Rare gastric outlet obstruction owing to ectopic pancreas: a case report and literature review

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Abstract: The ectopic pancreas is often observed in the gastrointestinal tract, and is typically asymptomatic. A 28-year-old woman was referred to our hospital following repeated vomiting after every meal. Following gastroscopy, contrast-enhanced computed tomography (CE-CT), and endoscopic ultrasonography (EUS), she was diagnosed with gastric outlet obstruction, also known as pyloric obstruction, caused by a giant submucosal cystic tumor. The condition was successfully treated with EUS-guided cystic drainage, and she was diagnosed with a cystic tumor originating from the ectopic pancreas. The tumor shrank following EUS-guided cystic drainage, and her obstructive symptoms resolved. In cases with overgrowth of the ectopic pancreas, the differential diagnosis of malignancy is important but challenging. Herein, we report a unique final pathology of this rare case and discuss the findings with a literature review.

Keywords: Case report; gastric outlet obstruction, endoscopic ultrasonography (EUS); ectopic pancreas

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Introduction

The ectopic pancreas is a congenital anomaly in which the pancreatic tissue is located outside its normal anatomical location and lacks vascular or ductal continuity. The ectopic pancreas is often visible in various organs, including the gastrointestinal tract, and is typically asymptomatic (1). In this report, we describe a rare case of cystic tumor involving the ectopic pancreas, which caused the gastric outlet obstruction, also known as pyloric obstruction. Final pathology results were obtained, and the unique histology is discussed with a literature review.

Case presentation

A 28-year-old woman with no remarkable medical or family history was referred to our hospital following complaints of repeated vomiting after every meal for 3 months. Physical examination and laboratory tests revealed no specific findings. Gastroscopy revealed a giant submucosal tumor (SMT) in the gastric antrum, obstructing the pyloric ring (*Figure 1*). Contrast-enhanced computed tomography (CE-CT) revealed a cystic tumor with a diameter of 5.8 cm; furthermore, the septal wall was minimally enhanced (*Figure 2*). SMT was solitary, and no other lesions were detected in the duodenum as well.

Endoscopic ultrasonography (EUS) was performed. The first and second layer corresponding to mucosa was intact and a large cystic lesion originating from the third layer corresponded to the submucosal growth. Subsequently, EUS-guided fine cyst drainage (EUS-CD) was successfully performed (*Figure 3*), and a total of 30 mL of internal serous liquid was aspirated (*Figure 3* inset). There were no procedural adverse events. The amylase level in the fluid was elevated at 9,623 U/L; therefore, the gastric ectopic pancreas was diagnosed. The tumor shrank following EUS-CD, and her obstructive symptoms resolved.

Thereafter, laparoscopic surgical resection was performed to achieve a histological diagnosis and prevent recurrence

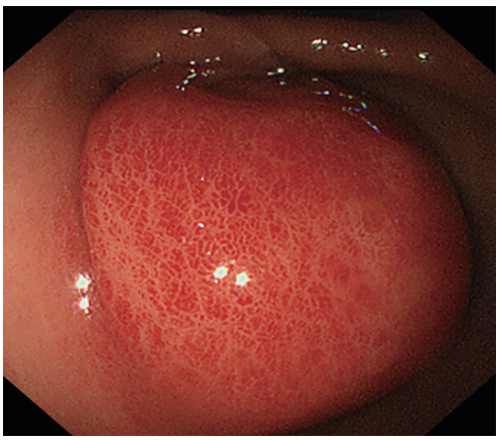

Figure 1 Gastroscopy image revealing a giant submucosal tumor obstructing the pyloric ring.

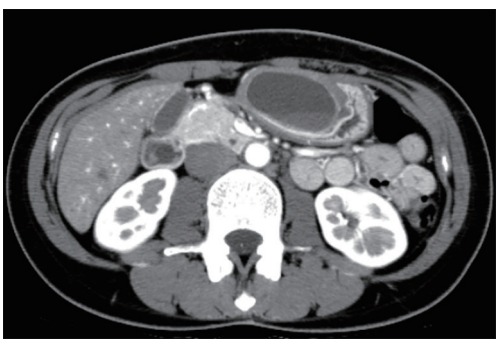

Figure 2 Contrast-enhanced computed tomography scan showing a cystic tumor with a diameter of 5.8 cm, with minimal enhancement of the septal wall.

