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Duodenal Gastrointestinal Stromal Tumor: A Rare Case

Duodenal Gastrointestinal Stromal Tümör: Nadir Bir Olgu

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Abstract: Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal system and are usually located in the stomach and small intestine. However, GISTs localized in the duodenum are quite rare and require special techniques for surgical treatment. In this case report, the surgical treatment and postoperative process applied to a 28-year-old female patient due to GIST detected in the second segment of the duodenum is discussed. The patient applied with complaints of loss of appetite, abdominal pain and intermittent nausea lasting approximately three months. Although no obvious pathology was detected in the physical examination, an ulcerated mass lesion was detected in the second segment of the duodenum in the upper gastrointestinal endoscopy and contrast-enhanced abdominal computed tomography (CT) examination and a decision was made for surgical exploration. During laparotomy, an approximately 4x5 cm ulcerated tumor was observed in the second part of the duodenum, adjacent to the pancreatic head, protruding into the duodenal lumen, and no obvious invasion of the surrounding tissues. Duodenal wedge resection and Roux-en-Y gastroenterostomy were performed. The patient was discharged without any postoperative complications. Histopathological examination revealed high mitotic activity and the tumor was reported to be a high-grade GIST. The patient was referred to oncology for adjuvant imatinib therapy. In conclusion, duodenal GISTs are rare but can be effectively managed with early diagnosis and surgical intervention. Given the high-grade nature of the tumor, surgical treatment remains the primary approach, and continued oncological follow-up improves survival significantly.

Keywords: Gastrointestinal tumor, duodenal tumor, mesenchymal tumor

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Özet: Gastrointestinal stromal tümörler (GİST), gastrointestinal sistemin en sık görülen mezenkimal kökenli tümörleridir ve genellikle mide ve ince bağırsakta yerleşim gösterirler. Ancak, duodenumda lokalize GİST'ler oldukça nadirdir ve cerrahi tedavileri özel teknikler gerektirir. Bu olgu sunumunda, 28 yaşında kadın hastada duodenumun ikinci segmentinde tespit edilen GİST nedeniyle uygulanan cerrahi tedavi ve postoperatif süreç ele alınmaktadır. Hasta, yaklaşık üç aydır süren iştahsızlık, karın ağrısı ve aralıklı bulantı şikayetleri ile başvurdu. Fizik muayenesinde belirgin bir patoloji saptanmamakla birlikte, üst gastrointestinal endoskopi ve kontrastlı batin bilgisayarlı tomografi (BT) incelemesinde duodenum ikinci segmentte ülsere bir kitle lezyonu tespit edildi ve cerrahi eksplorasyon kararı alındı. Laparotomi sırasında duodenumun ikinci kısmında, pankreas başına komşu, yaklaşık 4x5 cm boyutunda ülsere ve duodenal lümene protrüde, çevre dokulara belirgin invazyon gözlenmeyen tümör izlendi. Duodenum wedge rezeksiyon ve Roux-en-Y gastroenterostomi yapıldı. Ameliyat sonrası dönemde komplikasyon yaşanmayan hasta taburcu edildi. Histopatolojik incelemede, mitotik aktivitenin yüksek olduğu ve tümörün yüksek grade GİST olduğu rapor edildi. Hastaya adjuvan imatinib tedavisi amacıyla onkolojiye yönlendirildi. Sonuç olarak, duodenal GİST'ler nadirdir ancak erken tanı ve cerrahi müdahale ile etkili bir şekilde yönetilebilir. Tümörün yüksek dereceli doğası göz önüne alındığında cerrahi tedavi birincil yaklaşım olmaya devam etmekte olup, onkolojik takiplerin devamı sağ kalımını olumlu ölçüde iyileştirmektedir.

