

Justapapillar Duodenal Gastrointestinal Stromal Tumor (gist) Local Resection: A Case Report

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Introduction: Gastrointestinal stromal tumors (GISTs) are the most frequent non-epithelial tumors of the gastrointestinal tract (GIT). The most common location is the stomach, followed by small intestine, being very rare the cases of duodenal origin, where it can cause digestive bleeding and anemia. The surgical resection of the tumor is the gold-standard treatment and the definitive diagnosis is based on immunohistochemical analysis of the surgical specimen.

Case presentation: A case of exophytic and endophytic GIST located in the second portion of the duodenum, one centimeter below the duodenal papilla, was reported in a 33-year-old female patient.

Conclusion: The gold-standard treatment is surgical resection of the tumor with negative margins (R0), with no need for lymphadenectomy. Local lesion resection or duodenopancreatectomy can be performed. Duodenopancreatectomy, unlike local resection of the lesion, is associated with increased length of hospital stay and longer intraoperative time. Therefore, it should be reserved for lesions that cannot be resected locally. Fortunately, a local resection was performed, which have a more favorable prognosis.

Keywords: Gastrointestinal Stromal Tumors; Justapapillar duodenal GIST; Gastrointestinal neoplasms

Received: February 7, 2018; **Accepted:** March 3, 2018; **Published:** March 10, 2018

Competing Interests: The authors have declared that no competing interests exist.

Consent: Consent was taken from the patient for publication of this case report.

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Introduction

Gastrointestinal stromal tumors (GISTs) are the most frequent non-epithelial tumors of the gastrointestinal tract (GIT) [1] and represent approximately 0.1-3% of all gastrointestinal tumors [2]. Its histological origin is related to the interstitial cells of Cajal, which act in the physiological control of peristaltic activity, as an electric smooth muscle pacemaker. GISTs can affect any segment of the gastrointestinal tract, from the esophagus to the anus, where it has a worse prognosis [3]. The sites of higher incidence are the stomach (50-60%) and small intestine (25%), followed by colon and rectum (10%) and esophagus (<5%) [4,5]. GISTs originated in the duodenum are very rare and represent only about 5% of all GISTs [4]. They may also occur in the peritoneum, omentum and mesentery [4,6]. The majority of the tumors are asymptomatic or present inconsequential symptoms such as abdominal pain (43%), upper gastrointestinal bleeding (15%) and palpable abdominal mass (17%), in addition to chronic anemia [6,7]. They occur most commonly in men and after 50 years of age [6]. The definitive diagnosis is given by immunohistochemical analysis of the surgical specimen, with a search for the presence of CD117 receptors (c-Kit) [8]. The gold-standard treatment is a surgical resection of the tumor with negative margins (R0), without the need for lymphadenectomy [4]. In cases of malignant GIST, recurrence or when there is no possibility of resection, imatinib mesylate, a tyrosine kinase inhibitor can be used as an option of treatment [9].

Case presentation

A 33-year-old female patient, Caucasian, presenting history of digestive bleeding with hypovolemic shock, who needed blood transfusion. No comorbidities, no use of continuous medication and no drug atopies. On physical examination, she was in good general condition, lucid, attentive and coherent, with normocorrate and anicteric mucosa, BMI 38 kg/m². Abdominal auscultation with presence of normal hydro-aerial sounds, superficial and deep abdominal painless palpation, with no signs of peritoneal irritation. The laboratory tests were normal. Echoendoscopy showed the fourth-layer vascularized sub epithelial lesion in the second duodenal portion (GIST), approximately 1 cm below the duodenal papilla, in which fine needle aspiration (FNA) was performed. Pathologic examination revealed fusiform cell neoplasia (Figure 1 and 2). The immunohistochemical study was positive for CD117 and DOG1 and was negative to Actin ML, suggesting GIST. Computed Tomography (CT) scan of abdomen identified lesion close to the medial wall of the duodenum, involving the second portion of the duodenum and the transition from the second to third portion, nodular, ovoid, lobed, measuring 2.8 x 2.6 cm, with predominantly hyper vascular arterial and portal impregnation, by means of contrast, being still hyper dense in the late phase of indeterminate nature. No evidence of intra and extrahepatic biliary dilatation and pancreatic dilatation or ductal ectasia (Figure 3).

