Multiple Primary Gastrointestinal Stromal Tumors Presenting in Jejunum and Ileum

Marcelo A. Beltrán · Rodrigo A. Tapia ·
Víctor J. Cortés · Cristóbal Larraín · Vinka Koscina ·
María-Paz Rioseco · Matías Molina · Andrés Vera

Received: 31 July 2011 / Accepted: 21 June 2012 / Published online: 4 July 2012
© Association of Surgeons of India 2012

Abstract Multiple primary gastrointestinal stromal tumors (GIST) are an infrequent finding. Benign and malignant tumors could coexist in the same patient. We discuss one case of a benign jejunal GIST and a malignant ileal GIST coexisting in the same patient and present their radiological characteristics.

Keywords Gastrointestinal stromal tumor · GIST · Malignant GIST · Acute complications of GIST

Introduction

Gastrointestinal stromal tumors (GISTs) generally arise as solitary tumors in the stomach (50–60 %), duodenum, jejunum, and ileum (25–30 %), colon and rectum (5–10 %), esophagus (5 %), and other localizations (<1 %) [1, 2]. Multiple GISTs may be found in different clinical contexts—sporadic, familial GIST syndrome, as a component of Carney’s triad, Carney–Stratakis syndrome, von Recklinghausen disease or neurofibromatosis type I, and as metastatic disease in advanced malignant GIST [1, 3]. Multiple sporadic GISTs present as two or three lesions at the same localization or at different localizations and could be malignant or benign, often coexisting as both [1–3]. Herein,

we describe a patient with two primary tumors—the first small GIST located in the proximal jejunum and the second GIST located in the distal ileum.

Case Study

A 46-year-old man was admitted to the department of surgery at our institution with complaints of acute abdominal pain for 2 days. The pain was sharp and initiated over the hypogastria. Previously he was a healthy man without any known comorbidities. The patient was febrile and had tachycardia and normal blood pressure. The abdomen was tender and rigid. On palpation it had rebound tenderness, and bowel sounds were absent. Laboratory tests showed marked leukocytosis (20,500 mm³), and elevated levels of C-reactive protein (457 mg/l). An abdominal computed tomography (CT) scan was performed. At the terminal ileum, a cystic tumor with thick irregular borders, heterogeneous contents and cavitations, measuring 71 mm×75 mm was identified (Fig. 1a), and it was suspected of being complicated with perforation. Another GIST was also diagnosed in the proximal jejunum; this tumor was small, well delimited, homogeneous in appearance, measuring less than 3 cm, and was categorized as probably benign (Fig. 1b). A tridimensional reconstruction was performed clearly depicting both tumors, and the patient was submitted to emergency surgery (Fig. 2). The abdomen was approached by a midline incision, and after exploration and packing of the perforated ileal GIST, the jejunal GIST was wedge resected (Fig. 3a). Afterward, the perforated GIST was resected with a segmental resection of the terminal ileum and primary anastomosis (Fig. 3b). The surgical specimen showed a perforated tumor with necrotic and hemorrhagic contents. The postoperative period was uneventful and the

M. A. Beltrán · R. A. Tapia · V. J. Cortés · C. Larraín
Department of Surgery, Hospital De La Serena,
La Serena, Chile

M. A. Beltrán · C. Larraín · V. Koscina · M.-P. Rioseco ·
M. Molina · A. Vera
Medical School, Universidad Católica Del Norte,
La Serena, Chile

M. A. Beltrán (✉)
P.O. Box 912, La Serena, IV Región, Chile
e-mail: beltran_01@yahoo.com

Fig. 1 **a.** Computed tomography imaging. Coronal view depicting the terminal ileum where a tumor with thick walls measuring 71 mm was encircled with a yellow ring. **b.** Computed tomography coronal view showing a small jejunal GIST (yellow circle), measuring 23.7 mm × 21.8 mm. This tumor was considered benign and non-complicated

patient recovered well. Currently he is under treatment with imatinib mesilate with good response. The histopathological report described the ileal GIST with a thick wall (between 0.9 and 1.5 cm) composed of spindle-shaped cells in fascicles and epithelioid cells in a sheet-like growth pattern; the mitotic index was 15/50 high-power fields (HPF). Immunohistochemical stains showed myoid differentiation with positive reactions to CD117, CD34, α -smooth muscle actin (α -SMA), vimentin, and desmin, and a negative reaction to S100. The smaller jejunal GIST was a solid tumor with spindle and epithelioid cells with a low mitotic index (1/50 HPF) and

Fig. 2 Tridimensional reconstruction showing both GISTs encircled by blue rings

was considered benign. Its immunochemistry showed positive reactions to CD117, CD34, and α -SMA and negative reactions to vimentin, desmin, and S-100.

