Surgical Management of Duodenal Gastrointestinal Stromal Tumors: A Case Report

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Abstract. Gastrointestinal stromal tumors (GISTs) represent the most common mesenchymal tumor entity of the gastrointestinal tract. In fewer than 5% of cases, primary GISTs are located in the duodenum. Surgery represents the treatment of choice for localized tumor disease and remains challenging in GISTs located at the duodenum. The optimal surgical approach is currently discussed controversially in the literature due to the fact that extended resections in terms of pancreaticoduodenectomy may cause significant postoperative morbidity and mortality compared to limited resection. We report on a rare case of GIST located in the upper part of the duodenum treated by limited surgical resection. Avoidance of postoperative complications may lead to optimal oncological outcome in selected patients.

Gastrointestinal stromal tumor (GIST) of the duodenum represents a very rare tumor entity, being diagnosed in fewer than 5% of all primary GISTs in the gastrointestinal tract. Surgical treatment is difficult as pancreaticoduodenectomy (PD) might cause significant morbidity and mortality compared to the outcome of local resection (LR). In the literature, the extent of surgical resection is discussed controversially (1-3). We report on a rare case of a GIST located in the duodenum treated by organ-preserving surgery.

Case Report

In February 2015, a 68-year-old female patient was admitted to the Emergency Unit of the Division of Gastroenterology and Hepatology, Medical University of Graz because of abdominal pain and melaena, associated with hemodynamic instability. Her medical history included iron-deficient anemia of unknown origin for several months, coronary artery disease and the patient described having a heart attack several years earlier.

Emergency endoscopy due to hemodynamically relevant bleeding showed active bleeding type Forrest IIb of ulcerated tumor tissue located in the upper part of the duodenum (Figure 1). After primary management of the bleeding by endoscopy, tumor staging was initiated to plan further treatment strategies. Computed tomographic (CT) scan and magnetic resonance imaging (MRI) showed a solid tumor mass of the duodenum, 4.5 cm in diameter, with heterogeneous contrast enhancement (Figures 2 and 3). Distant metastases were excluded by positron-emission tomography–CT scan. With respect to these radiological findings, a primary GIST of the duodenum was suspected and surgical resection was planned as PD.

Intraoperatively, the tumor was located in the upper part of the duodenum and a limited surgical procedure by resection of the duodenal wall was performed. Histological examination of frozen section verified a tumor of mesenchymal origin and tumor-free circumferential resection margins. The duodenal wall was closed, hand-sewn by double-layer technique.

Histological examination of the specimen revealed mesenchymal tumor tissue within the submucosa and muscularis. The tumor was mainly composed of spindle cells with syncytial eosinophilic cytoplasm. The tumor cells strongly expressed CD117 (KIT) by immunohistochemistry. Eight mitoses per 5 mm² were counted. Mutational analysis revealed a deletion in KIT exon 11 (c.1702_1728del; p.Y568_L576del). The tumor was diagnosed as a GIST, of high risk according to the National Comprehensive Cancer Network guidelines (Figure 4).

The postoperative course was uneventful and the patient was discharged from hospital on the fifth postoperative day. Based on the decision of our Institutional Tumor Board, adjuvant therapy with imatinib was started and the treatment was well-tolerated by the patient.

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Discussion

GISTs represent the most common mesenchymal tumor entity in the gastrointestinal tract and in the majority of cases these tumors are located in the stomach (about 60%), small bowel (about 25%), colon or rectum (about 10%). The duodenum represents a very rare site of primary GIST, fewer than 5% of all cases, and the optimal surgical treatment remains challenging (1, 2).

Although symptoms may vary considerably, the most common clinical presentation of duodenal GIST is gastrointestinal bleeding in more than 60% of all patients, followed by anemia or abdominal pain, especially in the case of mucosal ulceration and when the tumor is located in the first or second part of the duodenum (3-5). Due to these predominating symptoms, gastrointestinal endoscopy remains the most commonly used diagnostic tool for duodenal GISTs (1). In our case, gastrointestinal bleeding led to emergency endoscopy, which detected an ulcerated tumor mass in the upper portion of the duodenum. The initial bleeding was managed by endoscopy and further investigations were performed based on the strong suspicion of a primary malignant tumor of the duodenum. CT scan and MRI showed a solid tumor mass with heterogeneous contrast enhancement and a primary GIST of the duodenum without distant metastases was suspected.

