

A CASE REPORT OF PEDUNCULATED EXTRALUMINAL GASTROINTESTINAL STROMAL TUMOR (GIST)

Le Ngoc Tu Tran, Le Ngoc Quy, Nguyen Anh Tuan, Dang Nhu Thanh, Hoang Minh Loi

SUMMARY

Gastrointestinal stromal tumors (GISTs) comprise a group of smooth muscle mesenchymal alimentary tract tumors of variable malignancy (1) GISTs may occur anywhere along the gastrointestinal tract, they are most common located in the stomach (50-60%) and the small intestine (30-35%), and less frequently in the colon, rectum and esophagus (2). More rarely, GISTs may arise from other intra-abdominal soft tissues within the abdominal cavity, usually in the omentum, mesentery or the retroperitoneum called extra-Gastrointestinal stromal tumors (EGIST). Three macroscopic growth patterns of GIST: intraluminal, exophytic (extraluminal), and transmural tumors with the distribution was even (3).

In this study, we describe case report of 70-year-old man was admitted to our hospital because of abdominal pain over 6 months with no pathological antecedent. Abdominal ultrasound (US) reveal a omental solid mass in left lumbar region, and on CT scan show a solid mass in left lumbar region in front of the descending colon, measuring around 50x87x133mm with well-circumscribed, slight enhance with intravenous contrast, no hemorrhage or necrosis area, no calcification and not clearly separated from small-bowel. Patient has laparoscopic surgeon, diagnosis was a pedunculated extraluminal gastrointestinal stromal tumor (GIST). Histopathological examination report and the CD-117 immunoreactive score is suitable of endoscopically invisible medium sized.

Keyword: *Pedunculated Extraluminal Gastrointestinal Stromal Tumor (GIST), Ultrasound and CT scan, Laparoscopic surgeon, CD-117 immunoreactive score.*

INTRODUCTION

The stomach is divided into the cardia, fundus, body, antrum, and pylorus. According to the World Health Organization, neoplasms of the stomach are classified into two large categories on the basis of the cell of origin: epithelial and nonepithelial (4).

- Epithelial neoplasms arise from the mucosa and account for the majority of gastric tumors, ranging from benign hyperplastic and adenomatous polyps to malignant adenocarcinomas.
- In contrast, Nonepithelial tumors arise deep to the mucosa, that is, from the submucosa, muscularis propria, or serosa. Nonepithelial lesions are commonly referred to as "submucosal" or "intramural". GIST is the most common of nonepithelial tumors.

Clinical manifestations of GISTs depend on their size and location. Small GISTs are usually discovered incidentally. Ulcerated lesions can manifest with symptoms of gastrointestinal bleeding, including hematemesis, melena, and iron-deficiency anemia (12). Patients with larger tumors can present with abdominal pain and early satiety. However, because of the exophytic growth pattern of these tumors, some patients remain asymptomatic until the tumor has become quite large. Bowel obstruction is rare.

GISTs are assessed for **risk of progressive** disease on the basis of their mitotic rate, size, and location (13, 14). Origin in the stomach is a favorable prognostic factor, and gastric GISTs less than 2 cm in size may have no or extremely low malignant potential (10,11).

Radiologic appearances of GISTs can vary widely depending on tumor size (16). Because they arise from

Departments of Radiology, Gastrointestinal surgery Department, Hue Central Hospital.

the deep muscularis propria, GISTs frequently have an exophytic or intramural pattern of growth; endoluminal growth is less common. Endoluminal tumors have classic features of intramural masses, being smoothly circumscribed with margins that form obtuse or right angles with the gastric wall. About 50% of lesions larger than 2 cm develop focal ulceration of the overlying mucosa because of pressure necrosis (17), a radiologic feature referred to as the bull's-eye sign. As they enlarge, exophytic GISTs may invade adjacent structures such as the pancreas, colon, or diaphragm.

Areas of hemorrhage, necrosis, or cystic degeneration are common, appearing as focal areas of low attenuation. Extensive hemorrhage or necrosis may result in formation of a cavity that communicates with the gastric lumen. In rare cases, clumps of calcifications are seen. Nearly half of patients present with metastatic disease, most commonly to the liver and peritoneum. Lymph node metastasis is infrequent (8).

