

# Limited Wedge Resection of a Duodenal Gist in a Patient with Intermittent Melena

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## ABSTRACT

Gastrointestinal Stromal Tumors account for 1% - 2% of all gastrointestinal (GI) tract tumors. Among GISTs, duodenal localization occurs in less than 5% and usually presents with upper GI bleeding. A 45-year-old man presented in the outpatient department with complaints of epigastric discomfort, intermittent melena and undocumented weight loss for the preceding 3 months. Initial upper GI endoscopy showed mild duodenitis and no other upper GI pathology. For unexplained symptoms, a CT Scan was performed which demonstrated a well-defined solid lesion along the second part of the duodenum. An endoscopic ultrasound (EUS)-guided biopsy of a subepithelial lesion at D2 was performed. Immunohistochemistry findings were suggestive of GIST. Wedge resection of the duodenal mass was done.

Duodenal GIST should be considered as a differential in cases of GI bleeding when other differentials have been ruled out. Limited resection of duodenal GIST should be considered over pancreaticoduodenectomy, in case of small size tumors.

**Keywords:** *Wedge resection, duodenal gastrointestinal stromal tumors, melena.*

## INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are believed to originate from the interstitial cells of Cajal or their precursors, located in the entire gastrointestinal tract [1]. Gastrointestinal stromal tumors (GISTs) are among the rare tumors of the digestive system, accounting for 1% to 2% of all gastrointestinal neoplasms [2]. Annual incidence varies from 4.3 to 22 per million population in different geographic regions, whereas the worldwide yearly incidence is 10 to 15 per million [3]. These neoplasms have genetic predilection and are seen in association with other familial oncological syndromes like Carney complex, Von Hippel-Lindau syndrome (VHL) and Multiple endocrine neoplasia Type 1 and Type 2 [4]. The most common location of Gastrointestinal stromal tumors in the gastrointestinal tract is the stomach (approximately 60%), followed by the small intestine (about 20–30%), and rarely in the duodenum (less than 5%) [1, 5]. Interestingly, duodenal gastrointestinal stromal tumor (DGIST) accounts for nearly 30% of all primary tumors of the duodenum [5, 6]. The most frequent site of these tumors in the duodenum is the second portion of the duodenum followed by the third, fourth and first portion. Duodenal gastrointestinal stromal tumors (DGISTs) present with gastrointestinal bleeding more commonly compared to GISTs at other locations in the alimentary tract due to increased submucosal vascularity [1]. Gastrointestinal stromal tumors (GISTs) pose malignant potential but only 10%-30% are clinically malignant [6]. Proto-oncogene proteins,

c-Kit mutations and platelet-derived growth factor (PDGF- $\alpha$ ) are characteristically present in GIST [4]. Duodenal GIST resection is difficult due to the complex relation of structures around it. Limited resection (LR) has comparable results to pancreaticoduodenectomy (PD). Given the low incidence, a comparison amongst wider and limited resection is difficult [6] and institutional practice dictates management in most cases.

## CASE PRESENTATION

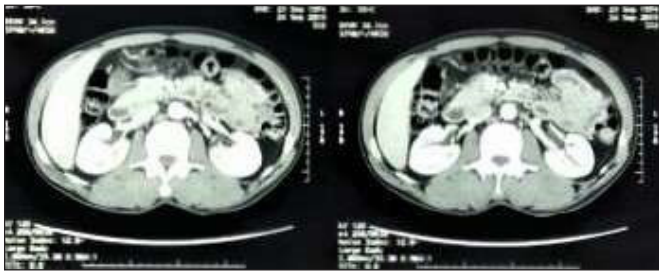
A 45-year-old man presented to the gastroenterology clinic with epigastric discomfort and intermittent episodes of lower gastrointestinal bleed for the last 3 months associated with undocumented weight loss.

