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Case report

Multiple primary GISTs: Unusual presentation as jejuno-jejunal intussusception

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Gastrointestinal stromal tumors (GISTs) are rare, occurring 10 - 15 per million, 0.2% of all gastrointestinal tumor, generally considered solitary tumor. This study presented a 57 year-old female with abdominal pain and intermittent vomiting for 6 months. Acute abdomen series didnot show any evidence of small bowel obstruction. Esophagogastroduodenoscopy (EGD) suggested partial small bowel obstruction. Contrast enhanced CT scan (CECT) of the abdomen demonstrated jejuno-jejunal intussusception induced by an intraluminal tumor and other two mesenteric tumors confirmed by histopathology and immunohistochemistry as GISTs. The patient had complete resection for the treatment. She recovered without complication and remained well. This study reported a rare case of multiple primary GISTs with unusual presentation as intussusception. CECT is able to detect tumors, its location, suggest diagnosis, define complication and metastasis.

Keywords: Multiple GISTs, jejuno-jejunal, CT, intussusception.

Gastrointestinal stromal tumors (GISTs) are rare in clinical conditions; globally, its incidence is 10 - 15 per million [1], representing 0.2% of all gastrointestinal tumors [2] , most of them commonly arise in the stomach.[2, 3] Generally, it is considered solitary tumor and the occurrence of multiple primary neoplasms and also regarded as exceptional event. [4] Intussusception and obstruction is also a very uncommon presentation of these lesions because of their tendency to grow in an extraluminal fashion. [5, 6]

This study reportes a rare case of multiple primary GISTs one of them causes jejuno-jejunal intussusception as a leading point, their CT findings and review literature.

Case report

A 57- year-old female presented with abdominal pain, intermittent vomiting for 6 months. She had persistent nausea and postpandrial vomiting for 2 months. She was diagnosed and treated as gastritis but the clinical didnot improve. No familial history of GISTs or neurofibromatosis Type1 (NF1) was identified. Physical examination showed palpable mass of 6 cm on the left paraumbilical region. Acute abdomen series showed no evidence of gut obstruction or definite mass. She underwent esophagogastroduodenoscopy (EGD) revealing that there was retained large amount of food and bile content suggestive of partial small bowel obstruction. Then CT scan of the upper abdomen (non-contrast and post contrast enhanced scans) was performed showed a bowel within bowel appearance on the left mid abdomen, a jejuno-jejunal intussusception containing a 4.0×4.0 cm^2 round enhancing mass with central necrosis at leading point, causing proximal small bowel obstruction.

A $5.0 \times 4.6 \times 4.4$ cm^3 mixed solid-cystic round mass with heterogeneous enhancement of the solid part was observed at the paramedian left upper abdomen. A $4.4 \times 4.1 \times 4.8$ cm^3 round- shaped, homogeneous enhancing mass was noted at left upper abdomen, anterior-inferior-medial to the aforementioned lesion, and just medial to the intussusception. No hepatic mass, peritoneal nodules, free fluid or enlarged lymph node was detected. (Figure 1, 2 and 3)

She underwent an exploratory laparotomy that identified a jejuno-jejunal intussusception causing by a 4.5 cm intraluminal jejunal tumor as a leading point. Another tumor, 5 cm diameter in the mesentery of the proximal jejunum was observed (Figure 4).

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Two mesenteric tumors were removed. Small bowel including mesentery resection around the intussusception with primary anastomosis was done.

Pathologic report showed three tumors: two jejunal mesenteric masses 5.0 cm in diameter each, and a 4.5 cm intraluminal jejunal mass. All tumors are confirmed by histopathology and immuno-histochemistry (positive CD 117) as GISTs with low mitotic figure (2 - 4/50 HPFs). She recovered well. Her follow up was without complication.

