Case report : duodenal stromal tumor

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Abstract

Tumors of the small intestine are rare lesions, but they should be kept in mind as possible causes of gastrointestinal symptoms. Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the digestive tract.

A 78 year-old woman complaining of abdominal pain, nausea and vomiting after meal and weight loss for three months was admitted to our clinic. On physical examination, there was only epigastric tenderness. No mass was palpated. She was anemic and total protein and albumin levels were low. Other laboratory tests were normal. A 9.0 × 7.5 cm heterogeneous mass was detected on the abdominal computerized tomography scan. Endoscopy confirmed a polypoid and vegetative mass in the second part of the duodenum. Histopathological diagnosis of endoscopic biopsy was gastrointestinal stromal tumor. Pancreaticoduodenectomy was performed. On the 11th postoperative day, relaparotomy was performed due to biliary leakage from the subhepatic drain. Biliary leakage was from the choledochojejunostomy. Choledochojejunostomy and pancreaticojejunostomy were revised. She was discharged on the postoperative 25th day. Histopathological examination of the resection specimen revealed duodenal stromal tumor.

Although stromal tumors are relatively rare in the duodenum, in the case of upper gastrointestinal obstruction and anemia, this type of tumors should be considered in differential diagnosis. (Acta gastroenterol. belg., **2005**, 68, **95-97**).

Key words : stromal tumor, duodenum, Whipple procedure.

Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the digestive tract (1). The concept of GIST has evolved significantly in recent years. GIST is now defined as KIT (CD117, stem cell factor receptor)-positive mesenchymal spindle cell or epithelioid neoplasms primary in the gastrointestinal tract, omentum and mesentery (2). The cell of origin considered to be the intestinal pacemaker cell, interstitial cells of Cajal (ICCs) (3-6). Duodenal stromal tumors, although not common, are most often found in the second part of the duodenum. Mesenchymal tumors are generally located within the muscularis mucosa or submucosa and may cause obstructive symptoms or may lead to ulceration and bleeding. They also may be noted when endoscopy is performed for unrelated symptoms (8).

The clinical presentation of duodenal tumors is nonspecific and insidious. The usual modes of presentation of these tumors are anemia related to occult or massive gastrointestinal bleeding, abdominal pain, weight loss and incomplete intestinal obstruction. As many as 60% of benign tumors of the small intestine may be asymptomatic and cannot be diagnosed easily (9).

Accurate diagnosis is difficult and special investigations may be required. Barium contrast radiography, computerized tomography (CT), endoscopic ultrasonography, magnetic resonance imaging (MRI), endoscopy and angiography are reported to be of use in diagnosis (10).

In the following case report, a giant duodenal stromal tumor causing upper gastrointestinal obstruction and anemia is reported.

Case report

A 78-year-old woman complaining of abdominal pain, nausea and vomiting after meal and weight loss for three months was admitted to our clinic. On physical examination, there was only epigastric tenderness. No mass was palpated. Rectal examination was normal, but fecal occult blood test was positive. Hemoglobin value was 9.7 g/dl, total protein was 6.4 g/dl and albumin was 3.3 g/dl. Other laboratory tests, including carcinoembriogenic antigen, alpha-fetoprotein, CA 15-3, and CA 19-9, were normal. The abdominal CT scan showed a 9.0 × 7.5 cm heterogeneous mass (Fig. 1). Endoscopy confirmed a polypoid and vegetative mass in the second part of the duodenum. Histopathological diagnosis of endoscopic biopsy was gastrointestinal stromal tumor. The celiac angiography revealed a probably malignant mass vascularizing from superior mesenteric artery and causing distortion of the branches of celiac and pancreaticoduodenal arteries (Fig. 2).

At operation, a 9 × 8 cm intraluminal mass extending from the second part to the fourth part of the duodenum was observed. The mass was fixed to the gallbladder and adjacent tissues. Pancreaticoduodenectomy (Whipple procedure) was performed. On the 11th postoperative day, relaparotomy was performed due to biliary leakage from the subhepatic drain. On exploration, it was seen that biliary leakage was from the choledochojejunostomy. Choledochojejunostomy and pancreaticojejunostomy were revised. She was discharged on the postoperative 25th day.