Differential diagnosis of GISTs includes schwannomas, true leiomyomas, and solitary (type 1) carcinoid tumors, particularly for smaller lesions. Occasionally, gastric adenocarcinoma or lymphoma may demonstrate intramural growth and mimic a GIST. However, advanced gastric carcinomas and lymphomas are usually associated with bulky perigastric or celiac lymphadenopathy, which is rare for GISTs. (5)

Histologic characteristics of GISTs usually consist of interlacing whorls of spindle-shaped cells with eosinophilic cytoplasm and elongated nuclei, although

tumors completely composed of ovoid cells are not uncommon. Immunoreactivity to c-KIT or DOG1 confirms the diagnosis (7,9).

CASE RERORT

A 70-year-old man was admitted to our hospital because of abdominal pain over 6 months with no pathological antecedent.

Barium imaging and endoscopically is normal. Abdominal ultrasound (US) describes an omental solid mass in the left lumbar region, measuring around 5cmx10cm with well-circumscribed.

CT scan shows a solid mass in the left lumbar region in front of the descending colon, measuring around 50x87x133mm with well-circumscribed, slightly enhance with intravenous contrast, no hemorrhage or necrosis area, no calcification, and not clearly separated from small-bowel. Initially, Radiologist omission a pedicle, and this case is misdiagnosis omental tumor.

The mass was excised using a laparoscopic procedure. The tumor was removed along with the gastric wall where the tumor was attached to by a pedicle. The external surface of the tumor showed a well-encapsulated appearance, clearly separate with small-bowel and the omental.

The final diagnosis was a pedunculated exoluminal gastrointestinal stromal tumor (GIST). The histopathological examination report and the CD-117 immunoreactive score are suitable.



Fig 1. Axial CT image show a well-circumscribed mass in front of the descending colon

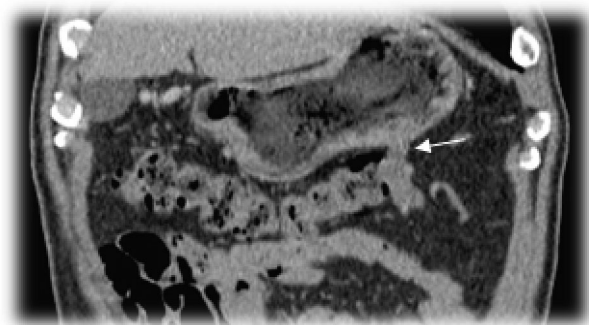


Fig 2. Coronal CT image show a pedicle (arrow): the tumor appeared to be connected with the wall of the gastric body by a pedicle.



Fig 3. Sagittal CT image show a pedicle (arrow) connect the tumor with the gastric.

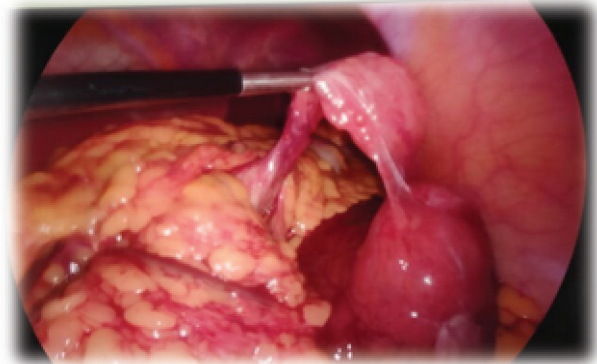


Fig 4. Laparoscopic show the pedicle clearly.

DISCUSSION

GISTs arise from the interstitial cells of Cajal, which are the most common nonepithelial tumors of the gastrointestinal tract. It is normally present in the myenteric plexus and is clearly distinct from other mesenchymal tumors, such as leiomyomas or leiomyosarcomas. (6)

Mesenchymal tumors are often well-circumscribed, with intact overlying mucosa. The CT features of GISTs vary greatly, depending on the size and aggressiveness of the tumor and the time of presentation during the course of the disease. Primary GISTs are typically large, hypervascular, enhancing masses on contrast-enhanced CT scans and are often heterogeneous because of necrosis, hemorrhage, or cystic degeneration at the time of presentation (16, 17). Ulceration and fistulization to the gastrointestinal lumen are also common features of GISTs (16, 17). Often, tumor vessels can be seen within the tumors. The masses usually displace adjacent organs and vessels, but the direct invasion of the adjacent structures is sometimes seen with advanced disease. It can be difficult to identify the origin of the mass because of its large size and prominent extraluminal location. Bowel

obstruction is rare.

