GIST. Keys for a fast radiologic identification

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**Learning objectives**

- Explain the most common radiological characteristics of this type of tumor, primarily radiological findings on CT according to location, schematically and simple, allowing a quick read and comprehension that allows easy memorization.

- Check the main differential diagnosis of these tumors, with emphasis on the radiological characteristics that differentiate them.

**Background**

**INTRODUCTION:**

The GIST (gastrointestinal stromal tumors intestinal) constitute 0.1-3% of tumors of the gastro-intestinal tract, but are the most common mesenchymal tumors at this level. They are epithelial tumors that grow in the muscular wall of the gastrointestinal tract. It is believed to originate from cells of Cajal, pacemaker cells that are involved in the regulation of gut motility.

It is defined by the expression of growth factor receptor tyrosine kinase, known as KIT or CD-117. Pharmacologically it is a therapeutic target with an inhibitor of the KIT tyrosine kinase receptor, known as STI-571 or imatinib.

They are different from other mesenchymal tumors (leiomyosarcomas, leiomyoblastomas, leiomyomas, schwannomas...) in their behavior and immunologically, although they share many similar characteristics by light microscopy. The latter do not express the antigen KIT.

The average age of onset is about 40-70 years. In young adults and children, its appearance has been associated with neurofibromatosis type I, familial GIST and Carney triad (gastric GIST, extra-adrenal paraganglioma, and pulmonary chondroma).

70-80% of them are benign constituting mostly incidental findings. Approximately 20-30% are malignant.

Radiographic features of GIST vary depending on the size of the tumor and the organ of origin.

**EPIDEMIOLOGY:**

GISTs have an approximate incidence of further 10-20 cases / million, with an average age of onset of about 40-70 years. They do not have sex predilection.
The most common site is the stomach, accounting for approximately 60-70%. Next in frequency the small intestine (20-30%), colon and rectum (5-10%) and esophagus (<5%). They account for 1-3% of gastric neoplasms, 20% of small bowel tumors and 0.2-1% of colon tumors. They can also be primary tumors of the omentum, mesentery or retroperitoneum.

CLINIC:

The clinic is often vague and depends on the size of the tumor, usually large symptomatic.

The most common symptoms are: abdominal pain or distention, or gastrointestinal bleeding unexplained anemia, mass, weight loss, nausea or vomiting. Ascites is very rare.

Duodenal tumors rarely cause duodenal obstruction or obstructive jaundice.

HISTOLOGY:

They tend to exophytic growth most involving the muscular, and in 50% of cases mucosal ulceration occurs.

Histologically classified into two main types, based on the predominant cell type: spindle cell (most common) or epithelioid.

Predicting malignant potential is difficult, but certain data are associated with poor prognosis:

- Location distal bowel (the gastric benign, are three times more frequent)
- Size: is the feature most indicative of malignancy or benignity, tumors smaller than 2 cm are usually benign and malignant larger than 2 cm.

High-mitotic activity.

- Growth or outside the gastrointestinal tract.

Indicate no malignant potential: necrosis, cystic changes, nuclear atypia, vascularization and the degree of staining for CD117.

Immunohistochemical features typical of GIST are:

- Positivity for KIT (CD117)
- 70% coexpressed CD34.
- They can be positive for smooth muscle actin and rarely for desmin and S-100 protein.
According to the consensus of pathologists, GIST terminology applies only to gastrointestinal mesenchymal neoplasms KIT immunoreactivity.

**TREATMENT**

The treatment of choice is surgical resection not requiring an intestinal wide excision and usually not lymphadenectomy.

Preoperative percutaneous biopsy presents the theoretical risk of peritoneal seeding or tumor rupture and is therefore only indicated if the tumor is unresectable.

If the disease is far advanced inhibitors of tyrosine kinase (imatinib mesylate) can be used and have produced good response and prolonged survival, currently considered molecular target therapy.

Different studies claim that approximately 59-69% of cases respond to therapy with STI-571 or imatinib, 19-26% and 11-13% are stabilized cases the disease progresses.

The systemic or intraperitoneal chemotherapy, arterial chemoembolization, surgery or radiation have not been effective in the treatment of patients with metastases or recurrence.

**EVOLUTION**

It is considered that there is a good response to treatment if the change is seen in lesions from being mass hiperatenuadas to hipoatenuadas and homogeneous, with decreased uptake heterogeneous and vascularization. Lesions may grow but it is accepted that there is no progression if decreased attenuation.

Can use in 2-3 years after treatment (spoken in some series up to 40% of cases). The most common recurrence patterns are local recurrence within the peritoneal cavity or liver metastases (Figura 1).

The malignant potential can not always be predicted by the use of conventional prognostic factors. Markers of malignant potential are major tumor size and mitotic rate per 50 high power fields.

Based on this are divided into four categories:

- **Very low risk of malignant potential**: tumors less than 2 cm and less than 5 mitoses per 50 high power fields.
- **Low risk of potential Malignant tumors**: 2 to 5 cm and less than 5 mitoses per 50 high power fields.
• Intermediate risk tumors smaller than 5 cm, with 6 to 10 mitoses per 50 high power fields or with a size between 5 and 10 cm and less than 5 mitoses per 50 high power fields.
• High risk: tumors larger than 5 cm and more than 5 mitoses or greater than 10 cm and any number of mitosis or any size and more than 10 mitoses per 50 high power fields.

