ANNALI ITALIANI DI CHIRURGIA

Digital Edition
e-publish on-line
ISSN 2239-253X
Direttore Nicola Picardi

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Ann Ital Chir, Digital Edition 2017, 6
pii: S2239253X17027220 - Epub, July 25
free reading: www.annitalchir.com

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Duodenal gastrointestinal stromal tumor. A case report

AIM: *To report an another case of duodenal GIST in patient that was treated with Whipple procedure.*
MATERIAL OF STUDY: *We report a case of 69 years old men that was admitted with melena. The preoperative investigations suggested a suspected duodenal GIST.A Whipple pancreatico-duodenectomy was performed. Only 3-5% of GISTs occur in the duodenum.*
DISCUSSION: *GIST were described as a separate entity by Mazur and Clark in 1983. These tumors are the most common among mesenchymal tumors of the gastrointeninal tract. The clinical presentations of duodenal GIST are highly variable. There is no consensus on the optimal surgical treatment of duodenal GIST.The aim was to achieve a R0-type surgery with complete en bloc surgical resection of the tumor and the surrounding tissue*
CONCLUSION: *In conclusion the duodenal GIST are fairly rare. The clinical symptomatology is very variable even if frequently occur with digestive bleeding. the purpose of the surgery is the complete removal of the tumor (R0) however looking to perform minimum resections.*

KEY WORDS: GI bleeding, GIST, Pancreaticoduodenectomy

Introduction

GIST were described as a separate entity by Mazur and Clark in 1983. These tumors are the mesenchymal most common of the gastrointestinal tract [1,2]. They can arise everywhere in the GI tract, even though they are more frequently found in the stomach (about 60%), small bowel (about 25%), colon and rectum (about 10%), while only 3-5% of GISTs occur in the duodenum. There are only sporadic reports of extra-gastrointestinal stromal tumors (e-GISTs) arising in the omentum, mesentery or retro- peritoneum [2-4]. Duodenal GISTs commonly present with GI bleeding (20-50%) and chronic anaemia, epigastric pain (50-70%), a palpable mass, and at a later stage, intestinal or biliary obstruction [3,5-8]. Actually there is no consensus on the surgical treatment of duodenal GISTs.Owing to the complex anatomy of the duodeno-pancreatic region, these tumors are often challenging for surgical management as radical resection may require extensive procedures such as pancreaticoduodenectomy [9-10]. We report a case of duodenal GIST that received follow-up care and treatment after upper gastrointestinal haemorrhage and after the performance of a Whipple procedure.

Case Presentation

A 69 years-old man was admitted in our hospital with melena, asthenia and dizziness. The blood tests revealed a severe anaemia. After the restoration of red blood cell mass, duodenal endoscopy was performed by an experienced endoscopist, who didn't found mucosal or submucosal abnormalities in the duodenum. Videocapsule endoscopy was performed that revealed a suspected Gist of the II portions of duodenum. CT scan confirm the presence of heterogeneous, hypervascular mass of 32 mm

Pervenuto in Redazione Aprile 2017. Accettato per la pubblicazione Luglio 2017
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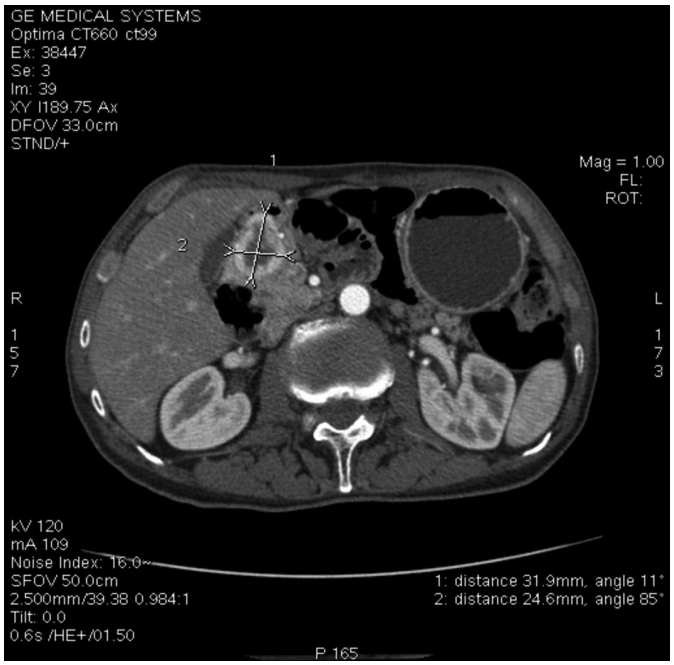

Fig. 1: Image CT scan.

