

A gastrointestinal stromal tumor of the third portion of the duodenum treated by wedge resection: A case report

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Core tip: Duodenal gastrointestinal stromal tumors (GISTs) are uncommon, with a relatively small subset of GISTs whose optimal surgical procedure has not been well defined. Because submucosal spread and local lymph node involvement is infrequent in GISTs, wide margins with routine lymph node dissection may not be required. Various techniques of limited resection for duodenal GISTs have been described, depending on the site and the size of the tumors. Herein, we present a case of GIST involving the third portion of the duodenum successfully treated by wedge resection with primary closure.

Abstract

A 65-year old woman was admitted to our hospital with abdominal pain. Computed tomography showed a tumor measuring about 3 cm in diameter with no metastatic lesion or signs of local infiltration. Gastroduodenal endoscopy revealed the presence of a submucosal tumor in the third portion of the duodenum and biopsy revealed tumor cells stained positive for c-kit. These findings were consistent with gastrointestinal stromal tumors (GISTs) and we performed a wedge resection of the duodenum, sparing the pancreas. The postoperative course was uneventful and she was discharged on day 6. Surgical margins were negative. Histology revealed a GIST with a diameter of 3.2 cm and < 5 mitoses/50 high power fields, indicating a low risk of malignancy. Therefore, adjuvant therapy with imatinib was not initiated. Wedge resection with primary closure is a surgical procedure that can be used to treat low malignant potential neoplasms of the duodenum and avoid extensive surgery, with significant morbidity and possible mortality, such as pancreatoduodenectomy.

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INTRODUCTION

Gastrointestinal tumors are the most common mesenchymal tumors arising within the gastrointestinal tract^[1] and the treatment of choice of these tumors is surgical resection^[2,3]. The small intestine is the second most common site of gastrointestinal stromal tumor (GIST), of which approximately 20% are found in the duodenum^[2]. The optimal surgical procedure for duodenal GIST, however, remains undefined^[4] because, while surgical resection clearly confers survival advantage, there is little submucosal spread in GIST and lymphatic involvement is rare. The few reports in the literature addressing the surgical procedures for duodenal GIST include pancreatoduodenectomy, pancreas-sparing duodenectomy, segmental duodenectomy or local resection^[4-6]. In this study, we



Figure 1 Computed tomography showed a well-demarcated enhancing tumor 4.0 cm in diameter in the third portion of the duodenum (white arrow).

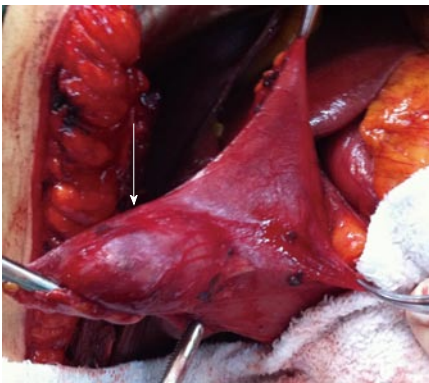


Figure 2 An endophytic gastrointestinal stromal tumor of the third portion of the duodenum (white arrow).

report a case of GIST involving the third portion of the duodenum successfully treated by wedge resection. This surgical technique is ideal when GIST does not involve the ampulla and has not been previously described for the management of this malignancy.

CASE REPORT

A 65-year old woman presenting with abdominal pain was referred to our hospital. Her medical and family history was unremarkable. She had no history of previous abdominal surgery. On physical examination, mild tenderness was complained of in the right upper quadrant area. Abdominal computed tomography (CT) showed a well-demarcated and enhanced tumor in the third portion of the duodenum, measuring approximately 3.0 cm in diameter. The mass appeared to compress the uncinate portion of the pancreas (Figure 1). From these radiographic findings, we diagnosed a submucosal tumor of the duodenum. She underwent an esophagogastroduodenoscopy, which revealed a submucosal tumor at the second and third portion of the duodenum. A biopsy obtained was reported as GIST. There was no evidence of metastases to her liver or lung. At laparotomy, a 3.0 cm sized solid mass was identified arising from the pancreatic border of the third portion of the duodenum

