Case Reports in Gastroenterology

Case Rep Gastroenterol 2022;16:546–551
DOI: 10.1159/000526135
Received: May 3, 2022
Accepted: July 15, 2022
Published online: September 16, 2022

Single Case

Heterotopic Pancreas in the Duodenum Diagnosed after Laparoscopic Biopsy

Ngoc Trinh Thi Pham[a] Anh Nguyet Thi Nguyen[a] Minh Tri Thi Vo[a]
Minh Duc Pham[b, c]

[a]School of Medicine and Pharmacy, The University of Danang, Da Nang, Vietnam; [b]Department of Surgery, Hue University of Medicine and Pharmacy, Hue University, Hue, Vietnam; [c]Department of Abdominal Emergency and Pediatric Surgery, Hue Central Hospital, Hue, Vietnam

Keywords
Ectopic pancreas · Heterotopic pancreas · Duodenal ectopic pancreas

Abstract
Heterotopic pancreas is defined as the presence of ectopic pancreatic tissue outside boundaries of the pancreas without vascular and duct system connection with the pancreas. Ectopic locations are mostly found anywhere in the gastrointestinal tract such as the stomach (24–38%), the duodenum (9–36%), and the jejunum (0.5–27%). Clinical manifestations are not specific, vague, and misdiagnosed another digestive disease. Most cases are incidentally detected by histological examination of specimens resected for different pathologies during endoscopy, surgery, or even autopsy. We report a case of a 31-year-old man who admitted to the hospital with the reason of epigastric pain for 3 days. Clinical examination showed mild epigastric tenderness. The past medical history of patient was unremarkable. A submucosal lesion was observed in the first part of the duodenum during endoscopy. Computed tomography and endoscopic ultrasonography findings were suspected to be heterotopic pancreatic tissue. After laparoscopic surgery for biopsy, it was histologically confirmed duodenal ectopic pancreas. It is difficult to differentiate gastrointestinal pancreatic heterotopia from gastrointestinal stromal tumors, leiomyoma, or lymphomas by using endoscopy because ectopic tissue is mostly located in the submucosal layer. In addition, rare cases of ectopic pancreatic tissue transform malignancy. Surgical treatment should be considered to take adequate tissue samples for biopsy or resect the lesions in symptomatic patients. Duodenal pancreatic heterotopia is an uncommon congenital malformation and most patients are asymptomatic. Histological examination is essential to exclude malignant lesions and to have an appropriate treatment.

Correspondence to:
Minh Duc Pham, pmduc@huemed-univ.edu.vn

Case Rep Gastroenterol 2022;16:546–551
DOI: 10.1159/000526135

Introduction

Heterotopic pancreas (HP), variably referred to as ectopic pancreas, is introduced as the pancreatic tissue in an abnormal anatomical position without vascular and ductal continuity with the main pancreas [1]. HP was first reported by Jean Schultz in 1729 and has been described since in the English literature, primarily as case reports or in small case series studies [2]. Heterotopic pancreatic tissue can be found in many sites, including gastrointestinal tract, mesentery, hepatobiliary system, spleen, mediastinum, lungs, fallopian tubes, umbilicus, and omentum [1]. The most common site is the upper gastrointestinal tract, such as the stomach (24–38%), duodenum (9–36%), and jejunum (0.5–27%) [1, 3]. The esophagus, ileum, and colon are the other less common sites [4]. The most common theory on the origin of HP involves the displacement of pancreatic tissue during development. The misplacement theory suggests that some components of the primitive pancreas are separated and form mature pancreatic tissue along the digestive tract length during the foregut rotation [1].

Accurate preoperative diagnosis of HP is difficult because of its location and nonspecific clinical symptoms [5]. In most cases, HP is incidentally detected in patients during endoscopy, surgery, or autopsy. The detection rate was 0.5% during laparotomies and at autopsy ranging from 0.5 to 13.7% [6]. The most common presentation of HP in the upper gastrointestinal tract is a small and submucosal lesion [5]. Previous studies evaluated that imaging findings were difficult to differentiate between gastrointestinal submucosal lesions [1, 7, 8]. Therefore, evidence of pathological finding is required to diagnose HP. We report a case in which a submucosal lesion of the duodenum was detected. Because there was no biopsy by endoscopic ultrasound, we performed a laparoscopic biopsy and diagnosed HP in the duodenum.

