

## Case Report

Open Access, Volume 3

# A Rare Adenocarcinoma of Ectopic Pancreatic from Duodenum: A Case Report

Zhiwei Chen; Kangshou Liu; Mingrong Cao\*

Department of General Surgery, The First Affiliated Hospital, Jinan University, Guangzhou, China.

## Abstract

Ectopic pancreas is a rare disease in which pancreatic tissue appears outside the pancreas. Malignant transformation of ectopic pancreatic tissue is uncommon. We present a case of ectopic pancreatic malignant transformation from the first part of the duodenum. A 68-year-old male was referred to our hospital with recurrent upper abdominal pain for 3 months. A massive mass in the right upper abdomen was revealed by computed tomography, which was closely related to the duodenum. The possibility of a stromal tumor was discussed. The pancreatic adenocarcinoma was surgically resected, and postoperative pathology revealed that it was moderately differentiated. Imaging examination and tumor-specific antigen testing revealed no obvious signs of tumor recurrence up to 6 months after surgery. Ectopic pancreatic malignant transformation is extremely rare, primarily occurring in the gastrointestinal tract's submucosa, and diagnosis is difficult. CT, MR, endoscopic ultrasonography, and tumor-specific antigen should be added in diagnosis, and once detected, surgery should be performed as soon as possible.

**Keywords:** Duodenum; Ectopic pancreas; Ductal adenocarcinoma.

**Abbreviations:** CT: Computed Tomography; MRI: Magnetic Resonance Imaging; CA 19-9: Carbohydrate Antigen 19-9; CA 125: Carbohydrate Antigen 125; EUS-FNA: Endoscopic Ultrasonography-Guided Fine-Needle Aspiration.

## Background

Ectopic pancreas, also known as abnormal pancreas, was first observed in 1727 in an ileal diverticulum [1]. Ectopic pancreas is a condition in which pancreatic tissue appears outside of the original pancreas and has no vascular or nerve connection to pancreas [2]. Ectopic pancreas is most commonly found in the gastrointestinal tract, but it can also be found in the biliary tract, liver, lung, brain, and other areas [3-7]. Malignant transformation of an ectopic pancreas is uncommon. According to a study, from 2000 to 2020, only 12 cases of ectopic pancreatic malignancy of

duodenum were reported in Pubmed [8]. We present a case of ectopic pancreatic cancer that originated in the first portion of the duodenum.

## Case introduction

A 68-year-old male presented to our hospital with epigastric pain and weight loss for three months. PET-CT in the other hospital indicated a large abdominal mass involving the gastric antrum and duodenum, which was considered to be gastrointestinal stromal tumor with right superior phrenic, portal vein and retro-

**Manuscript Information:** Received: Mar 15, 2023; Accepted: Apr 05, 2023; Published: Apr 14, 2023

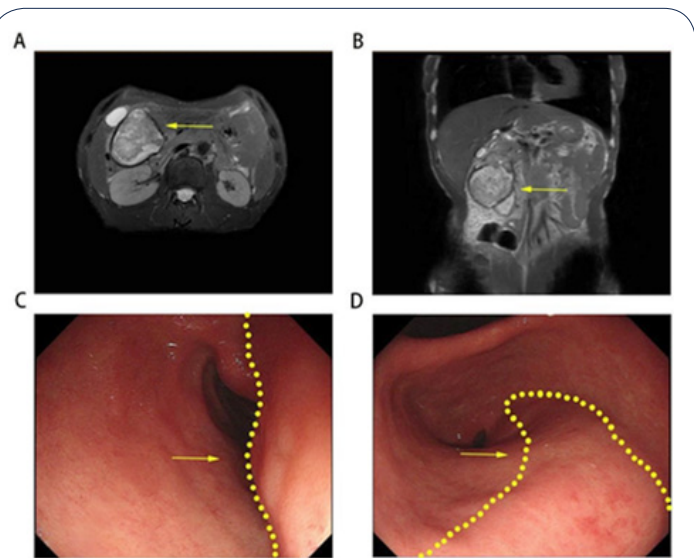
**Correspondance:** Mingrong Cao, Department of General Surgery, The First Affiliated Hospital, Jinan University, Guangzhou, China.

