

CASE REPORT

GIANT SMALL INTESTINE GIST IN A YOUNG MAN

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SUMMARY

Gastrointestinal stromal tumors (GISTs) are the most common type of mesenchymal tumors of the gastrointestinal tract. They are originating from the interstitial cells of Cajal, that are considered the GI pacemaker cells. The most common site is the stomach, only 23-38% are found in the small intestine. Giant GIST's (over 10 cm diameter) are associated with a high risk of complications and malignancy. We present a case of a giant GIST in a young man patient treated by surgical resection associated with postoperative adjuvant therapy (Imatinib).
Key words: GIST, small intestine, giant gastrointestinal tumor

RÉSUMÉ

Les tumeurs stromales gastro-intestinales (GIST) chez un jeune homme

Les tumeurs stromales gastro-intestinales (GIST) sont le type le plus commun de tumeurs mésoenchymateuses gastro-intestinales. Elles prennent leur origine dans les cellules interstitielles de Cajal, qui sont considérées comme les cellules stimulatrices du tractus gastro-intestinal. Le site le plus commun est l'estomac, seulement 23-38% se trouvent dans l'intestin grêle. Les tumeurs stromales géantes (plus de 10 cm de diamètre) sont associées à un risque élevé de complications et de malignité. Nous présentons le cas d'un jeune homme diagnostiqué avec une tumeur géante stromale traitée par résection chirurgicale associée à un traitement adjuvant (Imatinib).

Mots clés: GIST, l'intestin grêle, la tumeur gastro-intestinale géant

INTRODUCTION:

Gastrointestinal stromal tumors are the most common type of mesenchymal tumors of GI tract, originating from the parietal wall. They arise from the interstitial cells of Cajal, that coordinate the gut peristalsis. They are different from the other soft tissue tumors by c-kit proto-oncogene (CD117) expressions.

GISTs are preferentially located in the stomach (33-63%), small intestine (23-38%) and colon-rectum (5-32%).

Most GISTs are more than 5 cm in diameter at the time of diagnosis, because the clinical presentation is variable and nonspecific. Giant stromal tumors (over 10 cm in diameter) represent 20% of all tumors and they are associated with high risk of malignancy, of local and distal metastasis. In addition to tumor size, mitotic rate, location of the tumor and rupture of the tumor capsula is a prognostic factor.

CASE REPORT

A 46-year-old man patient was admitted to the Surgery Department due to melena, intermittent upper abdominal pain, nausea and bloating over the last 3 weeks.

His past medical history was unremarkable. Abdominal examination revealed a large palpable abdominal mass in the left hypocondrium, without signs of tenderness. Blood tests showed a haemoglobin level of 10.5g/dl.

Tumor markers carcinoembryonic antigen, alpha-feto-protein and carbohydrate antigen 19.9 were negative.

Upper endoscopy and abdominal X-rays did not reveal any abnormality.

Contrast-enhanced abdominal computed tomography revealed a 140x96x153 mm solid tumor with low-density area within, arising from the duodenojejunal junction, displacing gut and mesenteric vessels (Fig. 1a, b).

