

Duodenal tumors

Four case reports



Ann. Ital. Chir., 2014 85: 323-327
pii: S0003469X14021423

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Duodenal tumors. Four case reports

AIM: Reporting our experience in the treatment of duodenal neoplasms.

MATERIAL OF STUDY: Management and treatment of four patients presenting a duodenal neoplasm.

RESULTS: Three out of four patients died after surgery for dehiscence and/or bleeding. One, who underwent palliative bypass, died after four months from unknown causes.

DISCUSSION: Duodenal neoplasms are rare and their diagnosis is difficult and late. In literature, there is no standardized management of this kind of neoplasm; the commonest surgical approaches are pancreaticoduodenectomy, segmental duodenal resection and palliative operations. The surgical choice is based on site and stage, both in Literature than in our experience. Regardless of the surgical decision, the patients have a poor outcome because of an advanced stage at diagnosis.

CONCLUSIONS: It can be asserted that duodenal neoplasia offers many diagnostic and therapeutic difficulties. This may be due to its rarity, which does not allow the definition of a standard course of treatment, resulting in the inability to have a unique diagnostic and therapeutic approach. There is often a late diagnosis and the need to perform emergency surgery. Therefore prognosis is greatly aggravated by the high incidence of postoperative complications, in part due to the urgency in which the surgeries are carried out.

KEY WORDS: Duodenal neoplasms, Small bowel

Introduction

Small bowel malignancies are rare disease and represent less than 5% of malignant tumors of the digestive tract. They are most frequently found in the duodenum (52-55%), followed by jejunum (18-25%) and ileum (13%). The most common histologic subtype is adenocarcinoma but cases of neuroendocrine tumors, lymphomas, sarcomas and stromal tumors (GIST) can also be observed¹. Despite the extension of the small bowel compared with the large one, which is the main site of intestinal neo-

plasms, the overall low incidence of duodenal carcinoma may be due to the presence of several protective factors including rapid small bowel transit time and consequently low toxin exposure, immunological factors and an alkaline environment. On the contrary, food (diet rich in red meat, salt-cured or smoked foods), alcohol, tobacco, intestinal diseases, especially those with a genetic component (familial adenomatous polyposis-FAP, Crohn's disease, Peutz-Jeghers syndrome, adenomas) are considered the main risk factors for this type of cancer². However, it remains unexplained why the focal point of these neoplasms is the duodenum³.

Clinical signs and symptoms of these malignancies are often nonspecific and consist of abdominal pain, nausea, vomiting, anorexia, weight loss and bleeding or intestinal obstruction, this one being typical of advanced stages³. The shortage of clinical manifestations is responsible for a delayed diagnosis, although, nowadays the

Pervenuto in Redazione Febbraio 2013. Accettato per la pubblicazione Maggio 2013.

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development of endoscopic and radiologic imaging has allowed their early identification and has improved the identification of the true incidence of small duodenal cancers in the last years ^{4,6}.

Because of their rarity, there is no global consensus on their surgical treatment. Herein, we report our experience.

Case Reports

A 71-year-old man was admitted to the Emergency Room due to a poor general medical condition consisting in disorientation, subileus and laboratory tests showed anemia and cholestasis. His medical history was significant for a palliative GEA (gastro-entero anastomosis), carried out in another hospital for a stenosing lesion of the second and third portion of duodenum (D2-D3); it was uncleavable by pancreas, but not infiltrating mesenteric vessels. Despite symptoms, a neurological examination and cerebral computed tomography (CT) were negative for disease. The plain abdominal X-ray with Gastrografen[®] showed a faulty emptying of GEA. The patient underwent pancreaticoduodenectomy (DCP) with Child reconstruction and was transferred to ICU for hemodynamic instability, adverse to medical treatment. In addition tonic-clonic seizures appeared. A cerebral CT showed a picture of ongoing ischemia, probably of venous origin.

On the fourth day, the patient died. Histological examination indicated an adenocarcinoma G2 T4N1Mx.

An 86-year-old woman arrived presenting an intestinal obstruction. She had a medical history of rheumatic fever with aortic steno-insufficiency, atrial fibrillation, hiatal hernia and sigmoid diverticula. About four months before, the patient was admitted to the department of emergency medicine for pleural and pericardial effusion, acute anemia and infection by Varicella-Zoster Herpes. During this hospitalisation, because of persistent vomiting, the patient underwent esophagogastroduodenoscopy (EGDS) which showed a stenosis of the third portion of the duodenum. Abdominal CT identified a stenosis surrounding D3 and infiltrating the superior mesenteric vein. Because of intestinal obstruction and severe gastric dilatation, the patient underwent segmental resection with duodenum-jejunum end-to-end anastomosis. A postoperative abdominal X-ray with Gastrografen[®] showed a persistent dilatation of stomach and edema of the anastomosis. On the fourth day, the patient presented cardiovascular-respiratory failure and peritoneal reaction; she died on the sixth day.