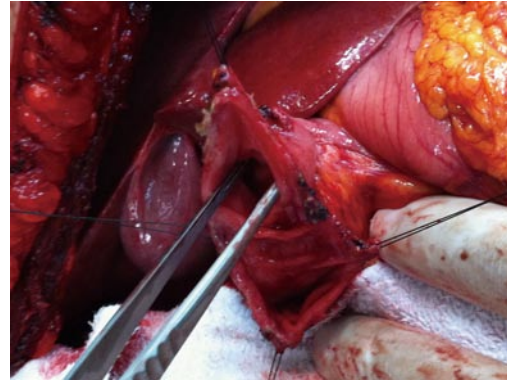


Figure 3 Local limited wedge resection was subsequently performed with clear margins. Surrounding bowel can be seen to be healthy, allowing for a primary anastomosis.

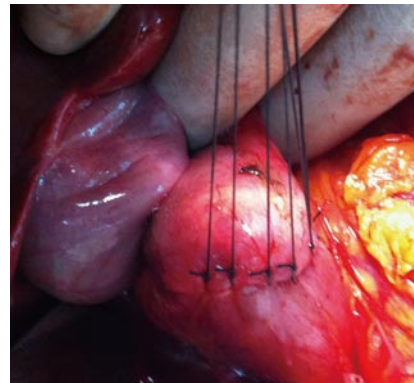


Figure 4 Wedge resection with primary closure.

(Figure 2). No evidence of local invasion of the pancreas or of distant metastases was found and the duodenal wall was carefully dissected from the inferior border of the pancreas. Considering that the pancreas and major papilla were not involved, a partial resection was performed, with a 1 cm disease-free margin (Figures 3 and 4). Operative time was 125 minutes and estimated blood loss was 50 mL. Histological examination revealed that the tumor was composed of spindle cells with a mitotic count < 5 mitoses/50 high power fields (Figure 5A and B). Immunohistochemical study revealed positive staining for CD 117 (c-kit) and S-100 (Figure 5C-E). Based on the above findings, the tumor was finally diagnosed as a GIST with low-grade malignancy originating from the duodenum. A molecular genetic analysis for KIT protein mutation was not performed because of its unavailability at our institute. The patient was doing very well with no evidence of disease recurrence when she was last seen, 4 mo after her operation.

DISCUSSION

GISTs are believed to originate from the interstitial cells of Cajal, which are intestinal pacemaker cells or mesenchymal stem cells^[7]. A typical feature of virtually all GISTs is a positivity at immunohistochemistry for