The patient described abdominal cramps and the passage of black stool, intermittently. The associated weight loss was undocumented but noted by the patient himself based on the loosening of his clothes. His past medical history and family history were unremarkable. On examination, the patient was mildly anemic with a soft, non-tender abdomen. His prior workup, performed at other institutions included upper GI endoscopy and duodenal biopsy that showed mild non-specific duodenitis. A CT scan was then performed to further evaluate unexplained symptoms, that showed a well-defined solid isodense lesion along the second and third part of the duodenum, abutting the pancreas, measuring 24x22 mm, likely arising from the duodenal wall and protruding into its lumen. The pancreas appeared unremarkable and no other bowel abnormality was noted either (Fig. 1). Repeat upper GI endoscopy was performed and it showed moderate gastritis and a duodenal submucosal bulge with linear superficial ulcer and a bleb. Colonoscopy was also done to exclude any other

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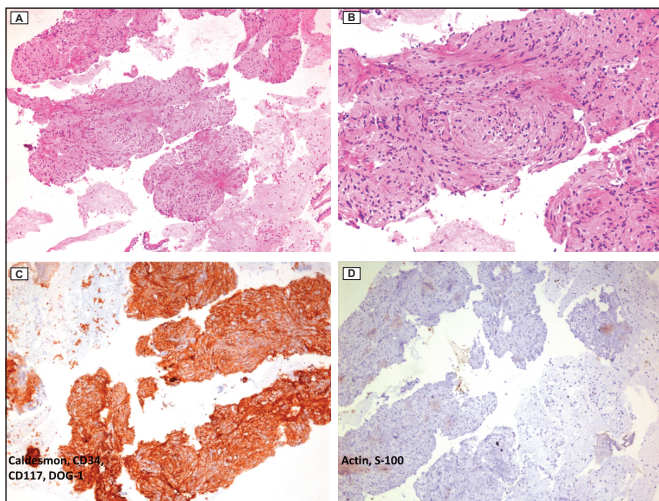
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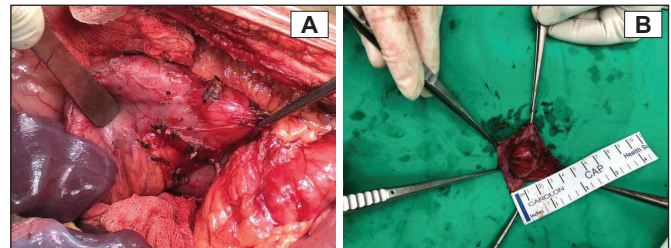
**Fig. (1):** The abdominal CT scan shows a soft tissue mass of size 24 x 22 mm located in second portion of duodenum.

cause of melena and was unremarkable. Subsequently, EUS was performed and it demonstrated a well-defined mass measuring 24x28 mm arising from the duodenal wall beyond the ampulla in D2/D3, along the medial wall. No vascular or pancreatic invasion or lymphadenopathy was seen. EUS guided biopsy of the subepithelial lesion at D2 was performed. Microscopic features showed fragments of glandular mucosa along with fragments of a neoplastic lesion composed of bland-looking spindle cells, containing pale to eosinophilic cytoplasm and oval to spindle nuclei. No necrosis and no mitosis were seen. Immunohistochemistry analysis showed that tumor stained positive for markers Caldesmon, CD117, DOG1, CD34 whereas it was negative for Actin and S-100. All these findings were suggestive of gastrointestinal stromal tumors (**Fig. 2**). Considering the small size of the lesion and low-grade characteristics on histopathology, a decision to perform a more conservative procedure instead of pancreaticoduodenectomy was made and the patient underwent wedge resection of the duodenal GIST following ERCP stent placement to avoid ampullary injury.



**Fig. (2):** Histopathological findings showing spindle shaped cells (**A, B**) and immunostaining showed that the tumor cells were positive for CD34, CD117, Caldesmon, DOG-1 (**C**) and negative for Actin, S-100 (**D**).