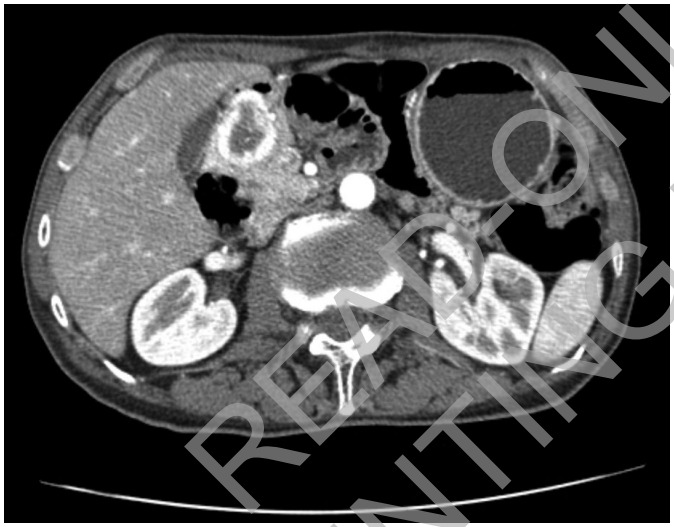

Fig. 2: Image CT scan.

x 25 mm. After completing the investigations, the patients was operated. The peritoneal cavity exploration confirmed the presence of a mass to the second portions of duodenum near the papilla, and therefore a pancreatico-duodenectomy according the Whipple procedure was performed. The histopathological exams conclude: duodenal GIST, originated from muscular layer and with extension in the submucosa and in pacreatic parenchyma, with spindle cells and a mitotic index 1 per 50 high-power field. The immunocytochemical revealed positive staining for c-kit protein and DOG1, but negative reactions for CD34, smooth muscle actin and S100. The margins were negative for tumor and no metastasis was

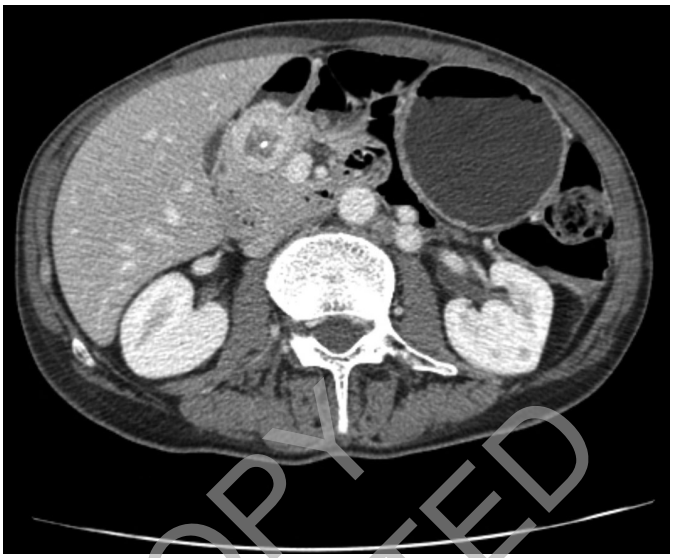

Fig. 3: Image CT scan.

noted in the regional lymph nodes which were excised. According to the small size of the tumor (2,2 cm of diameter) and the low mitotic index, the tumor was finally diagnosed as a GIST with low-grade malignancy. The patient had an uneventful postoperative recovery and was discharged after 14 days. After multidisciplinary team consultation and according to the stage of the disease, the patient was not administrated chemotherapy. He was well followed up at our department and she is alive 12 month after surgery.

