Introduction

Although the small intestine constitutes over 75% of the length and 90% of the mucosal surface of the gastrointestinal tract, small intestine cancer is rare and accounts for only 1% of gastrointestinal malignancies (1, 2). Adenocarcinoma together with carcinoid tumors are the most common histological types of primary malignant tumors of the small bowel but others, including lymphoma and leiomyosarcoma, may less frequently be encountered. Adenocarcinomas are predominantly located in the duodenum. Primary adenocarcinoma of the duodenum is a rare malignant tumor, accounting for 0.3-0.5% of all gastrointestinal malignancies. The diagnosis of primary adenocarcinoma of duodenum is often delayed because its symptoms and signs are nonspecific. In this work we want to focus on the diagnostic and therapeutic problems of duodenal adenocarcinoma, reporting a case report.

Case report

A 66-year old man recently came to our Unit (General and Thoracic Surgery Department of the University of Ferrara, Italy), with one and a half year history of weight loss, gastric and biliary vomit associated with abdominal pains without hematemesis and melena. Symptoms usually started after meals and were intermittently associated with dyspepsia. The patient had a history of a hiatal hernia treated with pantoprazole sodium, levosulpiride and sodium alginate-sodium bicarbonate. The alvus was open to gas and feces. A physical examination revealed no abdominal mass or tenderness. Blood tests, including tumour markers such as carcinoembryonic antigen (CEA), a-fetoprotein (AFP), carbohydrate antigen 19-9 (CA 19-9), and carbohydrate antigen 125 (CA 125), were within normal limits.

During one year and a half, because of his symptoms, the patient underwent esophagogastroduodenoscopy (EGDS) which showed “hiatal hernia, gastric hyperemia..."
and profuse secretion of bile in the stomach and duodenum”.

Colonoscopy, computed tomography of the abdomen and Magnetic Resonance Imaging of the abdomen and pelvis were negative. He was also subjected to complete digestive tract X-ray which showed the duodenal bulb regularly opaque.

One year after the onset of symptoms a gastrointestinal computed tomography revealed an irregular thick wall of the 3rd portion of the duodenum and no intraabdominal swollen lymph nodes (Figure 1).

A second endoscopy was performed which confirmed an intraluminal mass in the third and fourth part of the duodenum and the endoscopic biopsy specimen showed an infiltrative adenocarcinomas.

The patient underwent surgery. Intraoperatively, a solid mass in the third and fourth part of the duodenum was identified. Local excision of the tumour was meticulously investigated. Kocher’s manoeuvre, partial resection of the duodenum and jejunum accompanied with lymph node dissection along the superior mesenteric artery was performed.

No lymph node or distant metastasis was identified. Intestinal continuity was then restored by an end-to-end hand sewn duodenojejunal anastomosis.

Histology showed well-differentiated tubular adenocarcinoma with full-thickness invasion and extension beyond the wall infiltrating the perivisceral fat and one lymph node metastasis: pT3 pN1. The specimen’s margins were free of tumour.

Postoperative period was normal without complications. A complete postoperative digestive tract X-ray did not show stenosis or leakage. The patient resumed oral feeding in the 5th postoperative day and was discharged on the 8th postoperative day, excluding adjuvant chemotherapy after oncological advice.

A two years clinical-instrumental follow-up showed no recurrence of the disease.

Discussion

Primitive neoplasia of the duodenum is very rare (6, 7). The III and the IV duodenal portions are the most common sites (8-9) of 45% of tumors, 40% in the II and only 15% in the first. Thus, our case can be included in the first group. Adenocarcinoma of the third or fourth part of the duodenum presents a diagnostic challenge. There is an average delay of 2-15 months from the onset of symptoms to the time of diagnosis of adenocarcinoma of the duodenum (10).

Diagnosis is also often delayed due to the vague and non-specific symptoms and the subsequent difficulties in performing the relevant investigation, while most patients undergo a number of diagnostic tests before surgical exploration (2, 11-14).

The symptoms are not specific (6); 65% of cases are characterized by the association of intermittent abdominal pains with cramps and biliary vomit (15) as in our case.

Examining the entire duodenum using upper gastrointestinal endoscopy is challenging; adenocarcinomas of the 3rd and 4th portions of the duodenum are frequently inaccessible using endoscopy, and most cases require multiple investigations. In fact, the 1st endoscopic

![Fig. 1 - Gastrointestinal computed tomography reveals no intraabdominal swollen lymph nodes.](image)
examination in our patient could not identify the duo-
denal tumor. A definitive diagnosis of adenocarcinoma
was made 1 year and a half after the onset of his symp-
toms.

In the literature we read that the contrast enhanced
CT is useful to define malignancy and ability to excision
(11, 13, 16-18). However, tumours smaller than 2 cm
may not be seen (18). In our case only gastrointestinal
computed tomography was useful for diagnosis.

However, new modalities such as double-balloon
enteroscopy or capsule endoscopy can make diagno-
sis of small bowel or duodenal adenocarcinoma easier
(5, 19).

The difficulty of pathologist’s diagnosis was not the
malignant nature of the lesion but the need to obtain
an high impact clinical data with important surgical im-
lications by a little and superficial specimen obtained
with an endoscopic biopsy. In the first biopsy of our
case (Figs. 2, 3, 4) the pathologist signs out an high gra-
de displastic lesion with atypical glandular architectu-
re and an high pleomorfic nuclei with occasional aty-
ipical mitosis and suggestive aspects of overcoming of
basal membrane (Fig. 4) depicting infiltrative adenocar-
cinoma.

The surgical treatment is not yet well defined and co-
dified. Early stage duodenal carcinoma should be con-
sidered for endoscopic mucosal resection. Advanced sta-
ges of primary duodenal adenocarcinoma (PDA) require
surgical resection for cure.

The correct operation (pancreaticoduodenectomy, lo-
cal excision or segmental resection) has been debated.
Duodeno-cephalo-pancreatectomy (DCP) and seg-
mental resection of the duodenum are employed for trea-
ting PDA (20).

DCP remains the standard treatment for adenocar-
cinomas of the 1st and 2nd portion of the duodenum.
Segmental duodenectomy is the preferred resection
method for patients with adenocarcinoma of the 3rd and
4th portions of the duodenum (14). Our patient had a
T3 stage well-differentiated tubular adenocarcinoma with
one histological lymph node metastasis around the pan-
creas head and no macroscopic invasion of the pancreas
or surgical margins. We decided to treat our patient with
partial resection of the duodenum and jejunum and adju-
vant therapy with cisplatin and capecitabine.

The patient is doing well without any sign of recur-
rence two years later.

Little is known about the use of radiotherapy and che-
motherapy, but most physicians utilise therapeutic stra-
tegies like the management of large bowel cancer (2).

Cunningham observed no significant benefit of
adjuvant chemotherapy on survival (13). The progno-
sis is generally poor and depends on stage at presentation and surgical resectability (2, 11, 17).

Conclusion

Adenocarcinoma of the third and fourth part of the duodenum is very rare. The treatment of choice is radical surgical resection but the optimal surgical procedure, though, remains controversial.

In conclusion, taking into account the rarity of the adenocarcinoma of the duodenum, and that the patients typically present with a long history of variable, vague symptoms, we think that a precocious diagnosis of this cancer and its exact localization are crucial points. A higher degree of suspicion and a more aggressive, persistent investigation should lead to earlier treatment, higher curative resectability rate, and, therefore, better long-term results.

References