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Case Report

A rare case of duodenal adenocarcinoma [☆]

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ABSTRACT

Duodenal adenocarcinoma is very rare. Its clinical picture is nonspecific and the diagnosis is often accidental. The factors that affect survival are difficult to determine because the number of patients is not high. The common site of duodenal tumors and surgical removal are also debatable. The treatment guidelines published so far have mostly been evaluated in retrospective studies conducted over a 20-year period with relatively small sample sizes. The author presents a case of duodenal adenocarcinoma in a 62-year-old male patient with a clinical manifestation of melena. Duodeno-cephalo-pancreatectomy was the surgical option.

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Introduction

Small intestine cancer is a relatively rare disease, accounting for only 2% of all gastrointestinal cancers in the United States [1]. The primary location of small intestine cancer is the ileum, but the cancer is also found in the duodenum and the jejunum. Although small intestine cancer commonly arises in the duodenum, duodenal cancer makes up < 0.5% of gastrointestinal cancer cases [2]. Adenocarcinoma is the

most common type of cancer in the duodenum, whereas jejunal malignancies usually originate from neuroendocrine cells.

Clinical symptoms of small intestine cancer are atypical. Disease is usually diagnosed in the late stage. The 1-, 2-, and 5-year survival rate after surgery is 100%, 73.3%, and 31.6% respectively [3–5]. Familial adenomatous polyposis, Gardner's syndrome, Lynch syndrome, Muir–Torre syndrome, celiac disease, Peutz–Jeghers syndrome, juvenile polyposis syndrome, and Crohn's disease are risk factors for developing duodenal

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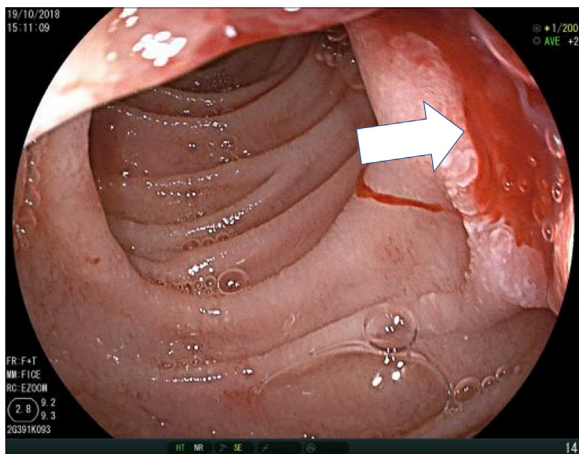


Fig. 1 – Upper gastrointestinal endoscopy showed an ulceration (arrow) 3 cm in diameter, with some small bleeding spots 5 cm from the papilla.

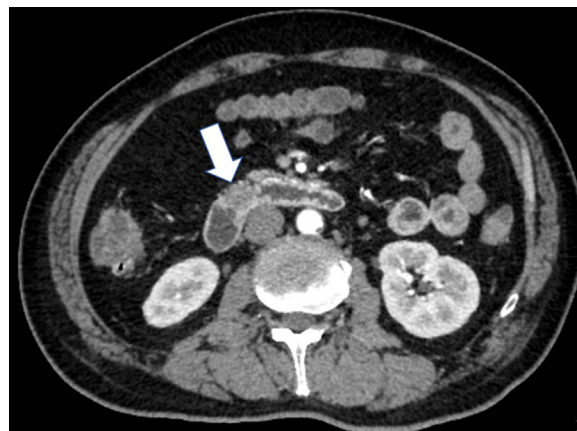


Fig. 2 – Abdominal computed tomography scan of the duodenal bulb. A low-density nodule, 10 x 4 mm in size and fatty in appearance, is seen in the submucosal layer. No abnormal lymph nodes are seen around the stomach.

cancer [6]. In this report, we intended to illustrate a rare case of duodenal adenocarcinoma.

Case description

The patient was a 62-year-old male with a normal medical history, with no alcohol abuse, no smoking, and no family factors related to gastrointestinal cancer. The patient presented with a 4-month history of self-limited melena of unknown causes. About 4 days before admission, the patient defecated a black tarry stool but denied vomiting up blood. On physical examination, the patient had anemia, and a digital rectal examination found melena.