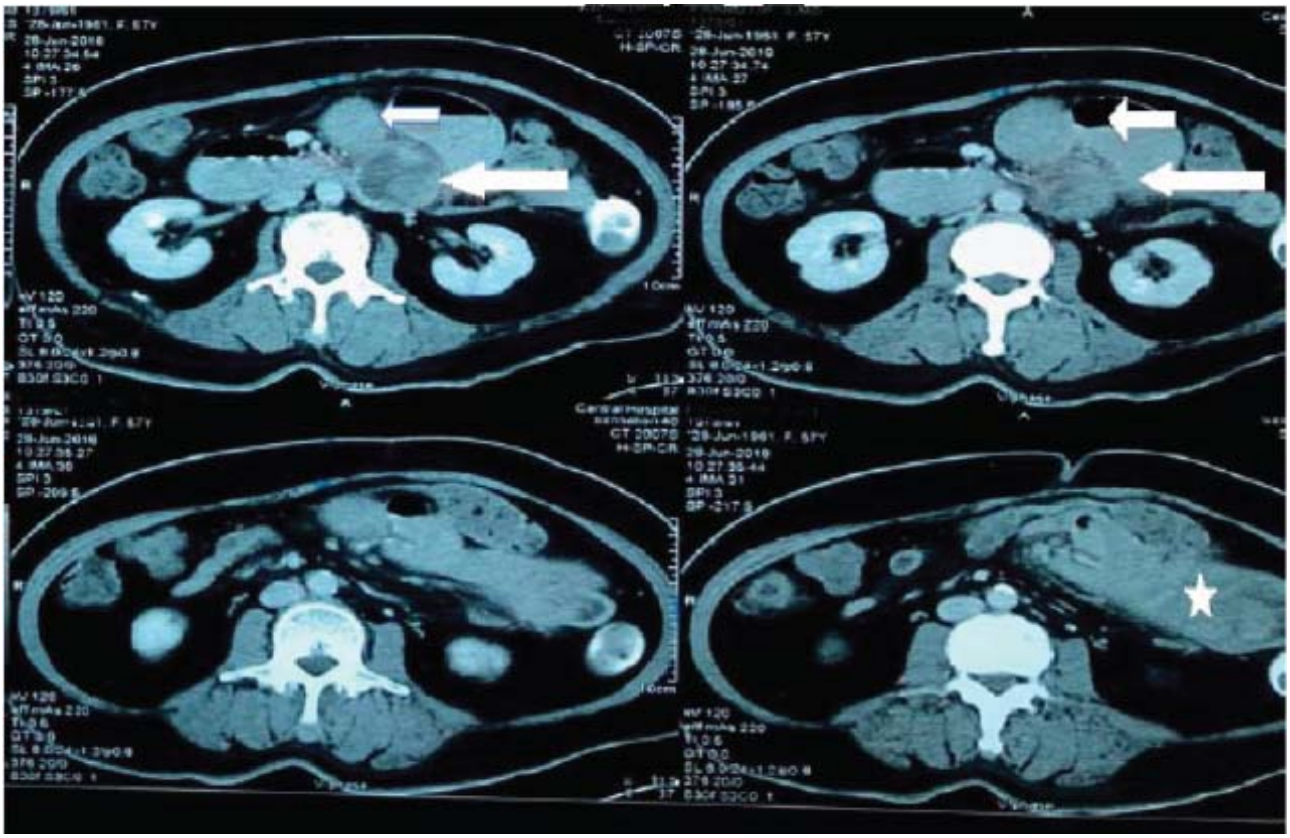

Figure 1. Axial CECT images: Two mesenteric GISTs: a 5.0 cm round solid-cystic one shows heterogeneous enhancement of the solid part and non-enhancing hypodensity area of cystic or necrotic component (long white arrows) and a 4.4 cm round homogeneous enhancing mass located anteriorly (short white arrows). A jejuno-jejunal intussusception was just inferior to mesenteric tumors. (star)