Case Presentation

A 31-year-old male patient was admitted to the hospital because of epigastric pain for 3 days. Upper gastrointestinal endoscopy found a submucosal lesion at the first part (D1) of duodenum (Fig. 1a). The lesion measured approximately 10 × 15 mm and appeared as protuberance in the lumen, oval shape, smooth inner surface. There was no history of nausea, weight loss, hematemesis, or melena. Body mass index was 24.2 kg/m^2. Vital signs were revealed

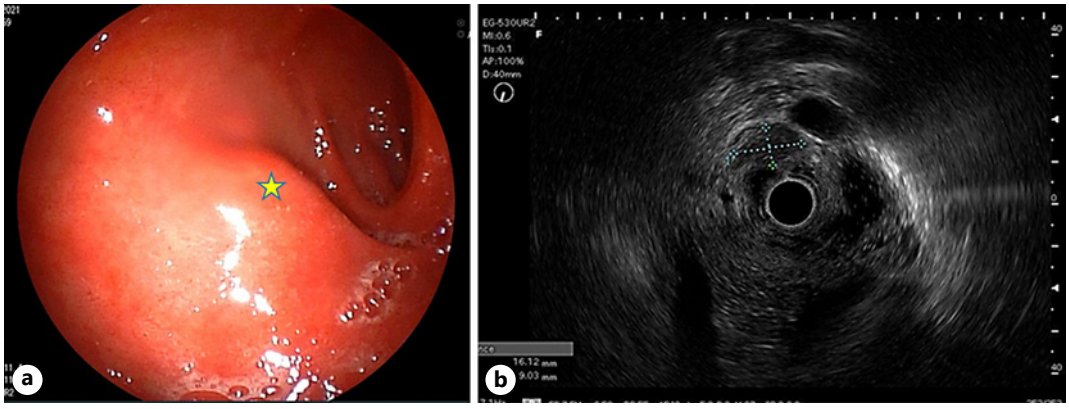

Fig. 1. a Endoscopic image revealing a submucosal lesion (star) with oval shape at the first part of duodenum. The lesion with smooth inner surface appears as protuberance in the lumen. **b** EUS view of the lesion revealing an about 9 × 16 mm hypoechoic mass originating from the submucosa. The mass has a regular border, well-defined limits, and no surrounding invasion.

Case Rep Gastroenterol 2022;16:546–551
DOI: 10.1159/000526135

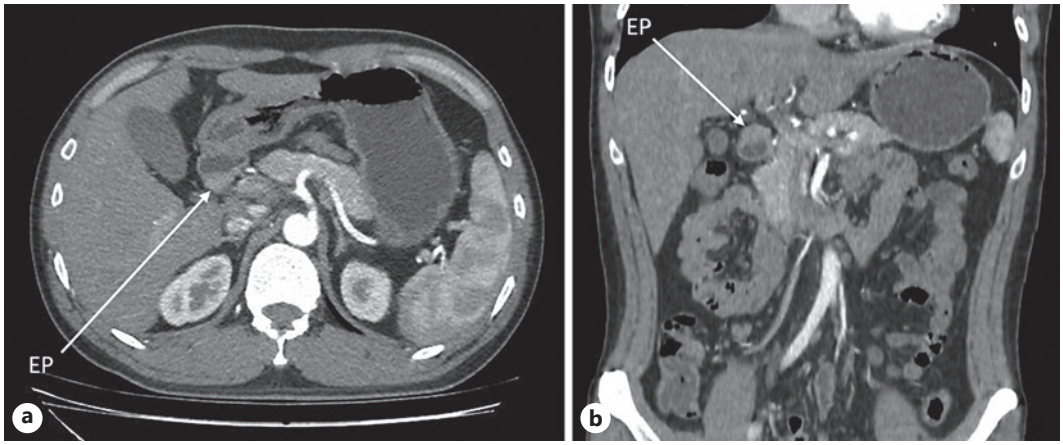

Fig. 2. A contrast-enhanced CT findings. **a** Axial view showing moderate enhancing lesion similar to pancreatic parenchyma at the posterior wall of the first part of duodenum. **b** Coronal view showing the same mass at the superior wall of the first part of duodenum. This mass is completely separate from pancreatic parenchyma. EP, ectopic pancreas.

within normal ranges. Physical examination typically showed mild epigastric tenderness. The hematological and biochemical tests were unremarkable. Past history was no known comorbidities, nonsmoker, and nondrinker.

