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Large gastrointestinal stromal tumour of the proximal jejunum with internal fistula

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ABSTRACT:

Gastrointestinal stromal tumours (GISTs) are malignant, rare neoplasms that comprise 0.2% of gastrointestinal tumours. The small intestine is the second most common location after the stomach. The authors present a 63-year-old female patient with an history of abdominal pain and weight loss. The image exams revealed a jejunal tumor, with a saccular pattern, 4cm parietal thickness, and central necrosis with apparent communication to the jejunal loop lumen. Surgical resection was performed, and operative findings revealed an 18×15cm tumor located 10cm from the angle of Treitz. The histology confirmed it to be a high-grade GIST, and imatinib mesylate 400mg once daily was given as adjuvant chemotherapy.

Keywords: Abdominal mass. GIST. Jejunum. Surgical resection.

INTRODUCTION

With an incidence of 2/100.000, GISTs comprise a distinct group of rare gastrointestinal tract tumours that originate from the interstitial cells of Cajal, which are involved in the regulation of gastrointestinal motility by pacemaker activity. Men and women are equally affected, and the incidence peak occurs around 50-60 years of age [1].

They can occur anywhere in the gastrointestinal tract, being most frequent in the stomach (60%) followed by small bowel (30%). Jejunal GIST are extremely rare accounting for 0.1-3% of all gastrointestinal tumours [1]. Because GISTs in the small bowel tend to have an exophytic grow pattern, they are usually asymptomatic. As they grow, can lead to a variability of symptoms such abdominal pain, a palpable mass or weight loss [2].

Complete resection is the only potential curative treatment for GISTs. Caution with the dissection is essential to not injure the pseudocapsule, which can leave to dissemination of the tumor cells. In the case of large GISTs with central necrosis, dissection with extremely care is necessary because the pseudocapsule usually is fragile. Lymphadenectomy is not necessary because lymph nodes metastasis is quite rare [2].

GISTs are categorized as low, intermediate or high risk of recurrence, based on tumour size, mitotic rate, location and presence of rupture [2].

Over 95% of GIST cells have mutations in one of the two genes, called cKIT (CD117) and PDGFRα. Imatinib is a tyrosine kinase inhibitor that targets both genes. Until 2001, when imatinib was introduced, the overall 5-year

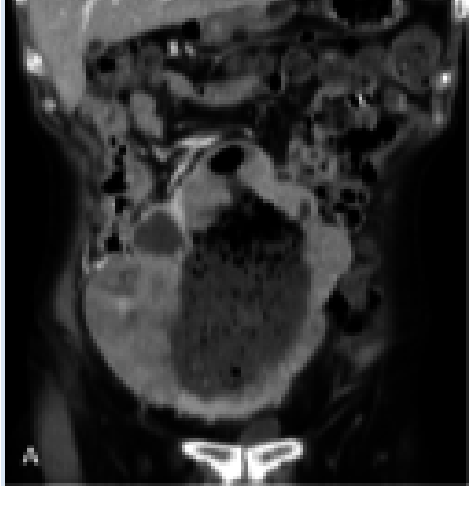
survival rate was 45–55%. Fortunately, the recurrence rate was significantly reduced, achieving 97% recurrence-free survival [3]. According National Cancer Institute recommendations, patients with GISTs categorized as high-risk, benefited more from treatment with imatinib [3].

