

PICTURES IN DIGESTIVE PATHOLOGY

Small bowel mesenchymal tumors: description of two unusual cases

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INTRODUCCIÓN

GISTs (gastrointestinal stromal tumors) are usually paucisymptomatic but may present complications requiring emergency surgery. We present two cases of complicated ileal GIST and discuss their management.

CASE REPORTS

Case report 1

A 42-year-old male consulted for rectal bleeding and melena of 24 hours of evolution. Laboratory showed leukocytosis and hemoglobin 5.2 g/dL. CT demonstrated a desmoid tumor in the mesenteric root. Emergency laparotomy was performed and a small bowel tumor, highly vascularized, was found, located 130 cm from the ileocecal valve (Figs. 1 and 2). Resection of the lesion with wide margins was performed. Histological analysis showed positivity for c-Kit (Fig. 3), with Ki-67 < 10% compatible with GIST. The postoperative course was uneventful and there is no evidence of recurrence at 12 months of follow up.

Case report 2

A 46-year-old female, in gynecological study for pelvic mass, was admitted for abdominal distension, pain, fever, and vomits of 4 hours of evolution. Physical examination revealed generalized abdominal peritonism and CT evidenced a large mass with neovascularized areas, necrosis and contrast extravasation, suggestive of intestinal perforation (Fig. 4). She underwent surgery, and diffuse purulent peritonitis and a large and lobulated tumor with cerebroid appearance and areas of necrosis were found, 35 cm from ileocecal valve and included in a conglomerate of perforated bowel loops (Figs. 5 and 6). The affected segment was excised and histological analysis showed a neoplasm of 37 x 22 x 17 cm and 3,600 g and 30 cm of ileum (c-Kit positive and Ki-67 = 20%) concordant with GIST. The patient received adjuvant chemotherapy and is disease-free after 8 months of follow up.



Fig. 1. Lobulated tumor of small intestine.



Fig. 2. Highly vascularized tumor of the small intestine.

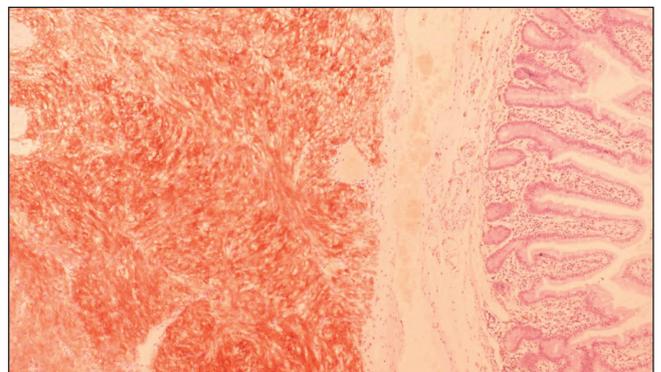


Fig. 3. Wall of small intestine with submucosal tumor vividly positive for c-Kit (CD117).

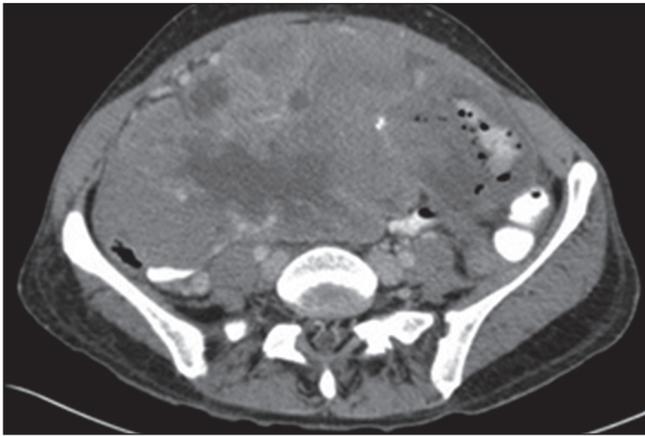


Fig. 4. Large neovascularized abdominal-pelvic mass with necrosis areas.

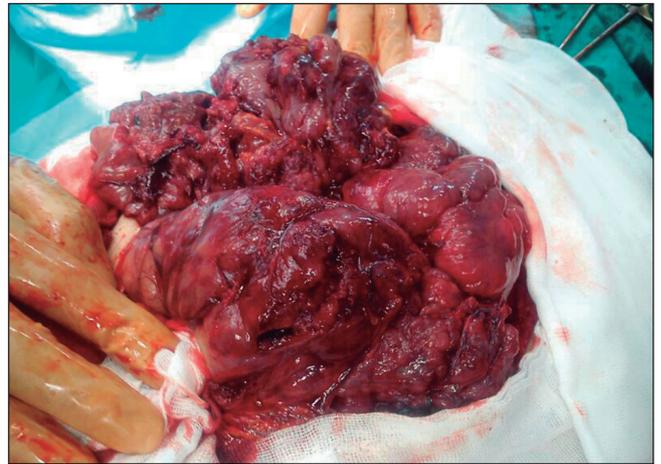


Fig. 6. Small bowel tumor with signs of macroscopic hemorrhage.

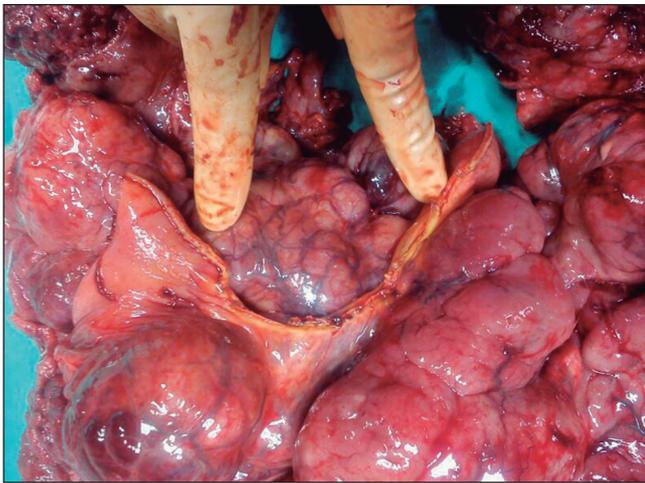


Fig. 5. Resection margins of the small bowel.

DISCUSSION

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract (1), accounting for 1-3% of gastrointestinal cancers. They use to be located at stomach (50%) and small intestine (25%) (2), and usually appear between the 4th and 6th decade of life.

This tumors usually express KIT receptor (CD117), and show mutations in KIT or PDGFRA (2) genes. They may present with abdominal pain, anemia, gastrointestinal hemorrhage or metastases, and urgent presentation is increasing.

Endoscopic procedures may not be always diagnostic; therefore other diagnostic tools such as CT or MRI are essential. Preoperative biopsy carries a high risk of bleeding and is only recommended prior to neoadjuvant therapy, or in uncertain diagnosis (3). They are considered as malignant when they exceed 5-10 cm, show high rate of cell division or metastasize. Treatment of choice is surgery; tyrosine kinase inhibitors such as imatinib are used for neoadjuvant therapy in potentially resectable tumors, in high risk tumors after surgery, or as palliative treatment. The prognosis is related to the size, the degree of proliferation and the preservation of pseudocapsule (3). Both cases described were categorized as “intermediate” and “high” risk of progression respectively, therefore a close monitoring of these patients is essential.

A high index of suspicion of this disease is therefore required since early diagnosis and treatment are imperative.

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