Gastric Heterotopic Pancreas: an Unusual Case and Review of the Literature

Anca Trifan1,2, Eugen Târcoveanu1,3, Mihai Danciu1, Cătălin Huțanașu1,2, Camelia Cojocariu1,2, Carol Stanciu2

1) „Gr. T. Popa” University of Medicine and Pharmacy; 2) Gastroenterology and Hepatology Center; 3) Department of Surgery, „St. Spiridon” University Hospital, Iasi, Romania

Abstract

Heterotopic pancreas is a rare condition and its preoperative diagnosis is difficult. It is generally asymptomatic, but it may become clinically evident depending on the size, location and the pathological changes. Heterotopic pancreas can occur anywhere in the gastrointestinal tract, but most commonly is found in the antrum of the stomach. We report an unusual case of a 31-year-old male patient with gastric outlet obstruction and chronic pancreatitis caused by a submucosal tumor at the pre-pyloric area revealed at endoscopy. Distal gastrectomy was performed and a histological diagnosis of heterotopic pancreas was established. The patient had an uneventful postoperative course and remained symptom free in the follow-up of six months. The difficulty of making an accurate preoperative diagnosis is highlighted and a review of the literature on this pathology is hereby presented. Although heterotopic pancreas is rare, it should be considered in the differential diagnosis of pancreatitis and gastric outlet obstruction caused by a submucosal gastric tumor.

Key words

Heterotopic pancreas – gastric outlet obstruction – pancreatitis – endoscopy - surgery

Introduction

Heterotopic pancreas (HP), also known as ectopic, aberrant or accessory pancreas is defined as the presence of pancreatic tissue outside its normal location and without anatomic and vascular continuity with the main body of the pancreas [1]. Heterotopic pancreas is most commonly found in the stomach, duodenum and proximal jejunum [2]; also it can occur anywhere in the gastrointestinal tract, pelvis, liver, biliary tract, spleen, omentum, mesentery, Meckel’s diverticulum, and thorax [3-6]. Heterotopic pancreas is usually found incidentally and is generally asymptomatic, but it may become symptomatic when complicated by inflammation, bleeding, obstruction or malignant transformation [7-9]. It has been found in approximately one of every 500 surgical operations involving the upper gastrointestinal tract and in 0.6% - 13% of necropsies [10, 11]. We report an unusual case of gastric outlet obstruction and chronic pancreatitis caused by HP in a young adult man.

Case report

A 31-year-old man without any past medical history has developed epigastric pain, nausea and postprandial vomiting over the last 10 weeks. He had a weight loss of 15 kg during this time. Epigastric pain was refractory to treatment with proton pump inhibitors. He had never smoked, and drunk alcohol only occasionally. One week before admission to our hospital, the patient undertook an upper gastrointestinal endoscopy in another hospital which showed a pre-pyloric polypoid mass.

On examination, the patient was in good health, his abdomen was soft and non-tender, with no palpable mass. He was afebrile, and his vital signs, respiratory and vascular systems were normal. Hematological examination and biochemical tests were within normal limits, except for a 3-fold increase in the serum amylase (307 IU/L, reference range 20-100 IU/L).

Abdominal ultrasound revealed a hypechoic solid lesion of 3x3 cm in gastric antral region; pancreas, liver and spleen were normal. Upper gastrointestinal endoscopy showed a round-shaped, sessile polypoid submucosal lesion in the posterior wall of the gastric antrum, within a few mm of the pylorus, with normal overlying mucosa (Fig. 1), measuring ~3 cm in diameter. The endoscope was passed with difficulty around the lesion into the pylorus and duodenum which were found to be normal. Biopsy of the lesion with standard biopsy forceps was unremarkable.

The patient refused further investigations (endoscopic
ultrasound-EUS, computed tomography-CT), and because of his persistent symptoms (particularly vomiting) he opted for surgery. Thus, with a presumed diagnosis of stromal tumor determining gastric outlet obstruction, a decision was made to proceed with surgery. At surgery, a submucosal tumor 3.5x3 cm located in pre-pyloric area, with a distal limit of 3 mm from the pylorus was found. The underlying mucosa was intact. The surgeon considered the tumor to be a gastrointestinal stromal tumor (GIST) and a distal gastrectomy with Billroth I anastomosis was carried out. Gross examination of the resected specimen showed a well-delimited mass with normal overlying mucosa, located on the posterior wall of the pre-pyloric antrum; longitudinal section of the tumor revealed a pancreatic-like tissue with hemorrhagic areas within muscularis propria and subserosa (Fig. 2). Microscopic examination of the lesion showed pancreatic tissue with ducts, acini, and islets distributed from submucosa to serosa (Fig. 3), and features of chronic pancreatitis with fibrosis, diffuse chronic lymphoplasmocytic infiltrate and remnant islets (Fig. 4).

The postoperative course was uneventful and the patient was discharged 6 days after surgery. He has remained free of symptoms and gained 10 kg of weight over a six months period, with serum amylase and lipase within normal limits.

Discussion

The first case of HP, developed in an ileal diverticulum, was reported by Jean Schultz in 1727, but the first histological confirmation was described by Klob in 1859 [12]. It is a rare entity, defined as the presence of pancreatic tissue outside its normal location that lacks anatomical or vascular connections with the normal body of the pancreas. Several theories have been proposed to explain the occurrence of HP. The most tenable implicates that during the embryonic rotation of the dorsal and ventral buds, fragments of the pancreas become separated from the main body and are deposited at ectopic sites [13].