A contrast-enhanced computed tomography (CT) of the abdomen demonstrated focal wall thickening at the D1 portion of the duodenum, like a mass with 12 × 16 mm in size, well-defined limits. This mass was completely separate from pancreatic parenchyma, moderate enhancement after injection, and partial enhancement similar to pancreatic parenchyma (Fig. 2). No progressive infiltrates or surrounding lymph nodes were seen. Serum markers including carcinoembryonic antigen, cancer antigen 19-9, cancer antigen 72-4, and cancer antigen 125 were within normal limits. Endoscopic ultrasonography (EUS) revealed a hypoechoic mass originating from the submucosa with 9 × 16 mm in size. This mass has a regular border, well-defined limits, and no surrounding invasion (Fig. 1b). EUS and CT findings were suspected to be heterotopic pancreatic tissue in the duodenum D1.

Our hospital did not have a needle for biopsy by EUS and the patient wanted to confirm by pathology. Therefore, we planned to perform laparoscopy for diagnosis and biopsies. The exploratory operation was performed by an experienced surgeon. During laparoscopy, we dissected the posterosuperior region of duodenal bulb and found the mass similar to that of the pancreatic parenchyma (Fig. 3a). Biopsies were performed at three sites. Histological examination identified normal-appearing pancreatic structures with some fibromuscular tissue (Fig. 3b). Therefore, the diagnosis was concluded to be HP at the D1 portion of the duodenum. The patient was discharged on the fifth postoperative day. The patient was followed up for 6 months and no progression of symptoms was noted.

Discussion

HP is an uncommon abnormality that is usually diagnosed accidentally in clinical practice. The incidence of HP has been estimated to be one instance every 500 upper abdominal surgeries and 0.5–13.7% of autopsy [5, 6] while EUS recorded about 9–11% of the gastrointestinal submucosal lesions as HP [9, 10]. Endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) for the diagnosis of HP in the gastrointestinal tract is indicated only in a

Case Rep Gastroenterol 2022;16:546–551
DOI: 10.1159/000526135

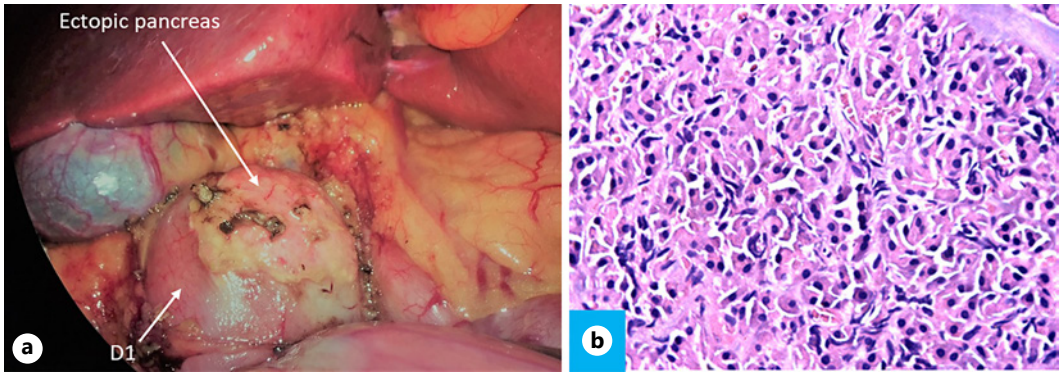

Fig. 3. a Intraoperative photographs showing the mass at posterosuperior wall of the first part of duodenum. **b** Histology of biopsy specimen revealing pancreatic acinar tissue (H&E. ×40). EP, ectopic pancreas; D1, the first part of duodenum.

small number of cases. A German study, including 63 patients with ectopic pancreas, performed EUS-FNA in 4 cases (6.3%) [10]. Many sites have been detected with heterotopic pancreatic tissue, most frequently in the upper gastrointestinal tract [2]. In the duodenum, HP is mainly found in the second segment, especially in the periampullary area, and less common in the duodenal bulb [1, 3]. HP in the duodenum was found in about 5–6% of cases after pancreatic and/or duodenal resections [7, 11].

