



Duodenal Stromal Tumor: A Rare Cause of Upper Gastrointestinal Bleeding

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Abstract:

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract. Surgery is the primary choice for GIST treatment. However, there is no consensus on the best primary treatment for duodenal stromal tumors. In this study, we discuss the surgical treatment options for duodenal stromal tumors and the clinical outcomes of these treatments with this sample case.

Key words: Duodenum, Gastrointestinal Stromal Tumors, Gastrointestinal Hemorrhage, Gastrointestinal Neoplasms.

Introduction

Gastrointestinal stromal tumors (GISTs) are mesenchymal tumors that originate in interstitial Cajal cells (located in the intestinal wall) or their precursor cells [1]. GISTs are extremely rare and constitute only 1% of all primary gastrointestinal tumors [2]. Although GISTs may occur at any location within the gastrointestinal tract, the most common locations are the stomach (50-60%) and the intestines (20-30%) [3]. One third of all stromal tumors are located in the duodenum [4]. Although most of these are asymptomatic, some are accompanied by abdominal pain (50-70%), gastrointestinal bleeding (20-50%) or abdominal mass [1]. Surgical resection is the primary treatment option for nonmetastatic cases. Because submucosal invasion and lymph node metastases are uncommon, surgical resection without lymph node dissection

is the most suitable surgical treatment [5]. Some authors maintain that pancreaticoduodenectomy is the best treatment option for stromal tumors, whereas others believe that a limited resection is sufficient because this treatment is associated with lower morbidity and mortality rates [5].

Case Report

A 42-year-old female was admitted to the emergency department with symptoms of nausea, vertigo, and dark-colored feces, which lasted for one week. Her arterial blood pressure was 100/60 mmHg, and her heart rate was 110 bpm. She was experiencing tenderness in the epigastric region, and 300 cc of blood was aspirated with nasogastric decompression. During endoscopic

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examination, an approximately 1.5 cm submucosal lesion was observed in the posterior region of the bulb. Abdominal tomography showed a well-circumscribed lesion, including intraluminal and extraluminal components with a size of 18×15 mm located in the duodenal bulb [Fig.1]. Following preoperative preparation, the patient was taken in for surgery. The aforementioned lesion was observed in the second part of the duodenum which was removed with negative margins [Fig.2]. There were no other pathological findings during surgical exploration. Following resection, the duodenal space was closed with primary sutures. Pathological examination revealed a gastrointestinal tumor with a low mitosis rate. The patient was discharged six days after surgery with no complications.

Discussion

Gastrointestinal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract, and have previously been classified as being either leiomyoma (benign) or leiomyosarcoma (malignant) [5]. GISTs were first defined by Mazur *et al.* (1983), after they examined a smooth muscle tumor using an electron microscope and observed no neural or

muscular activity [3].

Although the exact prevalence of GISTs is still unknown, it is assumed that incidence of the tumors is approximately 10 per million per year. Very small GISTs have been observed during autopsies of individuals aged over 50 years. This indicates that these tumors are slow growing [6]. GISTs can occur at any age and in any part of the gastrointestinal tract. Mean age of diagnosis is 60 years, and they are rarely seen in the elderly or children. The most common locations of GISTs are the stomach (50-60%) and intestines (20-30%) [3]. One third of all stromal tumors are located in the duodenum [7].

Most GISTs located in the duodenum are <5 cm. Tumors <2 cm are asymptomatic and are usually diagnosed incidentally while evaluating other medical conditions with tomography, endoscopy or surgery. Gastrointestinal stromal tumors may be submucosal, intramucosal, or subserosal. Ulcerations leading to bleeding may occasionally arise on the surface of the tumor. Tumors may grow toward the intestinal lumen or may show an exophytic growth



Fig.1: CECT showing well-circumscribed lesion located in the duodenal bulb.

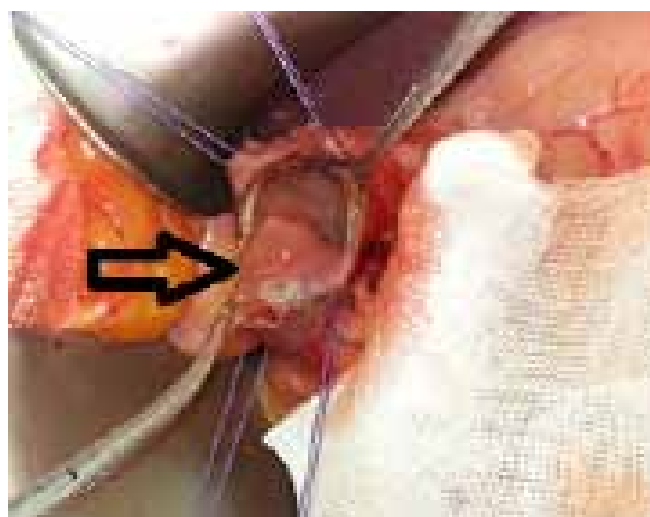


Fig.2: The appearance of the mass after duodenotomy.

pattern. Tumor size varies from 0.8 to 23 cm (mean 6–7 cm) [8].