During the autopsy suppurative peritonitis with septic state, ischemic necrosis of intestinal loops with areas of drilling near the anastomosis, incomplete obstructive atherosclerosis and neoplastic infiltration of adventitia of the superior mesenteric artery were found.

Histopathology confirmed a diagnosis of tubulo-papillary

adenocarcinoma G2 T3N0Mx stage IIA without lymph node involvement.

A 79-year-old woman was admitted to the Emergency Room with melena and severe anemia. She had a family history of gastrointestinal cancer and cardiovascular disease; she was a former smoker and her medical history was also significant for subtotal gastrectomy, for peptic disease and hypothyroidism. She was treated with oral anticoagulants due to surgical implantation of aortic valve, hypertension and stroke.

About 2 months before, the patient was admitted to another hospital with melena and anemia. An angiodysplasia, near gastro-jejunal anastomosis, was diagnosed and treated endoscopically. Once in our department, the patient underwent EGDS which showed the presence of a fungating and excavated lesion with stigmata of bleeding (probably by afferent loop, ignorant of the type of gastro-jejunal reconstruction performed before). During endoscopy, a biopsy was done.

Due to the persistence of anemia, despite blood transfusion therapy, we proceeded to emergency laparotomy and a duodenal lesion (D2-D3) was found. Considering patient's age and clinical status (post-hemorrhagic anemia), pancreaticoduodenectomy was non-existent, so we performed a segmental resection of the duodenum with duodenum-jejunum end-to-side anastomosis. The surgery was completed with cholecystectomy and placement of transcystic drainage (Bracci's catheter) to the mouth of Vater's papilla.

The postoperative course was complicated by an episode of atrial fibrillation, regressed by pharmacological therapy, and bilateral pleural effusion, which decreased temporarily by chest drainage and diuretic therapy. On the seventh postoperative day, anticoagulant therapy was restarted. A trans-catheter cholangiography was performed which showed a leakage probably from anastomotic site. On the eighth postoperative day severe anemia with a dark red bleeding from the drain tube was observed and it was decided to carry out a restoration of plasma volume and non-invasive ventilation (NIV). During the following days, hemodynamic was unstable. Abdominal CT showed anastomotic dehiscence and a presence of blood in the abdomen; CT scan also highlighted bilateral pleural effusions with consensual disventilation.

Due to the poor condition of the patient, we refrained from reoperating. However, her general condition gradually got worse until she died on the twenty-fifth day. The histopathological diagnosis was adenocarcinoma with a medium degree of differentiation T2 N0 Mx.

An 80-year-old woman was referred to our hospital for anemia and vomiting. She had a medical history of hypertension under drug treatment. The endoscopy showed a hiatal hernia and a fungating ulcerated lesion near the second portion of the duodenum which involved the papilla.

Due to progressive anemia, we opted for emergency

surgery. On the operating table, a neoplasm which embraced pancreas and the root of the mesentery was discovered, in the absence of liver metastases and peritoneal carcinomatosis. Owing to the advanced stage of the tumor and the age of the patient, it was decided to perform a palliative bypass with hepatic-jejunostomy and duodenum-jejunostomy on Roux-en-Y.

The postoperative course was regular. On the sixth day radiographic examination showed no signs of anastomotic dehiscence or alteration of bowel canalization. Discharge occurred on the thirteenth day. The patient died after four months for unknown reasons.

Discussion

Neoplasms of the small bowel are rare, representing less than 5% of gastrointestinal cancers, despite the fact that it constitutes about 75% of the length of the whole alimentary tract and about 90% of its mucosal surface². The particular "resistance" to the onset of neoplasms of the small intestine has been explained by several theories:²

- Rapid turnover of mucosal cells;
- Low bacterial flora load;
- Rapid small bowel transit time that reduces exposure to carcinogens;
- Malted chyme which could reduce mechanical trauma and protect small bowel from carcinogens;
- Alkaline environment which prevents the production of nitrosamines;
- Protective local IgA-mediated immune system;
- Low rate of activate precarcinogenic enzymes;
- Fewer stem cells compared to colon and stomach;
- Presence in the duodenum of a water-soluble component inhibiting the onset of tumors.

It is still unexplained why the core of these neoplasms is the duodenum. A possible explanation could be the presence of substances with cancerogenic action in pancreatic juice and bile, although this has not been confirmed by experimental studies³.