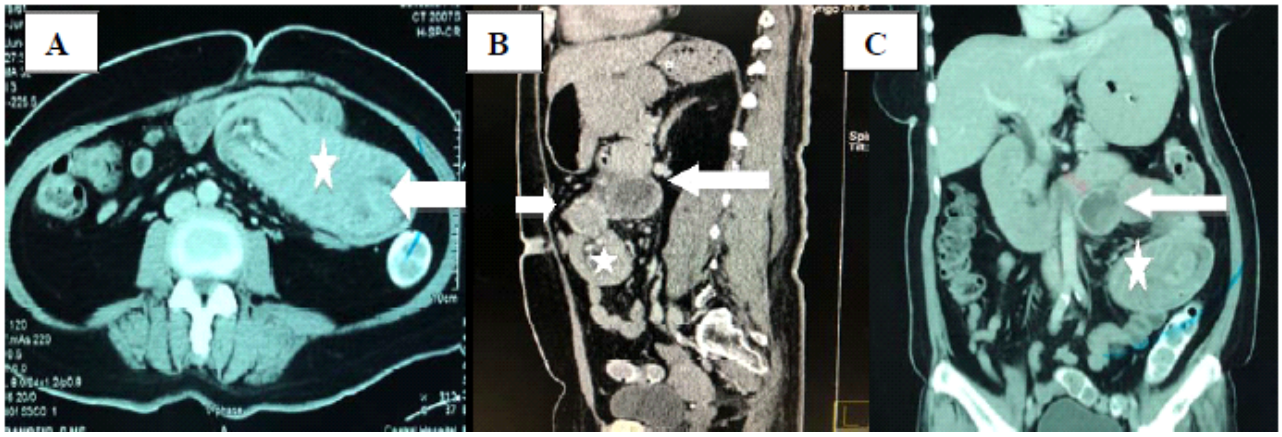

Figure 2. CECT: (A) Axial, (B) Sagittal, (C) Coronal images: A jejuno-jejunal intussusception with an intraluminal GIST as a leading point: A bowel within bowel appearance (stars) at left sided abdomen and a 4.0 cm round enhancing mass with central hypodensity area of necrosis or cystic degeneration as a leading point (white arrow in A) was seen. Other two mesenteric GISTs located just superior-medial to the intussusception (white arrows in B and C).

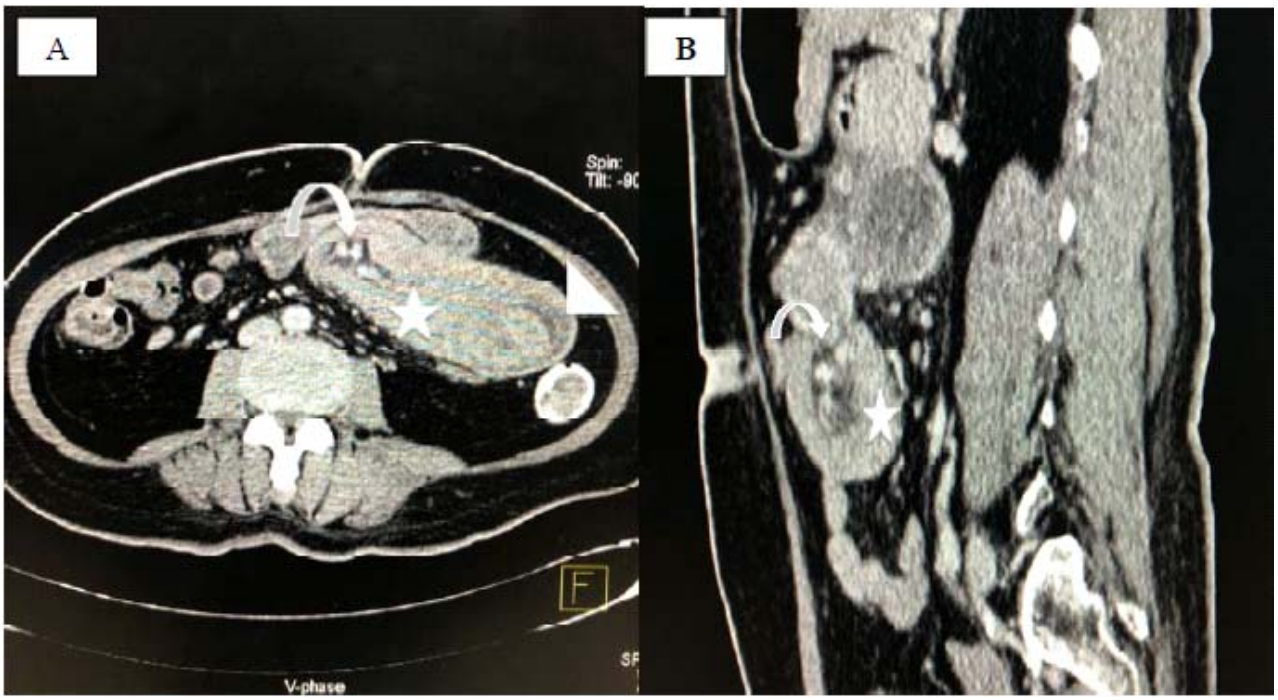

Figure 3. CECT: (A) Axial, (B) Sagittal images: A bowel within bowel appearance seen as a sausage-shaped mass in (A) (star) and atypical target-liked mass in (B) (star) showed hypodensity of mesenteric fat and mesenteric vessel (curve arrows) within thick- walled intussuscipiens (white arrowhead in A). Differentiation between intussusceptum and intussuscipiens was difficult due to bowel wall edema.

