CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 6 (2015) 58–62

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Peritonitis secondary to spontaneous perforation of a primary gastrointestinal stromal tumour of the small intestine: A case report and a literature review

Mario Alessiani[a,e], Marco Gianola[a], Sabina Rossi[b], Vittorio Perfetti[c], Piero Serra[a], Daniela Zelaschi[a], Enzo Magnani[a], Lorenzo Cobianchi[d,e,*]

[a] Department of General Surgery, Varzi Hospital, Italy
[b] Radiology Service, Voghera Hospital, Italy
[c] Department of Oncology, IRCCS San Matteo Hospital Foundation, Pavia, Italy
[d] Unit of General Surgery 1, IRCCS San Matteo Hospital Foundation, Pavia, Italy
[e] Department of Clinical, Surgical, Diagnostic and Paediatric Sciences, University of Pavia, Italy

ARTICLE INFO

Article history:
Received 4 October 2014
Received in revised form 3 December 2014
Accepted 5 December 2014

Keywords:
Gastrointestinal stromal tumours
Small intestine
Peritonitis

ABSTRACT

INTRODUCTION: A few cases of acute abdomen caused by perforation of small-intestinal gastrointestinal stromal tumours (GISTs) have been reported in the literature.
PRESENTATION OF CASE: Together with a review of the published cases, here we report a case of an elderly patient with peritonitis due to spontaneous perforation of a GIST of the jejunum. An 82-year-old man was admitted to the emergency unit of our hospital with fever and severe abdominal pain. An abdominal enhanced computed tomography scan detected a 6 cm solid mass in the left upper quadrant adherent to a jejunal loop and surrounded by free fluid and free air. Due to the radiological features of the mass, the diagnosis of a perforation of a GIST arising from the jejunum wall was suspected. The patient underwent emergency laparotomy. Intraoperative findings confirmed diffuse peritonitis secondary to jejunal tumour perforation. A segmental resection of the jejunum containing the mass was performed followed by a mechanical end-to-side anastomosis. The histopathologic examination of the mass confirmed the diagnosis of a perforated GIST of the small intestine (high-risk category). The post-operative course was uneventful and the patient was treated with adjuvant imatinib therapy.
DISCUSSION: Twenty-one other cases of spontaneous perforation of small intestine GISTs are reported in the literature and are summarized in the present review.
CONCLUSION: The described case is the tip of the iceberg and spontaneous rupture or perforation of GISTs are a far more frequent first presentation of this rare tumour.

1. Introduction

Gastrointestinal stromal tumours (GISTs) are mesenchymal neoplasms of the gastrointestinal tract (GI) that can arise anywhere from the oesophagus to the rectum [1]. GISTs accounts for less than 1% of all GI tumours; however they are the most common mesenchymal neoplasms of the GI tract. Population-based studies from different European countries report annual incidence rates ranging from 6.5 to 20 cases per million [2–6]. These studies also report an increase in the incidence of GISTs over the last two decades due to the improvement in their diagnosis and registration [2,3].

Few GISTs manifest as abdominal emergencies, such as GI haemorrhage, intestinal obstruction, or tumour perforation [7,8]. Although acute abdomen due to GIST perforation into the peritoneal cavity is rare, a few cases of peritonitis caused by perforation of small-intestinal GISTs have been reported in the literature [8–29]. Together with a review of the published cases, here we report a case of an elderly patient with acute abdomen due to spontaneous perforation of a GIST located in the jejunum.

2. Case presentation

An 82-year-old man was admitted to the emergency unit of our hospital with fever, vomiting, diarrhoea and diffuse abdominal pain that had been experienced for the previous 24 h. The patient

Abbreviations: GIST(s), Gastrointestinal stromal tumours; GI, Gastrointestinal; CT, Computed tomography; H&E, Hematoxylin and Eosin.

* Corresponding author at: Unit of General Surgery 1, IRCCS San Matteo Hospital Foundation, Pavia, Italy. Tel.: +393394421128; fax: +390382502447.
E-mail addresses: v.perfetti@smatteo.pv.it (V. Perfetti), piero_serra@ospedali.pavia.it (P. Serra), daniela_zelaschi@ospedali.pavia.it (D. Zelaschi), enzo_magnani@ospedali.pavia.it (E. Magnani), l.cobianchi@smatteo.pv.it (L. Cobianchi).