The patient underwent an upper gastrointestinal endoscopy, which showed an ulceration 3 cm in diameter, with some small bleeding spots 5 cm from the papilla (Fig. 1). The patient's hemoglobin level was 7.5 g/dL (normal range: 13–17 g/dL) and red blood cell count was 2.49 G/L (normal range: 4–10 G/L). Serum carbohydrate antigen (CA) 19.9, CA 125, and carcinoembryonic antigen were within normal limits. Upper abdominal computed tomography examination revealed a hypodense nodule, 15 x 4 mm in size and fatty in appearance, in the submucosal layer of the duodenum; no abnormal lymph nodes were seen around the stomach (Fig. 2). A biopsy was then performed. Histopathological examinations of the material collected from the duodenum enabled a diagnosis of adenocarcinoma (Fig. 3).

The patient's preoperative diagnosis was duodenal adenocarcinoma. The operation was performed using the Whipple method. A tumor, 2 x 2 cm in size, was detected in the duodenal bulb. Other parts of the intestine were normal. An intraoperative frozen biopsy of 16 lymph node stations revealed no malignant cells. Downstream cholecystectomy was performed, during which the common bile duct was removed. The antrum of the stomach and the duodenum (to the jejunum) were removed. The neck of the pancreas anterior to the superior mesenteric vein and the uncinate process near

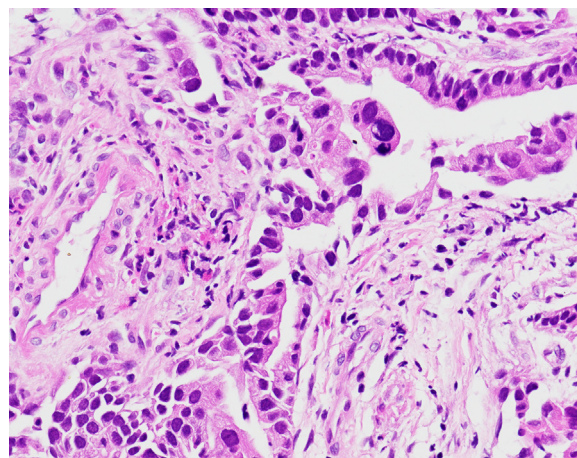


Fig. 3 – Biopsy results for the tumor in the duodenum. Tumor cells have large, atypical nuclei and form irregular glands. The stroma is infiltrated by chronic inflammatory cells. All findings are consistent with adenocarcinoma.

the superior mesenteric artery were removed. The lymph nodes surrounding the hepatic hilum and the pancreatic head were dissected. *Child's reconstruction* was performed, followed by layered closure (individual closure of the anatomic layers).

After 6 weeks, chemotherapy was administered in accordance with accepted standards, resulting in stabilization and gradual improvement of the general condition of the patient. However, because of the COVID-19 pandemic, we lost contact with the patient after this treatment.

Discussion

Duodenal cancer is very rare, making up <0.5% of gastrointestinal cancer cases [2]. Jejunal malignancies usually originate from neuroendocrine cells, whereas adenocarcinoma is

the most common type of cancer in the duodenum. According to data from the National Cancer Database (NCDB), which includes 67,843 cases of small intestine cancer from 1985 to 2005, 64% of patients with duodenal cancer had adenocarcinoma, 21% had neuroendocrine tumors, 10% had lymphoma, and 4% had sarcoma [7].

Because duodenal adenocarcinoma is an uncommon disease, the current treatment guidelines are relatively limited. In general, surgery is the treatment of choice and involves 2 surgical methods: duodeno-cephalo-pancreatectomy (DCP) and segmental resection of the duodenum (SRD). Previous studies have shown that patients with duodenal adenocarcinoma have a better prognosis than those with other tumors around the ampulla of Vater (such as pancreatic cancer and cholangiocarcinoma) [8].