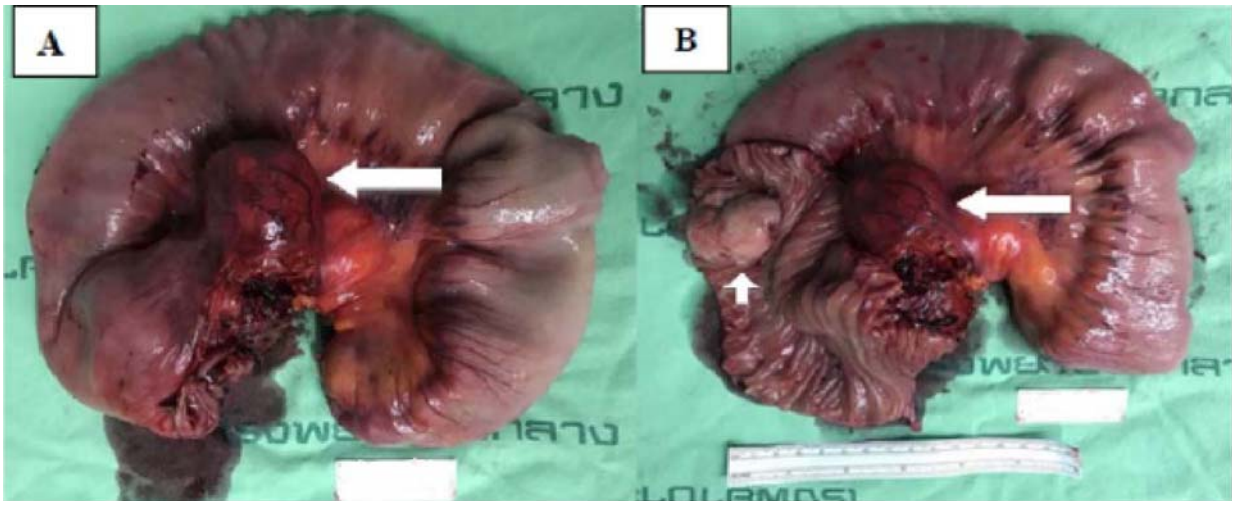

Figure 4. (A) A 5.0 cm GIST at mesentery of the proximal jejunum (long white arrows in A and B) was noted. (B) A 4.5 cm intraluminal jejunal GIST (short white arrows) was identified as a leading point of intussusception. The other mesenteric mass was not shown in these pictures.

Discussion

GISTs are mesenchymal tumors occurring anywhere along the gastrointestinal tract and are believed to originate from the interstitial cells of Cajal. [2, 3, 7] They generally arise as solitary tumors in the stomach (50 – 60 %), small bowel (25 – 30 %), colon and rectum (5 – 10 %), esophagus (5 %), and other localizations (<1 %). [1, 8]

They may occur primarily in the omentum, mesentery, and retroperitoneum. [7, 9] Multiple primary GISTS seem to be a less common scenario which may be found in different clinical contexts; sporadic, familial GISTs syndrome, as a component of Carney's triad, Carney-Stratakis syndrome, Von Recklinghausen disease or neurofibromatosis type I, and as metastatic disease in advanced malignant GISTs. [8, 10]

CT findings of GISTs: The enhancement pattern can vary from homogeneous enhancement to heterogeneous enhancement, with or without ulceration. GISTs was found to be well-defined tumors with homogenous enhancement [11] and also found as large heterogeneously enhancing masses due to areas of necrosis, hemorrhage or cystic degeneration. [7] Cystic changes are not seen commonly, but have been reported in rapidly growing primary GISTs. [12] There is no correlation between degree of necrosis, hemorrhage, cyst formation or contrast enhancement on CT imaging and malignant potential. [7] This reported case showed both enhancement patterns: homogeneous enhancing pattern of a mesenteric mass and heterogeneous enhancement with necrosis or cystic degeneration of the other two masses.

CT findings suggested that malignant GISTs are lesion with size larger than 5 cm, metastasis, invasion of adjacent organs [9,13] and irregular contour. [13]

Differentiation between multiple primary and metastatic GISTs are important due to the different treatment of each condition. Multiple sporadic GISTs are generally characterized by the presence of two or three lesions, at the same site or in different sites [8] as found in this case. Metastatic GISTs are found 50 - 65% at presentation. [14 - 16] The liver, mesentery and omentum are the most frequent sites of metastasis. [16] Liver metastasis is explained by hematogenous spread. [17] Mesenteric and peritoneal metastasis are shown as multiple masses diffusely involved peritoneal surface and mesentery which explained by tumor extending beyond the serosa and become serosal tumor seeding and peritoneal implants and often show accompanying liver metastasis. [16]

Differential diagnosis tumors of GIST include adenocarcinoma, lymphoma, peritoneal carcinomatosis, carcinoid, metastasis, and other mesenchymal neoplasm, e.g. leiomyoma. [18]

This case was shown to be sporadic primary three GISTs: one was intraluminal jejunal tumor and two jejunal mesenteric tumors without hepatic lesion, peritoneal seeding, familial history of GISTs or tumor of other organs.

