LOCAL SURGICAL RESECTION OF A CASE OF GASTRO INTESTINAL STROMAL TUMOR OF DUODENUM

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Abstract

Gastrointestinal stromal tumors of the duodenum are uncommon. We present the case of a 62-year-old woman with a gastrointestinal stromal tumor of the duodenum. Her complaint was melena. A local surgical resection of first part of duodenum with the GIST was done. She was doing very well with no evidence of disease recurrence. Gastrointestinal stromal tumors of the duodenum should be suspected in any patient with a duodenal wall mass. GIST may arise rarely in the duodenum, appendix, gallbladder, and urinary bladder. Preoperative biopsy is not routinely necessary for a primary, resectable neoplasm suspicious for GIST. In fact, preoperative biopsy may rupture a suspected GIST and increase the risk of dissemination. The ideal margin of resection is unknown. The extent of surgery is usually a wedge or segmental resection of the involved bowel without the wide margins necessary for adeno carcinoma.

INTRODUCTION:

Gastrointestinal stromal tumors (GISTs) occur rarely in the duodenum. Because of their low incidence, data on long-term survival and prognostic factors are limited. Gastrointestinal stromal tumors (GISTs) arising in the duodenum represent a rare entity and can be very demanding to manage. The diagnosis can be difficult (as these tumors can be misdiagnosed as pancreatic head tumors), and to treat owing to the complex anatomy of the duodenum and of the pancreatic head. Complete resection may require extensive procedures such as pancreaticoduodenectomy.

Diagnosis can be elusive and managing them can be difficult. GISTs are rare and mitotic rate, tumor size, type of mutation and number of chromosomal aberrations have prognostic implications.

CASE PRESENTATION:

A 62 year female was admitted with complaint of passing black stools for past 1 month, painless, No mass descending or bleeding per rectum or any GI disturbances. She is a known diabetic on oral medication for past 2 yrs with no major surgical or medical illness in the past or any chronic analgesic intake. Abdominal examination was normal and in per rectal examination melena was present with no palpable deposits. Upper GI endoscopy revealed an umbilicated lesion at D1 with minimal oozing. Biopsy not taken due to oozing.

Contrast enhanced CT showed an Exophytic lesion with heterogenous enhancement 4.5x2.7cms in first part of duodenum with the probability of duodenal GIST. We proceeded with limited resection of first part of duodenum with anterior gastrojejunostomy.

Figure 1: Exophytic lesion with heterogenous enhancement 4.5x2.7cms in first part of duodenum

Histopathology showed Submucosal tumour shows well differentiated proliferation of spindle cell with plump vesicular nuclei. Tumour replaced entire muscular layer. Very mild nuclear Anisonucleosis and (Mitosis more than 5/HPF).
Figure 2: well differentiated proliferation of spindle cell with plump vesicular nuclei.

Figure 3: C kit positive GIST.

Figure 4: Desmin negative GIST.

Immunohistochemistry showed c kit positive and desmin negative. Final diagnosis of Gastrointestinal stomal tumour of Duodenum. STAGE II (pT2NxMx) was made and the patient was started on C. IMATINIB 400mg bid.

Patient is on regular follow up.

DISCUSSION:

Most GISTs occur sporadically; about 5-10% occur in hereditary familial syndrome (type I Neurofibromatosis, Carney triad). (1,3). The clinical presentations of duodenal GISTs are highly variable, and they are related to their origin, growth (intramural or extramural), and size (4,7). The most common clinical presentations are gastrointestinal bleeding (when mucosal ulceration is present), and abdominal pain. (2,5,8). These tumors are often diagnosed incidentally, as they can be asymptomatic, especially for small tumors, or when the tumor growth is extra luminal. Some authors described back pain, jaundice (compression of common bile duct), or bowel obstruction, but these events are very rare. At diagnosis, most authors report a smaller size tumor (4.0 - 5.0 cm) for duodenal GISTs when compared with gastric or small bowel GISTs. However, this is still controversial, as these tumors are rare.

Histologically duodenal GISTs do not differ from other GISTs. These tumors usually present with spindle cell differentiation, which is more frequent than epithelioid cell differentiation or mixed types. There is no apparent correlation between histologic subtype and risk of tumor recurrence. The mitotic count has been found to be lower in duodenal GISTs, with a median count <5/50 HPF, which is lower than gastric and small bowel tumors (where a mitotic count >5/50 HPF has been found in more than 30% of cases). However, these data need to be interpreted carefully due to the small number of tumors examined. As described in the medical literature, GISTs in the duodenum do not differ from other GISTs in immunohistochemical reaction. Most of them express CD-117 (c-kit) and CD-34, which has been found to be associated with worse prognosis. PDGFRA expression is less frequent, and there is no relation to a different prognosis. However, these data need to be confirmed by larger series, and there is a probability that a more benign behavior was related to the smaller size and lower mitotic count at diagnosis. For these reasons, duodenal GISTs usually have better prognosis as compared with gastric and small bowel GISTs. According to the NIH risk stratification, they are more frequently identified as very low risk and low risk tumors (about 60 to 70% of reported duodenal GISTs have been classified as very low or low risk tumors).

The size at diagnosis, prognostic indicators, and risk stratification of duodenal GISTs should guide the surgeon to a limited resection, which should always be a R0 resection. However, until now, there is still lack of knowledge on the preoperative indicators of bad prognosis and high risk of recurrence.

Gastrointestinal endoscopy remains the most common diagnostic procedure in duodenal GISTs, especially in patients with intramural growth or mucosal ulceration and...
bleeding. It allows forceps biopsy, which is not helpful in extraluminal tumor. In the latter, the most used diagnostic test remains CT scan or MRI. On CT, GISTs are hypervascular with cystic and necrotic components.