Risk factors for duodenal adenocarcinoma have not been clearly defined. Diets rich in red meat and sugar and low in vegetables and fruits are risk factors for small intestine cancer as well as colon cancer [9]. In addition, the abuse of alcohol, caffeine, or tobacco is also a risk factor [10]. However, the impact of these risk factors on the development of duodenal adenocarcinoma is unclear. In addition, most cases of duodenal adenocarcinoma are not related to any risk factors. Our patient had neither a history of alcohol abuse nor a history of smoking.

Most duodenal adenocarcinomas arise in the second part of the duodenum (D2) and some in the third and fourth part (D3/D4). Tumors are rarely seen in the first part of the duodenum (D1), especially the ampulla of Vater. In the reported case, the tumor was located in the second part of the duodenum (D2; 4-5 cm from the papilla) [11].

Clinical manifestations of duodenal adenocarcinoma are nonspecific. Common symptoms are continuous dull abdominal pain, nausea, and gastrointestinal bleeding. Sixty-five percent of cases of duodenal adenocarcinoma present with abdominal pain and nausea. The reason for admission of our patient was recurrent gastrointestinal hemorrhage.

Esophagogastroduodenoscopy combined with biopsy is the gold standard for diagnosing duodenal adenocarcinoma. However, surgical methods have not been specifically advocated nor systematized. DCP with extended lymphadenectomy (of hepatic hilar lymph nodes and paraaortic lymph nodes) appears to be the preferred treatment option. However, SRD has recently been supported [11]. SRD is recommended for tumors located in the distal part of the duodenum, whereas DCP is prioritized for tumors located in the proximal segment, as in our case [12]. Although indications for DCP and SRD differ to some extent, the 2 methods have similar rates of complications (27% after DCP and 18% after SRD). Common complications are pancreatic fistula (9%-25%), abdominal abscess (16%-20%), peritonitis (8%-9%), and abdominal hemorrhage (3%-13%) [13].

Only a few studies have compared the 2 surgical methods. In 1 study, the 5-year survival rate of patients after DCP was 27.8%, higher than that of patients undergoing SRD (16.9%) [3]. The 2 methods had a similar survival rate in other studies. However, the surgical method was not a factor that improved survival prognosis [14]. The roles of chemotherapy and radiation therapy in the treatment of duodenal adenocarcinoma are not clear [15].

Compared to other cancers located around the ampulla of Vater (such as pancreatic cancer and cholangiocarcinoma), duodenal adenocarcinoma is associated with a significantly better prognosis. A cohort study of 150 patients from 6 hepatopancreato centers in the United Kingdom showed a 1-year, 3-year, and 5-year survival rate of 83.9%, 66.7%, and 51.2%, respectively. Median disease-free survival was 53 months [16]. Onkendi et al. [17] study of 124 patients with duodenal adenocarcinoma from 1994 to 2009 showed a 5-year survival rate of 37%. Factors associated with the better prognosis of patients with duodenal adenocarcinoma include age, distant metastasis, regional lymph node metastasis, tumor size, and general status. Lymph node metastasis is the most important prognostic factor [13].

Conclusion

Duodenal adenocarcinoma is a rare disease. Clinical symptoms are nonspecific, and patients are often diagnosed inadvertently. Compared to cancers around the ampulla of Vater, duodenal adenocarcinoma is associated with a better prognosis. Surgery is the treatment of choice and involves 2 surgical methods: DCP and SRD. DCP is prioritized for tumors located in the proximal segment of the duodenum, while SRD is recommended for tumors located in the distal part. The roles of chemotherapy and radiation therapy in treating duodenal adenocarcinoma are unclear. Lymph node metastasis is the most important prognostic factor.

Author's contributions

Ho XT and Nguyen MD: Case file retrieval and case summary preparation. Ho XT and Nguyen MD: preparation of manuscript and editing. All authors read and approved the final manuscript.

Availability of data and materials

Data and materials used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Ethics approval and consent to participate

Our institution does not require ethical approval for reporting individual cases or case series. Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

Consent for publication

Not applicable.

Patient consent

Informed consent for patient information to be published in this article was obtained.

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