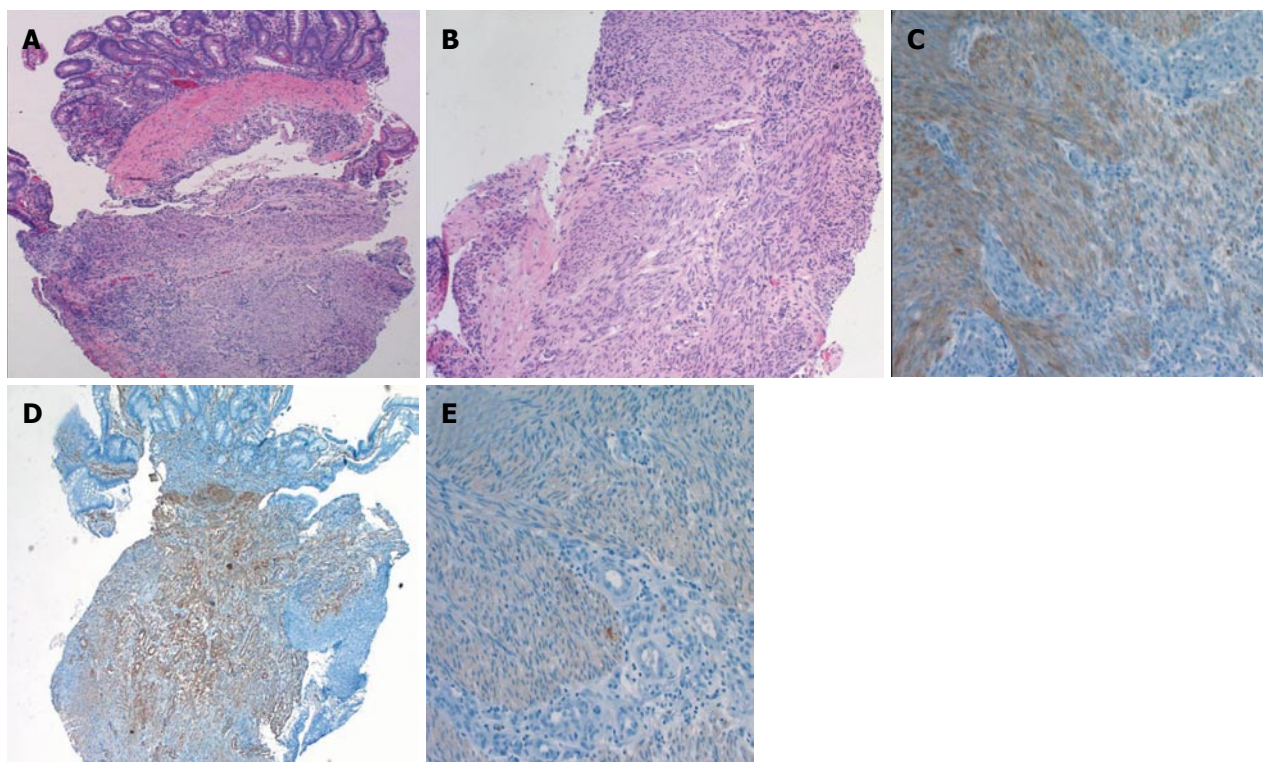


Figure 5 Histology. A: Submucosal tumor tissue is located (hematoxylin-eosin stain, original magnification, $\times 5$); B: Spindle tumor tissue is composed of cells (hematoxylin-eosin stain, original magnification, $\times 10$); C: Tumor tissue widely seen moderately strong staining of CD117 (CD117, original magnification, $\times 20$); D: Tumor tissue widely seen SMA staining (SMA, original magnification, $\times 5$); E: Tumor tissue, common, poor, S-100 staining is observed (S-100, original magnification, $\times 20$).

the KIT protein (CD117), a transmembrane receptor linked to an intracytoplasmic tyrosine kinase^[8]. Duodenal GISTs are mainly located in the second portion of the duodenum^[9]. The tumors are frequently located in close relationship to the ampulla of Vater, this determining surgical treatment strategy. In the case presented here, the tumor was located 3 cm distal of the papilla. Most duodenal GISTs present with GI bleeding, usually associated with melena and occasionally with massive acute bleeding^[9]. Other symptoms like abdominal pain, early satiety, bloating or obstructive jaundice due to involvement of the papilla of Vater were present in our patient. Diagnosis can be made with upper gastrointestinal endoscopy^[10]. The tumor is usually exophytic and appears as a submucosal swelling. Sometimes it presents only as an endophytic tumor, as in our case. The biopsy should be deep but may not always be diagnostic. Endoscopic ultrasound can help in delineating the submucosal tumor. Alternative diagnostic means include CT, magnetic resonance imaging (MRI), barium study or ultrasonography^[11]. However, CT and MRI seem to be the best imaging modalities for assessment of the primary lesion and detection of metastases^[12], although CT scans are not always helpful in specifying the origin of the mass. In several cases reported in the literature, the mass was misdiagnosed as arising from the head of the pancreas^[13].

There is currently uniform agreement that the surgical treatment of choice for GISTs is resection of the tumor with clear surgical margins, including adjacent