Small bowel cancers have a higher incidence in males and the average age of the patients at the time of diagnosis is about 60, although lymphomas occur earlier². In our experience, in contrast to the literature, we observed more cases of duodenal neoplasms in females and an average of diagnosis of over 70.

In literature risk factors and predisposing conditions are food (animal fats, animal protein, red meat, sausages and smoked foods, sugary soft drinks) and a particular lifestyle (intake of tobacco and alcohol);² these tumors are also associated with other diseases, such as adenomas of the small intestine, according to an adenoma-carcinoma sequence, familial adenomatous polyposis, Peutz-Jeghers syndrome, Lynch syndrome, exposure to ionizing radiation, chronic inflammatory disorders such as Crohn's disease and autoimmune disorders such as celiac disease².

Our patients didn't have any illness as predisposing this type of tumors; their nutritional habits and lifestyles were unknown to us.

The main histological type is adenocarcinoma (32-63%), followed by carcinoid (6-35%), lymphoma (7-18%), sarcoma (10-20%) and GIST¹. Among our patients, adenocarcinomas were exclusively found.

Clinical status is often nonspecific, being characterized by the association of intermittent abdominal pains (60%), anemia (50%), nausea and vomiting (50%), weight loss (40%), diarrhea (30%) and intestinal obstruction (30%). In other cases the disease may have a silent course until its final stage⁴.

This nonspecific symptomatology is responsible for a delayed diagnosis, also associated with the difficulties of imaging; direct consequence is a more severe prognosis⁴. On admission our patients showed severe anemia, sometimes associated with nausea, vomiting and subileus; all symptoms emerged suddenly, suggesting a completely asymptomatic course of the disease.

Diagnosis can be made by: endoscopic and radiographic imaging. Among the endoscopic procedures, that allow a direct identification of the pathological process, there are EGDS, extended to the first portion of the small intestine, and capsule endoscopy (VCE), for a more detailed inspection. A further advantage of the EGDS is to allow the execution of biopsies for histopathological typing⁴.

Until recently, barium enema was considered the most accurate radiological procedure. This technique is poorly tolerated by patients and provides a very unreliable assessment of intra- and extramural extension of the neoplasm⁴. Computed tomography (CT) and magnetic resonance imaging (MRI) are extremely suitable for the early detection of both inflammatory and neoplastic small bowel diseases⁴.

Ultrasound and CT are important for staging, especially for the detection of distant metastases, while endoscopic ultrasound is useful in the determination of T (100% diagnostic accuracy) and N (66% accuracy)^{3,5}.

In all our cases the diagnosis was obtained through endoscopy with biopsies, often showing a condition of duodenal stenosis. The other diagnostic techniques, in particular the CT, were used in stable patients, who did not require emergency surgery. CT images have been used mainly to confirm the endoscopic report, in order to stage the disease and to allow the choice of the most appropriate surgical technique to be adopted.

Sometimes laparoscopy can be useful, however it's contraindicated in intestinal obstruction or major bleeding, which is why none of our patients were submitted⁷.

Regarding treatment, in literature there is no yet well defined and codified surgical approach. Surgery may be either radical or palliative, the choice is made especially considering the invasion of the superior mesenteric vessels, the retroperitoneum and the presence of peritoneal carcinomatosis⁸.

Palliative surgical interventions include: biliary, gastric and jejunal bypass and jejunalstomy ⁷.

These treatments should be reserved for patients with advanced disease or patients with poor clinical condition or geriatric patients with high operative risk. In any case these interventions are preferable to non-surgical palliative procedures, as they improve survival ⁹.

Among radical choices, the most practiced are DCP and duodenal segmental resection. Some authors propose as preferential treatment DCP with en bloc resection, regardless of the duodenal portion involved¹⁰. Others choose the procedure based on the tract involved, opting for a segmental duodenal resection in case of the third and fourth portion implication, and others only at the level of D4, reserving the DCP in case of involvement of D1-D2 and D1-D2-D3, respectively ¹¹.

In literature, there would be no difference in terms of oncological radicality and survival between a DCP according to Whipple procedure and the one according to Traverso-Longmire ¹⁰. However, despite attempts, it is often impossible to obtain a curative resection due to the late diagnosis, which allows to act only on a very advanced stage ¹⁰.

With regards to our patients, surgical treatment has been varied according to general medical conditions, tumor side and involvement of pancreas and vessels.

Indeed, in the first case a DCP was performed based on CT images which showed a lesion at the level of the second and third duodenal portion inseparable from the pancreas but without infiltration of the superior mesenteric vessels.

