Pedunculated giant GISTs of the stomach with exophytic growth: Report of two cases

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Introduction

GISTs arise from mesenchymal precursor cells of the gastrointestinal tract that may differentiate towards the interstitial cells of Cajal, pacemaker cells regulating autonomous motility of G.I. tract.

About 60% of GISTs are found in the stomach, 20–50% in the small bowel (one third in the duodenum), and 5–15% in colon and rectum; GISTs, however, may rarely be found also in the oesophagus, omentum, mesentery or the retroperitoneum.

The distribution of these tumors in the stomach is: pars media, 40%; antrum, 25%; pylorus, 20%. In less than 15%, GISTs location is next to the EGJ, in the cardia and in the fundus.1,2

Gastric GISTs may also present as pedunculated, exophytic masses. This pattern of growth is rare, and diagnosis is often made after surgery, since the lack of specific symptoms.1–7

The Authors report two cases of pedunculated gastric GIST with totally extraluminal growth.

Case reports

Case 1

A 72-year-old man suffering from arterial hypertension and type II diabetes mellitus was referred to our institution for abdominal pain and weight loss. Clinical examination revealed a large palpable mass located in the epigastric region. Upper G-I endoscopy showed no abnormalities. CT scan revealed a large abdominal mass extending from the diaphragm to the left flank, displacing the small bowel to the right and the stomach anteriorly (Fig. 1).

At surgery a large mass occupying the abdominal cavity was found. The tumor was located between the stomach and the transverse mesocolon. The mass was isolated from the surrounding structures, and it was found to take origin from the larger gastric curvature, having a short pedicle. The tumor was completely excised with a small portion of the gastric wall surrounding the pedicle.
The specimen was 15.5 × 13 × 9.5 cm in size. The cut surface revealed the presence of necrotic and hemorrhagic areas. Histology showed the presence of epithelioid cells arranged in interlacing bundles, with round nuclei. There was high cellularity and pleomorphism with focal necrosis and hemorrhage. Mitotic index was high (10–15/50 hpf). Immunohistochemistry showed positive reaction to CD117 (c-kit), CD34, Vimentin. So the tumor was diagnosed as high-risk GIST. The gastric wall excised showed no infiltration. Postoperative course was uneventful, and the patient was discharged on p.o. day 10.

The patient did not receive adjuvant chemotherapy or Imatinib, since the lack of disease residual. Follow-up was carried out by clinical examination at 1 month and US-scan at 1, 6 and 12 months after surgery, and CT-scan at 3 and 12 months after surgery. After one-year follow-up the patient is doing well and he has no signs of disease recurrence.

Case 2

A 63-year-old man was referred to our institution for evidence of abdominal mass. CT scan revealed a sub-diaphragmatic mass extended from the left hepatic lobe to the pelvis, with small bowel and stomach dislocation (Fig. 2). Upper G-I endoscopy did not show abnormalities.

At surgery a large mass (Fig. 3) occupying the abdominal cavity was found. This tumor displaced the transverse colon and small bowel on the left side, and the stomach anteriorly. The lesion originated from the posterior gastric wall, by a short pedicle. It was completely excised with the greater omentum and a small portion of the gastric wall. The specimen was 19 × 17.5 × 12.5 cm in size. The cut surface revealed the presence of necrotic and hemorrhagic areas.

Histology showed of spindle-shaped cells arranged in interlacing bundles. There was high cellularity and pleomorphism with focal necrosis and hemorrhage. Mitotic index was high (18/50 hpf). Immunohistochemistry showed reaction to CD117 (c-kit) and CD34 (Fig. 4). So the tumor was diagnosed as high-risk GIST.

Postoperative course was uneventful, and the patient was discharged on p.o. day 8.

The patient did not receive adjuvant chemotherapy or Imatinib, due to the lack of disease residual. Follow-up was carried out by clinical examination at 1 month and US-scan at 1, 6 and 12 months after surgery, and CT-scan at 3, 12 and 18 months after surgery. After 18-months follow-up the patient is doing well and he has no signs of disease recurrence.

Discussion

GIST account for approximately 1% of gastric neoplasms.8,9 About 60% of GISTs are submucosal; 30% are subserosal and 10% are intramural. Sometimes GISTs may present as large pedunculated extragastric lesion.

Some of these lesions present cystic change, and this may be the consequence of a rapid tumor growth, with congestion and intratumoral hemorrhage.6,7
Diagnosis is often made only after surgery, since the presence of large abdominal mass can rarely be identified preoperatively as a gastric tumor.

Instrumental examination may include Upper G-I endoscopy, CT scan, MRI and even endoscopic ultrasonography. This technique permits to distinguish mesenchymal tumors from other kinds of tumors, and even benign from malignant mesenchymal lesions. Furthermore, EUS-guided fine-needle aspiration biopsy can lead to a precise clinical diagnosis.

According to the current pathologic and prognostic classifications, these pedunculated lesions are often classified as lesions having high risk of malignancy (due to the large dimension and the high number of mitoses) even if they use to have only expansive growth, without infiltration of surrounding structures.

For this reason the treatment of choice is extensive surgical resection of the tumor with a wedge resection of the stomach at the level of the tumor pedicle. Patients who have complete tumor resection (R0) have more favorable outcome than those with less complete surgery.

Incomplete surgical resection should be performed only for the palliation of symptoms due to bleeding, pain or mass effect. Non-resectable or metastatic GIST is a fatal disease that resist conventional chemotherapy or radiotherapy.

Treatment with Imatinib seems to be the best systemic therapy for metastatic and locally advanced GISTs.

The high response rates with Imatinib in advanced and metastatic setting have fostered interest in its role even in the adjuvant or preoperative settings. There may be an improvement in surgical outcome in patients treated with Imatinib preoperatively. Currently, there are several trials to address combining surgery and Imatinib.

In conclusion the clinical presentation of pedunculated gastric GISTs as large abdominal masses should request extensive R0 surgery, and strict post-operative follow-up, and even post-surgical treatment with selective kit-inhibitors, due to the elevated risk of malignancy.

Figure 4 Immunohistochemistry shows positive reaction to CD-117 (c-kit).

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References