http://dx.doi.org/10.1016/j.ijscr.2014.12.012

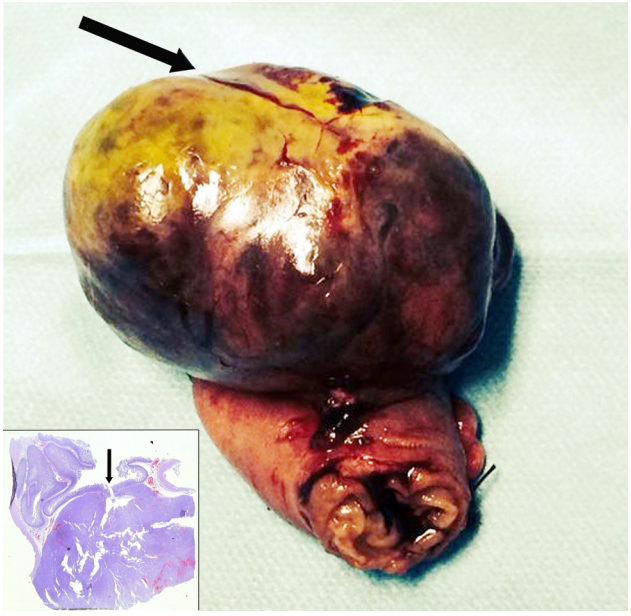

Fig. 1. Surgical specimen. The resected specimen includes the jejunal loop and the perforated GIST arising from the antimesenteric side of the loop. The arrow indicates the ruptured margin of the tumour. The insert shows the histology of the specimen: the perforation to the lumen of the jejunum is indicated by the arrow.

had a history of arterial hypertension. No other co-morbidities were reported. Vital signs were within normal limits and the rectal temperature was 38.6 °C. On examination, the abdomen was distended with generalized tenderness and muscular defence. Laboratory tests showed elevated white cell count (13.430/mm3, 92.7% neutrophils) and hyper-glycaemia (221 mg dl^{-1}).

Abdominal plain radiography showed gas-fluid levels in the small bowel without free intraperitoneal air. An abdominal enhanced computed tomography (CT) scan was urgently performed and revealed the presence of a 6 × 5.5 × 5.5 cm solid mass in the left upper quadrant towards the umbilicus, adherent to a jejunal loop, and surrounded by free fluid. A small layer of free extraluminal air was also seen close to the margin of the mass (Fig. 1). Due to the radiological features of the mass, the diagnosis of a perforation of a GIST located in the jejunum wall was suspected.

The patient underwent emergency laparotomy. Intraoperative findings showed free purulent exudate in the abdominal cavity and a diffuse inflammatory reaction of the peritoneum. The mass was located 10 cm away from the Treitz ligament, on the antimesenteric side of the first jejunal loop (Fig. 2). The perforation was identified on the external border of the mass. A segmental resection of the jejunum containing the mass was performed followed by a mechanical end-to-side anastomosis, peritoneal irrigation and toilet, and finally the placement of multiple drainages. A search for other gastrointestinal or peritoneal abnormalities was negative.

The post-operative course was uneventful and the patient was discharged 10 days after surgery. On histological examination, the specimen was described as a solid smooth grey and white mass (maximum diameter of 7 cm) arising from the wall of the resected segment of small bowel. H&E staining showed spindle-shaped cell proliferation (Fig. 3a). The mitotic index was 16/50 high-power field (HPF) and the Ki-67 value was 15%. The immunohistochemical staining was positive for C-KIT (Fig. 3b), focal positive for h-caldesmon, negative for smooth muscle actin, desmin, CD34, S-100, EMA and cytocheratine. All these findings led to the diagnosis of GIST of the jejunum, which was classified as high-risk using the prognostic classification by Fletcher et al. [30]. The surgical margins were disease-free. As soon as the diagnosis was confirmed by histology, the patient was started on imatinib mesylate therapy at the daily oral dose of 400 mg and he is currently under follow-up after six months of therapy.

3. Discussion

Nearly a third of patients with GISTs are asymptomatic and the diagnosis is made incidentally during surgical, endoscopic, radiologic procedures or at autopsy [1,31]. Most of the symptomatic patients present with vague, nonspecific abdominal pain or discomfort, sometimes associated with nausea and vomiting. Patients may also complain of early satiety or a sensation of abdominal fullness. More frequently, GISTs larger than 4 cm may produce symptoms secondary to obstruction or GI bleeding. The bleeding can be either chronic, often leading to anaemia, or acute with episodes of haematemesis or melena. Very few cases manifest as other abdominal emergencies, such as haemoperitoneum secondary to intra-abdominal tumour rupture, or peritonitis secondary to tumour perforation.

This latter emergency has been reported for all gastrointestinal tracts [32–34]. However GIST perforation seems to occur more frequently in the small bowel compared to other anatomic sites [35]. Our literature search revealed 21 cases of acute abdomen with diffuse or localized peritonitis caused by spontaneous perforation of small intestine GISTs (Table 1) [9–29]. Two more cases are reported in the case-series of Mansour and Coll [8]. They are not included in Table 1 because the details of the cases are not available. In another two cases not listed in Table 1, spontaneous ruptures of the small intestinal GISTs were associated with an intraperitoneal abscess without perforation to the intestinal lumen [35,36].

Of the 22 cases reported in Table 1, including this case, 16 patients were male and five were female. GIST perforation can occur irrespective of the age, since the patients' ages ranged from 22 up

Fig. 2. Preoperative computed tomography (CT). (a) CT image showing air in the perforation track of the tumour (arrow); (b) CT image showing free air and intraperitoneal free fluid (arrow) around the jejunal tumour.