Tel: +8613247377319 & +8613392692226; Email: [tcaomr@jnu.edu.cn](mailto:tcaomr@jnu.edu.cn)

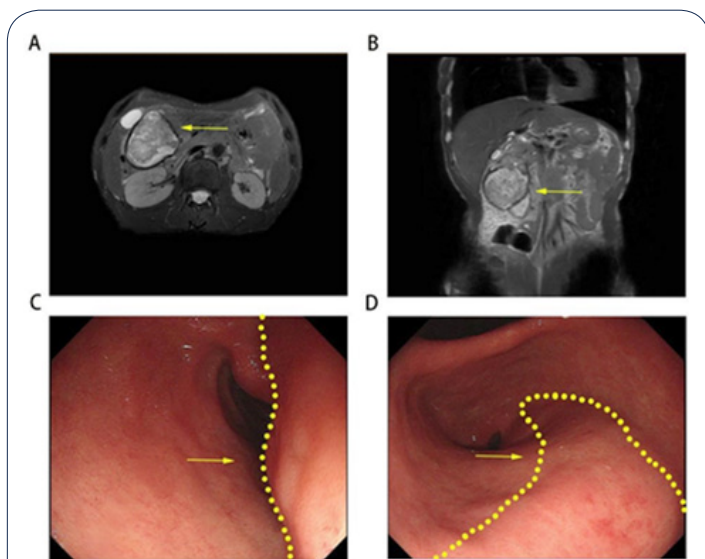
**Citation:** Chen Z, Liu K, Cao M. A Rare Adenocarcinoma of Ectopic Pancreatic from Duodenum: A Case Report. *J Surgery*. 2023; 3(1): 1088.

**Copyright:** © Cao M 2023. Content published in the journal follows creative common attribution license.

peritoneal lymph node metastasis. Following admission, relevant examinations were performed, and the patient's serum CA19-9 were 153.2 IU/L and CA125 were 101.3 IU/L. Combined with upper abdominal MRI (Figure 1 A,B) and gastroscopy (Figure 1 C,D), the possibility of gastrointestinal stromal tumor was still considered to be high. With the consent of the family, the operation was performed. During the operation, it was discovered that the tumor originated from the first portion of the duodenum and was clearly not associated with the pancreas. Then, a distal gastrectomy was performed along with regional lymph node dissection. Pathological findings revealed that it was a moderately differentiated ductal adenocarcinoma infiltrating the muscular layer of the duodenum, with vascular invasion and lymph node metastasis of the duodenal ligament, but no nerve invasion (Figure 2). Following surgery, the patient underwent two courses of gemcitabine chemotherapy and then refused further treatment. Six months after surgery, abdominal CT (Figure 3) revealed no signs of tumor recurrence, and serum CA19-9 and CA125 levels remained normal.



**Figure 2:** Microscopically, the tumor was diagnosed as moderately differentiated adenocarcinoma extending from the submucosa to the muscoli propria of the duodenum. Normal pancreatic tissue was observed near the tumor, suggesting the presence of an ectopic pancreas.

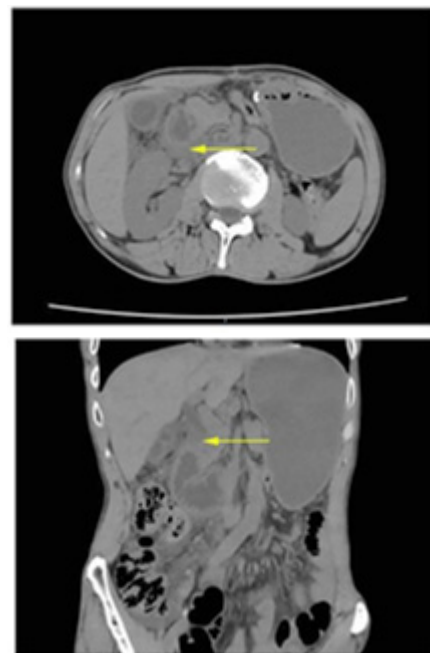


**Figure 1A,B:** Preoperative abdominal magnetic resonance imaging showed a huge mass in the right upper abdomen, which was closely related to the duodenum and gastric antrum. **C,D** Preoperative gastroscopy showed protruding lesions in the first part of duodenum and gastric antrum, which was considered as lateral wall compression.

