Gastrointestinal stromal tumor: Typical and atypical manifestations

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Authors: S.-W. Jang, H. J. Kim; Seoul/KR
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Learning objectives

Identify typical and atypical radiologic manifestations of gastrointestinal stromal tumors (GISTs).

Discuss useful imaging findings for differentiation of GISTs from other tumors and making a correct diagnosis.
Background

GISTs are the most common mesenchymal tumors of the gastrointestinal (GI) tract. They are defined as KIT (CD117, stem cell factor receptor)-positive mesenchymal spindle cell or epithelioid neoplasms.

Most individuals are over 50 years of age at the time of presentation.

Presenting signs, symptoms and radiologic features depend on the size and anatomic location of the tumor. As a result, GISTs have a wide spectrum of radiologic appearances.

Surgical resection is the treatment of choice for potentially resectable tumors. For patients with completely resected primary high-risk GISTs or advanced unresectable GISTs, tyrosine kinase inhibitor therapy can be effective.
Typical manifestation

Location

GISTs occur mostly in the stomach and small bowel, but can occur anywhere along the gastrointestinal tract and rarely in the omentum, mesentery, and retroperitoneum.

Imaging features

Typically GISTs seem to be well-defined extraluminal or intramural masses, with varying attenuation based on size (Fig. 1,2, 3). Small tumors tend to appear homogeneous. The larger tumors frequently show central areas of necrosis or hemorrhage (Fig. 4,5,6,7,8). This description applies to GISTs in all locations. Nevertheless, there are features of GISTs that may be unique to location.

Gastric GISTs commonly demonstrate extension into the gastrohepatic ligament, gastroepiploic ligament, and lesser sac; frequently, the bulk of the tumor is seen in an extragastric location. Cavities that develop from central necrosis or hemorrhage in the larger tumors may communicate with the gastric lumen and thus contain fluid, air, or contrast material.

Most gastric GISTs are located in the gastric body, and rarely located in the antrum. This may help to distinguish gastric GISTs from solitary gastric carcinoids, which are most commonly seen in the antrum.

Small intestinal GISTs may appear as intramural masses or intraluminal polyps and may show extension into adjacent mesentery. Encasement of adjacent small bowel, colon, and bladder can be seen. The appearance is different from small bowel adenocarcinoma, which typically appears as an annular lesion in the proximal small intestine.

Anorectal GISTs most commonly present as a well-defined, eccentric, mural masses that expand the rectal wall and may contain mucosal ulceration (Fig. 9,10,11,12). The mass spreads via extension into the ischiorectal fossa, prostate, or vagina. Intraluminal polypoid masses are rarely seen in anorectal GISTs, but are more common in leiomyosarcoma.
Colonic GISTs are described as transmural tumors that involve the intraluminal and extraserosal surfaces of the colon. Colonic GISTs have been seen to exhibit circumferential growth and secondary dilatation of the affected bowel lumen. Rarely, large colonic GISTs may communicate with the colonic lumen due to central necrosis (Fig. 13,14,15,16,17).

Metastasis

Most GISTs (70%-80%) are benign. There is, however, a continuum from benign to malignant that can be predicted, although not absolutely, according to tumor size and mitotic frequency.

The most common site for GIST metastasis is the liver (Fig. 18,19,20) followed by the peritoneum. Lymph node or lung metastasis is very rare.

Differential diagnosis

The differentiation from other primary gastrointestinal malignancies can often be made on the basis of these specific findings, since lymphomas tend to cause circumferential mural thickening with homogeneous enhancement and/or lymph node enlargement. Carcinoids are mainly found in the terminal ileum and can be seen a desmoplastic reaction, while carcinomas are likely to demonstrate local infiltration and visceral obstruction, especially if large. Metastases to the bowel are typically multifocal, unlike GISTs, and often come with an appropriate history of primary malignancy

Atypical manifestation

Calcification (Fig. 21,22,23)

Calcifications are rare manifestation of gastrointestinal tumors. Calcifications in a GIST are also rare, and present in 0~10 % of GISTs. It may occur in a mottled pattern or be present extensively throughout the tumor.

In one study, GISTs containing calcifications were seen only in intermediate and high risk tumors.

Other gastrointestinal tumors that may contain calcifications include hemangiomas, calcified gastric carcinomas, glomus tumors and lymphomas. Calcifications in hamangioma typically manifest as clusters of phleboliths in the gastrointestinal wall, and in gastric carcinoma, especially mucinous adenocarcinoma, they typically manifest as military and punctuate calcifications. Calcifications in lymphoma are nearly always associated with previous treatment.
Multifocal lesion (Fig. 24,25,26)

Most GISTs occur sporadically and multiplicity is very rare. Multifocal GISTs usually arise in specific settings like in patients with KIT or PDGFRA germline mutations and in patients with NF-1. These patients usually develop tumors in the small intestine.