Discussion

Gastrointestinal stromal tumors are low-grade malignant mesenchymal tumors of gastrointestinal tract and are believed to originate from the neoplastic transformation of the interstitial cells of Cajal from their precursors [11]. These tumors are characterized by the expression of a transmembrane receptor tyrosine kinase kit, a product of the c-kit proto-oncogene and identified by the expression of CD117 which is present in over 95% of these patients [12]. Moreover, 70-90% also express CD34, 20–30 % actin, 8-10% S-100 and desmin in 2-4% [12]. They are more frequently found in the stomach (about 60%) and only less than 4% are in the duodenum where frequently involve the 2nd portion followed by the 3rd ,4th and 1st portions [3]. Generally metastases are intraabdominal, involving the peritoneum and liver. The majority of GIST are sporadic but there are many syndromes associated with GIST (Carney Triad, Neurofibromatosis Type I, Carney –Stratakis Syndrome) [13-15]. The clinical presentations of duodenal GIST are highly variable according to their size and the existence of mucosal ulceration. According to the previous report, duodenal GISTs

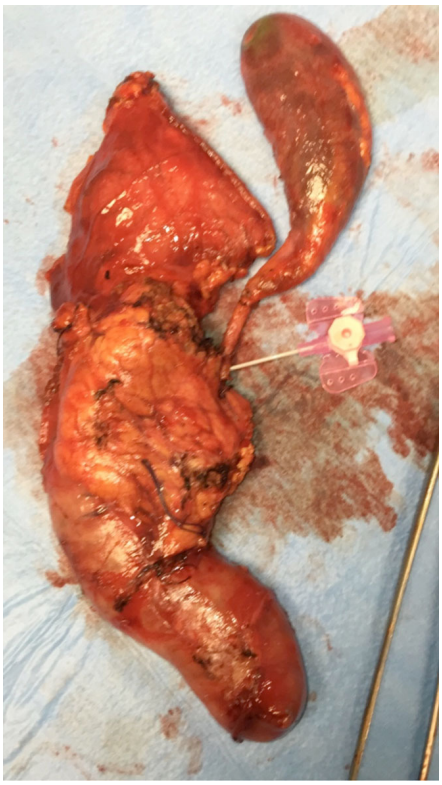

Fig. 4: Whipple procedure specimen.

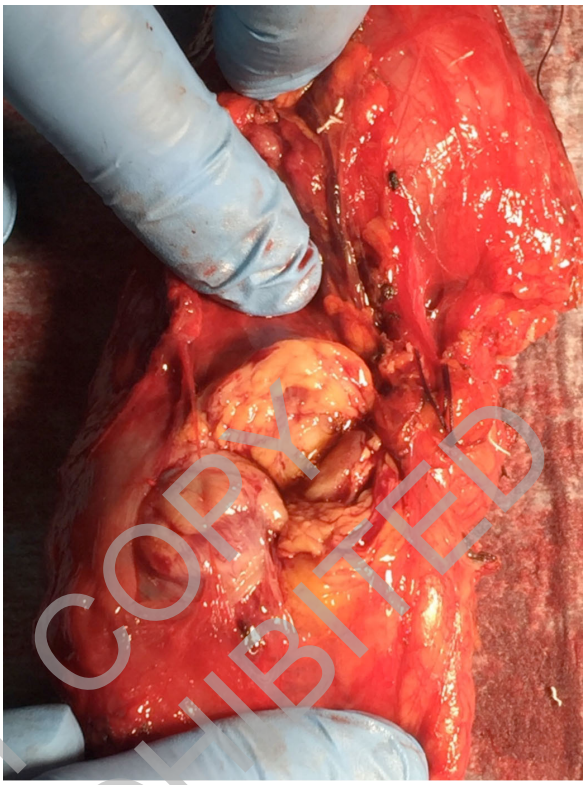

Fig. 5: Section of duodenal tumor.