CASE STUDY

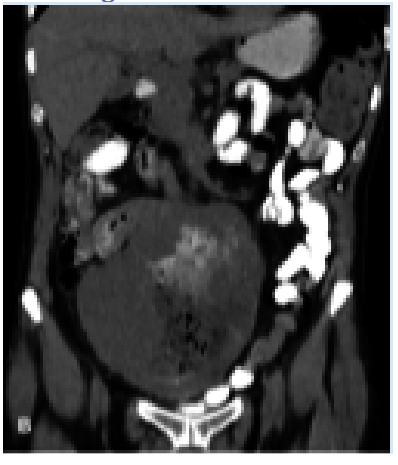
A 63-year-old female patient presented with an history of abdominal pain and weight loss in the last 2 months. The physical examination revealed a hard mass in the right quadrants of the abdomen. The abdominopelvic ultrasonography showed a large tumour probably related to the right ovarium. For better characterization, a computed tomography (CT) scan was performed and revealed that the tumour was originated from the proximal jejunal, characterized as an intestinal dilatation with a saccular pattern and a 4 cm parietal thickness (Figure 1). This exophytic saccular dilation showed alimentary luminal content as well as gas and oral contrast projection. The entry point and the exit point of the described lesion were adjacent to each other which resulted in an exophytic saccular aspect of the lesion (Figure 1).

Figure 1:

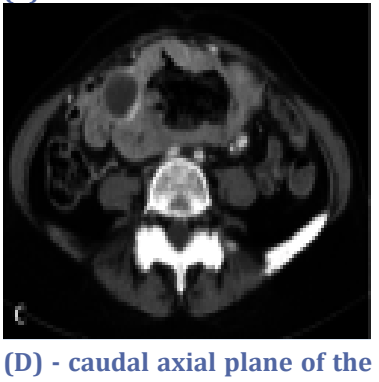
(A) - Coronal plane of the CT scan showing a tumor originated from the proximal jejunum, characterized by a marked intestinal dilation with a saccular configuration, with a very thick wall (4 cm of maximum parietal thickness).



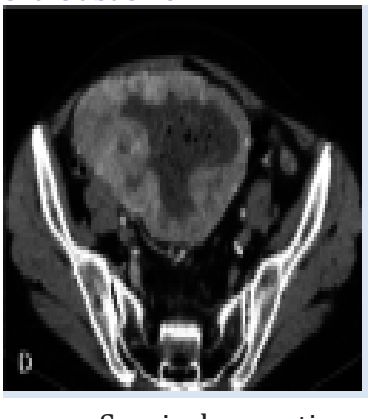
(B) - Coronal plane of the CT scan showing a saccular dilation with luminal food content as well as gas. It is also possible to observe some contrast projection for the lumen to this pathological segment of intestinal loop. The entry point and the exit point of the described lesion are adjacent to each other which results in a saccular aspect of the lesion with respect to these segments.



(C) - Cranial axial cut of the CT scan



(D) - caudal axial plane of the CT scan showing a lesion measuring 16.5 x 14.5 x 7.5 cm that occupies a large part of the mesogastric and hypogastric region of the abdomen



Surgical resection of the tumour was carried out. Intraoperatively, a well vascularized saccular tumor with almost 17x15 cm was in the jejunum, 10 cm from the treitz angle. An enterectomy was performed with removal of the tumor (Figure 2).

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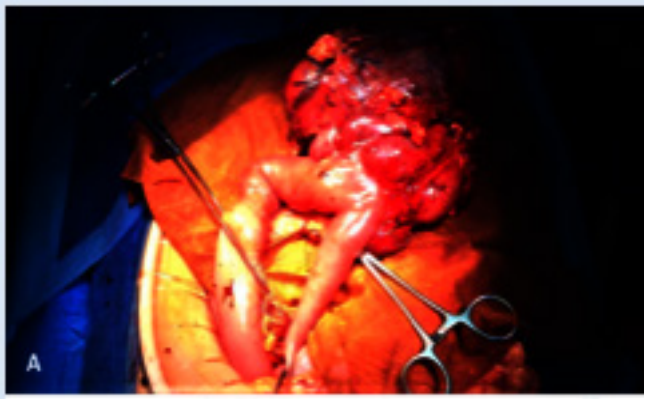
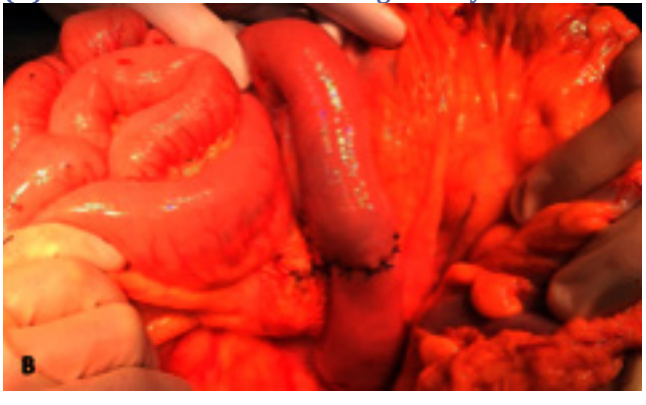
Figure 2:
(A) – Intraoperative pictures showing a tumor arising from the antimesenteric wall of the jejunum.

