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## Asymptomatic large duodenal GIST – An incidental finding in abdominopelvic ultrasonography: A case report

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## ABSTRACT

**INTRODUCTION:** Duodenal GISTs are rare mesenchymal tumors of the digestive tract. They usually present as gastrointestinal bleeding or non-specific abdominal pain, but they can also be asymptomatic. High-risk GISTs have malignant potential. Surgical resection is the only curative option and its extension depends on different factors. Tyrosine kinase inhibitors are of the utmost importance in high-risk and metastatic disease.

**CASE DESCRIPTION:** We herein report the case of a 75-year-old male, referred to our general surgery outpatient clinic, with an asymptomatic duodenal mass incidentally diagnosed during an abdominopelvic ultrasonography. Abdominal CT suggested GIST. Since no other lesions were found, a segmental duodenectomy was performed and the patient recovery was uneventful. Histologic exam confirmed the presence of a high-risk GIST, therefore adjuvant therapy with imatinib was initiated.

**DISCUSSION:** Duodenal involvement by GISTs is rare, even more in its third/fourth portions. The widespread clinical use of radiological exams such as ultrasonography may lead to the diagnosis of asymptomatic masses. Surgical treatment is the gold-standard giving the malignant potential of these neoplasms. There are risk stratification scores that may be used to assess the need for adjuvant therapy. **CONCLUSION:** Even large GISTs can be asymptomatic. Giving their malignant potential, early diagnosis and treatment are crucial. The histologic findings and genetic profile have an impact on the patient's survival.

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## 1. Introduction

Duodenal GISTs (gastrointestinal stromal tumors) are mesenchymal neoplasms that arise from the interstitial cells of Cajal or their precursors [1]. They can appear at any site in the gastrointestinal tract but they are more common in the stomach (60%), with only 5% originating in the duodenum [2,3], usually in its second portion (D2) [4,5]. Gastrointestinal bleeding and unspecific abdominal pain are the most common clinical presentations of these tumors [2–4]. Small gastric GISTs are sometimes diagnosed as incidental findings in routine endoscopic imaging [5]. According to the current evidence, surgery is the only potentially curative treatment for non-metastatic GISTs, as long as a R0 resection with negative margins is performed [2,3]. The type of surgery may vary from a segmental duodenectomy to a pancreatoduodenectomy, depend-

ing on the site of the tumor and its histopathological characteristics [4].

These neoplasms are the consequence of activating mutations either in one protein tyrosine kinase receptor: KIT (CD117) or in the platelet-derived growth factor receptor alpha (PDGFRA) [6]. Knowing this allowed the introduction of tyrosine kinase inhibitors (TKIs), like imatinib and sunitinib, as a therapeutic option or even the standard one, in patients with high-risk or metastatic disease [6,7].

We herein present a case of an asymptomatic large duodenal GIST in an unusual location incidentally diagnosed during an abdominopelvic ultrasonography.

The present work has been reported in line with the SCARE criteria [8].

## 2. Case description

A 75-year-old male, with history of arterial hypertension, type 2 diabetes, thyroid nodules and benign prostatic hyperplasia, was referred to our general surgery outpatient clinic after undergoing a

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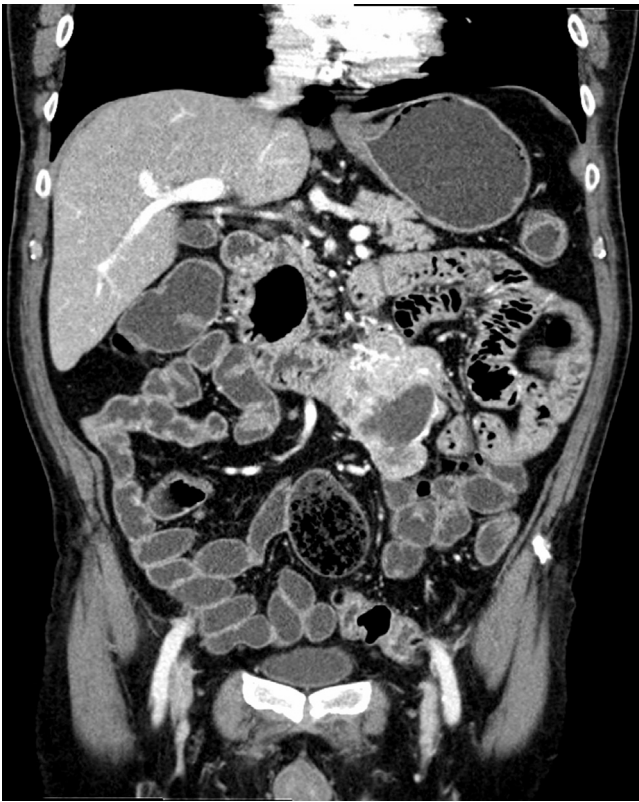


Fig. 1. CT-enterography image of the lesion.

pelvic ultrasonography in October 2019, for prostatic evaluation. During this exam, a reniform-shaped mass, measuring approximately  $92 \times 37$  mm with a cystic area of 29 mm, was incidentally found in the middle third of the abdomen, left to the midline.