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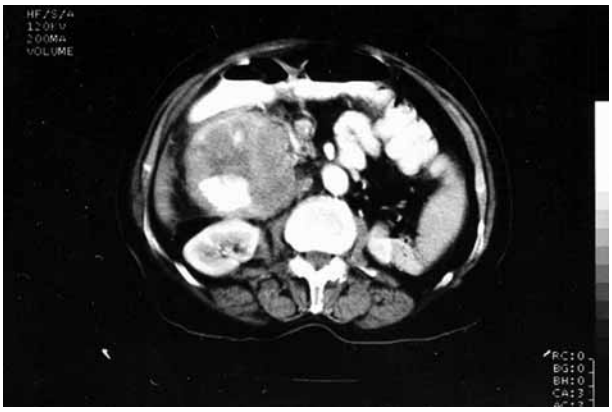

Fig. 1. — Contrast-enhanced computerized tomography demonstrating heterogeneous mass.

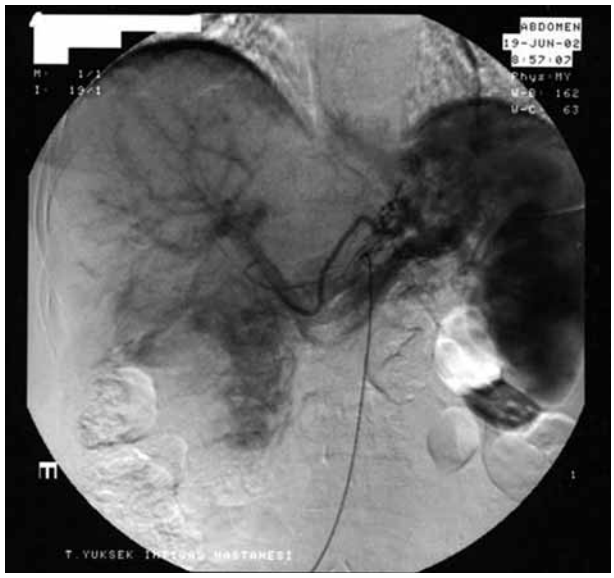

Fig. 2. — Celiac angiography showing a probably malignant mass vascularizing from superior mesenteric artery.

In gross pathologic examination, intramural polypoid mass with ill-defined borders, 8 × 7 × 3 cm in size, showing protrusion into the lumen of small intestine with central ulceration was observed. The cut surface was smooth, gray-white in color and showed no necrosis or hemorrhage. In addition to this, a cholecystectomy material with nonspecific macroscopic findings was noticed. All the resected specimens were fixed in 10% buffered formalin solution for a few days. After fixation 2-mm slices were taken from tumor, adjacent tissues, surgical borders and cholecystectomy material for routine processing. Each paraffin block was sectioned at 5-μm for hematoxylin and eosin staining. Also, tumor blocks (total of 10 blocks) were sectioned at 5-μm for immunohistochemistry.

Under light microscopy, tumor, beginning from the submucosa and reaching but not invading the serosal layer that consists of bundles of spindle cells crossing each other with cigar shaped nuclei and perinuclear halo was observed. Pressure atrophy has caused ulceration of the intestinal mucosa overlying the tumor tissue (Fig. 3 and 4). There was no mitotic activity or cellular atypia within tumor cells. The surgical borders were all intact and there was no lymph node involvement. Immunostaining demonstrated wide-spread tumor cell reaction for vimentin, desmin and smooth muscle actin, but there was no reaction for S-100 protein. So, the tumor was diagnosed as stromal tumor.

The patient has been symptom-free during 18 months of follow-up.

Discussion

Gastrointestinal stromal tumors (GISTs) constitute the largest group of mesenchymal tumors of the gastrointestinal tract. GISTs comprise the vast majority of the combined group of smooth muscle and stromal tumors of the duodenum. Since older case reports or small series are difficult to translate into modern classification, the relative frequency of duodenal GISTs is unknown. GISTs are most common in the stomach and small intestine (11).

The presentation of benign duodenal tumors may mimic a vast array of gastrointestinal pathologies (12). The most common symptom at presentation is anemia (83-100%) due to acute gastrointestinal bleeding or occult blood loss (9,10). Other presenting symptoms are abdominal discomfort and pain, weight loss, obstruction and jaundice (12). Our patient was admitted due to abdominal pain, nausea and vomiting after meal and weight loss. She was anemic and fecal occult blood test was positive.