Discussion

Multiple GISTs are a common occurrence in certain clinical conditions such as neurofibromatosis type I and Carney's

Fig. 3 **a.** A wedge resection was carried-out and was considered sufficient for the jejunal GIST (black circle). **b.** The GIST localized in the terminal ileum was perforated, covered by the greater omentum with some fibrin and pus. A segmental resection with primary anastomosis was performed (black circle)

triad among others [1–4]; however, multiple sporadic primary GISTs seem to be a less common clinical scenario. The case reported here illustrates some frequent characteristics of sporadic primary GISTs since the patient presented with an acute complication of his tumor. Acute presentation of GISTs has been reported to account for most cases in some underdeveloped regions of the world such as ours [5, 6]. Approximately 40–50 % of small intestinal GISTs are clinically malignant [1]. The cause might be the late presentation when tumors are bigger and frequently complicated because they go unnoticed by the patient due to the lack of symptoms when the tumors were small. Acute perforation of the GIST or iatrogenic perforation during surgery with spillage of its contents is a factor associated with metastatic disease and poor outcome in these patients, and constitutes an indication of imatinib treatment. Besides being perforated, the ileal GIST of this patient was bigger than 5 cm and had a high mitotic index, conditions that categorized the tumor as malignant and prompted imatinib treatment. In this patient both coexisted, a malignant and a benign GIST. The small jejunal GIST was benign. Generally these small tumors are incidental findings detected on endoscopy, radiologic imaging, or during surgery [2, 7].

The wide availability of CT scan has permitted the accurate diagnosis of GISTs from very small lesions to giant tumors and its related complications including metastatic disease. Typically, primary GISTs are well-circumscribed highly vascular tumors arising from any viscera in the gastrointestinal tract and growing in an intraluminal or extraluminal manner; most GISTs, however, are predominantly exophytic [8, 9]. The tumor capsule may have smooth, lobular, irregular, or invasive margins; nonetheless, most GISTs are small and have a well-delimited capsule with smooth or lobular margins [8–12]. Malignant GISTs might have invaded adjacent organs and mesenteric fat [12]. Vascular encasement is not a habitual feature of GISTs [8, 9]. GISTs do not disseminate via lymphatic vessels. Consequently CT scans do not identify any lymphadenopathy [8–10, 12]. Smaller GISTs have a homogeneous appearance; while larger GISTs are heterogeneous, have a cystic configuration with enhancing borders of variable thickness and irregular central areas of fluid, air, necrosis, or cavitations that communicate with the gastrointestinal lumen [2, 8, 10–12]. The ileal GIST of this patient presented with characteristics of malignancy such as cystic degeneration with hemorrhagic and necrotic contents and perforation into the abdominal cavity with spillage of its contents. Differentiation of GISTs from other gastrointestinal tumors can be made on the basis of the specific findings related to GISTs;

that is tumors with a well-circumscribed capsule, heterogeneous or homogeneous content depending on size, and absence of other nodes [8–10]. The diagnoses that are more difficult to differentiate from GISTs are those of other soft tissue tumors, including leiomyoma, leiomyosarcoma, fibrosing mesenteritis, lymphangioma, plexiform neurofibromatosis, and rare malignant tumors of nerve sheath and vascular origin [8, 12]. Taking into account all of these characteristics, the preoperative CT scan performed in this patient yielded an excellent diagnosis well correlated with the surgical findings.

Conflict of Interest No conflict of interests declared – Study funding by the authors

References

1. Díaz-Delgado M, Hernández-Amate A, Sánchez-León M, Pereira-Gallardo S, Prieto-Sánchez E, Jiménez-Sáenz M, González-Campora R (2010) Multiple non-metastatic gastrointestinal stromal tumors. Differential features. *Rev Esp Enferm Dig* 102:489–497
2. van der Zwan SM, DeMatteo RP (2005) Gastrointestinal stromal tumor: 5 years later. *Cancer* 104:1781–1788
3. Beltrán MA, Cruces KS, Barria C, Verdugo G (2006) Multiple gastrointestinal stromal tumors of the ileum and neurofibromatosis type I. *J Gastrointest Surg* 10:297–301
4. Beltrán MA, Barria C, Contreras MA, Wilson CS, Cruces KS (2009) Tumor del estroma gastrointestinal (GIST) en una paciente con neurofibromatosis tipo I. *Rev Med Chil* 137:1201–1204
5. Beltrán MA, Cruces KS (2007) Primary tumors of jejunum and ileum as a cause of intestinal obstruction: a case control study. *Int J Surg* 5:183–191
6. Beltrán MA, Vicencio AO, Barra MM, Contreras MA, Wilson CS, Cruces KS (2011) Resultados del tratamiento quirúrgico de los tumores del estroma gastrointestinal (GIST) en la IV Región de Chile. *Rev Chil Cir* 63:290–296
7. Beltrán MA, Pujado B, Méndez PE, González FJ, Margulis DI, Contreras MA, Cruces KS (2010) Gastric gastrointestinal stromal tumor (GIST) incidentally found and resected during laparoscopic sleeve gastrectomy. *Obes Surg* 20:393–396
8. Hersh MR, Choi J, Garrett C, Clark R (2005) Imaging gastrointestinal stromal tumors. *Cancer Control* 12:111–115
9. Lee CM, Chen HC, Leung TK, Chen YY (2004) Gastrointestinal stromal tumor: computed tomographic features. *World J Gastroenterol* 10:2417–2418
10. Lupescu IG, Grasu M, Boros M, Gheorghe C, Ionescu M, Popescu I, Herlea V, Georgescu SA (2007) Gastrointestinal stromal tumors: retrospective analysis of the computer-tomographic aspects. *J Gastrointest Liver Dis* 16:147–151
11. Logrono R, Bhanot P, Chaya C, Cao L, Waxman I, Bhutani MS (2006) Imaging, morphologic, and immunohistochemical correlation in gastrointestinal stromal tumors. *Cancer* 108:257–266
12. Ulsan S, Koc Z, Kayaselcuk F (2008) Gastrointestinal stromal tumors: CT findings. *Br J Radiol* 81:618–623