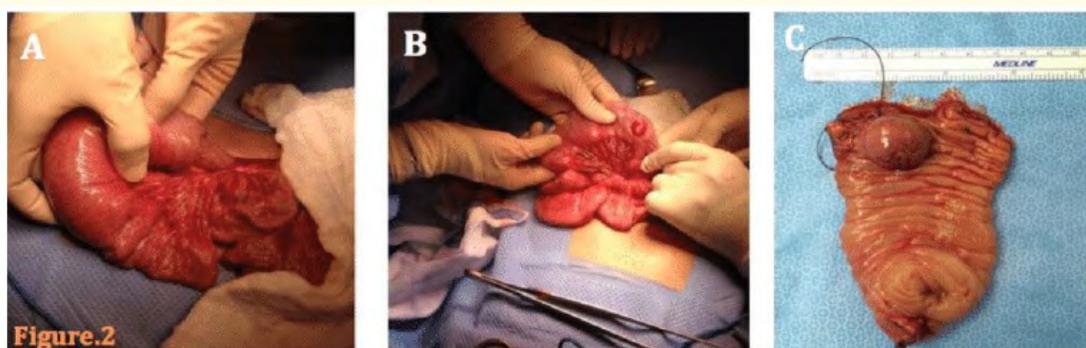


Figure 2: Laparotomy findings showing A. proximal jejunal intussusception. B. After reduction of the intussusception, the jejunal loop containing the tumor located about 20 cm distal to tritz. C. Gross specimen with free margin containing 3.0 X 2.4 X 2.3 cm jejunal GIST.

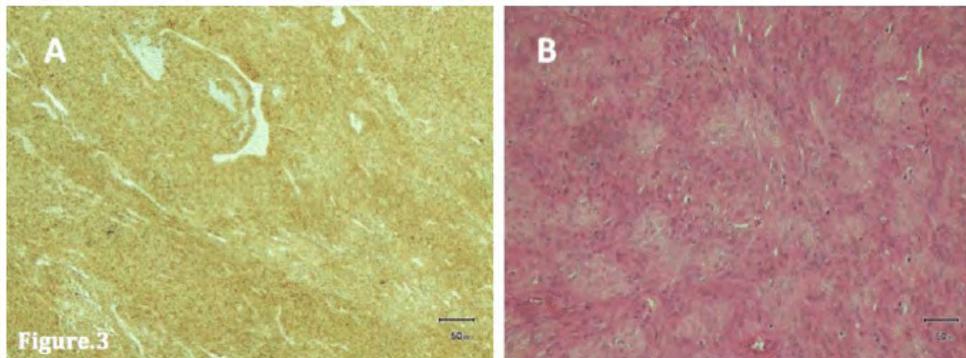


Figure 3: Histopathology findings showing A. Positive for Kit (CD117). B. Spindle cell proliferation with nuclear atypia, high mitosis, prominent nuclei and focal epithelioid differentiation.

Discussion

Gastrointestinal stromal tumors (GIST) represent the most common mesenchymal tumor of the gastrointestinal tract which arises from multipotential mesenchymal stem cells [1]. The term was first coined by Mazur and Clark in 1983 in order to describe a heterogeneous group of gastrointestinal non-epithelial neoplasms arising from the connective tissue of the digestive tract with an incidence level of 0.1 - 1% of gastrointestinal malignancies. In 1998, Hirota reported that GISTs contained activating c-kit mutations, which play a central role in its pathogenesis [2].

The incidence of GIST is estimated to be approximately 10 - 20 per million people [1]. Around 90% of GISTs occurs in age group of greater than 40 years with a Male dominance [2,3]. Only 2.7% of tumors occurred under the age of 21 years and 9.1% under the age of 40 years [4]. The most common location of GIST is stomach (50 - 60%) and small intestine (30% - 40%), (5% - 10%) arises from the colon and rectum, and 5% is located in the esophagus [1]. The percentages of GISTs found in other portions of GI tract are reported as 30% in jejunum and ileum, 5% in duodenum [5] the remaining may arise within the omentum or within the peritoneal layers (Extra-Gastrointestinal Stromal Tumors, EGISTs) [6].

