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Case Report

Multidisciplinary approach and segmental resection of the third portion of the duodenum for a gastrointestinal stromal tumour: a case report

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Abstract. Gastrointestinal stromal tumours (GIST) are known as the most frequent mesenchymal tumours of the gastrointestinal tract. In literature it is described that only 4.5% of GISTs are located in the duodenum. There is no exact consensus, if duodenal GISTs should be operated by a duodenopancreactectomy (DPC) or by a segmental resection of the duodenum. A patient of 49 years old presented to the emergency department with an epigastralgia associated with melena that appeared since 24 hours. Preoperative examinations diagnosed the presence of a tumor of 25mm in size on the anterior face of the D3, without visible ulcerations. After an elective radiologic embolisation, a segmental resection of the third portion of the duodenum with duodeno-jenunal anastomosis was performed. Histology was in favour of a GIST. GISTs of the duodenum are an origin of gastrointestinal bleeding. Limited resection of the duodenum can serve as a less extensive surgical intervention for GIST situated in the third portion of the duodenum.

Key Words: gastrointestinal stromal tumours, melena, duodenum resection

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Introduction

Gastrointestinal stromal tumours (GISTs) represent the most common mesenchymal tumours arising in the gastrointestinal tract (Fletcher et al 2002; Rubin et al 2007). The location of GISTs can be anywhere in the gastrointestinal tract, stomach (40-60%) and small intestine (30-40%) and duodenal GIST amounts to 4.5% of all GISTs (Pidhorechly et al 2000; Goettsch et al 2005). Surgical resection is the treatment of choice for gastrointestinal stromal tumours (Pidhorechly et al 2000; Dematteo et al 2000; Goh et al 2008). Whenever possible an en-bloc resection is recommended. The most frequent clinical presentations are gastrointestinal bleeding when there is mucosal ulceration, and abdominal pain (Goh et al 2008; Winfield et al 2006). Due to the low incidence of GIST located in the duodenum, the best treatment for duodenal GISTs is not described (Mennigen et al 2008; Kwon et al 2007). In duodenal GISTs regional lymph node metastases is rare (Gervaz et al 2009; Goldblum et al 1995), and duodenopancreactectomy, which has been used to treat about 40% of reported duodenal GISTs (Uehara et al 2001; Goh et al 2008), may be an extreme measure of treating this pathology.
In view of this we report a case in which segmental resection of the third duodenum and duodeno-jejunal anastomosis was efficacious in treating a duodenal GIST.

Case report

A 49-year old male patient without a history of known pre-existing diseases presented to the emergency department with an epigastralgia associated with melena with a debut since 24 hours. The systematic blood test revealed a haemoglobin level of 9.8g/dl. The patient was admitted in the intensive care unit for monitorisation.
Once the patient was stable several examinations consisting of gastroscopy, abdominal CT-scan and ileocolonoscopy were realised. It was diagnosed the presence of a polyp about 25mm in size on the anterior face of the D3, without visible ulcerations, but totally pathological macroscopically. The patient continued to have a haemorrhage with a haemoglobin level of 7.2g/dl, which could not be controlled by endoscopic interventional approach. This led to blood and platelets derivatives transfusion in the intensive care unit.
An ultra selective arteriography, allowed the discovery of an angiodysplasia of the D3, which was most likely the source of the haemorrhage. The radiologic interventional department performed an elective embolisation (Figure1, Figure2), and a marking by methylene blue for the subsequent surgical intervention to be performed.
Under general anaesthesia and thoracic epidural anaesthesia the abdominal cavity was approached through a bi-sub costal incision. Intraoperative no secondary lesions were observed intra