Anahtar Kelimeler: Gastrointestinal tümör, duodenal tümör, mezenkimal tümör

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1. Introduction

Gastrointestinal stromal tumors (GIST) are rare mesenchymal tumors, accounting for less than 1% of all gastrointestinal tumors (1, 2). Although they can occur in any part of the gastrointestinal tract, they are most commonly found in the stomach (40-60%), followed by the small intestine (30%) and colon (15%), with duodenal GISTs comprising only about 5% (3, 4). Initially believed to originate from smooth muscle cells, GISTs are now known to arise from the interstitial cells of Cajal in the intestinal wall (5, 6). Histologically, GISTs are classified into three subtypes: spindle cell type (77%), epithelioid type (8%), and mixed type (15%). While GISTs are most commonly diagnosed in the fifth decade of life, they occur equally in men and female (4).

Approximately 70% of GIST patients are symptomatic, though symptoms are non-specific. Most patients present with gastrointestinal symptoms such as nausea, vomiting, abdominal tenderness, weight loss, and early satiety. Symptoms vary based on tumor location, with abdominal pain (60-70%) being the most common, followed by gastrointestinal bleeding (30-40%), which typically results from tumor surface erosion. Less commonly, depending on

tumor location, dysphagia, jaundice, or small bowel intussusception may occur (7, 8). Lymph node metastasis in GISTs is rare, while distant metastases most commonly affect the peritoneum, omentum, mesentery, and liver. Due to the high mitotic rate, GISTs can reach significant sizes (9).

2. Case Report

A 28-year-old female patient presented with a three-month history of loss of appetite, abdominal distension, intermittent nausea, and pain localized in the left upper quadrant. The patient had no known medical history. Physical examination revealed tenderness in the left upper quadrant but no other significant findings. Routine blood tests were within normal limits. Abdominal computed tomography (CT-portal venous phase) revealed a centrally cystic, peripherally enhancing mass lesion measuring 4.2×5.2 cm at the pancreaticoduodenal level, in close relation to the pancreatic head, the second part of the duodenum, and segment 5 of the liver (Figure 1). Upper gastrointestinal endoscopy showed a mucosally ulcerated lesion, approximately 2 cm in diameter, narrowing but not obstructing the lumen of the second part of the duodenum due to extrinsic compression (Figure 2).

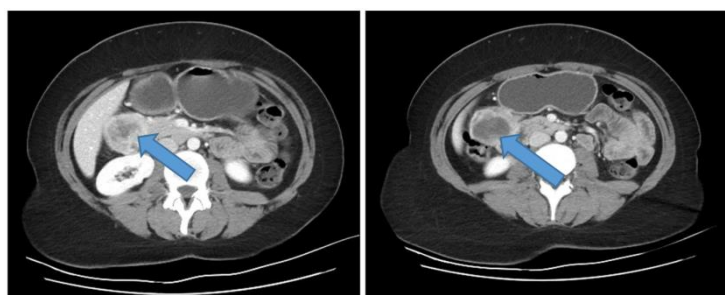


Figure 1. Peripheral contrast-enhancing cystic mass lesion at the pancreaticoduodenal level in portal venous phase contrast CT, adjacent to the pancreatic head and duodenum (shown with arrow)

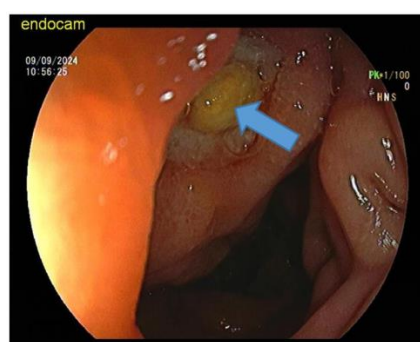


Figure 2. Intraluminal view of a lesion with an ulcerative mucosa and a lumen-narrowing lesion of approximately 2 cm in diameter in the second segment of the duodenum in upper gastrointestinal endoscopy

Surgical exploration was planned, and after obtaining informed consent, the patient underwent surgery. Intraoperatively, a mass measuring approximately 4×5 cm was identified originating

from the duodenum. The tumor extended from the second part of the duodenum towards the pancreatic head, with close proximity to it (Figure 3).