The patient did not present any relevant gastrointestinal symptoms and had no complaints of fatigue or weight loss. No blood loss was documented and the physical exam was normal.

Taking this into consideration, the patient underwent an abdominal CT, which revealed a well-defined solid and cystic mass, oval-shaped, in the third/fourth portion of the duodenum (D3/D4), with central necrosis and small calcifications, measuring  $6 \times 4,6$  cm, suggesting GIST.

An upper endoscopy could not reach the lesion therefore it was not possible to perform an endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) biopsy.

A CT-enterography was obtained in January of 2020. The report described “in the transition between D3 and D4, an exophytic lobulated mass, apparently originating from the duodenal wall, heterogeneous, with hypodense central regions (of necrosis) and peripheral contrast-enhancement areas, measuring  $64 \times 53 \times 73$  mm, molding its shape to conform to the adjacent structures, without clear invasion (...)” (Fig. 1). Once again, the most likely diagnosis was duodenal GIST.

The patient was therefore proposed for open surgical resection of the lesion. A supra-umbilical vertical midline incision was made to access the abdominal cavity. Then, the right white line of Toldt was incised allowing the liberation of the right colon and the exposure of D2 and D3. No abnormal mesenteric rotation was observed. The gonadal vessels and right ureter were identified and preserved and the inferior vena cava and aorta were exposed. We found a bulky neoplasm compatible with GIST, with approximately 10 cm in diameter and prominent neovascularization, involving D4 with extension to the Treitz’s angle (Fig. 2). We separated the duodenum from the pancreas, ligating the neovessels and the pan-

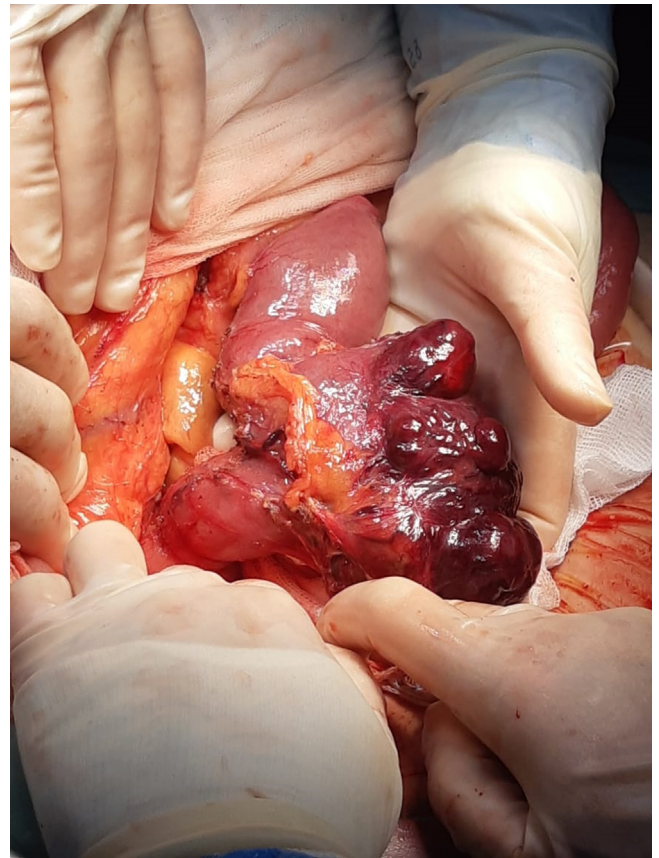


Fig. 2. GIST in the fourth duodenal portion.

creaticoduodenal vascular pedicles to the distal portion of D3 and D4. The superior and inferior mesenteric vessels were identified. A segmental duodenectomy (D3–D4) with a two-layer interrupted end-to-end anastomosis (Fig. 3) was then performed. The procedure was performed by a specialist with appropriate training and experience and it was uneventful. The operative specimen is presented in Fig. 4.

The patient recovered well and was discharged by the eighth day. On the first and fourth months post-surgery, upon follow-up evaluations, he was feeling well, tolerating a normal diet, with no symptoms whatsoever.

The histologic examination confirmed the diagnosis of a spindle cell tumor, with morphological characteristics compatible with GIST of predominantly epithelioid cell type and mitotic rate of more than 5 per  $5 \text{ mm}^2$ . This classifies the tumor as a high-risk GIST, with a rate of progression of 85%, according to the AFIP (Armed Forces Institute of Pathology) criteria [9,10]. The microscopic margins were negative and there was no evidence of tumor rupture. Hence, it was classified as a pT3, according to the TNM classification. In what concerns the immunohistochemical features, this GIST was positive for CD117 and SMA (smooth muscle actin) and negative for CD34, PS100, Desmin and Cam 5.2.

