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Unusual presentation of a giant jejunal gastrointestinal stromal tumor

AIM: *Gastrointestinal stromal tumours (GISTs) are rare tumours. Tumour rupture is an additional adverse prognostic factor and should be recorded, regardless of whether it took place before or during surgery.*
CASE REPORT: *A case report of hemoperitoneum from spontaneous rupture of a gastrointestinal stromal tumor of the jejunum is presented. The patient underwent an urgent laparotomy. An "en bloc" resection was performed.*
CONCLUSION: *The information in the literature is examined. Spontaneous rupture of the tumor with concomitant hemoperitoneum is an important prognostic factor in these patients. The hemoperitoneum contributes to a worse prognosis because of its ability to produce peritoneal seeding*

KEY WORDS: Gist

Introduction

Gastrointestinal stromal tumours (GISTs) are rare tumours [1]. Although quite rare, GISTs are the 0.1-3% of all digestive tract cancers [2]. GISTs are neoplasia originating from the stromal tissue of the digestive tract. They represent the most common subgroup of non-epithelial primary tumours of the gastro-intestinal tract. Their clinical and histological placement is, however, recently acquired because the definition of these tumours has been, for a long time, the subject of disputes and debates. In fact, the GISTs are currently still quite heterogeneous regarding biological behaviour, histogenesis, diagnostic criteria and prognostic factors [3].
There is a slight prevalence in males [4]. The median age is around 60–65 years, with a wide range. Prognostic factors are the mitotic rate, tumour size and tumour site (gastric GISTs have a better prognosis than small bowel or rectal GISTs) [3]. Spontaneous tumour rupture is an additional adverse prognostic factor and should be recorded, regardless of whether it took place before or during surgery. A case report of hemoperitoneum from rupture of a gastrointestinal stromal tumor of the jejunum is presented.

Case Report

A 52–year-old woman with no known comorbidities was brought to the clinic with bowel obstruction. She had no previous history of any surgery or any chronic disease. She denied any previous medication, hematemesis, melena, or weight loss. Her medical and family histories were unremarkable. Physical examination revealed a palpable abdominal mass. The abdomen was distended, with diffuse tenderness and without rebound tenderness. Blood analysis revealed an initial hemoglobin level of 6.0

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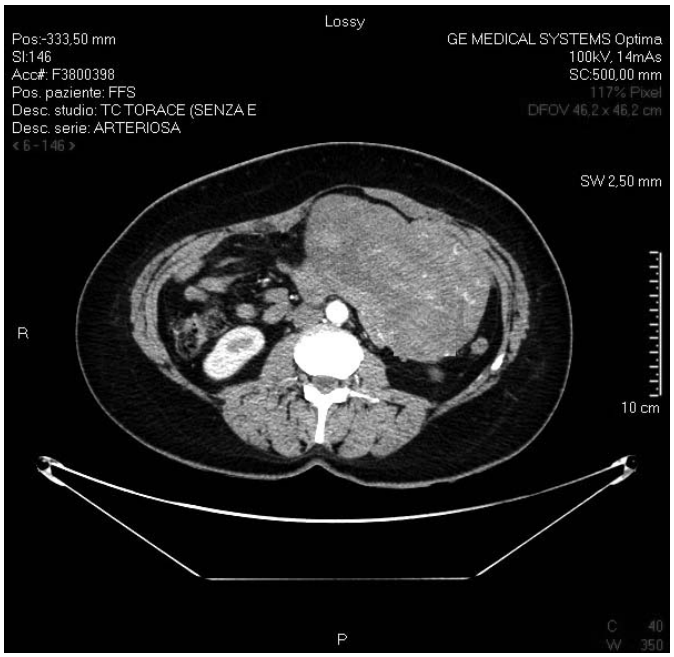

Fig. 1: Abdominal enhanced CT scan.

Fig. 2: Intraoperative findings.

g/dL. The laboratory tests for blood glucose, electrolytes, urea, creatinine, and amylase were normal, as were the liver function tests. An abdominal enhanced CT scan revealed the presence of a heterogeneous solid mass in the left quadrant and intraperitoneal free fluid collection (Fig. 1). The mass was radiologically suspected as a GIST arising from the jejunum and the fluid in the peritoneum indicated tumor rupture.