The location in the small intestine has been linked with poor development.

GISTs have malignant disease free survival without metastasis five years and only 10 to 20 months if they are accompanied.

Images for this section:

Fig. 1: Display A rounded multiple heterogeneous multiple hepatic lesions and gastric exophytic mass (same patient as in Figure 4): gastric GIST with liver metastasis. Image B: Good response after treatment (3 months later) with Imatinib, with decreased density and size of the gastric mass and multiple hepatic lesions. Image C: Relapse (12 months
after initiation of treatment) with increased density of the liver metastases, one with inside hyperdense nodule (arrow).
Imaging findings OR Procedure details

GENERAL RADIOLOGY CHARACTERISTICS

GISTs tend to wrap the outer muscle layer showing exophytic growth.

The predominant radiographic pattern is enlarging mass of the wall and projecting into the abdominal cavity, capturing heterogeneously contrast. Occasionally may have an intraluminal component.

Typically, these are well-circumscribed masses that compress the adjacent tissue and do not have a true capsule. They may have areas of hemorrhage, cystic degeneration and necrosis. They range in size from several millimeters to over 30 cm. Not described vascular invasion or venous thrombosis associated.

The 50% of GISTs have gastric mucosal ulceration, introducing air or oral contrast within the tumor.

Usually affect the bowel wall concentrically therefore intestinal obstruction is rare and may cause aneurysmal dilatation of the affected.

Mesenteric masses are usually well defined, with smooth surface and show no spiculation of the mesentery. The major air can cavitate showing the dependent portion, what is known as Bernoulli sign-Toricelli.

Radiographic features of secondary involvement:

Nodal metastases or lymphadenopathy are rare, most studies did not describe them but pathologists say they can come to pass. We argue that if we are evaluating the tumor presents, we have to think of another diagnostic possibility.

Mesenteric metastases usually are recurrences, but may be the presentation of enteric large tumors.

Omental metastases are usually small (<2 cm), captan contrast evenly and can appear in different positions in the TC tracking due to movements of the omentum(Figure 2).

Liver metastases are usually hypervascular on CT and MRI before chemotherapy. They are hypodense in CT and can usually grasp homogeneously or heterogeneously contrast in the portal phase. In any case described in portal phase isodense metastases why the study would be useful in two phases. It is possible that not all metastases appearing on the same liver have equal vascularization.

In MRI, usually have low or intermediate signal intensity.
Necrosis is common in large masses, bleeding is rare. TC is very common in purely cystic metastases after chemotherapy, but rare before treatment (figure 3).

**Lung metastases** are extremely rare.

Based on the findings described in TC, characteristics useful for the differentiation between benign and malignant tumors, these features were found more frequently in GIST with high mitotic index, are as described below:

1) lesion larger than 5 cm

2) heterogeneous contrast enhancement

3) Location of the tumor

4) Presence of metastasis

5) Component necrotic-cystic

**RADIOLOGICAL CHARACTERISTICS BY LOCATION AND COMMON DIFFERENTIAL DIAGNOSES:**

**GASTRIC GIST:**

The stomach is the most common location for GIST, accounting for 2-3% of gastric tumors.

According to the series, the percentage of occurrence depending on the gastric area affected is: 75% in body, 4% in fundus and antrum 11%.

The most common radiographic finding is TC intramural mass component that may have spread to the extra-gastric ligament gastrohepatic, gastrosplenic or lesser sac. In some cases they may show a thin pedicle wall binding.

92% of the cases show a pattern of peripheral uptake, corresponding to areas of viable tumor with central areas of low attenuation due to hemorrhage, necrosis or cystic. Only a minority have homogeneous uptake.

Sometimes you can see cavities that communicate with light and contain air-fluid levels or oral contrast.

The masses larger than 10 cm or any recurring may show irregular margins and aggressiveness associated findings (figure 4).

There may be evidence of invasion of adjacent organs, omentum or peritoneal dissemination and liver metastases. The lymph nodes are rare.
In MRI have variable characteristics may exist solid portions with low signal intensity on T1 and T2 high. Show gadolinium enhancement.

The differential diagnosis of gastric GIST arises primarily:

- Other mesenchymal neoplasms:
  I. Leiomyomas or leiomyosarcoma: uncommon in stomach.
  II. Schwannomas: radiological features may be similar but are less common.
  III. Neurofibromas
    - Neuroendocrine neoplasms (solitary gastric carcinoid) are more common in the gastric antrum and often have central ulceration.
    - Adenocarcinoma and lymphoma: rarely have marked exophytic growth and often have associated lymphadenopathy.

Small bowel GIST:

Most often appear in jejunum and present an average size of 8.6 cm. (Figure 5 y 6).

The most characteristic imaging findings on CT are the intramural mass, intraluminal polyp or mass with large subserosal component. Usually, like the gastric heterogeneous mass, well defined, with low attenuation peripheral uptake and central. Less frequently display homogeneous, that is more typical of benign smaller than 5cm.