The histological classification is divided based on the predominant cell type into three types, spindle cell, epithelioid cell or mixed cell type, with the spindle cell variant accounting for about 75% of GISTs [3]. The definitive diagnosis of the majority of GISTs is revealed by histopathological examination of the specimen. Approximately 95% of GISTs expresses CD117, which is part of the KIT receptor tyrosine kinase [7]. The expression of CD117 also helps to differentiate GISTs from other mesenchymal tumours of the GI tract, such as leiomyomas and leiomyosarcomas, and guides treatment selection [8]. In our case, the mass histopathology showed unifocal, spindle cell type and immunocytochemistry was CD117 positive.

Most GISTs are clinically silent till they grow large, bleed, rupture, cause mechanical obstruction or act as a lead point for intussusception [9]. One of the most common clinical manifestation for symptomatic GISTs is occult GI bleeding from mucosal ulceration [10]. Approximately 40% of GIST cases causes intestinal bleeding [7]. The hemorrhagic potential of these tumors might be contributed to a number of factors. First, location at the small bowel is associated with the highest incidence of bleeding. In which 64% of small-bowel GISTs present with bleeding compared with < 50% incidence of bleeding gastric, colonic and rectal tumors. Second, although extraluminal in origin, GIST may ulcerate through the overlying mucosa, causing intraluminal bleeding. Third, stromal collagen is minimal in most GISTs, but delicate, thin-walled vessels may be prominent, making stromal hemorrhage a common feature of these tumors [11-13].

Adult intussusception is a rare entity representing less than 1% of all intestinal obstructions. Diagnosis of the jejunojunal intussusception is difficult requiring a high index of suspicion and the utilization of imaging studies especially CT scans [11]. Only 2 case reports jejunojunal intussusception in the literature were reported in adult [10,13].

It is difficult to diagnose a jejunal GIST preoperatively due to the nonspecific and variable clinical symptoms, and it is also difficult to distinguish the tumor based solely on images [7].

Abdominal CT and MRI play an important role in the diagnosis of small intestinal tumors, especially larger size tumors, and facilitate the evaluation of their extent, fixed tumor structures, staging and the detection of abdominal metastasis. However, the appearance of GISTs on enhanced CT varies depending on the size, location and aggressiveness of the tumor. Tumors > 5 cm often appear heterogeneous because of necrosis, hemorrhage, and myxoid degeneration while smaller GISTs typically appear as well-defined soft-tissue and low-density masses that are relatively homogeneous on enhanced CT [12]. CT scan with contrast has been shown to be the most accurate diagnostic tool for the evaluation of intussusceptions [9]. In our case, (CT) of Abdomen showed jejunal loop thickening with suspicious of focal enhancing lesion associated with intussusception.

The mainstay of management for jejunal GIST is a complete surgical excision. The surgical imperative is a complete gross resection with an intact pseudo-capsule and negative microscopic margins [14].

In such kind of tumors, outcome is strongly dependent on tumor size, mitotic activity, tumor necrosis and a Ki-67 (MIB-1) index of > 10. GISTs can be categorized as low or high risk tumors by taking into account the possibility of metastasis or recurrence [15]. The incidence of metastasis in tumors > 10 cm and > 5 mitoses/50 HPFs is 86% compared with only 2 - 3% in tumors < 10 cm and < 5 mitoses/50 HPFs. However, tumors >10 cm with mitotic activity < 5/50 HPFs and those <5 cm with mitoses >5/50 HPFs had a relatively low metastatic rate (11% and 15%) However, the main prognostic factor is the mitotic count [4].

Conclusion

In conclusion, jejunal GIST is an uncommon mesenchymal tumor that rarely present in population under age of 30. It's extremely rare to present with intussusception with massive GI bleeding at the same time with high mitotic count histological feature. To the best of our knowledge, this might be the first case to be reported with these findings.

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