The patient underwent an urgent laparotomy. Intraoperative findings revealed a 20 × 15 cm bleeding ruptured tumour with the jejunum wrapped around and inseparable from it (Fig. 2). A total of 1500 mL of blood with blood clots was evacuated from the abdominal cavity.

"En-bloc" resection and primary side-to-side anastomosis were performed. No regional lymphadenectomy was performed. The patient recovered well without postoperative complications and was discharged on the fifth postoperative day.

The pathological report confirmed the diagnosis of GIST arising from the jejunum, with evidence of tumor rupture. Histopathology showed a GIST of the jejunum with a mitotic index > 5 x 5 mm^2 high power field (HPF), Ki67 10-15%, high risk.

Patient underwent a post-operative treatment with Imatinib mesylate, 400 mg daily. During the follow up, the patient remains symptom-free with no evidence of recurrence or metastases at 20-months follow up.

Discussion

The combination of hemoperitoneum due to a spontaneous rupture and bowel obstruction is an uncommon presentation of a GIST.1,5-9 Preoperative diagnosis is difficult because of the absence of pathognomonic signs or symptoms [10].

GISTs are visceral tumors arising from any site of the gastrointestinal tract [11]. Approximately 30% of cases occur in the small intestine, and 10% in the jejunum [12,13]. Primary tumors are typically larger than 5 cm and non-homogeneously enhanced on CT scans.

Approximately 20% of patients with a GIST may be asymptomatic. GISTs are usually associated with abdominal pain, bleeding of the gastrointestinal tract, or a palpable mass [3,4,14]. However, a GIST in the small intestine rarely causes hemoperitoneum. Bleeding into the peritoneal cavity on account of a ruptured GIST can lead to acute abdominal pain presenting as a surgical emergency. In the present case, bleeding in the tumor leading to rupture of the capsule may have caused hemoperitoneum.

CT scan plays an important role in the diagnosis and identification of intraluminal or exophytic tumors [3,13, 15]. Tumor size, mitotic activity, pleomorphism of nuclei, degree of cellularity, nucleus/cytoplasm ratio, and mucosal invasion are pathological factors that determine malignancy [1]. In addition to the tumor size and mitotic rate, tumor rupture is believed to be a prognostic factor for the outcome of patients with a GIST [1]. The prognosis is dismal when the tumor presents with symptoms or signs such as perforation or rupture. Our patient was diagnosed with a high-risk malignant GIST because the tumor size and mitotic activity. Also, rupture of the tumor with concomitant hemoperitoneum is an important prognostic factor in our patient. The hemoperitoneum contributes to a worse prognosis because of its ability to produce peritoneal seeding [16]; however, CT scans and intraoperative findings did not reveal macroscopic evidence of distal spread.

The standard treatment of localised GISTs is complete surgical excision of the lesion, with no dissection of clin-

ically negative lymph nodes [1,3,4]. If laparoscopic excision is planned, the technique needs to follow the principles of oncological surgery [13,17]. A laparoscopic approach is clearly discouraged in patients who have large tumours, because of the risk of tumour rupture, which is associated with a very high risk of relapse. R0 excision is the goal (i.e. an excision whose margins are clear of tumour cells). When R0 surgery implies major functional sequelae, and preoperative medical treatment is not effective, the decision can be made with the patient to accept possible R1 (microscopically positive) margins. Surgical resection is the gold standard treatment for a localized, non-metastatic GIST. A laparoscopic procedure is considered to be useful for the treatment of select cases with a GIST in the small intestine. However, in our case, neither diagnostic nor therapeutic laparoscopy was performed because the tumor size and the presence of massive fluid in the peritoneal cavity indicated laparotomy to be more appropriate.

Riassunto

Descriviamo il caso clinico di un tumore stromale del digiuno, giunto alla nostra osservazione in emergenza per un quadro di emoperitoneo. L'emoperitoneo era secondario alla rottura spontanea del tumore. Il caso è stato trattato chirurgicamente. Riteniamo che la chirurgia R0 rappresenti la terapia ottimale nei pazienti affetti da queste rare neoplasie. Riteniamo che la laparoscopia debba essere riservata al trattamento delle neoplasie di dimensioni contenute.

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