Mesentery and can spread to affect other segments of the small intestine, bladder, ureters, colon and abdominal wall.

Intestinal malignant GISTs usually accompanied by secondary liver involvement in omentum or peritoneum (Figure 7).

In magnetic resonance have a similar appearance to gastric GIST.

Differential diagnosis of intestinal GIST:

- Adenocarcinomas: Annular lesions are usual.
- Lymphoma accompanied by lymphadenopathy.
- Neoplasms of the mesentery which secondarily affect small intestine and mesenteric fibromatosis are, inflammatory pseudotumor, sclerosing mesenteritis and metastasis.

ANO-RECTAL GIST

Account for less than 5% of GIST.
The most characteristic imaging findings on CT are well circumscribed mass focal wall that extends to the wall of the rectum, less frequently appear as a polypoid intraluminal mass. They may have mucosal ulceration and spread to the ischio-rectal fossa, prostate or vagina.

In the present MRI signal intensity and uniform intermediate sequences T1, and still more heterogeneous hyperintense on T2-weighted sequences. Gadolinium usually capture heterogeneously.

**The differential diagnosis of anorectal GIST** is done:

- Adenocarcinoma usually have irregular margins and peri-rectal lymph nodes.
- Anal squamous cell carcinoma.
- Lymphoma.
- Malignant Melanoma.
- Carcinoid.
- Leiomyoma / leiomyosarcoma.
- Adenocarcinoma # prostate, prostate or peritoneal sarcomas: usually present significant perirectal extension.

**ESOPHAGEAL GIST**

They are very rare, the tumor in this location are the most common mesenchymal leiomyomas (75%).

Most often located in the distal third of the esophagus, appearing in patients with a mean age of 35 years.

There is little information on the appearance at this level because of its rarity.

The findings described in TC are homogeneous or heterogeneous intramural masses with areas of necrosis, hemorrhage or cystic. They can occur as polypoid masses extending to the esophageal lumen.

**The differential diagnosis of esophageal GIST** is mainly done with:

- If they are small tumors: leiomyoma, duplication cysts, lipomas, granular cell tumors and hemangiomas.

- If they present as polypoid masses: papillomas, adenomas, polyps, inflammatory, fibrovascular polyps and carcinomas.

- If they are as big and aggressive masses ranging mediastinum: carcinomas, melanomas, lymphomas, leiomyosarcomas.
MESENTERIC OR OMENTAL GIST

Omental or mesenteric GISTs usually present as large masses (16–5 cm medium size).

The most characteristic imaging findings are: mass large, complex and heterogeneous with necrosis, hemorrhage and cystic changes. In most cases the contrast picks solid peripheral portion. They tend to be well-defined and smooth surface.

The differential diagnosis of mesenteric or omental GISTs is done with:

• Leiomyosarcoma

• malignant fibrous histiocytoma

• Fibrosarcoma

• Liposarcoma

• If you are shown as homogeneous mass: mesenteric fibromatosis, inflammatory pseudotumor

• Metastatic GIST in other locations, which are usually multiple. In these cases the differential diagnosis fundamental, we will do so: peritoneal carcinomatosis, lymphomatosis and disseminated peritoneal leiomyomatosis.

Images for this section:
Fig. 2: CT with oral and intravenous contrast. Omental mass very well defined and heterogeneous without associated lymphadenopathy corresponding to recurrence of gastric GIST.
Fig. 3: CT with oral and intravenous contrast. Image A: multiple hypodense hepatic lesions, well defined. Image B: ureterohydronephrosis right flank mass izdo mesenteric and retroperitoneal lymphadenopathy. C and D: Multiple solid masses, heterogeneous, well-defined, with peripheral contrast uptake in mesentery and retroperitoneum. Intestinal GIST with liver metastases, mesenteric and retroperitoneal.
Fig. 4: CT with oral and intravenous contrast. Presence of heterogeneous mass containing air cavity widely dependent on the gastric fundus level (blue arrow) which causes thickening of the wall. It is associated with the presence of multiple heterogeneous liver masses and well defined. The result was gastric GIST with liver metastasis.
**Fig. 5:** CT with intravenous and oral contrast, arterial phase (A) and portal (B). Exophytic, well-defined and heterogeneous air inside, under the wall of the jejunum. Pathology: jejunal GIST.
Fig. 6: CT with oral and intravenous contrast. Heterogeneous mass is seen well defined dependent duodenum wall. Duodenal GIST.
Fig. 7: CT with oral and intravenous contrast. It is appreciated dependent exophytic jejunal wall (yellow arrows), hypodense and well defined. Multiple well-defined and multiple hepatic lesions and lymph hypodense soft tissue density in omentum (red arrow). Jejunal GIST with liver metastasis and omental.
Conclusion

GISTs are the most common mesenchymal tumors of the digestive tract and although radiological findings vary depending on your location, keep in mind that the most common site is the stomach followed by the small intestine, where its presentation is as unique exophytic large, with well-defined boundaries and heterogeneous contrast enhancement.

References


Personal Information