HP is usually asymptomatic and incidentally detected during endoscopy, CT, or unrelated surgery [4]. In the symptomatic patients, they may present with nonspecific symptoms depending on the location and diameter of the lesion [11]. HP in the duodenum may have manifestations such as abdominal pain, upper gastrointestinal bleeding, ulceration, duodenal obstruction, and pseudocyst. These nonspecific symptoms result from inflammation, necrosis, or malignant transformation [4, 12]. Clinical diagnosis and imaging modalities remain challenging. Although endoscopy may reveal HP in the duodenum, this lesion presents as a mass covered by normal mucosa and protrudes in the lumen [7]. Heterotopic pancreatic tissue is typically located in the submucosa (76%), the muscular layer (15%), or the subserosa (9%) [11]. Therefore, endoscopic surface biopsies cannot be diagnosed. EUS is useful for determining which layers of gastrointestinal wall an HP is involved [4]. Especially, EUS-FNA is also essential for histopathology examination with an accuracy of 78% [9]. However, EUS-FNA is not routinely used in previous literature [10, 11, 13].

On contrast-enhanced CT images, HP is seen as round or oval lesion, ill-defined margins, and prominent enhancement of the overlying mucosa [4]. In addition, the enhancement of HP is similar to normal pancreatic tissue. However, the degree of enhancement and homogeneity of HP depends on its histologic composition. HP with predominantly acini is showed greater enhancement than normal pancreas, whereas ductal predominant lesions are showed lower enhancement [1, 14]. These findings are often nonspecific, making the diagnosis difficult. It is still easily misdiagnosed with other submucosal tumors such as gastrointestinal stroma tumor, leiomyoma, or lymphomas [7]. Histologically, HP has been categorized into three types according to Heinrich's classification: type I (all components of pancreatic ducts, acini, and islet cells), type II (including acini and ducts), and type III (only ducts) [11]. HP also has the same pathological conditions as normal pancreas, and the malignant transformation is extremely rare. Several studies have reported that a few cases of malignant transformation arising from heterotopic pancreatic tissue have been reported from 0.7 to 1.8% [8, 11, 13]. Kaneko et al. [15] reviewed 14 cases of malignant transformation arising from an HP in the duodenum. The mean age was 70.2 years (range 56–86 years), and the mean tumor size was 27.9 mm (range 12–50 mm).

Case Rep Gastroenterol 2022;16:546–551
DOI: 10.1159/000526135

In our case, the 31-year-old patient had a 9 × 16 mm lesion with no signs of malignancy. However, this suspected HP must be differentiated and excluded malignancy in the diagnosis.

Histologically proven HP without any complications or symptoms can be treated conservatively [7, 11]. In addition, malignant lesions must also be excluded. In case of symptomatic duodenal HP, it may be indicated to perform local surgical excision or partial duodenal resection. If malignancy is suspected, extensive oncological resection with lymph node dissection should be performed [11, 13]. Therefore, it is important to have clear pathological evidence before considering surgical treatment. Pathological examination should be performed by EUS with FNA or core biopsy, or surgical biopsy [9]. In our hospital, we did not have the instruments for deep biopsy by EUS. For submucosal tumors in the gastrointestinal tract (including the esophagus, stomach, and duodenum), we indicated laparoscopy/thoracoscopy to remove the tumor or biopsy. Small tumors have been performed with a combination of laparoscopy/thoracoscopy and gastrointestinal endoscopy. In this case, we planned to perform an exploratory laparoscopy. The case was a young patient with no significant symptoms. During laparoscopy, we easily exposed the duodenal bulb and found the tissue similar to the pancreas. There were no suspicious signs of malignancy and we performed biopsies at three sites. Histopathological results showed benign heterotopic pancreatic tissue. The patient was then be followed up. If severe symptoms or complications are present, surgical resection will be considered the treatment of choice.