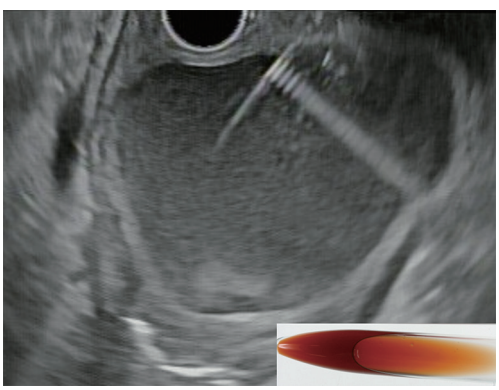

Figure 3 Endoscopic ultrasonography-guided fine cyst drainage successfully performed and aspiration of serous fluid with high amylase levels (inset).

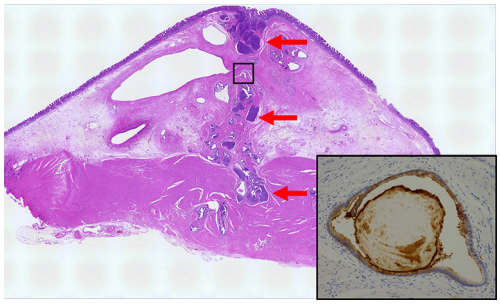

Figure 4 The resected specimen showing a pseudocyst on the ectopic pancreas. The heterotopic pancreas is characterized by a large number of ducts and acini (red arrows). The area of the black box is magnified in the inset (CEA staining, ×400 magnification).

upon the patient's request. On final pathology, a pseudocyst was detected on the ectopic pancreas. The ectopic pancreas had a microscopic appearance consistent with Heinrich's class II pancreatic tissue (1), with the majority of the heterotopic pancreas characterized by a large number of ducts and acini (*Figure 4*, red arrow). The involvement of the islets of Langerhans was not noted. Malignant components were not observed in the specimen. During one year of follow-up using gastroscopy, no recurrence was observed. Written informed consent was obtained from the patient.

Discussion

In this report, we describe a rare case of a giant submucosal cystic tumor originating from the ectopic pancreas that was successfully treated by EUS-guided cystic drainage and present the final pathology of this rare case.

Ectopic pancreas involving the stomach is most frequently seen in the antrum (2). Therefore, clinically, gastric outlet obstruction can be caused by a large cystic tumor on the ectopic pancreas. Thus, the ectopic pancreas should be considered in the differential diagnosis of pyloric flow obstruction owing to SMTs. Pathologically, cystic tumors on the ectopic pancreas are caused by the external secretion of mucin and the associated inflammation, which can lead to occlusion of the capillary lumen. EUS and, in succession, EUS-CD were reported to be effective for the diagnosis of SMTs, and treatment of symptomatic cases of cystic tumors on the ectopic pancreas (3,4); however, the long-term treatment efficacy remains unknown.

Furthermore, malignant transformation is rare but may be detected during long-term follow-up (5-9); therefore, periodic examination with EUS or CE-CT is necessary. The indication of surgical resection is a symptomatic condition, and it may be critical particularly in recurrent and malignant cases.

The literature of the ectopic pancreas in the stomach, which had been histologically confirmed, is summarized in *Table 1*. The presence of a symptomatic or asymptomatic condition is not considered useful to differentiate cases of malignancy. In contrast, EUS has the potential to diagnose malignancy, and enlargement with time, extramural growth with marginal irregularity, and heterogeneous or multilobulated appearance are suggestive but not specific for malignancy. Inflammation owing to acute pancreatitis shows a high echo signal on EUS; therefore, the mass may look heterogeneous. In patients with pancreatitis on the ectopic pancreas, we also need to carefully consider the symptoms and elevation in serum pancreatic enzymes. Cases wherein the entire mass cannot be observed should be dealt with cautiously, and there should be an objective interpretation of the findings by an expert. Depending on the size and depth of invasion of the tumor, endoscopic submucosal dissection may be used as an alternative approach for histological examination and curative treatment (21,27). Another differential diagnosis is the rare entity of gastric and duodenal cystic dystrophy in heterotopic pancreas (28,29), which is a development of multiple cystic changes in heterotopic pancreatic tissues generally in the duodenum, and in original pancreas; chronic pancreatitis was observed in the majority of them. However, only case reports and case series are available in literature, thereby lacking comparative studies to assess the efficacy of diagnosis and treatment methods, which can be considered a limitation of this study.

In conclusion, the ectopic pancreas should be considered in the differential diagnosis of gastric outlet obstruction. EUS and EUS-CD are effective for the diagnosis and treatment of symptomatic cases, although the indication of surgical resection should be considered according to individual cases.