Multifocal GISTs may be misinterpreted as recurrent or metastatic disease, leading to inappropriate treatment.

Associated with neurofibromatosis type-1 (Fig. 27,28,29)

Neurofibromatosis 1 (von Reckling-hausen’s disease) is an autosomal-dominant genetic disorder with mutation of the NF1 gene. The hallmarks of NF1 are abnormal skin pigmentation (café au lait spots and axillary freckling), cutaneous and plexiform neurofibromas, skeletal dysplasias, and Lisch nodules (pigmented iris hamartomas).

The genetic abnormality in neurofibromatosis has been localized to the neurofibromatosis 1 gene on chromosome 17, which is a tumor suppressor gene in some cell types. Mutations in the NF1 gene result in loss of functional protein, causing the wide spectrum of clinical findings, including NF1-associated tumors. Abnormal tumor suppression increases the incidence of benign and malignant neoplasia in children and adults with neurofibromatosis 1.

GISTs in patients with neurofibromatosis 1 are histologically and immunophenotypically identical to those in patients without neurofibromatosis 1. But, as previously mentioned, GISTs that occur in patients with neurofibromatosis are often multiple and commonly originate from the proximal small intestine.

Pedunculation (Fig. 30)

Pedunculated GISTs have been reported by several authors. They usually present in exophytic masses of stomach.

Pedunculated GISTs have high risk of malignancy due to its large size and high number of mitoses even if they use to have only expansive growth, without infiltration of surrounding structures.

Intussusception (Fig. 31,32)

Intussusception is a condition that a portion of gastrointestinal tract into the lumen of the immediate adjacent intestine, leading to intestinal obstruction in most cases. Neoplasm is the most common cause and is found in approximately 65% of adult intussusceptions.
GISTs are usually asymptomatic, but they may present with bleeding, obstruction or perforation in some cases. Intussusception is a very rare complication of GISTs.

GISTs may be complicated with gastroduodenal, intestinal and colonic intussusceptions.

Formation of ulcer (Fig. 33,34,35)

Mucosal ulceration is seen on the luminal surface of the tumor in up to 50% of cases in which patients present with symptoms and signs associated with gastrointestinal bleeding.

Large ulceration of GISTs may mimic other tumors such as carcinoma, lymphoma, and metastasis. Ulcers are usually associated with relatively large tumors which accompany a poor prognosis.

Intraperitoneal rupture with hemoperitoneum (Fig. 36,37)

Symptomatic GISTs present most often with gastrointestinal bleeding. The majority of bleeding occurs in the central ulceration, but rarely due to angiodysplasia of GISTs.

We should consider the rupture when GISTs present with ascites in an appropriated clinical context.
Fig. 0: A 61-year old female with incidentally detected gastric GIST. Axial contrast-enhanced CT image shows a poorly enhancing mass (arrow) abutting to the stomach with intact gastric mucosa.

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**Fig. 0:** Coronal contrast-enhanced CT image shows that the mass (arrow) originates from lesser curvature of the gastric angle

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**Fig. 0:** Photograph from resected specimen shows the mass with necrosis and hemorrhage.

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Fig. 0: A 50-year old man with a jejunal GIST. Axial CT scan during arterial phase shows an exophytic growing enhancing mass (arrow) in the jejunum.

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Fig. 0: Coronal CT scans during arterial (Fig.5) and portal phase (Fig.6) show the enhancing mass (arrow) and extravasation of the contrast in the jejunal lumen.

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**Fig. 0:** Coronal CT scans during arterial (Fig.5) and portal phase (Fig.6) show the enhancing mass (arrow) and extravasation of the contrast in the jejunal lumen.

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**Fig. 0:** DSA shows a hypervascular tumor staining (arrow) originating in the jejunal branches of the superior mesenteric artery.

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Fig. 0: Photograph from resected specimen shows the mass with hemorrhage.

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**Fig. 0:** Contrast enhanced CT scan (Fig.9) shows a 6.5 cm submucosal mass (arrow) with large central necrosis in the left lateral wall of the distal rectum. Necrosis within mass (arrow) appears low-signal-intensity on T1-weighted image (Fig.10), and high-signal-intensity on T2-weighted image (Fig.11).

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**Fig. 0:** Coronal T2-weighted image shows that the well-demarcated mass (arrow) originates from the distal rectum.