Intraoperatively tumor was identified, following extended mobilization of the duodenum, along the distal half of its second part, placed posterolaterally with a rim of a normal duodenal wall separating it from the pancreas. The tumor was thus removed *via* wedge resection,



**Fig. (3):** At laparotomy, the wedge resection of tumor was done from second portion of duodenum and resected specimen measures about 2x4 cm.

with 1 cm of gross resection margin, including 3/4<sup>th</sup>s of the circumferential wall (**Fig. 3**). A frozen section of tumor margin was sent, and it came out to be negative. Initially medial or pancreatic side of the duodenal wall was primarily anastomosed from inside followed by the lateral or anti-mesenteric wall with external knotting in an interrupted fashion, using a 4/0 PDS suture. The 2x4cm mass was sent to histopathology for diagnosis which confirmed the diagnosis of Duodenal GIST and adequate resection. The postoperative recovery was uneventful and the patient was discharged on the 7<sup>th</sup> post-operative day.

## DISCUSSION

The clinical presentation of DGISTs depends upon the size and mucosal ulceration only and not on tumor location. Symptoms commonly seen are melena or abdominal pain [6]. The source of bleeding was a linear ulcer on the submucosal duodenal mass causing melena as the primary symptom in this case.

Histologically, GISTs are composed of spindle-shaped cells that are diagnosed on basis of immunohistochemical analysis. EUS-FNA establishes the pre-operative diagnosis of different lesions with low risk of complications (1.6%) [1] and image-guided percutaneous biopsy of resectable small-sized tumors is not preferred anymore due to concern for rupture of pseudo capsule and spillage of tumor cells in the peritoneum. However, CT and MRI are the imaging modalities of choice for the assessment of both primary lesion and potential secondaries [6]. En-bloc resection with its pseudo capsule (R0) is the curative treatment of the tumor, however, due to the low incidence, variable virulence of the disease and anatomy of the duodenum, the optimal surgical approach is still controversial. Three surgical approaches that can be performed either laparoscopically or by open approach include wedge resection (WR), segmental resection (SR) and pancreaticoduodenectomy (PD) [5]. The selection of surgical approach depends upon the size and location of the tumor, distance from the duodenal papilla and surgeon's expertise [6]. Limited resection is preferred over PD in duodenal GIST as it is linked with a shorter duration of surgery, comparable survival benefits [6] and has fewer postoperative complications as pancreatic function is reserved [5]. Roux-en-Y duodenojejunostomy can be performed after wedge resection in cases of antimesenteric duodenal GIST whereas for mesenteric

duodenal GIST the defect can be repaired by translocation of the common bile duct. Duodenal GISTs present in D1, D3 and D4 can be managed with gastrojejunostomy and end to end duodenojejunostomy after segmental resection. Side to end duodenojejunostomy is associated with stump syndrome and therefore is not preferred. Pancreaticoduodenectomy is indicated in cases of large-sized tumors in D2 segment and with malignant potential [6]. CT guided radiofrequency ablation (RF) can be considered in patients who are not fit for surgery [2]. DGIST tumors have a favorable prognosis due to early presentation and small size and have a good five-year disease-free survival rate compared to other small intestine GIST [5]. Pharmacological treatments (tyrosine kinase inhibitor) are widely prescribed as adjuvant therapy in high-risk local tumors and metastatic GIST [6]. Common sites of metastasis are mesentery and omentum (30%), liver (28%), lungs (7%), lymph nodes (4.7%) and bones (2.3%). Follow up serial CT scan is advised every three months for five years in patients who are at high risk for metastasis [2] .

### CONCLUSION

This is a case of a 45-year-old man with intermittent melena who was found to have a small-sized duodenal GIST. Imaging studies play an important role along with histopathological confirmation of the lesion in the diagnosis of this rare pathology and limited resection is a preferred surgical treatment in small-sized duodenal GIST.

### CONSENT FOR PUBLICATION

Informed consent was taken from the patient for data sharing.

### CONFLICT OF INTEREST

The authors declare no conflict of interest.

### ACKNOWLEDGEMENTS

None.

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