The case was ultimately discussed in a multidisciplinary meeting and the patient referred to a medical oncologist in order to initiate adjuvant therapy with imatinib.

### 3. Discussion

Despite being the most common mesenchymal neoplasms of the gastrointestinal tract [6], GISTs are rare tumors with an estimated annual incidence of 1/100.000 diagnosed cases [7]. They affect mostly people in their sixties with a slightly higher preva-





Fig. 3. End-to-end duodenojejunal anastomosis.



Fig. 4. Final operative specimen.

lence in the male gender [4,7]. In this case, the age of diagnosis was higher than average. Duodenal involvement by this kind of tumor is rare, but when it occurs, it usually affects D2.

Gastrointestinal bleeding is the main clinical manifestation of GISTs [2–4], that was not found in this patient, who was incidentally diagnosed during an abdominopelvic ultrasound. Median size of GISTs is 4 cm [2], whereas this patient's tumor was twice that size.

The diagnostic workup usually starts with an endoscopic examination, especially when investigating the source of an alimentary tract hemorrhage. Nonetheless, more distal lesions, as the one described here, may not be reached by a conventional endoscope and therefore subsequent image exams are usually necessary.

Abdominal ultrasound may play a role in diagnosing the mass but a CT scan or a MRI is mandatory for better lesion characterization and staging of the disease [4].

The management of GISTs should be conducted by a multidisciplinary team [7]. Current evidence states that surgery is the gold standard for local disease, but the best surgical technique remains controversial and may vary according to tumor size and location as well as invasion of adjacent structures [2,4,11]. In this case, the patient underwent a segmental duodenectomy, since the lesion was located on D4, with no involvement of the Vater's papilla or the pancreas, excluding the need for a pancreatectomy (the usual approach for D2 lesions) [4]. Several studies and the meta-analysis published by Chock et al. showed no statistically significant differences in the survival rate or recurrence among patients who underwent limited resections and the ones in whom a pancreatoduodenectomy was performed [4]. In fact, the recurrence of GISTs appears to have a stronger connection to the tumor biology rather than the kind of surgery performed [3].

All GISTs have malignant potential, which appears to be higher in those arising in the small intestine comparing to the gastric ones [5,6]. Histologically, the best known prognostic factors are tumor size and the mitotic index (number of mitoses per 50 high power fields or 5 mm<sup>2</sup>) [6,9,10]. Tumor rupture is also a poor prognosis factor [4].

There are several risk stratification scores available for GISTs [10]. We used the AFIP criteria, the one recommended by the European Society for Medical Oncology (ESMO) [7]. This score has the advantage of including not only tumor size and mitotic rate, but also the location of the tumor in estimating the rate of disease progression.

Moreover, GISTs have a characteristic immunohistochemical staining profile, which helps confirming the diagnosis. About 95% stain positive for KIT (CD117) [6], which was observed in this case. Other markers include the CD34 antigen, which was negative, since it usually has a stronger expression in esophageal, gastric and rectal GISTs. The SMA staining was positive, which is also frequently reported in the literature, especially in small bowel tumors [6].

Taking into consideration that this patient presented with a high-risk GIST, there is a formal indication for adjuvant therapy with a TKI. In fact, since 2010, the adjuvancy with imatinib, in a daily dose of 400 mg, has been recommended for patients with high-risk GISTs [6,11]. Sunitinib may be used as an alternative for patients with imatinib-resistant disease [6]. The duration of this treatment should be three years, as some trials reported a higher disease-free survival rate in these patients, comparing to those who underwent only one year of therapy [7].

The optimal follow-up length is still unknown for patients with local disease, in whom a R0 resection was performed. Some hospitals choose to order abdominal CTs or MRIs every 3–6 months in the first 3 years of imatinib therapy for patients with high-risk GISTs, with successively larger intervals in the following years [7].

#### 4. Conclusion

This case report shows that, despite being rare and often diagnosed in the context of a gastrointestinal bleeding, GISTs can be asymptomatic, even when they have already reached a considerable size. En bloc resection is the gold-standard treatment for local disease, considering all GISTs have a malignant potential, especially those arising in the duodenum. The histologic examination of the operative specimen plays a key role in assessing the need of adjuvant therapy, with a significant impact on the patients' survival.

**Declaration of Competing Interest**

Nothing to declare.

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**Ethical approval**

This study is exempt from ethical approval at our institution.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

**Author contribution**

Andreia J. Santos – data collection, data analysis and interpretation and writing the paper.

André Tojal – data collection and data analysis.

Liliana Duarte – operated the patient, data analysis and interpretation and manuscript review.

Conceição Marques, Luís F. Pinheiro and Carlos Casimiro – manuscript review.

**Registration of research studies**

Clinical case report, not a formal research project.

**Guarantor**

Liliana Duarte.

**Provenance and peer review**

Not commissioned, externally peer-reviewed.

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