The diagnosis of duodenal tumors is difficult and special investigations may be required. Barium contrast radiography, CT, MRI, endoscopy and angiography are diagnostic tools (10). There has been an increasing use of endoscopic ultrasonograpy (EUS) and CT-guided fine-needle aspiration in the evaluation of intraabdominal masses (8,13-15). In our case, endoscopy confirmed a polypoid and vegetative mass in the second part of the duodenum. CT and angiography were used to distinguish the nature and spread of the lesion. On the CT scan a 9.0 × 7.5 cm heterogeneous mass was detected. The celiac angiography revealed a probably malignant mass. Histopathological diagnosis of endoscopic biopsy was gastrointestinal stromal tumor.

Surgery remains the standard treatment for primary duodenal tumors (16). Even in the presence of a single mass without evidence of contiguous organ involvement or metastatic disease, the lesion should be treated as malignant. GISTs can behave as a malignant tumor with delayed recurrence even when histologic features indicate a benign nature (1). In our patient, a 9 × 8 cm intraluminal mass extending from the second part to the fourth part of the duodenum and causing intestinal

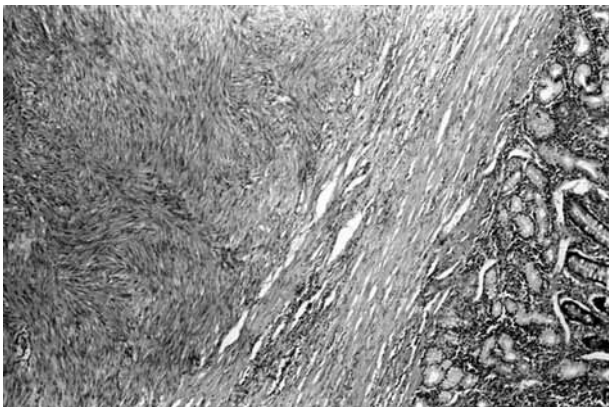

Fig. 3. — Tumoral tissue which consists of smooth muscle fibers crossing each other, is located just underneath the mucosa and replace the muscular layer (hematoxylin and eosin stain, ×20).

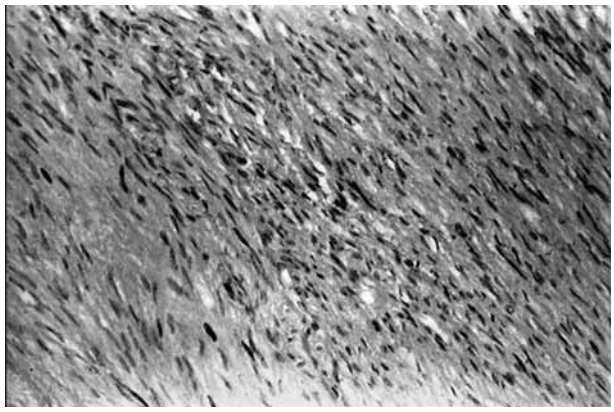

Fig. 4. — A higher magnification of tumoral tissue that does not show atypia, pleomorphism or mitotic activity (hematoxylin and eosin stain, ×200).

obstruction was observed. The mass was fixed to the gallbladder and adjacent tissues. Although histopathological diagnosis was benign stromal tumor, we performed pancreaticoduodenectomy (Whipple procedure) due to the explorative findings. On the postoperative 11th day, the patient was reoperated because of biliary leakage. Choledochojejunostomy and pancreaticojejunostomy were revised. After the second operation, our patient was recovered and she has been symptom-free for 18 months.

The concept of GISTs was introduced several years ago but the cellular origin of GISTs was not known (17). The protooncogene *c-kit* is the transmembrane tyrosine kinase receptor (18). Hirota *et al.* (5) showed that the majority (94%) of GISTs stained positively with a polyclonal *c-kit* antibody, CD117. Kindblom *et al.* (3) found that 100% of GISTs in their serial were CD117 positive. Since interstitial cells of Cajal (ICCs) also stained with antibodies to both CD34 and CD117, they postulated that GISTs might originate from ICCs. In another study, Miettinen *et al.* (11) demonstrated that 54% of GISTs were positive for CD34 and 39% were positive for smooth muscle actin, but none was positive for desmin.

Although duodenal stromal tumor is a very rare pathology, we advise to consider this clinical condition in the differential diagnosis of patients with anemia and upper gastrointestinal obstruction.

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