Based on echo-endoscopic, computed tomographic, anatomopathological and immunohistochemical findings, the patient underwent laparotomy with bilateral subcostal transverse incision. Tumor lesion was identified on topography of the second duodenal portion distal to the duodenal papilla by about 1.2 cm in the anteromedial wall of the duodenum, with exophytic and endophytic growth and ulceration in the inner part of the tumor. Extensive Kocher maneuver, cholecystectomy with choledochal duct catheterization and exit of the catheter by duodenal papilla and identification of the same was performed (Figure 4). From the evaluation of the local resectability of the lesion (being considered resectable locally), partial duodenal release of the pancreas head was performed, followed by complete local resection of the tumor lesion with negative margins (confirmed by intraoperative frozen section) and primary closure of the duodenum with suture in two planes

(Figure 5 and 6). Anatomopathological examination identified oval cells neoplasia, measuring 3 x 2.8 cm, and presence of 2 mitoses per 50 high power field (HPF). No tumor necrosis, angiolymphatic and perineural invasion were identified and the surgical limits were confirmed free of neoplasia (Figure 7). Immunohistochemical study was performed using the indirect immunoperoxidase method with diaminobenzidine (DAB). Microscopically, neoplastic cells exhibited positivity with antibodies DOG1, C-kit and Ki-67 (3%). There was not positivity with antibodies Actina, CD34 e S100. Therefore, the exam concluded the diagnosis of gastrointestinal stromal tumor with fusocellular and epithelioid pattern, with proliferative index of 3%.



Figure 1 Upper Digestive Endoscopy showing justapapillar ulcerated subepithelial tumor lesion