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Fig. 0: A 69-year-old female with a colonic GIST. Contrast enhanced CT shows a poorly enhancing mass (white arrow) with central necrosis (black arrow) originates from the descending colon (red arrow) and this mass is abutting to the stomach (blue arrow).

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Fig. 0: Coronal enhanced CT shows fistula tract (black arrow) between the necrotic mass and the descending colon.

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Fig. 0: Fistula tract (white arrow) is observed between the mass and the descending colon.

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Fig. 0: Small bowel follow-through demonstrates that fistula tract (black arrow) between the necrotic space of GIST (red arrows) and the descending colon (white arrow).

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**Fig. 0:** Photograph from resected specimen shows the necrotic mass (white arrow) and fistula tract (black arrow) between the mass and descending colon.

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Fig. 0: A 50-year old male with a gastric GIST. Coronal contrast-enhanced CT image shows a heterogeneously enhancing mass with central deep ulceration (arrow) along lesser curvature of the stomach.

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Fig. 0: Axial contrast-enhanced CT image shows a poorly enhancing nodule (arrow) in the hepatic segment VIII, suggesting hepatic metastasis.

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Fig. 0: Photograph from resected specimen shows the huge mass with deep central ulceration (arrow).

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Fig. 0: A 37-year old female with a gastric GIST. Contrast enhanced CT scan shows a well-demarcated mass (arrow) with calcifications in the gastric cardia.

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Fig. 0: Coronal reconstructed CT scan shows the mass (arrow) with multiple punctuated calcifications.

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Fig. 0: Photograph from the resected specimen shows the fragmented mass with calcifications.

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Fig. 0: A 56-year old woman with incidentally detected multiple GISTs in the small intestine. Enhanced CT scans show three homogenously enhancing masses (arrow) in the duodenum (Fig. 24,25) and proximal jejunum (Fig. 26).

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**Fig. 0:** Enhanced CT scans show three homogenously enhancing masses (arrow) in the duodenum (Fig. 24,25) and proximal jejunum (Fig. 26).

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Fig. 0: Enhanced CT scans show three homogenously enhancing masses (arrow) in the duodenum (Fig. 24,25) and proximal jejunum (Fig. 26).

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Fig. 0: A 63-year old male with neurofibromatosis type-1 and an ileal GIST. Contrast enhanced CT shows a well-defined, subcutaneous nodule (arrow) in the left anterior chest wall, suggesting neurofibroma.

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**Fig. 0**: Contrast enhanced CT shows a well-defined, homogeneously enhancing mass in the distal ileum (arrow).

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**Fig. 0:** Coronal reconstructed CT scan shows the ovoid shaped mass (arrow) in the distal ileum.

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**Fig. 0:** A 67-year old male with a gastric GIST. Coronal reconstructed CT scan shows a heterogeneously enhancing mass (white arrow) with stalk (black arrow) from greater curvature of the stomach.

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**Fig. 0:** A 40-year-old man with a jejunal GIST. Dynamic CT scan demonstrates a small bowel intussusception (arrow) caused by a GIST on arterial phase (Fig. 31) and spontaneous reduction on portal phase (Fig. 32).

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**Fig. 0:** Dynamic CT scan demonstrates a small bowel intussusception (arrow) caused by a GIST on arterial phase (Fig. 31) and spontaneous reduction on portal phase (Fig. 32).

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**Fig. 0:** A 62-year old man with a duodenal GIST. Contrast enhanced CT scan shows an enhancing, intraluminal mass with ulceration (arrow) in the medial of the duodenum.

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**Fig. 0:** Coronal CT scan shows that the mass (arrow) originates from 2nd portion of the duodenum.

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**Fig. 0:** Photograph from endoscopy show the ulcerated mass in the duodenum.

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**Fig. 0:** A 60-year old male with an ileal GIST. Axial contrast-enhanced CT image obtained during the arterial phase shows an enhancing mass (white arrow) surrounding hemoperitoneum (black arrow) in the pelvic cavity.

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Fig. 0: Coronal contrast-enhanced CT image shows that the mass (arrow) originates from pelvic ileal loop.

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Conclusion

GISTs are most common gastrointestinal tumors with various imaging findings. Typical imaging findings of GISTs images can make diagnosis easily.

Imaging findings of GISTs can be atypical depending on the location, tumor components, multiplicity, and the presence of combined complications.

Understanding the atypical imaging manifestation of GISTs is useful for differentiating these lesions from other gastrointestinal tumors and in formulating the correct diagnosis.
Personal Information


Department of Radiology and Research Institute of Radiology,
University of Ulsan College of Medicine, Asan Medical Center,
388-1 Poongnap-2dong, Songpa-gu, Seoul 138-736, Korea
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