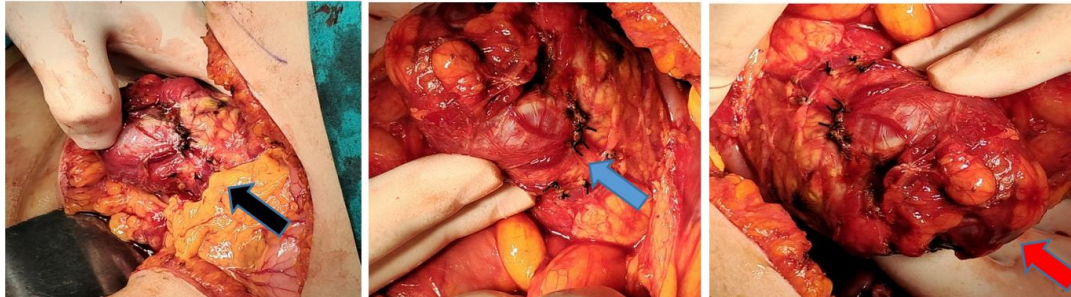


Figure 3. Macroscopic appearance of the lesion originating from the duodenum and in close relation to the head of the pancreas during laparotomy

The lesion was excised by wedge resection with careful blunt and sharp dissection. The ampulla of Vater was preserved, and the defect was closed in two layers. Due to postoperative luminal narrowing, a Roux-en-Y gastrojejunostomy was performed. The patient had an uneventful recovery and resumed oral intake.

Histopathological analysis confirmed a high-grade GIST with high mitotic activity and negative margins. Immunohistochemical staining was positive for c-KIT (CD117). The tumor was staged as T3. The patient was referred for adjuvant imatinib mesylate therapy. Long-term outcome data are currently unavailable.

3. Discussion

GISTs are rare, accounting for less than 1% of all primary gastrointestinal neoplasms (1). However, they represent the most common mesenchymal tumor type of the GI tract (2). Most GISTs are positive for CD117 and primarily arise due to activating mutations in the KIT or platelet-derived growth factor receptor alpha (PDGFRA) genes (6). Symptoms are non-specific and often include nausea, vomiting, abdominal discomfort, weight loss, or early satiety. Clinical presentation is usually related to tumor location rather than specific disease symptoms (10).

Contrast-enhanced CT is the preferred imaging modality for lesion characterization, staging, and metastasis assessment. It also plays a role in monitoring treatment response and recurrence (10). Diagnosis is confirmed via endoscopic biopsy, but due to the submucosal nature of GISTs, standard endoscopic biopsies may be

inconclusive. Endoscopic ultrasound (EUS) with fine-needle aspiration is increasingly utilized, providing an 80-85% diagnostic accuracy (9). Positron emission tomography (PET) can differentiate between active tumor tissue and necrotic/inactive scar tissue and is useful in distinguishing benign from malignant lesions (11). While CT is widely used to assess primary tumor size, location, and metastases, it may not detect small metastases, as it only visualizes structural changes (12). PET-CT, by evaluating metabolic activity, improves detection of small and microscopic metastases, aiding in early-stage diagnosis and monitoring treatment response (13, 14).

Surgical resection remains the gold standard treatment for localized disease. The primary goal is complete tumor removal without rupture or dissemination. Unlike adenocarcinoma, GISTs rarely metastasize to regional lymph nodes; thus, lymphadenectomy is only necessary in cases of apparent nodal involvement (15). Prognostic factors include tumor size and mitotic activity. Small tumors (≤ 2 cm) with low mitotic activity ($<5/50$ HPF) have a good prognosis, while larger tumors (>5 cm) with high mitotic activity are associated with poor outcomes (1). Despite complete surgical resection, GISTs carry a high risk of metastatic recurrence, particularly in the liver (65%) and peritoneal surfaces (50%) (9). When surgical resection is incomplete, imatinib mesylate is the preferred treatment for advanced or metastatic GISTs (16).

In conclusion, duodenal GISTs are rare but can be effectively managed with early diagnosis and

surgical intervention. In this case, complete tumor resection was achieved without complications. Given the high-grade nature of the tumor, long-term oncological follow-up is essential. Surgical

treatment remains the primary approach, and a multidisciplinary strategy improves patient survival.

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