There is no standard imaging method for diagnosing GISTs, and the only reliable method is biopsy of suspicious tissue; however, this is not advised for resectable tumors as interruption of the capsule of the tumor may lead to tumoral seeding to the surrounding zones. Therefore, preoperative biopsy should be considered for unresectable or high risk surgeries, to provide neoadjuvant therapy. One of the most important malignancy criteria for GISTs is mitosis count. However, preoperative needle biopsies do not give perfectly accurate results; therefore, mitosis rate cannot be used as a prognostic criterion [9]. Theoretically, any GIST has the potential to become malignant. It would be more accurate to classify these tumors as very low, low, intermediate, or high risk tumors rather than benign or malign. Furthermore, tumors <2 cm are considered to be benign. The most common criteria used to investigate risk of malignancy are tumor size and mitosis rate. For our patient, the tumor diameter was 1.5 cm and mitosis rate was <1 under 50X magnification zone; therefore, the tumor was considered to be low risk.

Fletcher *et al.* classified GISTs according to their tumor size and mitosis count (Table 1) [10]. In

contrast, Miettinen and Lasota suggested that tumor location is an equally important factor for prognosis, and that tumors located in the stomach have a worse prognosis (Table 2) [11]. Two other factors that increase risk of recurrence are the existence of metastases and perforation of the tumor [11].

Surgical resection should be the preferred treatment for GISTs. Surgical procedures are well-defined for tumors located in the stomach and intestines [12]. However, there is no certain consensus on treatment of tumors located in the duodenum. There are different opinions about the respective benefits of pancreatico-duodenectomy and wedge resection [13].

There are three conditions under which preoperative medical treatment should be considered over surgery: (i) Tumors which are not suitable for total excision because of local invasion or metastases, (ii) Patients who are at risk for surgery, and (iii) If surgery is associated with high morbidity or mortality rates for patients. The three main aims of surgery are achieving a negative surgical margin, prevention of tumoral perforation and achieving a total R0 resection. Although GISTs are large tumors, they could easily be resected with negative tumor-free margins because of their noninvasive nature

Table 1: Risk stratification of primary GISTs based on mitotic index, tumor size and anatomic site

Mitotic Index	Size, cm	Risk of progressive disease (%)			
		Gastric	Duodenal	Jejunum/ Ileum	Rectum
≤ 5 per hmf	≤2	None	None	None	None
	>2 and ≤5	Very low	Low	Low	Low
	>5 and ≤10 Low	Insufficient Data	Moderate	Insufficient Data	
	>10	Moderate	High	High	High
>5 per 50 hmf	≤2	None	Insufficient Data	High	High
	>2 and ≤ 5	Moderate	High	High	High
	>5 and ≤10 High	Insufficient Data	High	Insufficient Data	
	>10	High	High	High	High

Table 2: Estimated malignancy potential

Malignancy Risk	Size (cm)	Mitotic (50 hmf)
Very low	<2	<5
Low	2-5	<5
Intermediate	<5	6-10
	5-10	<5
High	>5	>5
	>10	Any index
	Any size	>10

[14]. Because of this, a simple resection is sufficient treatment for tumors located in the stomach or intestines.

There is still no consensus on the optimal treatment method for duodenal stromal tumors. Similar survival rates have been reported for patients treated with pancreatico-duodenectomy and simple resection [13]. Therefore, simple resection with negative margins should be the preferred treatment, when possible [15]. Moreover, there is further discussion about treatment of cases with signs of positive microscopic residual tumor after resection. Re-resection, medical treatment or follow-up without treatment are considered to be the options. As lymph node metastases are rare for GISTs, usually lymph node dissection is unnecessary, unlike for intestinal adenocarcinomas [14]. Furthermore, special attention should be paid to protect the integrity of the tumor, as tumor perforation is a poor prognostic factor.

Exploration in our case revealed palpable mass had no relations with the vital organs that were close to the duodenum. The mass was excised with clear margins and no lymphadenectomy was required. It is our opinion that the excision with clear margins that was performed on the patient who has lower malignant potential due to the low mytosis rate and small size of the tumor was up to date.

In cases where surgery is not possible due to a known

medical condition, imatinib is the drug of choice for neoadjuvant therapy [16]. Some studies have reported that, in cases of unresectable infiltrative tumors treated with imatinib, pathological response was 86% and the ratio of complete resection was 89% [17]. In addition to these benefits, other studies demonstrate that postoperative treatment with imatinib decreases recurrence rates [18]. Therefore, large infiltrative tumors should be re-evaluated for surgical removal after neoadjuvant therapy [19]. Further questions have arisen about when surgical treatment should be planned after neoadjuvant treatment. Many authors report that 6 to 12 months is the most suitable period for surgery. As resistance to drug therapy develops after 2 years, surgical evaluation must be done before 2 years [20].

Recurrences are usually seen in the peritoneum, liver or both. Peritonectomy and intraperitoneal chemotherapy might be preferred under these conditions [20]. Liver metastases must be removed if possible. However, diffuse peritoneal implants and liver metastases usually prevent surgical intervention. Therefore, other treatment modalities, such as hepatic artery embolization, chemoembolization, or radiofrequency ablation, should be kept in mind.

Conclusion

GISTs are infrequently located in the duodenum and usually present with bleeding. To achieve lower

morbidity and mortality rates, partial resection should be selected over pancreatoduodenectomy for nonmetastatic duodenal stromal tumors.

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