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Oncology | Case report

Primary Duodenal Adenocarcinoma: Case report and literature review

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Abstract

Introduction
Primary adenocarcinoma of the duodenum is a rare clinical condition, accounting for only 5% of all gastrointestinal neoplasms and 0.5% of all intestinal neoplasms. Nonspecific clinical presentation contributes to the majority of cases being diagnosed in advanced stages. The prognosis is poor, with a median survival of five years, varying from 23% to 57% in patients undergoing curative surgery (cephalic duodenopancreatectomy).

Methods
Review of medical records and bibliographic research in databases Lilacs, Scielo, Science Direct, ResearchGate; and the Sabiston treatise on surgery.

Results
There are few case reports found in the literature on primary adenocarcinoma of the duodenum. The present study presents the case of a male patient, 69 years old, with primary adenocarcinoma of the stage IV duodenum, submitted to palliative surgery. The patient evolved to death on the 15th postoperative day.

Conclusion
This case report is intended to alert clinicians about the importance and early diagnostic difficulty of primary adenocarcinoma of the duodenum.

Keywords: Primary tumor of the duodenum, Adenocarcinoma, Cancer

Introduction

Despite constituting 80% of the gastrointestinal tract and comprising 90% of the mucosal surface, small bowel neo-formations are rare, corresponding to only 5% of all gastrointestinal neoplasms. Primary adenocarcinoma of the duodenum represents only 0.5% of all intestinal neoplasms and constitutes approximately 50% of malignant lesions of the small intestine [1]. Two North American series published in 2004 and 2010, demonstrated, respectively, that 65% and 57% of cases of adenocarcinoma of the small intestine appeared in the duodenum [2, 3].

The carcinogenesis of primary small intestine adenocarcinoma is not yet fully understood. However, it is believed to be related to mutations of the k-rass gene, involving allelic losses in tumor suppressor genes such as APC (Adenomatous polyposis coli), p53 and SMAD4 (Mothers against decapentaplegic homolog 4). The factors related to its low incidence come from theories related to the liquid content with low bacterial load and the rapid emptying in this intestinal segment, contributing to a lower exposure of the mucosa to carcinogenic agents. In addition to protective factors such as alkaline pH and high level of IgA [1].

Duodenal neoplasia can be classified according to its conformation in relation to the Vater ampoule, and it can be supra, peri or infra-ampullary [4]. Most of these tumors have a periampular location, probably due to the possible interaction of pancreatobiliary secretions with carcinogenic substances in food [5]. He has a predilection for males, with a peak of involvement in the seventh decade of life. The main risk factors include: familial adenomatous polyposis, hereditary non-polypoid colorectal cancer (CCNPH), Peutz-Jeghers syndrome, Crohn's disease, gluten-sensitive enteropathy and altered bile flow [1].

Clinical case

Male, 69 years old, white, with a history of type II diabetes mellitus, arterial hypertension and three ischemic strokes, went to the emergency department with complaints of abdominal pain, hematemesis and arrest feces elimination. He reported having undergone cholecystectomy 6 months ago, with normal histopathology. He denied weight loss. On objective examination, the patient was pale, normocardial, normotensive and diaphoretic. The abdominal palpation revealed a diffusely painful abdomen, distended, with no signs of peritoneal irritation, with a subcostal scar, Kocher type. Laboratory tests showed microcytic and hypochromic anemia with a hemoglobin of 12.6 g / dl and a hematocrit of 38.2%, slightly increased transaminases (AST 156 IU / L and ALT 246 IU / L), alkaline phosphatase of 200 IU / L and GGT of 354 IU / L. In addition to E.A.S. with 12 to 15 leukocytes per field and 3 to 6 red blood cells per field. Abdominal-pelvic Computed Tomography (CT) without contrast was requested.

CT revealed a mass in the hepatic hilum to the pancreatoduodenal groove, causing extrinsic obstruction of choledoccus and gastrectasis. No other injuries were found in Organs abdominal-pelvic organs.