Small GISTs can be endoluminal and polypoid in appearance. Small, localized GISTs are usually homogeneous and may be an incidental finding at CT or endoscopy. Visibility with endoscopy is limited to smaller tumors, and results of endoscopic biopsy often can be nondiagnostic primarily because of insufficient tissue collection (10,11). Therefore, CT should be performed to follow up on suspected GISTs identified with endoscopy.

Our patient has a tumor with, no hemorrhage or necrosis area. it is explained because the overlying mucosa is not ulcerated. That is demonstrated by barium imaging and endoscopically.

These tumors can grow in an endoluminal, exophytic, or mixed (dumbbell-shaped) pattern. Smaller masses and lesions arising in the submucosa often protrude into the lumen of the stomach. Larger masses and tumors that arise from the deep muscularis propria, such as GISTs, often demonstrate an exophytic pattern of growth, toward the peritoneal cavity.

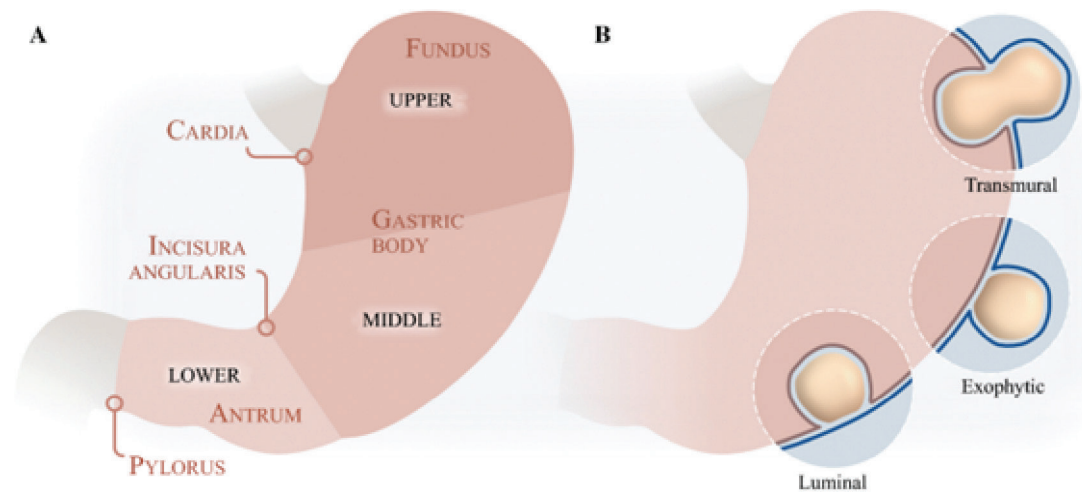


Fig 5. A. The partition of the stomach into the upper, middle, and lower thirds. B. Macroscopic growth pattern: luminal, exophytic, and transmural (mixed) tumors.

Shoko's report summarizes 12 reported cases of adult extraluminally pedunculated gastric GISTs. These tumors tended to be large (the median size was 12 cm), but the case report have characteristic of GISTs like a mixture of solid and cystic areas, calcification, hemorrhage, and necrosis. (15)

In our case report, the GISTs grow exophytic with a long pedicle, nonspecific with no calcification, no hemorrhage, and no necrosis. It can be mimic with the omental tumor in ultrasound and CT images.

Because of these clinical symptoms, the normal report of the ultrasound and barium image, the nonspecific of CT scans and the exophytic growth of the tumors, GISTs are often not detected until late in their progression. Histopathologic analysis, including immunohistochemistry is usually required for diagnosis.

