Clinical and Radiologic Features of the Gastrointestinal Stromal Tumours: The experience of a Portuguese Hospital from 2004 to 2011

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Purpose

Gastrointestinal stromal tumours (GISTs) are the most common mesenchymal neoplasms in the gastrointestinal tract. In the last few years they became more recognized as a distinct set of tumors, with different sites of involvement, clinical presentations and radiologic features.

Therefore the aim of this study was to investigate and describe clinical, radiologic and pathologic features of the gastrointestinal tumours.
Methods and Materials

We reviewed retrospectively 36 cases of GISTs, between the years of 2004 and 2011, followed at our institution and confirmed hystopathologically and describe their main clinical, imaging and hystopathological features.

The following variables were taken into account: sex, age, clinical presentation, size, morphology, contours, enhancement pattern, evidence of bleeding, necrosis, calcification, lymphadenopathies, local invasion and distant metastases, immunohistochemical markers and mitotic index.
Results

Background

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract (GI). (1) They account for less than 1% of all GI neoplasms and have an estimated incidence of 10-20 per million people per year. (2)

GISTs are more frequent between 40 and 80 years of age with a median age around 60 years, but have been reported in all ages. They affect both gender equally. (1,4)

This type of tumors commonly arise from the stomach (60%) and small bowel (30%). Colon, rectum and esophagus account for 5% of cases, (1,5,6) and rarely they can be found outside the GI tract (e.g. uterus, vagina or peritoneum). (1)

Clinically, GISTs present in a variety of ways according to their location and size. (1,3) About 70% of GISTs are symptomatic and tend to present as GI bleeding, weight loss, abdominal mass, abdominal pain and, rarely, as intestinal occlusion. (1) 20% are asymptomatic, particularly if smaller than 2cm, and are incidentally diagnosed by endoscopy, imaging or laparotomy. (7) About 10% of GISTs are only found at autopsy. (1)

GISTs are non-epithelial tumors typically originating from the Cajal cells located in the intestinal pacemaker tissue of the muscularis propria. (1,3,4)

Immunohistochemically, 95% of GISTs are positive for the expression of the KIT protein (CD117), a receptor tyrosin kinase type III, acting as a specific tumoral marker, as well as for DOG1 (discovered on GIST 1)(6). 60 to 70% are positive for the expression of CD34+; 30 to 40% are positive for smooth muscle actin (SMA) and about 5% for the expression of protein S-100. (1,5)

Results

In our study, 19 female patients (53%) and 17 male patients were included, with ages between 28 and 88 years and a mean age of 65 years.

Clinically, the most frequent symptom was GI bleeding (47%), followed by abdominal discomfort or overt pain (19%), palpable pelvic mass (3%) or intestinal occlusion (3%). 11% of cases were associated with anemia and 17% were incidentalomas.

The small bowel was the most frequent site of involvement (50%, n=18), followed by the stomach (n=12), peritoneum (n=4), colon (n=1) and rectum (n=1). The mean tumoral size was 5.7cm, ranging from 1 to 19 cm, with two thirds of cases below 5cm (n=23; 66%).
Most cases presented on CT as a rounded mass (n=19; 50%), with well defined borders (n=20; 53%) and an heterogeneous enhancement (n=23; 61%). Signs of bleeding were found in 53% of cases (n=19) and 42% showed low central attenuation values (n=15). Calcifications were found in only one patient (3%).

Two patients had liver metastases at the time of diagnosis and no patients showed signs of local invasion or lymphadenopathies.

All GIST cases were positive for CD117 while one third were simultaneously positive for CD34+ (n=13). Most cases also revealed a mitotic index of 5 / 50 high-power fields.

Uncommon radiological findings that were documented during this study were:

- Presence of calcifications (n=1);
- Active bleeding from the GIST during CT exam (n=1);
- Perforated GIST with gas inside (n=1);

Simultaneously nine patients had synchronous findings:

- Colonic adenocarcinoma (n=3);
- Gastric adenocarcinoma (n=1);
- Melanoma (n=1);
- Colonic adenoma (n=2);
- Myelodisplastic syndrome (n=1);
- Solitary lung nodule (n=1);

**Discussion**

While being rare tumors, recent research on GISTs has allowed for certain typical features of these tumors to be recognized. In our case series we analyse and document them and report that, in our case series:

1. GISTs affect both sexes in fairly equal proportions.

2. The mean age at diagnosis is around 65 years old.
3. Clinically, most patients present with GI bleeding or abdominal discomfort / overt pain. GIST as an incidentalloma at imaging or laparotomy is the third most common presentation.