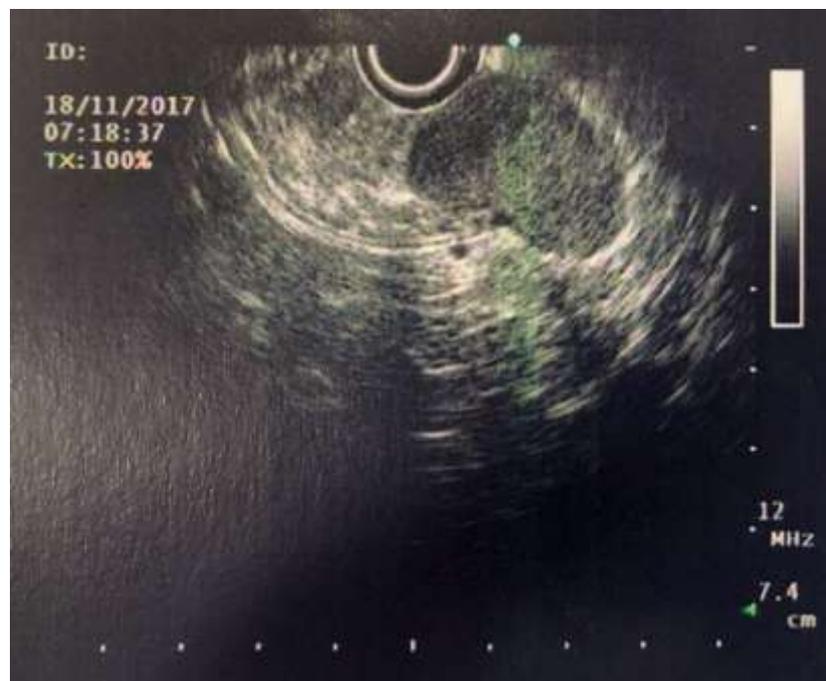


Figure 2 Echoendoscopy showing lesion in the fourth duodenal layer

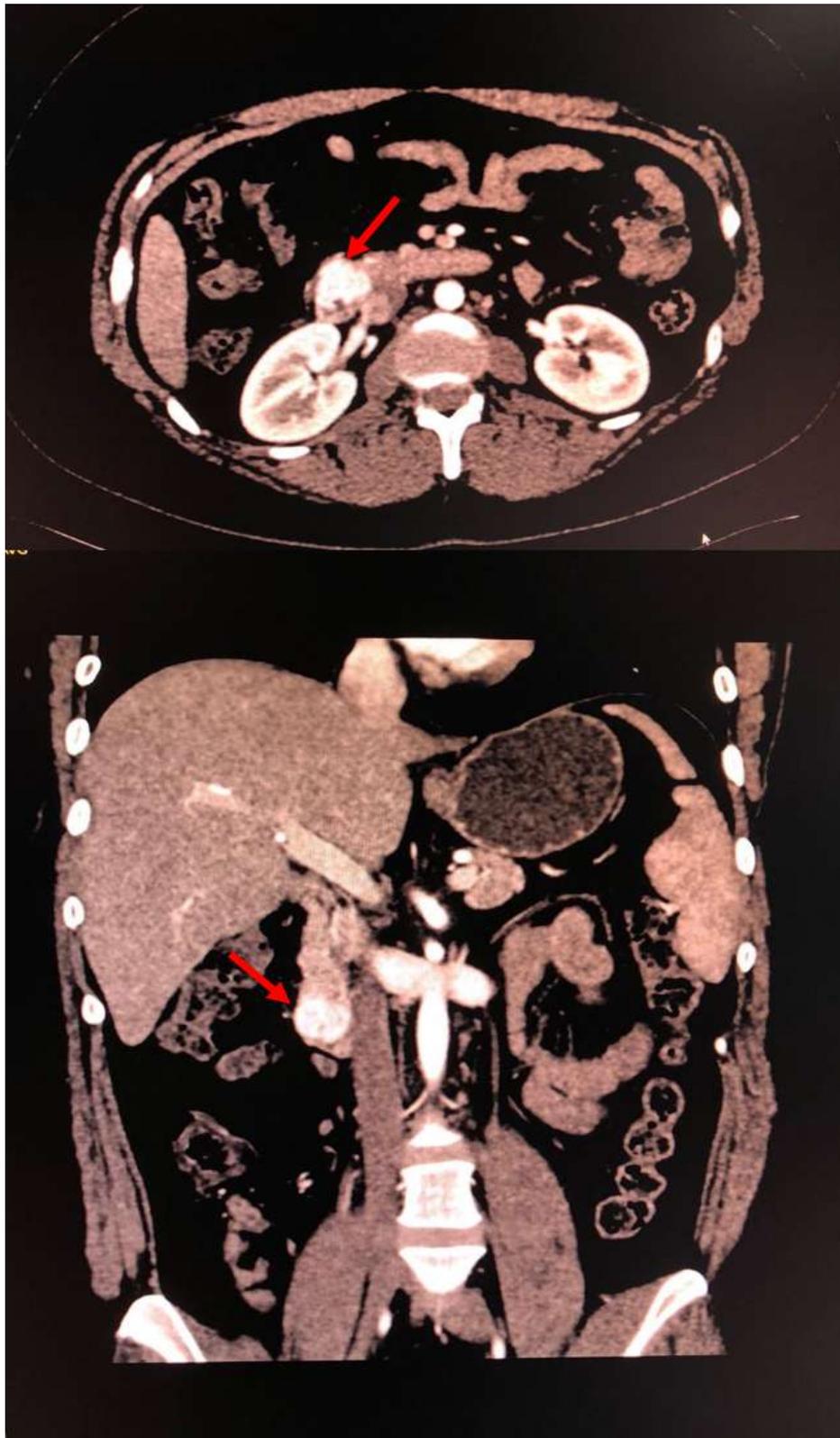


Figure 3 Computed tomography showing lesion in the second duodenal portion

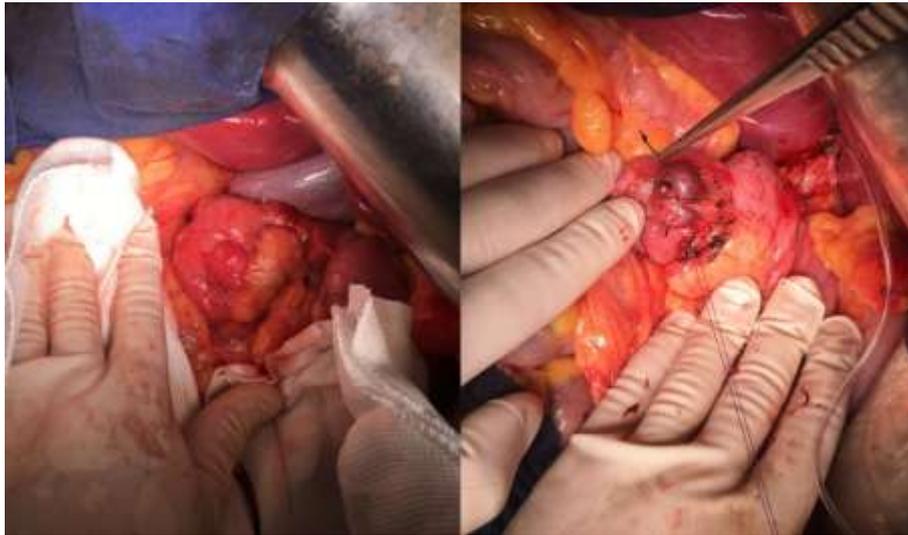


Figure 4 Lesion before and after duodenal release

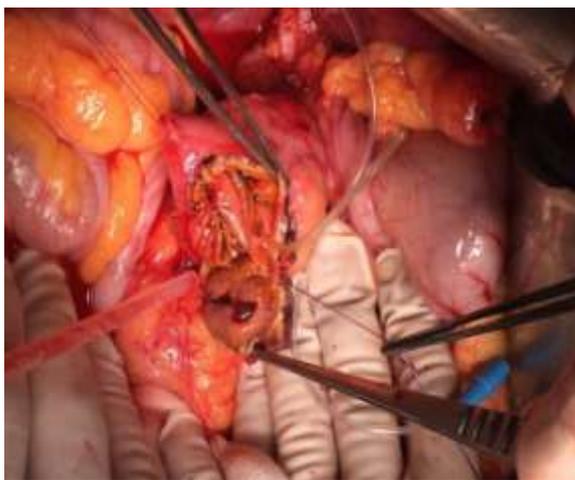


Figure 5 Resection with catheterized duodenal papilla



Figure 6 Duodenal closure



Figure 7 Resected tumor

Discussion

GISTs can occur at any part of the GIT [3] and could be originated at any age, with its higher incidence at 60 years of age. In addition, they affect the male and female population equally [10,11]. The main morphological parameters of differentiation of biological behavior related to malignancy and benignity are based on the size of the lesion and the number of mitoses per high power field (HPF) [6]. Regarding the immunohistochemical characteristics, the CD117 protein (KIT), which is expressed in about 95% of the GISTs, is helpful to differentiate them from the other sarcomatous tumors of the gastrointestinal tract. On KIT-negative lesions, the marker DOG1 is useful in the diagnosis, since it is identified in approximately 97% of the cases. Other immunohistochemical markers such as SMA, S100 Protein, PKC theta and Desmin can help in the diagnosis as well [6]. When originated in the small intestine, the jejunum is the most affected part (30 to 45%), followed by the ileum (17 to 27%) and duodenum (5 to 10%). The gold-standard treatment is surgical resection of the tumor with negative margins (R0), with no need for lymphadenectomy, since lymph node involvement is very rare [4]. As for the surgical technique, a resection with a margin of 1-2 cm from the lesion is usually recommended, although microscopic margins that are negative for tumor cells are