On the other hand the second patient has undergone a duodenal segmental resection with duodenum-jejunum end-to-end anastomosis, according to CT showing a neoplasm of D3 and infiltrating the superior mesenteric vessels.

Moreover in the third case, we opted for a segmental duodenal resection with duodenum-jejunum end-to-side anastomosis, mainly because of the serious condition of the patient which required an emergency surgery.

Finally in the last patient, we decided to perform a palliative surgery consisting of hepatic-jejunostomy and duodenum-jejunostomy because of a tumor in the second portion of the duodenum involving pancreatic head and superior mesenteric vessels documented by CT.

In literature, the incidence of post-operative complications after DCP is about 40-50%. More frequent are pancreatic fistulas (2-20%), delayed gastric emptying, intra-abdominal abscess, hemorrhage, wound infection and metabolic complications such as diabetes and exocrine pancreatic insufficiency. In contrast, biliary fistulas, duodenal and gastric ulcer, organ failure, pancreatitis and anastomotic ulcer are more rare ¹¹.

In case of segmental resections of the duodenum the most frequent complication is represented by the anastomotic dehiscence ⁸.

In our series there was a high incidence of post-opera-

tive complications of both medical and surgical interest. In fact anastomotic dehiscence was the most common one, causing death in 2 cases out of 4. The only patient in which there were no post-operative complications, had undergone palliative bypass.

In addition to complications, there are other factors affecting survival, but they are not well defined because of many discrepancies in literature. As a result, some authors consider as the main independent prognostic factor lymph node involvement ¹². For this reason, there is no clear agreement on the extension of lymphadenectomy. According to the AJCC the minimum number of lymph nodes needed for a good staging is 6. Probably, patient outcome is particularly unfavorable when more than 20% of examined lymph nodes is involved ¹².

Others think that the main prognostic factors are radical surgery and stage of disease with a median survival of 36% in stages I-III and 6% in stage IV ¹.

According to other authors survival is not affected by any specific factor such as age, gender, location of the primary tumor, loss of weight, clinical stage and histological type ⁷.

The 5-year survival appears to be greater in tumors localized in the distal end. In fact, the tumors of the I and II duodenal portion have a greater tendency to infiltration of the adjacent structures, such as the pancreas and bile ducts, while those of the III and IV portion have a lower aggressiveness, compared to the remaining small intestine tumors ³.

Conclusion

In conclusion, considering our experience and data from literature, we can assert that duodenal adenocarcinoma presents many diagnostic and therapeutic difficulties. This may be due to its rarity, which does not allow the definition of a standard course of treatment, resulting in the inability to have a unique diagnostic and therapeutic approach. That is why there is often a late diagnosis and the need, as in our experience, to perform emergency surgery. Therefore prognosis of this tumor is greatly aggravated by the high incidence of postoperative complications, in part due to urgency in which the surgeries are carried out.

Riassunto

SCOPO: Riportiamo la nostra esperienza nel trattamento delle neoplasie duodenali.

MATERIALI: Abbiamo analizzato la gestione ed il trattamento di quattro pazienti affetti da neoplasia duodenale.

RISULTATI: Tre dei quattro pazienti sono morti nel post-operatorio per deiscenza anastomotica e/o emorragia. Il paziente, sottoposto a bypass palliativo, è deceduto quattro mesi dopo la dimissione per cause a noi sconosciute.

DISCUSSIONE: Le neoplasie duodenali sono rare e sono di difficile e spesso tardivo inquadramento diagnostico. In letteratura non esiste un trattamento standardizzato di questa tipologia di neoplasie; le tecniche chirurgiche più comunemente utilizzate sono la duodeno-cefalo-pancrea-sectomia, la resezione segmentaria del duodeno e gli interventi di tipo palliativo. La scelta chirurgica si basa sulla localizzazione e la stadiazione sia in letteratura che nella nostra esperienza. Indipendentemente dalla scelta chirurgica, i pazienti hanno scarsa prognosi per la presenza di uno stadio avanzato della malattia alla diagnosi.

CONCLUSIONI: Si può affermare che il cancro duodenale presenta diverse difficoltà diagnostiche e terapeutiche. Ciò sarebbe imputabile alla sua rarità, che non consentirebbe la definizione di un percorso standard di trattamento. Spesso la diagnosi è tardiva e spesso c'è la necessità di ricorrere ad un intervento chirurgico in regime di urgenza. Pertanto la prognosi è aggravata dall'alta incidenza di complicanze postoperatorie, in parte dovuta all'esecuzione del trattamento chirurgico in regime di urgenza.

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