### Discussion

Patients with ectopic pancreas usually have no specific symptoms, though a few may experience non-specific symptoms such as abdominal pain, dyspepsia, or digestive tract obstruction due to inflammation and tumor [9]. Ectopic pancreas is typically found in the submucosa, making imaging and general endoscopy difficult to diagnose. Although endoscopic ultrasound can help in diagnosis, a definitive diagnosis still requires a fine-needle aspiration biopsy or post-operative pathological biopsy [6].

The risk of malignant transformation of ectopic pancreas is very low, estimated to be between 0.7% and 1.8% in the literature [10,11]. About one-third of all cases of ectopic pancreatic cancer occur in the stomach, followed by the duodenum and jejunum, with cases also occurring in the esophagus, spleen, rectum, mesentery, liver, Meckel diverticulum, and brain [6]. Because of the



**Figure 3:** No tumor recurrence or metastasis was found in the whole abdominal CT reexamination 6 months after surgery (the arrow indicates the original tumor location).

disease's different location, ectopic pancreatic cancer can cause a variety of nonspecific clinical symptoms, the most common of which are gastrointestinal and systemic manifestations, such as abdominal pain, nausea, dyspepsia, and weight loss, and a few patients may experience abnormal defecation, gastrointestinal bleeding, hypoglycemia, and so on [9]. About half of the patients had elevated tumor markers, with the most common being an increase in serum CA 19-9, but the positive rate of tumor markers were lower than pancreatic cancer [12]. In our case, the patient experienced abdominal pain, weight loss, and gastrointestinal

bleeding, as well as an increase in the serum CA 19-9 and CA 125. However, these differences are insufficient to differentiate it from gastrointestinal stromal tumor, gastrointestinal neuroendocrine tumor, gastric carcinoid, gastric lymphoma, and gastric cancer. Imaging, such as abdominal CT and MRI, can only pinpoint the location of the tumor and cannot provide a definitive diagnosis. Endoscopy has the function of differential diagnosis, but tissue biopsy, even if performed, is not diagnostic because the sampling location is usually superficial. Endoscopic ultrasonography is considered to be an effective method for the diagnosis of ectopic pancreas. Submucosal lesions can be cytologically evaluated when combined with fine needle aspiration biopsy, making the diagnosis of ectopic pancreatic malignant transformation more reliable [5]. The EUS-FNA was not performed in our case because the patient's abdominal pain was severe and required curative or palliative surgery. Pathological diagnosis of surgical resection is typically used as the gold standard for ectopic pancreatic cancer diagnosis. Some researchers proposed diagnostic criteria for ectopic pancreatic cancer as early as 1974 [9]: (1) The tumor must be in or near the ectopic pancreas; (2) the transition zone between pancreatic structure and cancer shall be observed, while metastatic tumor or adjacent malignant tumor infiltration ought to be excluded; and (3) non-neoplastic ectopic pancreatic tissue was supposed to include well-developed acini and ductal structures.

Because of its rarity, treatment for ectopic pancreatic cancer is limited. Surgical resection is usually the first option, and different surgical methods, such as pancreaticoduodenectomy, distal gastrectomy, pylorus-preserving pancreaticoduodenectomy, and subtotal stomach-preserving pancreaticoduodenectomy, can be chosen depending on the specific location of the tumor [13-15]. If the intraoperative frozen pathological examination reveals that the tumor is malignant, regional lymph node dissection should be performed routinely. There is currently no evidence that chemotherapy is effective in the treatment of ectopic pancreatic cancer [6,15]. According to studies, the overall survival time of ectopic pancreatic cancer after surgery is longer than that of pancreatic cancer, and approximately 39% of patients have a survival time of more than one year after operation, with a low probability of recurrence after surgical resection, which may be related to the fact that ectopic pancreatic cancer manifests clinical symptoms earlier than pancreatic cancer [16].