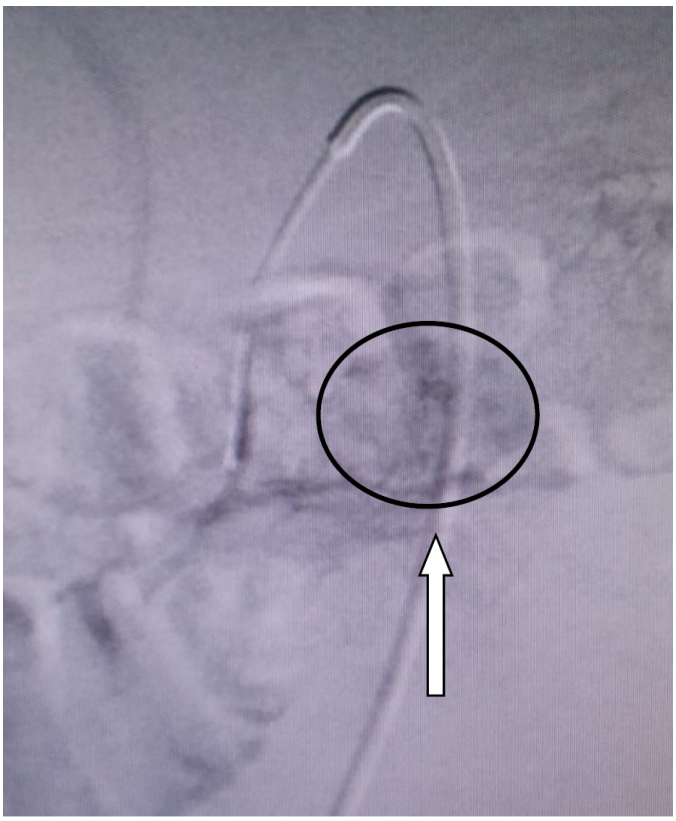

Figure 1. Tumoral mass during arteriography

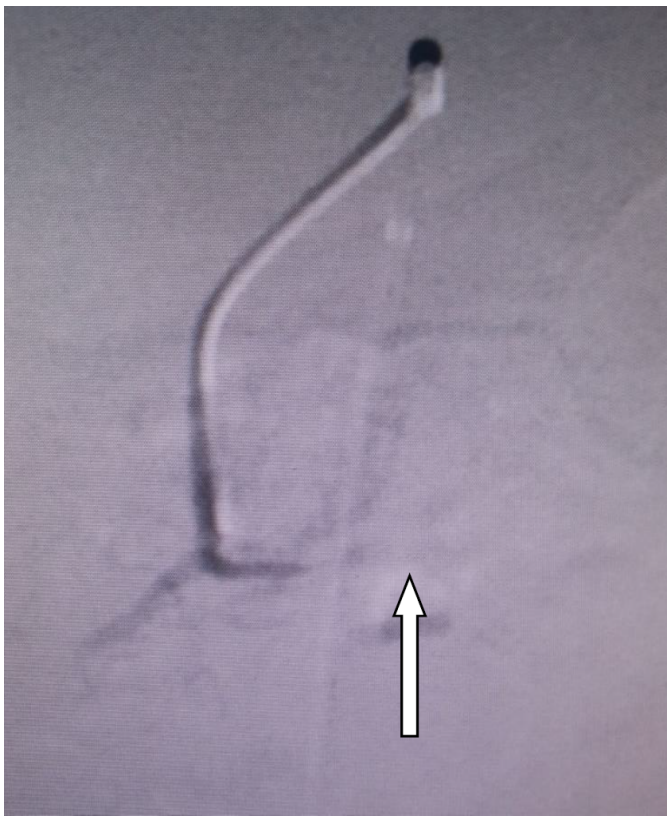

Figure 2. Post transarterial embolisation

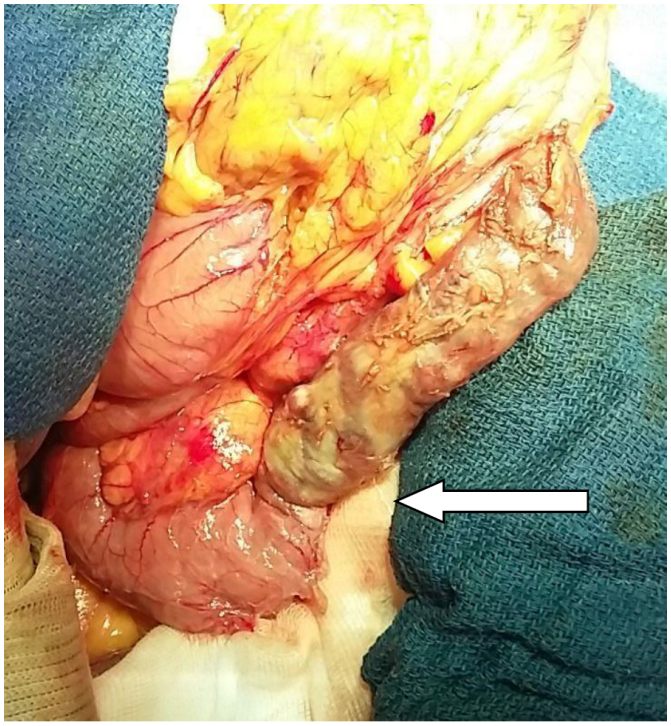

Figure 3. Dissection and complete mobilisation of D3

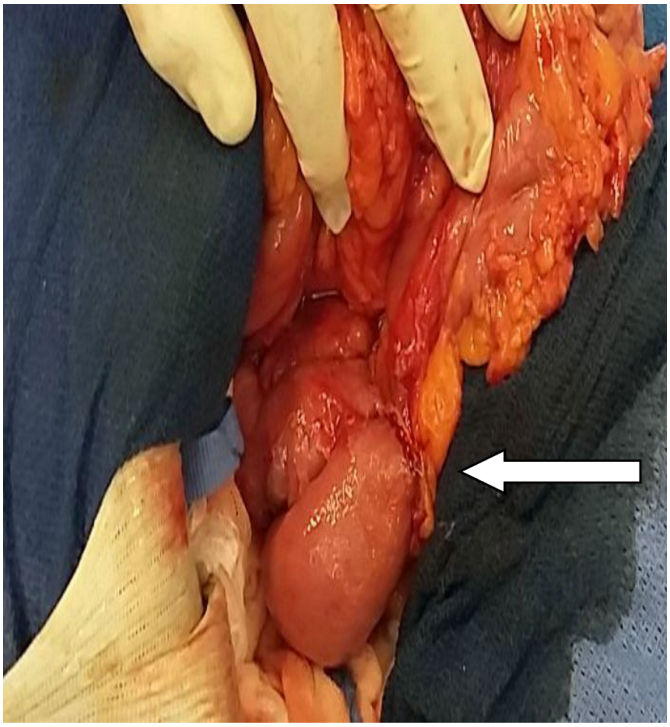

Figure 4. Duodeno-jejunal latero-lateral iso-peristaltic anastomosis

peritoneal, absence of ascitic fluid and the hepatic parenchyma was normal. We proceeded with a cholecystectomy, and systematically a specimen of bile was sent for bacteriology analysis. Furthermore dissection was done at the angle of Treitz, but due to difficulty accessing the lesion, the Kocher manoeuvre was mandatory. This allowed a complete pancreatico-duodeno en bloc mobilisation. Mechanical sectioning of the duodenum at 12 cm from angle of Treitz was performed and dissection of the mesentery in contact with the superior mesentery. With a complete dissection and margin free of 2cm from the lesion, we

decided for a segmental duodenectomy instead of a DPC. We completed the intervention with a mechanical duodeno-jejunal latero-lateral iso-peristaltic anastomosis (Figure3, Figure 4). The operation time was 4h, and blood loss was 20 ml.

The post-operative course was uneventful, and the patient was discharged 10 days after surgery. The histo-pathology confirmed a chronic cholecystitis and the immunohistochemistry fully compatible with a GIST. Based on the Miettinen classification it was classified as grade I.