MRI findings of GIST are extremely variable, depending on tumor necrosis, hemorrhage and cavitation, which can affect signal intensity. The difficulties in preoperative diagnosis make a preoperative diagnosis of GIST rare. In most cases, GISTs is only “suspected”. Recently, endoscopic ultrasound (EUS) has been found to be very helpful for esophago-gastro-duodenal GISTs, with high sensitivity and specificity rates. Furthermore, EUS is also capable of guiding fine needle aspiration (FNA) under real-time ultrasound using a through-the-scope needle aspiration device to obtain specimens for cytologic examination and immunocytochemical evaluation for CD-117 and CD-34 positivity First, we should aim at a complete surgical resection with clear margins (R0 resection). Second, as sub mucosal spread, and local lymph node involvement are frequent for GISTs, wide resection margins with routine lymph node dissection are not necessary as for epithelial tumors.

There is no consensus on the optimal surgical treatment for GISTs arising from the duodenum. Operations which vary from tumor enucleation (for extramural GISTs) to pancreaticoduodenectomy for infiltrating or larger tumors have been advocated. Limited resections (LR) can be performed in small tumors not infiltrating the surrounding structures, and when the papilla of Vater can be preserved. In large series, duodenal wedge resection has been performed for small tumors (<1 cm in diameter) which were located on the duodenal wall at least 2 cm from the papilla, so that a safe resection and anastomosis can be carried out with minimal risk of stenosis.

Segmental duodenectomy, with end-to-end or side-to-end duodeno-jejunal anastomosis can be performed for larger tumors which are located below the papilla of Vater in the third or fourth duodenal portions. Recently, partial duodenectomy with a Roux-en-Y duodeno-jejunal anastomosis has been proposed for large tumors which involved the antimesenteric border of the second and third portions of the duodenum, in order to avoid carrying out pancreaticoduodenectomy.

The main concern on limited resection is the theoretical increased risk of tumor recurrence. As malignancy is not established at the time of surgery in most cases, the data in the medical literature are not conclusive. The data seems to suggest that local recurrence is influenced by the presence of positive resection margins,(2,5,8), but not by the kind of surgical resection (limited resection vs. pancreaticoduodenectomy), as the two procedures had similar disease-free survival rates,(5). There is also a tendency to perform pancreaticoduodenectomy for large tumors which have a higher risk of malignancy and recurrence, and limited resection for small tumors which have a benign behavior. The disease-free survival rate at 1-3 years after surgical complete resection, in all kinds of resection, has been reported to vary from 86 to 100%,,(6,7), indicating that duodenal GISTs have a slightly more favorable prognosis than gastric or small bowel GISTs.

Imatinib mesylate, a tyrosine kinase inhibitor, plays a key role in the management of GISTs. Its use in neoadjuvant therapy, adjuvant therapy and in tumor recurrence,(9,10), has dramatically changed the natural history of metastatic and recurrent GISTs. In the neoadjuvant setting for GISTs located in the second portion of the duodenum, Imatinib mesylate has been used for tumor downstaging in order to perform a less extensive surgery with free resection margins,(6). However, this requires a precise preoperative diagnosis of GIST which is not always easy to obtain. The use of Imatinib as adjuvant therapy, or for local or metastatic recurrence of duodenal GIST, should not differ from that for other GISTs, and the treatment should be continued indefinitely (usually at a dose of 400 mg/day). Interruption of treatment is generally followed by rapid tumor progression. (6,11). The major limitation of Imatinib is the development of tumor resistance, which is related to the acquisition of additional c-kit mutations.

Gastrointestinal stromal tumors (GISTs) are rare neoplasms. Although they represent only 0.1-3%of all gastrointestinal malignancies, they account for 80% of gastrointestinal mesenchymal neoplasms.GISTs commonly arise in the stomach (50–70%), small intestine (25–35%), colon and rectum (5–10%), mesentery or omentum (7%), and esophagus (<5%).

GIST may arise rarely in the duodenum, appendix, gallbladder, and urinary bladder. Preoperative biopsy is not routinely necessary for a primary, resectable neoplasm suspicious for GIST.

In fact, preoperative biopsy may rupture a suspected GIST and increase the risk of dissemination.

The ideal margin of resection is unknown. There are no data to confirm that a positive microscopic margin (R1 resection) impacts survival.

The extent of surgery is usually a wedge or segmental
resection of the involved bowel without the wide margins necessary for adenocarcinoma.

Duodenal GISTs constitutes <5% of entire gastrointestinal GIST, 30% of primary duodenal tumors. No lymph node metastasis or infiltration occur. Hence local resection is adequate. Prognostic factors are Size, Site, and Mitotic rate (12).

CONCLUSION:

Duodenal GIST,

• Always presents as solitary lesion, Surgical resection is first choice of treatment.

• Surgical modalities include Local resection, Segmental duodenectomy, Pylorus preserving duodenopancreatectomy.

• Frequency of occurrence follows the order PART 2, 3, 4, 1.

• Best surgical procedure – Controversial due to lower incidence and fewer data and depends on site of tumor (13),

• Part 1—Local resection

• Part2—Duodeno pancreatectomy

• Part3—Segmental duodenectomy

• Part4—Segmental duodenectomy

• Local resection promises very good result in terms of local and systemic Disease Free Survival, irrespective of margin status. Due to complex/unique nature of pancreaticoduodenal region, <5mm after resection margin is adequate (14,15).

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