The prevalence of HP at necropsy is between 0.6% and 13.7% [11], and has been observed in one of every 500 upper abdominal operations [10]. Heterotopic pancreas may be present at any age, but most often is discovered in the fifth and sixth decades of life, with a male predominance [14]. It is most commonly found in the stomach (25-38%), duodenum (17-21%), and proximal jejunum (15-21%) [15], although it can occur anywhere in the gastrointestinal tract, pelvis, liver, biliary tract, spleen, omentum, mesentery, fallopian tube, Meckel’s diverticulum, mediastinum, and lung [3-6].

Heterotopic pancreas is usually asymptomatic and it
is found during endoscopy, imaging or surgery, but it may become clinically evident depending on its size and the pathological changes (acute or chronic pancreatitis, cyst formation, malignant degeneration) [7-9]. Any pathological processes associated with the normally placed pancreas can occur in the ectopic pancreas.

Gastric HP is located in the antrum in over 90% of the cases [15] and symptoms depending upon the anatomical location (e.g., gastric outlet obstruction in a pre-pyloric HP) originate from the mass effect of the tumor and its size (symptoms are more likely to occur with lesions larger than 2 cm in diameter) [16].

The diagnosis of HP is difficult as there are no specific diagnostic methods. Thus, in one series of 32 cases of symptomatic HP, none of the patients was diagnosed preoperatively [17]. Barium swallow studies may show a rounded filling defect, sometimes with central indentation. Imaging studies (EUS, CT) are frequently used for the diagnosis of gastrointestinal submucosal tumor and can be helpful in the diagnosis of gastric HP, but are not specific [18, 19]. However, recent studies [20, 21] have suggested that both EUS and CT help to distinguish HP from other submucosal tumors. Endoscopic appearance of HP is that of a well circumscribed submucosal mass with a normal overlying mucosa and a central dimpling which corresponds with the opening of a duct. The characteristic dimpling or umbilication is observed in less than half of the cases, and therefore, HP may easily be misinterpreted as another submucosal tumor such as stromal tumor, or leiomyoma at endoscopic examination. Because GISTs are by far the most common gastric submucosal tumors, HP can frequently be mistaken for GIST at endoscopy as happened in our case [21, 22]. Endoscopic biopsy performed by using standard biopsy forceps is most often unremarkable. However, a few reported cases of HP were diagnosed with biopsies obtained with the jumbo forceps [23].

The definitive diagnosis of HP is reached on the histopathological examination of the tissue. Histologically, Heinrich in 1909 classified the HP into 3 types [24]: type 1, the most common, with all the components of the pancreas including acini, ducts and islets; type 2, with acini and ducts and no islets; type 3, with ducts alone. Our case belongs to type 1. Later, the Heinrich classification was modified by Gaspar Fuentes et al [25] into 4 types: type 1, composed by acini, duct and islets similar to those seen in normal pancreas; type 2, composed of ducts only; type 3, consists of acini only (exocrine pancreas); type 4, composed of islets only (endocrine pancreas).

Surgical resection should be performed if HP is symptomatic or when the lesion is found accidentally during surgery and is larger than 3 cm in size, in order to prevent complications [26]. Laparoscopic surgery to gastric HP appears to be technically feasible and safe and should be considered a viable alternative to open surgery [27]. Several reports describing various methods of endoscopic resection of gastric HP have been published [28, 29].

A few cases of HP as a cause of gastric outlet obstruction in infant [30], child [31], and adult [1, 7, 10, 14, 32-35] have been published. In our case, like in almost all others, the diagnosis of HP was established at surgery. The clinical picture of our patient dominated by postprandial vomiting and weight loss associated with the endoscopic finding of a pre-pyloric submucosal tumor with an intact mucosal surface has suggested the diagnosis of stromal tumor which seemed to explain the typically symptoms of gastric outlet obstruction. It should be mentioned that the diagnosis of HP was difficult even intraoperatively, due to the gross similarity with GIST.

Ectopic pancreatitis has been reported in patients with HP located in the stomach and other sites of gastrointestinal tract [36-46]. All reported cases were successfully treated with surgery. There have been a few reported cases of histologically proven pancreatitis in gastric HP [40, 42, 44-46]. Burke et al [42] reported a case of gastric antral HP causing gastric outlet obstruction secondary to pancreatitis and pancreatic pseudocyst. To our knowledge, our case is the first one of HP causing chronic pancreatitis and gastric outlet obstruction. Although there was a mild increase of serum amylase and the patient had abdominal pain, we missed the diagnosis of pancreatitis (normal echogenicity of the pancreatic parenchyma at the abdominal US, no history of alcohol abuse) which was proved microscopically in the resected specimen. The particularity of our case is the fact that the gastric outlet obstruction was due to the location and size of the HP and not to complications of pancreatitis such as a pseudocyst formation.

Conclusion

Cases of heterotopic pancreas causing both gastric outlet obstruction and pancreatitis are very rare and the preoperative diagnosis is difficult. However, heterotopic pancreas should be considered in the differential diagnosis of pancreatitis and gastric outlet obstruction caused by a submucosal gastric antral tumor.

References