Ectopic pancreas is becoming increasingly common as endoscopic detection and ultrasonic endoscopic technology become more widely available. Although the likelihood of malignant transformation of an ectopic pancreas is low, the possibility of malignant transformation should not be overlooked. Specialists should have an understanding of the common sites, clinical characteristics, histological and pathological features, as well as diagnosis and treatment methods for ectopic pancreatic cancer.

## References

1. Elfving G, Hästbacka J. Pancreatic heterotopia and its clinical importance. *Acta chirurgica Scandinavica*. 1965; 130: 593-602.
2. Liu X, Wu X, Tuo B, Wu H. Ectopic pancreas appearing as a giant gastric cyst mimicking gastric lymphangioma: a case report and a brief review. *BMC gastroenterology*. 2021; 21: 151.
3. Kim JY, Lee JM, Kim KW, Park HS, Choi JY, et al. Ectopic pancreas: CT findings with emphasis on differentiation from small gastrointestinal stromal tumor and leiomyoma. *Radiology*. 2009; 252: 92-100.
4. Filip R, Walczak E, Huk J, Radzki Rp, Bieńko M, et al. Heterotopic pancreatic tissue in the gastric cardia: a case report and literature review. *World J Gastroenterol*. 2014; 20: 16779-16781.
5. Christodoulidis G, Zacharoulis D, Barbanis S, Katsogridakis E, Hatzitheofilou K, et al. Heterotopic pancreas in the stomach: a case report and literature review. *World J Gastroenterol*. 2007; 13: 6098-6100.
6. Cazacu IM, Iuzuriaga Chavez AA, Noguera Gonzalez GM, Saftoiu A, Bhutani MS, et al. Malignant Transformation of Ectopic Pancreas. *Digestive diseases and sciences*. 2019; 64: 655-668.
7. Alqahtani A, Aljohani E, Almadi F, Billa S, Alqahtani M, et al. Heterotopic pancreatic tissue in the gastric antrum an incidental finding during bariatric surgery: A case report and literature review. *International journal of surgery case reports*. 2020; 67: 39-41.
8. Minami T, Terada T, Mitsui T, Nakanuma Y. Adenocarcinoma arising from a heterotopic pancreas in the first portion of the duodenum: a case report. *Surgical case reports*. 2020; 6: 141.
9. Ourô S, Taré F, MONIZ L. Pancreatic ectopia. *Acta medica portuguesa*. 2011; 24: 361-366.
10. Guillou L, Nordback P, Gerber C, Schneider RP. Ductal adenocarcinoma arising in a heterotopic pancreas situated in a hiatal hernia. *Archives of pathology & laboratory medicine*. 1994; 118: 568-571.
11. Hickman DM, Frey CF, Carson JW. Adenocarcinoma arising in gastric heterotopic pancreas. *The Western journal of medicine*. 1981; 135: 57-62.
12. Makhlouf HR, Almeida JL, Sobin LH. Carcinoma in jejunal pancreatic heterotopia. *Archives of pathology & laboratory medicine*. 1999; 123: 707-11.
13. Kaneko T, Ohara M, Okamura K, Fujiwara-Kuroda A, Miyasaka D, et al. Adenocarcinoma arising from an ectopic pancreas in the duodenum: a case report. *Surgical case reports*. 2019; 5: 126.
14. Kinoshita H, Yamaguchi S, Shimizu A, Sakata Y, Arii K, et al. Adenocarcinoma arising from heterotopic pancreas in the duodenum. *International surgery*. 2012; 97: 351-355.
15. Fukino N, Oida T, Mimatsu K, Kuboi Y, Xida K, et al. Adenocarcinoma arising from heterotopic pancreas at the third portion of the duodenum. *World J Gastroenterol*. 2015; 21: 4082-4088.
16. Eisenberger Cf, Gocht A, Knoefel Wt, Busch CB, Peiper M, et al. Heterotopic pancreas--clinical presentation and pathology with review of the literature. *Hepato-gastroenterology*. 2004; 51: 854-858.