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Case Report

Rare Case of Heterotopic Pancreas in the Pylorus

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Abstract

Heterotopic Pancreas (HP) is an aberrant anatomic malformation that occurs outside the normal anatomical site, most commonly in the upper gastrointestinal tract. Detection of HP remains a major challenge due to its similarity to submucosal tumors, so it is often misdiagnosed. For this reason, pathological confirmation is necessary to diagnose HP.

A 62-year-old male patient was admitted with suspected gastric Gastrointestinal Stromal Tumor (GIST) located in the pylorus for distal resection. He reports that he first felt a palpable change under the skin of the abdominal cavity 2 years ago and since then he has been diagnosed with pancreatic and liver diseases.

Scintigraphy, Computed Tomography (CT) and abdominal Magnetic Resonance (MR) imaging indicated tumor in the anterior pyloric wall. Endoscopic Ultrasound (EUS) biopsy attempt-negative. The patient was qualified for distal gastrectomy (Billroth II technique). Histopathological examination, showed a histological appearance corresponding to a HP with the presence of pancreatic ducts.

The variability in presentation and difficulty with characterizing HP suggest that further observation and documentation of HP may prove beneficial in managing patients with radical resection.

Keywords: Heterotopic pancreas; Aberrant pancreas; GIST; Pylorus tumor; Distal gastrectomy

Introduction

Heterotopic Pancreas (HP) is an infrequent congenital condition defined as pancreatic tissue detected outside its normal location with no anatomic, neural, or vascular relationship to the main pancreas [1,2]. Other names include abnormal pancreas, ectopic pancreas, residual pancreas and pancreatic heterotopia. A case of HP was first described in 1727, the lesion was found in the diverticulum of the ileum [3].

The aberrant pancreatic tissue can be located at any part of the gastrointestinal tract, but most commonly occurs in the stomach (24% to 38%), duodenum (9% to 36%), and jejunum (0.5% to 27%) [1,4]. Ectopic pancreas located in the stomach poses a diagnostic challenge due to its similarity to other gastric tumors, caused by its frequent submucosal appearance [5].

In most cases, HP presents no characteristic clinical symptoms or signs, and the diagnosis is made on the basis of pathological material or at autopsy [4,6]. However, HP can cause nonspecific gastrointestinal symptoms, especially if it is complicated by inflammation, bleeding, obstruction or malignant transformation [7-9]. It has been proven that the most common symptom reported by HP patients is abdominal pain [10]. Symptoms are more likely to occur in lesions larger than 1.5 cm [11]. In some severe cases, HP may require surgical intervention [4]. Herein, we present a case of a patient with a tumor suspected as GIST, later diagnosed as HP.

Case Presentation

A 62-year-old male patient was admitted with suspected gastric GIST located in the pylorus for distal resection. He reports that he first felt a palpable change under the skin of the abdominal cavity 2 years ago (accompanied by dyspeptic symptoms and pain in the epigastrium) and since then he has been diagnosed with pancreatic and liver diseases. Scintigraphy, CT (Figure 1) and abdominal MR imaging indicated a tumor in the anterior pyloric wall. A EUS biopsy was attempted and a negative result was obtained. At the same time, the patient was diagnosed with prostate cancer. Due to the unclear picture of the stomach tumor, oncologists did not decide to start treatment for prostate cancer. An exploratory laparotomy and distal gastrectomy (using the Billroth II technique) were performed. During the procedure, a fragment of the pylorus measuring 8 × 4.5 × 1.5 cm was excised. Omega gastro-enteric and Braun entero-intestinal anastomoses were produced. Histopathological examination showed a histological appearance corresponding to HP with the presence of pancreatic ducts (Figure 2). The size of the tumor was 15 mm in each dimension and no neoplastic changes were found. Microscopic examination also revealed in the gastric mucosa signs of chronic inflammation with small focal areas of medium intensity, intense intestinal metaplasia and three lymph nodes with sinus histiocytosis. H. pylori infection was excluded.

Outcome and Follow-Up

Due to the stable condition the patient was discharged after a week of observation and pain treatment. The patient has been scheduled for a follow-up appointment. Endoscopic follow-up after treatment was recommended. Currently, the patient is in follow-up 11 months after surgery and reports no gastrointestinal symptoms.

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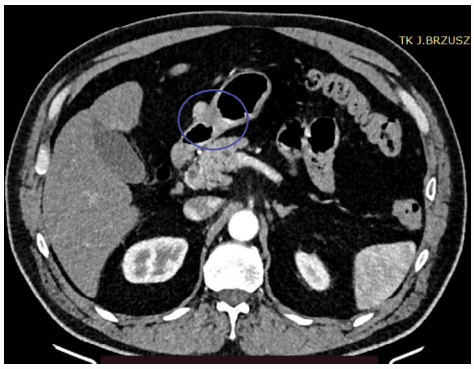

Figure 1: CT indicating tumor 17 × 15 mm in the anterior pyloric wall.