4. While the stomach is referred to, in the peer-reviewed literature, as the most common site of involvement, (Fig. 1-3) in our case series we found the small bowel to be the most commonly affected site (Fig. 4, 5). (9-11)

5. On CT imaging, GISTs may present with various sizes, according to its location. In our study population the mean size of the mass was 5.7cm, with smooth, well-defined borders, and heterogeneous enhancement. About half the cases showed signs of necrosis or internal bleeding (Fig. 6).

6. Lymphadenopathy, local invasion and distant metastases are uncommon.

7. GISTs are defined by the expression of CD117 and are frequently associated with other markers such as CD34+.

Our case series also allowed us to identify atypical findings such as:

1. Small bowel intussusception diagnosed by CT in patients presenting with small bowel occlusion (Fig. 7).

2. Large pelvic mass containing air on CT imaging, in a case presenting as a palpable mass (Fig. 8).

3. Active GI bleeding documented on CT. While GI bleeding is a frequent finding, active bleeding documented by CT is not.

4. Less common sites of primary involvement such as the colon, rectum and peritoneum (Fig. 9, 10, 11).

5. Evidence of tumoral calcifications at CT imaging (Fig. 12), being calcifications rare in GISTs and even in GI neoplasms as a whole.

6. While historically though of as mostly benign tumors (70 - 80%)(1,6), recent reports suggest distant metastases to the liver and peritoneum (8-12). We present two cases with multiple liver metastases (Fig. 13).
7. While the studied population is small and statically not significant, we believe the high number of cases with synchronous findings (n=9), specifically another GI neoplasm or pre-malignant conditions, to be significant. In our cases series we report concomitant diagnosis of adenocarcinoma of the colon (n=3), adenoma of the colon (n=2) and gastric adenocarcinoma (n=1).

8. Two other patients presented with simultaneous neoplasias (i.e. Mielodisblastic syndrome and melanoma). One patient presented with a solitary lung nodule but died before its nature could be clarified.
Fig. 1: Gastric GIST presenting as a nodular lesion (3 cm wide), with intra-lesional air.

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Fig. 2: Upper gastrointestinal series with an anteroposterior view of the stomach showing classic features of submucosal lesion

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**Fig. 3:** Axial CT shows exophytic tumor with irregular margins with cystic and necrotic components. The lesion was a gastric gist.

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Fig. 4: CT shows exophytic lesion with irregular margins with heterogeneous enhancement. The lesion was a small bowel gist.

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Fig. 5: Heterogeneously enhanced small bowel mass with areas of necrosis and air inside. It was proven to be a small bowel gist.

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Fig. 6: Axial CT shows a large mass (13cm) with central cavity, necrotic areas and air inside. The lesion was a small bowel gist.

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Fig. 7: Ileo-ileal intussusception as documented by CT in a patient with abdominal pain and a palpable mass. GIST was confirmed at the head of the intussusception.

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**Fig. 8:** Large oval mass (9cm large), with irregular borders and an intra-lesional air-fluid level with oral contrast. These features point to necrosis and fistulization.

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Fig. 9: Large mesenteric heterogeneously enhancing mass with areas of necrosis. Notice the contact with the left colon, the stomach and the pancreas.

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**Fig. 10:** Transmural lesion involving the anterior wall of the rectum. Notice both the endoluminal and the extra-luminal components.

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Fig. 11: Large retroperitoneal heterogeneously enhancing mass measuring 13cm wide. Notice ample contact with the liver, duodenum, pancreas and right renal vein

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Fig. 12: A 6cm wide heterogenously enhancing pelvic mass with an oval morphology and irregular borders. Notice the scattered calcifications and the air-fluid level.

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Fig. 13: Multiple liver metastases proven to be from a primary GIST.

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Conclusion

GISTs' growing interest as a topic of research has expanded our knowledge of its clinical features, which largely depend on its site of involvement. GI bleeding is its most frequent presentation.

Computed tomography allows a very good detection and radiological characterization of primary GISTs.

The most common imaging presentation is a rounded regular, well defined, heterogeneously enhancing mass.

This type of tumor rarely metastasises, but in our experience, may occur with other tumors simultaneously.
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