In literature, CT scan and MRI findings of GISTs are described as being variable, depending on tumor size,
necrosis, hemorrhage or cavitations. Based on these facts, an exact preoperative diagnosis remains challenging (1, 6-8).

Surgery represents the treatment of choice in patients with localized tumoral disease, but in literature there is no clear consensus about the optimal surgical treatment of GISTs arising from the duodenum. PD may lead to significant surgery-related morbidity and mortality rates in up to 50% and 3% of all cases, respectively (9, 10). Considering these morbidity and mortality rates, LR is discussed as a possible surgical approach for duodenal GISTs whenever feasible, depending on tumor location and tumor size with respect to oncological outcome (2, 4-6). Several studies investigated the postoperative outcome after PD compared to LR (2-6, 11-13).

Johnston et al. analyzed a cohort of 96 patients with duodenal GISTs undergoing PD or LR in terms of perioperative and long-term outcome. The authors found that patients undergoing PD had significantly greater number of postoperative complications compared to patients treated by LR (57.9% vs. 29.3%, \( p=0.005 \)), moreover the length of hospital stay was significantly longer in patients treated by PD (11 vs. 7 days, \( p=0.001 \)). Tumor size, mitotic count >10 mitosis/50 high-power fields, American Joint Committee on Cancer stage III disease and National Institutes of Health (NIH) high-risk classification were significantly associated with a shorter recurrence-free survival. The type of surgical technique was not associated with overall survival. Thus, it was concluded that LR is appropriate for duodenal GISTs whenever feasible (11).

Similar results were found by Zhou et al. in a retrospective study of 48 patients. Compared to LR, PD was associated with a significantly longer operating time and a significantly longer hospitalization. NIH high risk was a significant factor associated with a short tumor-free survival in a multivariate analysis (3). Goh et al. found a significantly shorter operating time in patients treated with LR (125 vs. 350 min) compared to patients undergoing PD, but similar morbidity rates (23% vs. 43%) in 22 patients (13). Concerning the oncological outcome, Tient et al. reported NIH high risk as significant predictor for disease recurrence in 25 patients (12). A larger cohort was analyzed by Colombo et al. The oncological outcome in duodenal GISTs in 48 patients was investigated in a prospective multicenter study. The authors recorded a 5-year overall and disease-free survival for the entire cohort of 89% and 64%, respectively. Patients undergoing PD had a significantly larger median tumor size (7 cm vs. 5 cm, \( p=0.024 \)) and the tumors displayed a higher mitotic count (39% vs. 19% >5/50 high power fields, \( p=0.005 \)) compared to patients treated by LR. With respect to overall and disease-free survival no difference between that the two groups was found. The study group concluded the type of surgical technique should be determined by the duodenal site of origin and tumor size (2). Several other studies proposed LR for duodenal GISTs as a safe and effective surgical technique leading to good local and systemic control and thereby extended disease-free results (4-6).
In our case, local resection was performed based on intraoperative conditions. The tumor was located in the duodenal wall without infiltrating the pancreatic head, mesenteric or hepatic vessels, and curative resection was confirmed by intraoperative histopathology on frozen-section material. In our opinion, LR should be performed whenever a resection for curative intention is possible based on intraoperative conditions. Oncological outcome is comparable to extended resections and intraoperative histological examination of resection margins may easily confirm a curative resection. The postoperative course in our case was uneventful and the patient was discharged on the fifth postoperative day. Possible avoidance of postoperative morbidity and mortality and short hospitalization should be kept in mind and LR should be preferred whenever possible. In conclusion, we believe that based on the reported results from the literature and our own experience, LR for duodenal GISTs may avoid postoperative morbidity and mortality associated with extended resection with comparable oncological outcome.

References


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