Discussion

GISTs are explained to derive from the interstitial cells of Cajal, which are mesenchymal stem cells or intestinal pacemaker cells (Kindblom et al 1998). GISTs represent the most frequent mesenchymal tumour entity of the gastrointestinal tract. In immunohistochemistry a sensitive marker for GISTs is the KIT protein CD117, a transmembrane receptor linked to an intracytoplasmatic tyrosine kinase (Sarlomo-Rikala et al 1998). A smooth muscle marker, h-caldesmon, is expressed in >70 % of GIST and smooth muscle antigen (SMA) in 30-40 %, whereas desmin positive staining is observed in <5 % and neural markers such as S100 are observed in <1 % of GIST (Joensuu 2006). GISTs can be located anywhere in the GI-tract. Most common sites are stomach (40–60%) and small intestine (30–40%) (Fletcher et al 2002; Goettsch et al 2005). Most patients presenting with a GIST have a mean age of 53 years. Only about 5% of GIST patients are younger than 30 years (Miettinen et al 2003). GISTs of the duodenum make up only 4.5% of all GISTs (Goettsch et al 2005) and therefore express a rare tumour location.

Miettinen published a clinicopathologic study on 156 duodenal GISTs in 2003 giving understanding about tumour characteristics and natural history (Miettinen et al 2003). Most duodenal GISTs present with GI bleeding usually associated with melena, occasionally with massive acute bleeding like in our patient (Goh et al 2008; Winfield et al 2006). Our patient did not present other symptoms like abdominal pain, early satiety, bloating, or obstructive jaundice due to involvement of the papilla of Vater. Endoscopic detection of duodenal GISTs is easily possible in case of a visual endoluminal mass. This was the case in our patient.

In our patient, acute gastrointestinal bleeding that could not be controlled by endoscopy led to an emergency supervision in the intensive care unit with red blood cells and platelets derivatives transfusions. After stabilisation of our patient we controlled the bleeding by transarterial embolisation, which is in accordance to the literature described by Kurihara (Kurihara et al 2005). This procedure should be given due consideration in case of acute bleeding from duodenal GIST, if angiography is available within reasonable time.

Surgery is the therapy of choice for localized GIST. Common sites of metastases from GISTs are in the peritoneal cavity and in the liver, and in a lesser frequency, in lung, and bones. In contrast, a lymph nodal spread is uncommon, and obligatory lymph node dissection has no proven value (Joensuu 2006; Miettinen et al 2003; Fujimoto et al 2003). In our patient, no suspicious peritumoral lymph nodes were present. Therefore, in order to minimize operative morbidity, no lymph node dissection was performed.

Several surgical techniques have to be considered for the therapy of duodenal GIST. Small tumours can be treated by local excision if the remaining lumen is adequate. Segmental resection of the duodenum with the need of a duodenojejunostomy, as performed in our patient, is another possibility. Tumours located near to the ampulla of Vater with local invasion may require a duodenopancreatectomy. In the study of Miettinen, about 20% of patients underwent duodenopancreatectomy, whereas segmental resection and local wedge resection were performed in 45% and 20%, respectively (Miettinen et al 2003). In another study done by Uehara 40% of patients treated by duodenopancreatectomy for duodenal GIST was reported (Uehara et al 2001). Taking into account that only 30% of duodenal GISTs show a malignant appearance, duodenopancreatectomy can be considered an extreme operation, especially as this procedure leads to a significant morbidity and mortality. There is limited documentation available on the choice of surgical procedures for duodenal GIST.

Conclusion

We present a case of a duodenal GIST located in the third portion of the duodenum treated by a segmental duodenal resection. Extensive lymph node dissection is not obligatory, due to the rarity of metastases in regional lymph nodes. In an emergency case of active bleeding transarterial embolisation is beneficial. We confirm the feasibility of segmental resection of the third portion of the duodenum, avoiding the higher morbidity and mortality of a duodenopancreatectomy.

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