organs as necessary^[12]. As local and regional lymph node involvement is infrequent in GIST, routine lymph node dissection is not advocated^[11,14,15] and limited resection is frequently performed. The surgical choice depends not only on the size of the tumor, but also on the location in the duodenal wall and the relationship to the ampulla of Vater^[12,16,17]. Patients with duodenal GISTs close to the papilla of Vater should be treated by pancreatoduodenectomy. Various techniques of limited resection for duodenal GISTs have been advocated, depending on the site and the size of the tumors. Wedge resection with primary closure can be performed for small lesions if the resulting lumen is adequate and the ampulla can be preserved^[9,18]. Segmental duodenectomy with side-to-end or end-to-end duodenojejunostomy can be performed for larger tumors located at the third and fourth portion of the duodenum^[18]. Partial duodenectomy with Roux-Y duodenojejunostomy can be performed for larger tumors involving the antimesenteric border of the second and third portion of the duodenum^[19]. Although a limited operation procedure, such as wedge or segmental resection, is relatively simple to perform, there is a risk of subsequent anastomotic leakage or stenosis development, as well as later tumor recurrence in patients treated by limited operation. By contrast, pancreatoduodenectomy as a treatment for duodenal GISTs can provide a wider surgical margin but may be associated with excessive morbidity, especially in patients with a tumor of low-grade malignancy^[20]. It is not clear what the optimal

Table 1 Risk of aggressive behavior in gastrointestinal stromal tumors

Risk	Size (cm)	Mitotic count (mitoses per 50 high powered fields)
Very low risk	< 2	< 5
Low risk	2-5	< 5
Intermediate risk	< 5	6-10
	5-10	< 5
High risk	> 5	> 5
	> 10	Any mitotic rate
	Any size	> 10

Adapted from Fletcher *et al*^[22].

surgical margin should be, but a negative one is essential to prevent local recurrence of the tumor. No lymph node dissection is required because they are very unlikely to be involved^[18,21]. The outcome depends on the pathological features of the tumor and the completeness of surgical resection. Local recurrence is higher in tumors not completely removed or with a positive microscopic margin. In our patient, no suspicious peritumoral lymph nodes were present. Therefore, in order to minimize operative morbidity, we did not perform a formal lymph node dissection.

Fletcher *et al*^[22] established a risk stratification based upon tumor diameter and mitotic activity (Table 1)^[22]. The tumor presented in this case belongs to the category determined by size between 2-5 cm and a mitotic count < 5/50 high power fields, which is classified as “low risk”. As we performed a wedge resection of the GIST, this indicates a good prognosis for our patient.

Imatinib (Gleevec, Novartis, Basel, Switzerland) is the treatment for locally advanced or metastatic GIST. Imatinib is a signal transduction inhibitor and in particular inhibits the binding of adenosine triphosphate to tyrosine kinase that includes PDGFRA and the c-Kit receptor expressed in GISTs^[23]. Recently sunitinib malate, an oral receptor tyrosine kinase inhibitor, was approved for the treatment of GISTs after progression or intolerance to imatinib mesylate. Sunitinib inhibits platelet-derived growth factor receptors and vascular endothelial growth factor receptors, which play key roles in tumor angiogenesis and tumor cell proliferation^[24]. As our patient was classified as “low risk”, we did not initiate an adjuvant treatment with imatinib.

In summary, we report a case of a duodenal GIST located 3 cm distal of the ampulla of Vater successfully treated by a wedge resection. Wedge resection with primary closure is a surgical procedure that can be used to treat low malignant potential neoplasms of the duodenum and avoid extensive surgery, with significant morbidity and possible mortality, such as pancreatoduodenectomy.

COMMENTS

Case characteristics

A 65 year old woman was admitted to hospital with abdominal pain.

Clinical diagnosis

Gastroduodenal endoscopy revealed the presence of a submucosal tumor in the third portion of the duodenum and biopsy revealed tumor cells stained positive for c-kit.

Imaging diagnosis

Abdominal computed tomography showed a well-demarcated and enhanced tumor in the third portion of the duodenum, measuring approximately 3.0 cm in diameter.

Treatment

The patient underwent an esophagogastroduodenoscopy which revealed a submucosal tumor at the second and third portion of the duodenum.

Related reports

There is currently uniform agreement that the surgical treatment of choice for gastrointestinal stromal tumors is resection of the tumor with clear surgical margins, including adjacent organs as necessary.

Experiences and lessons

Wedge resection with primary closure is a surgical procedure that can be used to treat low malignant potential neoplasms of the duodenum and avoid extensive surgery with significant morbidity and possible mortality such as pancreatoduodenectomy.

Peer review

The manuscript is in general a nice case report but the discussion of the article needs polished.

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