He was admitted to the General Surgery service for hemodynamic stabilization and etiological study. In this sense, in the face of the hypothesis of a duodenal neoformative process, the conduct was the passage of a nasogastric catheter in siphoning, zero diet and request for contrast imaging and endoscopic examination.

Abdominal-pelvic CT with oral contrast showed tumor lesion in the first and second duodenal portions, with invasion of adjacent organs: hepatic hilum, pancreas and stomach. Gastric lumen was narrowed, with no progression from oral contrast to the duodenum. The examination was interrupted due to the patient's intolerance to oral contrast, causing vomiting.

Upper Digestive Endoscopy (EDA) revealed an enlarged stomach due to gastroparesis. The gastric lake had dark liquid with residual bleeding in coffee grounds. In addition, the organ was deviated in the antral region, making it difficult to examine the region, but with a transposable pylorus, where a huge clot was observed in the removed duodenal bulb, with an infiltrating lesion occupying the posterior wall. It was not possible to progress to the second duodenal portion. Likewise, the biopsy and the passage of an enteral tube in the second portion through stenosis was not performed.

To complete the staging of the lesion, a chest CT scan was performed, which showed no changes and CEA, which was positive. In a later laboratory evaluation, he revealed hemoglobin of 9.4 g / dl and hematocrit of 30.1%, total proteins of 6.4 g / dl with an albumin / globulin ratio of 1.0, INR 2.81 and a total bilirubin of 4.8 mg / dl, of which 2.7 mg / dl of direct bilirubin and 2.1 mg / dl of indirect bilirubin. Exclusive parenteral nutrition was initiated through deep venous access in the right jugular vein and vitamin K replacement on alternate days.

The patient underwent exploratory laparotomy, gastroenteroanastomosis posterior to 40 cm from the Treitz angle and a biopsy that showed adenocarcinoma of the duodenal mucosa. Subsequently, it was presented as a complication in the postoperative period of efferent loop syndrome. The patient evolved to death on the 15th postoperative day.

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Discussion

Clinical Presentation

The non-specificity of the clinical manifestations of primary duodenal adenocarcinoma predisposes to diagnostic difficulties [1]. Dyspeptic symptoms, anorexia, malaise, nonspecific abdominal pain, postprandial nausea and vomiting, anemia and jaundice (present in periampular tumors), can constitute the presentation of tumors in the duodenum [6].

Diagnostic Methods

Early diagnosis is uncommon, only 20% to 50% of symptomatic patients have a correct preoperative diagnosis. There are no consensual recommendations regarding the sequence of tests to be requested in the face of clinical suspicion of primary duodenal adenocarcinoma. It can be diagnosed by radiological methods, particularly contrasted examinations of the digestive tract, Computed Tomography, CT Enterography or Magnetic Resonance Imaging that provide an accurate diagnosis in 50% to 70% of patients and enteroclysis with a diagnostic precision of approximately 90% [1, 7]; and by endoscopic methods, specifically EDA for proximal lesions and double-balloon enteroscopy and video capsuleendoscopy for distal lesions. Simple abdominal radiography and ultrasound were not effective for preoperative diagnosis. Despite the wide variety of diagnostic resources, proof of the existence of a small bowel tumor is often obtained during exploratory laparotomy, whether elective or emergency [7].

Staging

The staging system used for duodenal adenocarcinomas is the TNM system. Where T describes the extent of tumor spread through the layers that form the wall of the small intestine (mucosa, submucosa, own muscle and serosa); N describes the existence of tumor spread to regional lymph nodes and M indicates metastatic spread to distant organs. The TNM system was developed in 2010 by AJCC (American Joint Committee on Cancer) and UICC (International Union Against Cancer) (Table 1) [8].

Considering that in the case presented there was evidence of metastasis, we classified the tumor as stage IV (Table 1). In the United States, 32% of patients with adenocarcinomas of the small intestine are already diagnosed in stage IV, 26% in stage III, 27% in stage II and only 12% and 3% in stages I and 0, respectively [9].