GISTs usually involve the outer muscular layer, they have a propensity for exophytic growth. Therefore, the most common appearance is that of a mass arising from the intestinal wall and projecting into the abdominal cavity. [11, 19, 20]

Intussusception and obstruction are a very uncommon presentation of these lesions because of their tendency to grow in an extraluminal fashion. [5, 6] Intussusception in adult is rather uncommon, accounting for 5% of all cases of intussusceptions and 1% - 5% of bowel obstruction. [21] In contrast to childhood where intussusception is idiopathic in 90% of cases, in adults a pathologic lesion is found in more than 90%; neoplasms is considered to be the cause in 65% of the cases. A few cases of small bowel intussusceptions from stromal tumor in adults have been described in medical literature. [2, 3, 5, 6, 22, 23]

Most adult patients with intussusception present with chronic and nonspecific symptoms suggestive of intestinal obstruction. Abdominal pain is the most common symptom followed by nausea, vomiting, and a palpable abdominal mass. [21, 24] Preoperative diagnosis is often difficult because of nonspecific symptoms. [2, 3, 8] Intussusception is correctly and preoperatively diagnosed in only one-third of the cases. [2, 5]

Intussusception is the invagination of a bowel loop with its mesenteric fold (intussusceptum) into the lumen of a contiguous portion of bowel (intussuscipiens) as a result of peristalsis. [24]

CT findings of intussusception: the presence of a bowel within bowel configuration with or without mesenteric fat and mesenteric vessels is a pathognomonic character which can be seen as target-liked or sausage-shaped mass. The presence of a lead point, the configuration of the lead mass, the degree of bowel wall edema, and the amount of invaginated mesenteric fat all affect the appearance of an intussusception. [24] Intussusception of this case was shown to be a bowel within bowel appearance but differentiation between intussusceptum and intussuscipiens was difficult due to bowel wall edema so typical multilayer of target sign was not identified; however sausage-shaped mass and atypical target-liked mass containing mesenteric fat and mesenteric vessel were evident which were pathognomonic characters of this condition. (Figure 2 and 3)

This case presented with abdominal pain, vomiting and abdominal mass and CT showed bowel within bowel appearance, also indicated the cause of intussusception as a round necrotic mass at the leading point and incidental findings of other two mesenteric tumors. (Figure 1 and 2)

Taking all of these characteristics, the preoperative CT scan performed in this patient yielded an excellent information which able to detect lesions, locate its site, suggest diagnosis, extension of disease, complication, metastasis and well correlated with the surgical findings.

Tumor size, tumor site and mitotic index are the main variables used in risk stratification systems. As this case tumor size was not larger than 5 cm, tumor located at small bowel and mitotic index was lesser than 5 per HPFs, according to risk stratification system developed by National Institute of Health (NIH), also called as Fletcher's criteria [25], this case belonged to low risk category.

Surgery is the primary therapeutic option with the goal being complete resection for nonmetastatic GISTs. Lymph node metastases are rare and routine removal of lymph nodes is typically not necessary.[26] This case underwent surgery, three GISTs, jejunal segment and its mesentery were removed with free margin. The treatment was complete by surgery without additional imatinib due to the low risk of these tumors.

In cases of high risk category who need imatinib for adjuvant therapy, CT can evaluate response of treatment as well. [27]

Conclusion

GISTs are subset of mesenchymal tumors occurring along GI tract and originating from interstitial cell of Cajal, usually considered single tumor. Multiple primary GISTs is less common. Clinical presentation of GISTs are not specific. Intussusception as presentation of GISTs is infrequent. Contrast enhanced CT scan of abdomen is able to detect tumors, its location, suggest diagnosis, define complication, metastasis which useful for design primary and adjuvant therapy and also evaluate response of treatment.

This study reported multiple primary GISTs with intussusception as an unusual presentation of GISTs.

Conflict of interest

The author has no potential conflict of interest to disclose.

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