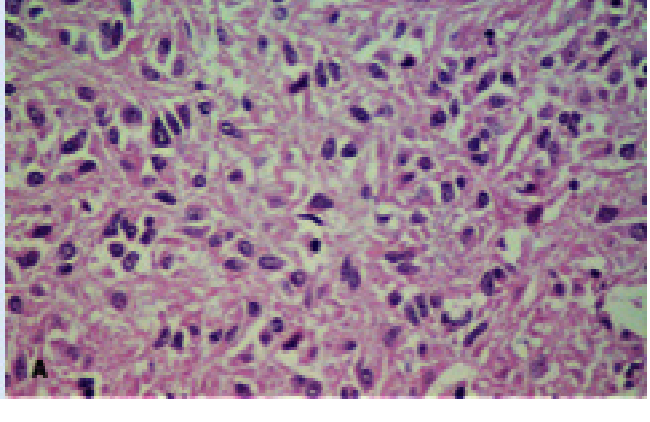


(B) – Manual anastomosis using two layers of suture.

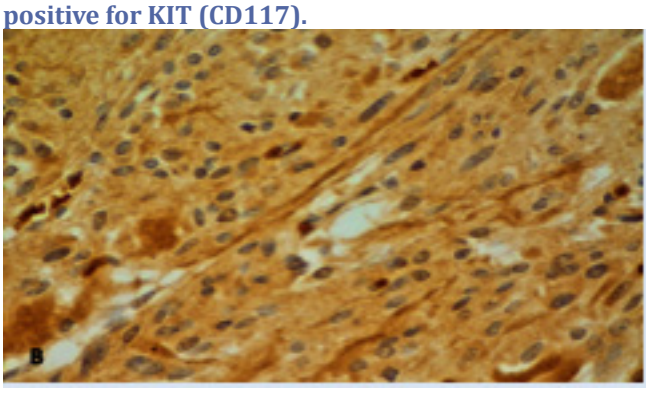


Macroscopically, the intestinal loop presented mucosa with preserved folds, a perforation area that communicated with the lesion described. Cut surface was greyish and the lesion had a wall 4 cm thick, with abundant blood content and food remains. A large area of central necrosis was observed, and the mitotic index was low (3 mitosis/50hpf). The lesion was composed of areas of spindle and epithelioid cells, and immunohistochemical analysis showed positive staining with BcL2 and CD117 (Figure 3).

Figure 3:
(A) - Image (H&E stain; original magnification ×400) showing oval to spindle cells arranged in fascicles



(B) - Image (IHC stain; original magnification ×400) of immunohistochemical study showing tumor cells positive for KIT (CD117).



The histopathological appearance and immunohistochemical profile confirmed it to be a high-grade GIST.

The patient had an uneventful postoperative course and was proposed to adjuvant chemotherapy with imatinib mesylate 400mg once daily. After 26 months, there is no evidence of recurrence.

DISCUSSION

Jejunal GIST are extremely rare accounting for 0.1-3% of all gastrointestinal tumours [1]. Usually these tumours are asymptomatic, however, with the mass growth the patient may become symptomatic. Complete resection is the only potential curative treatment for GISTs. The knowledge of the molecular behaviour of the GISTs allowed the discovery of the target therapy, such imatinib, which is known to have an important impact on the survival of these patients.

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