Treatment

The set of aspects mentioned, namely clinical manifestations, tumor dissemination and the pattern of distant ganglion and organ metastasis, allows the treatment of primary duodenal adenocarcinoma to be established. In tumor lesions of the first and second portion of the duodenum, cephalic duodenopancreatectomy (Whipple Surgery) is the surgery of choice. In third and fourth portion lesions, segmental resection can be performed [1]. Whipple surgery has, in general, a higher morbidity and mortality than segmental resection, despite this, it is the surgical treatment that satisfies the principles of curative oncological surgery, since it is able to perform an en bloc resection of the lesion with lymphadenectomy. Regional lymphadenectomy should be performed regardless of the preoperative stage of the tumor, as it has demonstrated a direct impact on patient survival in five years [6, 10].

In case of tumor unresectability or poor general condition of the patient, palliative surgery is indicated, with a deviation of the duodenal transit through bilio-digestive surgery or gastroenteroanastomosis. As described, the latter being the procedure performed in the case under analysis [11-13].

There is no standard protocol for adjuvant therapy in patients with adenocarcinoma of the small intestine. They are usually treated similarly to periampular tumors [1].

Prognosis

The median survival of patients with primary duodenal adenocarcinoma ranges from 23% to 57%, over five years. Patients undergoing palliative surgery have a survival of months [14]. Several theories are related to the poor prognosis of this neoplasm, among which are, imprecision of symptoms, absence of physical findings, lack of clinical suspicion and rarity of these lesions [1]. Increases in the life expectancy of these patients are believed to be related to performing curative surgery (cephalic duodenopancreatectomy), presence of negative resection margins, as well as tumor location in the first or second duodenal portions.

Conclusion

Primary adenocarcinoma of the duodenum is a rare neoplasm, with a high rate of morbidity and mortality, mainly due to the imprecision of its symptoms, thus culminating in delayed diagnosis. Contrast radiological examinations and endoscopic examinations are useful in its preoperative diagnosis. Regarding treatment, cephalic duodenopancreatectomy is indicated in proximal lesions and segmental resection in distal lesions. Regarding the average survival of these patients, it is directly proportional to the state of nodal involvement, degree of injury and surgical resection. And finally, the presentation of this case aims to alert clinicians to the importance and difficulty of early diagnosis of primary duodenal adenocarcinoma.

Conflict of Interest

The authors declare that there is no conflict of interest.

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Abbreviations

AST: Aspartate aminotransferase
ALT: Alanine aminotransferase
GGT: Glutamyl transferase range
EAS: Summary examination of urine or type 1 urine
INR: Reason for international standardization
CEA: Carcinoembryonic antigen

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Table 1: TNM system developed in 2010 by American Joint Committee on Cancer and International Union Against Cancer (8).

Primary tumor (T)			
TX	The primary tumor has not been identified		
T0	No evidence of primary tumor		
Tis	Carcinoma in situ		
T1a	The tumor invades the lamina propria		
T1b	The tumor invades the submucosa		
T2	The tumor invades the muscularis itself		
T3	The tumor goes beyond the muscularis propria to the subserosa or the perimuscular tissue in the non-peritoneal areas, with an extension less than or equal to 2 cm		
T4	The tumor extends from the visceral peritoneum to the subserosa or directly invades other organs or structures (pancreas, bile duct, retoperitoneum, mesentery or other areas of small intestine)		
Regional lymph nodes (N)			
NX	Regional glands not evaluated		
N0	No ganglion metastases		
N1	Metastases in 1-3 regional ganglia		
N2	Metastases in 4 or more regional ganglia		
Distant metastasis (M)			
M0	No distant metastases		
M1	With distant metastases		
Anatomical staging / prognosis groups			
Stage 0	Tis	N0	M0
Stage IA	T1	N0	M0
Stage IB	T2	N0	M0
Stage IIA	T3	N0	M0
Stage IIB	T4	N0	M0
Stage IIIA	Any T	N1	M0
Stage IIIB	Any T	N2	M0
Stage IV	Any T	Any N	M1

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