We have performed laparotomy which revealed a giant

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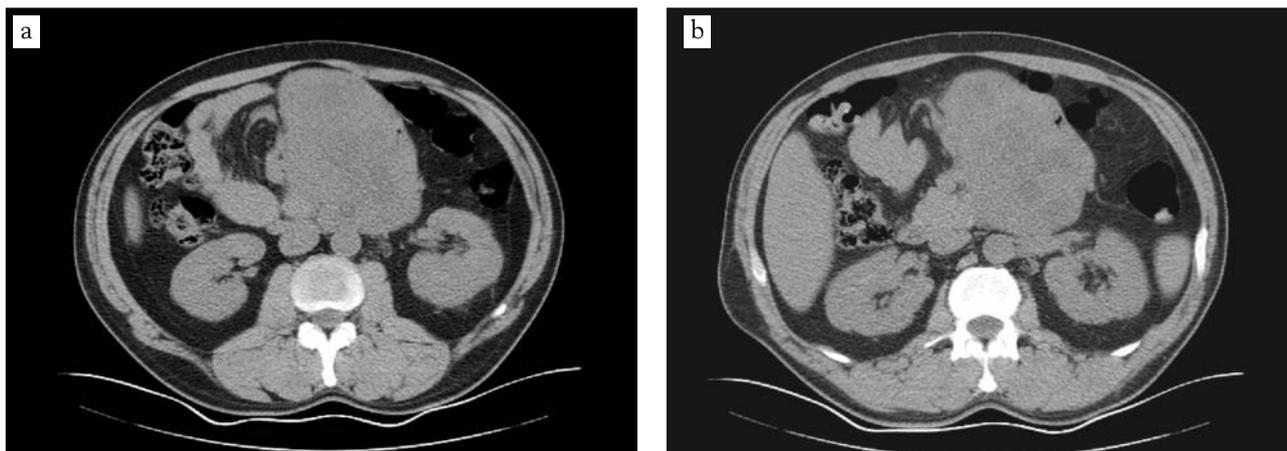


Figure 1 - a: Abdominal CT scan: giant tumor developed in the mesentery of duodenojejunal junction
b: Abdominal CT scan: tumor displacing gut and mesenteric vessels

encapsulated tumor (16x14cm) originating from the first jejunal loop, developed in the mesentery (Fig. 2 a, b). A segmental resection was performed with adequate gross margin followed by an end-to-end duodeno-jejunal hand-sewn anastomosis. No evidence of lymph node or intra-abdominal metastasis.

After an uneventful postoperative course, the patient was discharged in the 9th POD.

The resected mass was a well-circumscribed tumor. The solid parenchyma contained central necrosis and bleeding with a fistula to the lumen of the jejunum.

Histopathological examination of the tumor showed a typical aspect of a GIST composed with spindle cells. The mitotic index was lower than 5/50 high power fields; the proliferation index Ki67 was 2-3% and tumour cells were positive for CD117.

Based on tumor size (>10 cm), the patient has high risk for recurrence and he started an adjuvant therapy with Imatinib mesylate (400mg once daily).

12 months postoperatively the patient remains disease-free (abdominal IRM).

DISCUSSIONS

GISTs are rare mesenchymal tumors of the digestive tract, 0.2% of all neoplasms and 80% of all mesenchymal tumors. The most common site is the stomach (33-63%), the small intestine (23-38%), 5-15% the esophagus, colon or rectum, and only occasionally in the mesentery, peritoneum, appendix and omentum.

Preoperative diagnosis is difficult, the symptoms and signs are nonspecific: abdominal pain, palpable mass, nausea, bloating, early satiety. Bleeding is the most common symptom (30-40%). Ulceration of the mucosa of the small intestine or pressure necrosis may cause gastrointestinal bleeding, causing hematemesis, melena, anemia and fatigue. Imagistic investigations are used to confirm the diagnosis. CT scan is the most effective of all.

Radical surgery is the treatment of choice of primary, resectable GISTs: complete gross resection with negative margins (R0) and intact pseudocapsule. Laparoscopy can be an option in special cases (favourable anatomic sites). The introduction of the imatinib mesylate, a tyrosin kinase inhibitor, opened a new era in the treatment of GISTs, changing the natural history and the prognosis of the disease.



Figure 2 - a: Intraoperative aspect: en bloc resection of the tumor and first jejunal loop
b: Intraoperative aspect: giant tumor in contact with the first jejunal loop

CONCLUSIONS

Giant tumors represent only 20% of all cases and are associated with high risk of malignancy. The tumor size, tumor site and mitotic index are the most important prognostic factors. Additional risk factors are tumor rupture and surgical margins.

Conflicts of interest: None.

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