most commonly present GI bleeding, epigastric pain, palpable mass, and intestinal obstruction [3]. Diagnosis of GIST is similar to that of other digestive-tract tumors. Gastrontestinal endoscopy remains the most common diagnostic procedure in duodenal GISTs, especially in patients with intramural growth or mucosa ulceration and bleeding [16,17]. It allows forceps biopsy, which is not ever helpful in extraluminal tumor. In the latter, the most used diagnostic test remains CT scan or MRI. There is no consensus on the optimal surgical treatment of duodenal GIST.The aim was to achieve a R0-type surgery with complete en bloc surgical resection of the tumor and the surrounding tissue [3,17-21]. The ideal surgical procedure depends on the size, anatomical location of the tumor (relationship to the ampulla) and possible adjacent organ involvement [6]. Limited resection should be considered a viable treatment option for duodenal GISTs when technically feasible. Various techniques of limited resection for duodenal GISTs have been advocated. Duodenal Wedge resection can be performed for small tumors (1-2 cm) provided located on the duodenal wall at least 2 cm from the papilla [3,6,16]. A segmental duodenectomy with side-to-end or end-to-end duodenojejunostomy may be performed for larger tumors located in the 3rd and 4th portions of the duodenum [22]. Recently, partial duodenectomy with a Roux-en-Y-duodeno-jejunal anastomosis has been proposed for large tumors with involved ntimesenteric border of the second and third portions of the duodenum [6]. But when the tumor is located in the second part of the duodenum and involves the papilla, pancreas or the duodenum bulb, or if the tumor is large with high malignant potential, major resection as pancreaticoduodenectomy is indicated [3,21,23]. However, limited resection, when feasible, is perceived to contribute to a better quality of life, functional preservation of the pancreas and continuity of the gastrointestinal tract [17,18,19,21]. In contrast, a pancreaticoduodenectomy as a treatment for duodenal GISTs can provide a wider tumor clearance but may be associated with excessive morbidity especially in patients with a tumor of low grade malignancy. Imatinib, which is an inhibitor of the tyrosine kinase activity of C-Kit, has been added as a line of treatment of GISTs [24]. Neoadjuvant and adjuvant therapy with imatinib has been shown to reduce the risk of recurrence and improve the survival [25]. Imatinib can be used as neoadjuvant therapy, for large size GISTs to reduce tumor size, increase the rate of complete resection of the tumor, and help to improve prognosis [26,27]. The prognoses of GISTs vary widely according to tumor size, mitotic index, organs from which the tumor originates, and c-KIT mutation. Fletcher et al. [28] showed that tumors have a stronger chance of being malignant in cases where they are more than 5 cm in size or in tumors that are less than 5 cm but have a number of mitosis on the 5 mitosis /50 (>5–10 mitoses per 50 high power fields) [29].

Conclusion

The duodenal GIST are fairly rare. The clinical symptomatology is very variable even if frequently occur with digestive bleeding. Thepurpose of the surgery is the complete removal of the tumor (R0) however looking to perform minimum resections. Local resections, when technically feasible, are preferred for the lower incidence of morbidity and complications. But for larger tumors, or for tumor localized in the second duodenal portion, more extensive resections are necessary as the pancreaticoduodenectomy. In the case that we reported, the tumor was less than 1 cm from the duodenal papilla so we performed a pancreaticoduodenectomy according to Whipple. The patient has not undergone adjuvant imatinib therapy since it has been classified as a gist low-grade malignancy

Riassunto

I GIST, descritti per la prima volta come entità separate da Mazur e Clark nel 1983, sono i tumori mesenchimali più comuni del tratto gastrointestinale. Possono interessare qualsiasi porzione del tratto GI, ma più frequentemente si trovano nello stomaco (circa il 60%), dell'intestino tenue (circa il 25%), colon e retto (circa il 10%), mentre solo il 3-5% dei GIST si verificano nel duodeno. Dal punto di vista clinico la sintomatologia può essere estremamente variabile anche se più frequentemente si manifestano con emorragie digestive, anemia cronica, dolore epigastrico o massa palpabile. La terapia è chirurgica e ha come obiettivo l'asportazione completa del tumore con resezioni R0. Allo stato attuale non vi è consenso sulla strategia chirurgica da adottare. Quando possibile e tecnicamente fattibile, si preferiscono delle resezioni locali e contenute, ma vista la complessità anatomica della regione cefalo-pancreatica, a volte si rendono necessarie resezioni più estese quali anche la duodenocefalopancreasectomia soprattutto per i tumori più grandi e/o localizzati nella seconda porzione duodenale. In questo articolo riportiamo il caso di un paziente giunto alla nostra osservazione per melena da GIST duodenale sottoposto ad una duodenocefalopancreasectomia.Il paziente non è stato sottoposto a terapia adiuvante con imatinib poiché è stato classificata come un GIST a basso rischio.

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