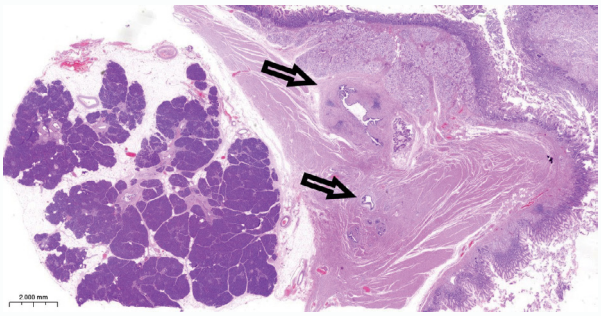

Figure 2: The Heterotopic pancreas in subserosal and muscularis layers of the stomach. The pancreatic ducts (marked with arrows) seem to play a key role in the transport of pancreatic secretions to the stomach. H&E, mag 60x.

Discussion

HP is a rather rare congenital abnormality with an occurrence of 1/500 in surgical specimens [2,6]. A retrospective cohort study from 2022 identified 1762 described cases in total [6]. Although pathogenesis of HP is unclear, several mechanisms have been proposed to explain its occurrence including the mislocation theory, the chemotaxis theory, and the totipotent cell theory [2,12]. The most tenable mislocation theory implicates that during embryonic rotation, dorsal and ventral deposits of pancreatic tissue separate and migrate from the main body of the pancreas to different ectopic sites [2,12-14]. However, this mechanism does not fully demonstrate the development of HP in all cases as aberrant pancreatic tissue has also been found outside the gastrointestinal tract. Uncommon locations of HP include the common bile duct, gallbladder [15], Meckel's diverticulum [16], mesentery of the intestine [17], and umbilicus [18], esophagus [19], fallopian tube, mediastinum, omentum, mesentery and spleen [20,21], brain [22], rectum [23], papilla of Vater, urinary bladder, lung and lymph nodes [24]. Another possible cause of HP formation is explained by the chemotaxis theory, which posits that embryonic tissue migrates to the submucosa during development and then transforms into pancreatic tissue. An alternative explanation suggests that totipotent endodermal tissues develop into pancreatic tissue during embryologic development and can be present in various tissues throughout the body [2,6].

Heinrich's classification of ectopic pancreas distinguishes three types: type 1 is characterised by the presence of acini, ducts and Langerhans islands, in type 2 only acini and ducts are present, and type 3 includes only ectopic proliferating ductal tissue. On pathological examination, the macroscopic image of HP reveals the characteristic central opening of the duct [25]. HP is usually located in the submucosal layer (76%), muscularis (15%) or subserosal (9%) layer [26].

HP is often mistaken for other tumors because of its appearance and location. In the differentiation of heterotopic pancreatic tissue, the main considerations should be gastrointestinal lining tumors, gastrointestinal autonomic nerve tumors, gastric carcinomas, lymphoma or gastric cancer [27,28]. The literature also reports rarer cases were HP mimics neoplastic metastases [27]. Distinction from other submucosal tumors may be challenging. CT imaging can help put forward a suspicion of HP, as there are a number of features indicative of this condition: submucosal masses, ill-defined borders, endoluminal growth, prominently enhancing overlying mucosa, bright enhancement similar to the normal pancreas, surface dimpling and low intralesional attenuation, a long diameter to short diameter ratio greater than 1.4 [28-30].

Another helpful imaging method is MR. The most important feature of HP on MR is the isointensity present in the orthotopic pancreas in all sequences. In addition, it usually shows high SI at T1-WI, there is no diffusion restriction or hypervascular behavior, which is similar to native pancreas [1].

Despite the fact that endoscopic ultrasonography has become a major tool in detecting submucosal tumors, it should be noted that histological confirmation is required for a definitive diagnosis. It has been shown that echogenicity can vary depending on the layers in which HP is present, therefore cytology or histology is necessary [25].

Unfortunately, biopsies taken too superficially are non-diagnostic [15], so the procedure should be performed very precisely. Another challenge for pathologists is the fact that tissue samples are often obtained using conventional endoscopic biopsy forceps [31]. Some sources recommend Endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) to obtain histopathological material of the highest possible quality. In case of ineffective EUS-FDA, a deep tunnel (exposure) forceps biopsy may be considered [32].

Although most HP are asymptomatic, they sometimes cause serious complications, including obstruction, inflammation, ulceration and perforation, intussusception and bleeding [33].

HP should be considered as a source of potentially malignant lesions [34]. Currently there are no standardized guidelines for treatment [35]. The decision to treat the lesion surgically depends on the decision of doctors, who take into account the location and size, the age and condition of the patient, the presence of symptoms [36]. Some authors suggest resection of asymptomatic HP to prevent potential complications, but this is still under debate [37,38]. In symptomatic HP, excision of the entire tumor with safe margins is the recommended treatment [39]. In most cases, laparoscopy becomes the preferred treatment method [35,40]. In the presented case, the need to perform the procedure was determined by the location of the tumor within the pylorus and, additionally, obtaining a histopathological result before starting treatment of the second tumor.

Regular follow-up examinations after surgery are important because up to 12.7% of pancreatic rests can develop malignant transformation [41].

Conclusions

HP is a rare lesion, but should be considered in the differential diagnosis in patients with massive gastric lesions, as well as when there are unclear upper gastrointestinal symptoms. The variability

in presentation and difficulty with characterizing HP suggest that further observation and documentation of HP may prove beneficial in managing patients and preventing future morbidity associated with radical resection. The role of histopathologists in diagnosing HP and distinguishing it from potential tumours remains very important.

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