sufficient for a complete resection and curative possibility [13]. However, based on the duodenal location of the lesion in our patient's case, it should be remembered that, unlike resections in the stomach, there is a greater complexity in locally resecting lesions in the duodenum, due to direct proximity to the pancreatic head, duodenal papilla and mesenteric roots [4]. In relation to surgical techniques, local lesion resection or duodenopancreatectomy can be performed. Duodenopancreatectomy, unlike local resection of the lesion, is associated with increased length of hospital stay and longer intraoperative time. Therefore, it should be reserved for lesions that cannot be resected locally [14]. The prognosis of patients with GIST varies according to the location of the lesion. Although small bowel lesions present a worse prognosis than gastric lesions, they have a more favorable prognosis than those with colorectal and extragastrointestinal GISTs [15].

Conclusion

Gastrointestinal stromal tumors (GISTs) are the most frequent non-epithelial tumors of the gastrointestinal tract (GIT) [1]. The gold-standard treatment is surgical resection of the tumor with negative margins (R0), with no need for lymphadenectomy [4]. In relation to surgical techniques, local lesion resection or duodenopancreatectomy can be performed. Duodenopancreatectomy, unlike local resection of the lesion, is associated with increased length of hospital stay and longer intraoperative time. Therefore, it should be reserved for lesions that cannot be resected locally [14]. Fortunately, a local resection was performed, which have a more favorable prognosis.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Acknowledgements

The authors reported no conflict of interest and no funding was received for this work.

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