Table 1 Summary of cases of ectopic pancreas in the stomach confirmed histologically

Authors	Reporting year	Age (years)	Sex	Symptoms	EUS findings	Malignancy on histology
Goldfarb WB (10)	1963	55	F	Abdominal pain, vomiting	N/A	Adenocarcinoma (S)
Tanimura A (11)	1979	55	F	Gastric discomfort	N/A	Adenocarcinoma, 5×20×20 mm^3 (S)
Hickman DM (12)	1981	58	M	Epigastric pain, vomiting	N/A	Adenocarcinoma, 23 mm (S)
Yasuda K (3)	1989 (n=7)			Not written	12×6-24×12 mm^2, Hypoechoic~intermediate	No malignancy (S and endoscopic biopsy)
Jeng KS (13)	1991	27	F	Epigastric fullness	N/A	Adenocarcinoma, 25×20 mm^2 (S)
Guillou L (14)	1994	60	M	Epigastric pain, dysphagia	N/A	Adenocarcinoma, 60×45×40 mm^3 (S)
Herold G (15)	1995	73	F	Abdominal discomfort	N/A	Adenocarcinoma (S)
		48	M	Epigastric pain,	N/A	Adenocarcinoma (S)
Matsushita M (16)	1997	33	M	Abdominal pain	Heterogenous hypoechoic tumor	No malignancy (S)
Ura H (17)	1998	60	F	No symptoms	Increased to 33×30 mm^2, extramural growth with marginal irregularity	Adenocarcinoma (S)
Osanai M (18)	2001	57	M	Epigastric discomfort, nausea	N/A	Adenocarcinoma, 125×90 mm^2 (S)

Table 1 (*continued*)

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Authors	Reporting year	Age (years)	Sex	Symptoms	EUS findings	Malignancy on histology
Jeong HY (9)	2002	64	M	Dyspepsia, vomiting	N/A	Adenocarcinoma, 30×30×15 mm^3 (S)
Emerson L (5)	2004	52	M	Abdominal pain, distention	N/A	Adenocarcinoma, 40×25×15 mm^3 (S)
Matsuki M (6)	2005	58	F	Vomiting	Difficult to scan the lesion appropriately due to stenosis	Adenocarcinoma 25×17 mm^2 (S)
Hirasaki S (19)	2005	32	M	Abdominal pain	30mm, hypoechoic heterogeneous mass	No malignancy
Trifan A (2)	2012	31	M	Epigastric pain, vomiting	30×30 mm^2, hypoechoic solid lesion (US)	No malignancy, 35×30 mm^2 (S)
Endo S (20)	2014	73	M	Epigastric pain, abdominal fullness	32×24 mm^2, hypoechoic mass with a cystic component (spotty high echo)	Mucinous adenocarcinoma (EMR-C)
Gong EJ (21)	2015	29	M	Abdominal pain, vomiting	Cystic lesion	No malignancy (ESD)
Matsumoto T (22)	2015	21	F	Epigastric pain, vomiting	Complex low and high echogenicity	No malignancy, 26×20 mm^2 (S)
Attwell A (23,24)	2014, 2015 (n=10)	52 (26–64)	M:F3:7	One: recurrent epigastric pain and presumed ectopic pancreatitis; all others: asymptomatic.	Hypoechoic: isoechoic =9:1. Homogenous:heterogenous =5:5	No malignancy
Jin HB (4)	2017	40	M	Abdominal pain, nausea and vomiting	21×25 mm^2, cystic lesion	No malignancy by EUS-FNA, 4 year follow up
Parra V (8)	2017	24	F	Epigastric pain	24×20 mm^2, heterogeneous and multilobulated	Mucinous cystic neoplasm (S)
Flores A (25)	2018	43	F	Abdominal pain vomiting	14×9.7 mm^2, homogeneous and hypoechoic	No malignancy (FNA)
Calcagno P (26)	2018	21	M	Vomiting, dyspepsia	80×60 mm^2, a capsulated, mixed echoic lesion, with a nodular hypoechoic portion	No malignancy (S)
		57	M	Epigastric pain, vomiting	Mixed echoic mass without a clear distal margin	Ectopic pancreas with signs of chronic inflammation (S)

M, Male; F, Female; S, surgical resection; EUS, endoscopic ultrasonography; N/A, not available; FNA, fine needle aspiration; EMR-C, endoscopic mucosal resection-using a Cap fitted endoscope; ESD, endoscopic submucosal dissection.

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Footnote

Conflicts of Interest: The authors have no conflicts of interest

to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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