CONCLUSION

In the exophytic growth pattern of these tumors, some patients remain asymptomatic until the tumor has become quite large. The tumor with a pedicle sometimes made the radiologist mistake with Intra-abdomen soft tissues from the omentum, mesentery, or the retroperitoneum..

On the other hand, the characteristics of GISTs have many things the same as other mesenchymal tumors. Differentiating between various gastric intramural tumors may be challenging because these lesions often have overlapping radiologic appearances (5). Attention to a combination of features, such as tumor margin, location, growth pattern, attenuation, and enhancement, may help one suggest a particular diagnosis.

REFERENCES

1. D Michael King . The radiology of gastrointestinal stromal tumours (GIST)
2. Miettinen M, Lasota J. Gastrointestinal stromal tumors (GISTs): definition, occurrence, pathology, differential diagnosis and molecular genetics. *Pol J Pathol* 2003; 54:3–24.
3. Sandrasegaran K, Rajesh A, Rushing DA, Rydberg J, Akisik FM, Henley JD. Gastrointestinal stromal tumors: CT and MRI findings. *Eur Radiol.* 2005 Jul; 15(7):1407-14.
4. Hamilton SR, Aaltonen LA. World Health Organization classification of tumors: pathology and genetics of tumours of the digestive system. Lyon, France: IARC, 2000. Google Scholar

5. Hyunseon C. Kang, MD, PhD, corresponding author Christine O. Menias, MD, Ayman H. Gaballah, MD, FRCR, Stuti Shroff, MD, PhD, Melissa W. Taggart, MD, Naveen Garg, MD, and Khaled M. Elsayes, MD. Beyond the GIST: Mesenchymal Tumors of the Stomach. Received 2013 Feb 18; Revisions requested 2013 Mar 20; Revised 2013 Apr 10; Accepted 2013 Apr 15.
6. Nishida T, Hirota S. Biological and clinical review of stromal tumors in the gastrointestinal tract. *Histol Histopathol* 2000;15:1293–1301. 4. DeMatteo RP.
7. Sarlomo-Rikala M, Kovatich AJ, Barusevicius A, Miettinen M. CD117: a sensitive marker for gastrointestinal stromal tumors that is more specific than CD34. *Mod Pathol* 1998;11(8):728–734. Medline, Google Scholar
8. DeMatteo RP, Lewis JJ, Leung D, Mudan SS, Woodruff JM, Brennan MF. Two hundred gastrointestinal stromal tumors: recurrence patterns and prognostic factors for survival. *Ann Surg* 2000;231(1):51–58
9. Espinosa I, Lee CH, Kim MK et al.. A novel monoclonal antibody against DOG1 is a sensitive and specific marker for gastrointestinal stromal tumors. *Am J Surg Pathol* 2008;32(2):210–218. Crossref, Medline, Google Scholar
10. Agaimy A. Gastrointestinal stromal tumors (GIST) from risk stratification systems to the new TNM proposal: more questions than answers? A review emphasizing the need for a standardized GIST reporting. *Int J Clin Exp Pathol* 2010;3(5):461–471
11. Miettinen M, Lasota J. Gastrointestinal stromal tumors: definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. *Virchows Arch* 2001;438(1):1–12
12. Levy AD, Remotti HE, Thompson WM, Sobin LH, Miettinen M. Gastrointestinal stromal tumors: radiologic features with pathologic correlation. *RadioGraphics* 2003;23(2):283–304
13. Franquemont DW. Differentiation and risk assessment of gastrointestinal stromal tumors. *Am J Clin Pathol* 1995;103(1)
14. Fletcher CD, Berman JJ, Corless C, et al. Diagnosis of gastrointestinal stromal tumors: a consensus approach. *Hum Pathol* 2002;33(5):459–465
15. Shoko O, Masayoshi N, Takaya N, Tamaki M, Seiichi H. A rare case of extraluminally pedunculated gastrointestinal stromal tumor with postoperative metastasis to pancreas. *rjab42*.

Correspondent: Le Ngoc Tu Tran. Email: lengoctutran@gmail.com

Received: 15/10/2021. Assessed: 21/10/2021. Reviewed: 26/10/2021. Accepted: 15/11/2021