In summary, HP in the duodenum is a rare case. Most of the diagnoses are incidental because the symptoms are nonspecific. The accurate diagnosis of HP remains challenging. Histopathological examination should be considered in the differential diagnosis and excluded malignancy. If in case EUS-FNA cannot be performed and there is no indication for resection, surgical biopsy may be a choice for diagnosis.

Statement of Ethics

Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Funding Sources

No funding received.

Author Contributions

Ngoc Trinh Thi Pham: data collection, conception and design of the manuscript, and writing the paper. Anh Nguyet Thi Nguyen and Minh Tri Thi Vo: conception and design of the manuscript and writing the paper. Minh Duc Pham: critical revision and final approval. All authors approved the final manuscript.

Case Rep Gastroenterol 2022;16:546–551
DOI: 10.1159/000526135

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

References

1 Rezvani M, Menias C, Sandrasegaran K, Olpin JD, Elsayes KM, Shaaban AM. Heterotopic pancreas: histopathologic features, imaging findings, and complications. Radiographics. 2017 Mar;37(2):484–99.
2 Farah A, Mansour S, Khuri S. Gastrointestinal tract heterotopic pancreas: asymptomatic pathology? Gastroenterol Res. 2021;14(1):45–7.
3 Thoeni RF, Gedgaudas RK. Ectopic pancreas: usual and unusual features. Gastrointest Radiol. 1980 Dec;5(1):37–42.
4 Subramanian M, Wee E, Desai V, Peh WCG. Clinics in diagnostic imaging. 158. Duodenal ectopic pancreas. Singapore Med J. 2014 Dec;55(12):629–33; quiz 633.
5 Yang C-W, Che F, Liu X-J, Yin Y, Zhang B, Song B. Insight into gastrointestinal heterotopic pancreas: imaging evaluation and differential diagnosis. Insights Imaging. 2021 Dec;12(1):144.
6 Dolan RV, ReMine WH, Dockerty MB. The fate of heterotopic pancreatic tissue. A study of 212 cases. Arch Surg. 1974 Dec;109(6):762–5.
7 Distler M, Rückert F, Aust D, Saeger H-D, Grützmann R. Pancreatic heterotopia of the duodenum: anatomic anomaly or clinical challenge? J Gastrointest Surg. 2011 Apr;15(4):631–6.
8 Mehra R, Pujahari AK, Jaiswal SS. Duodenal heterotopic pancreatic tissue: a case report and literature review. Gastroenterol Rep. 2015 Aug;3(3):262–5.
9 Attwell A, Sams S, Fukami N. Diagnosis of ectopic pancreas by endoscopic ultrasound with fine-needle aspiration. World J Gastroenterol. 2015;21(8):2367–73.
10 Gottschalk U, Dietrich CF, Jenssen C. Ectopic pancreas in the upper gastrointestinal tract: is endosonographic diagnosis reliable? Data from the German Endoscopic Ultrasound Registry and review of the literature. Endosc Ultrasound. 2018;7(4):270–8.
11 Betzler A, Mees ST, Pump J, Schölch S, Zimmermann C, Aust DE, et al. Clinical impact of duodenal pancreatic heterotopia: is there a need for surgical treatment? BMC Surg. 2017 Dec;17(1):53.
12 Minami T, Terada T, Mitsui T, Nakanuma Y. Adenocarcinoma arising from a heterotopic pancreas in the first portion of the duodenum: a case report. Surg Case Rep. 2020 Dec;6(1):141.
13 Cazacu IM, Luzuriaga Chavez AA, Nogueras Gonzalez GM, Saftoiu A, Bhutani MS. Malignant transformation of ectopic pancreas. Dig Dis Sci. 2019 Mar;64(3):655–68.
14 Kim JY, Lee JM, Kim KW, Park HS, Choi JY, Kim SH, et al. Ectopic pancreas: CT findings with emphasis on differentiation from small gastrointestinal stromal tumor and leiomyoma. Radiology. 2009 Jul;252(1):92–100.
15 Kaneko T, Ohara M, Okamura K, Fujiwara-Kuroda A, Miyasaka D, Yamabuki T, et al. Adenocarcinoma arising from an ectopic pancreas in the duodenum: a case report. Surg Case Rep. 2019 Dec;5(1):126.