Ectopic Pancreatic Tissue (EPT) in a cholecystectomy specimen: A rare incidental pathologic finding

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Introduction

Ectopic pancreas tissue (EPT) or pancreatic heterotopia is a rarely observed congenital abnormality defined as the presence of pancreatic tissue in another organ without any anatomical or vascular connection to the pancreas. The term consists of the two Greek words "hetero-" which means "other" and "-topia" which means "site", pointing out the unique location of pancreatic cells. EPT's favored sites are the stomach, duodenum, colon, jejunum, and Meckel’s diverticula (1). The gallbladder is a highly infrequent location for EPT (2). Almost all cases are detected incidentally during the histopathological examination after cholecystectomy for other pathologies. The prevalence of EPT in the gastrointestinal tract varies from 0.6% to 13.7% in autopsy series and 0.2% in laparotomies (3, 4). Although the malignant transformation of this tissue is not frequently expected, pathologists must be aware of it to ensure no malignant pathology is present and prevent further misdiagnosis. In this study, we present a case of EPT that we found incidentally during the histopathological examination of the specimen from the gallbladder in a patient who underwent cholecystectomy due to acute cholecystitis.

Case History

A 40-year-old male was referred to the emergency department for acute pain in the right upper quadrant of the abdomen. The pain was constant and initiated hours before and intensified when he had dinner. He also reported fever, anorexia, nausea, and one involuntary vomiting episode containing only stomach contents. Besides intellectual disability and epilepsy, patient’s medical history was unremarkable otherwise. He was on Valproate(200mg/D), Risperidone(1mg/BD) and Clonazepam(1mg/HS). He denied smoking and any recreational drug use. No allergies were reported. During the clinical examination, his vitals were slightly above normal ranges (Blood pressure: 115/70mmHg; Heart rate: 108bpm; Respiratory rate: 19 breaths/min; Temperature: 37.9°C; Oxygen saturation: 99% without supplemental oxygen). The palpation of the abdomen revealed tenderness in the right upper quadrant and a positive Murphy sign with no rebound tenderness or guarding. The laboratory evaluation was within normal range except for a WBC count of 10300, ESR of 32, and a 2+ CRP. Additionally, he underwent an ultrasonographic evaluation of the upper abdomen, reporting a thickened wall gallbladder containing multiple stones (measuring up to 8x10mm). The patient was scheduled for emergency laparoscopic cholecystectomy under the diagnosis of acute calculous cholecystitis. During surgery, the gallbladder was found to be gangrenous. The patient had a complicated postoperative period due to surgical wound infections and was hospitalized for a week to receive intravenous antibiotics; however, he did not report any delayed complications or symptoms after discharge.

Investigations

During pathologic examination, gallbladder measured 9cm x 4cm containing multiple small yellowish stones. The mucosa was green, and the maximum wall thickness was 2mm. An area of wall thickening measuring...
7mm x 4mm x 3mm was noted in the neck region of the gallbladder. Microscopic examination revealed characteristics indicative of acute gangrenous cholecystitis. Regarding the wall thickening in the neck area described macroscopically, histopathology confirmed the presence of heterotopic pancreatic tissue composed of acini and ducts (exocrine pancreatic tissue only; no islets of Langerhans were detected). There was no evidence of malignancy or dysplasia in any of the sections examined.

Discussion

EPT is a rare entity; however, its actual incidence in the gallbladder is unidentified, as a lack of clinical symptoms can complicate the diagnosis (5). It can occur anywhere, but the most common locations are the stomach (27.5%), duodenum (25.5%), colon (15.9%), jejunum, and spleen (6). By contrast, the presence of EPT in the gallbladder, lung mediastinum, liver, mesentery, and ileum is considered extraordinary (2, 7). EPT can be diagnosed in any age group, but most cases are between 40-60 years old. Albeit the male-to-female ratio of any other type of EPT is 3:1, there is a female predominance, specifically for EPT in the gallbladder, which may be due to the fact that cholecystitis-related cholecystectomies are more prevalent in women (2, 8). The etiologies of EPT are still unclear, but three hypotheses about its origin have been proposed. The first theory, which is widely accepted, suggests that EPT separates from the primitive pancreas gland during the rotation of the gastrointestinal tract in the embryogenic period. The second theory suggests that the longitudinal growth of the intestine from the lateral budding of the rudimentary pancreas tissue while penetrating the intestinal wall causes the irregular transportation of the pancreatic tissue (9, 10). The third theory supports that abnormalities in the notch signaling system can result in changes in different foregut endoderm tissue during embryogenesis (11). Jean Schultz first described the heterotropic pancreas in the 18th century; however, the first classification was made by Von Heinrich et al. in 1909, which was later modified by Fuentes in 1973 that consisted of four types (12):

1. Type one: acini with ducts and islet-like pancreatic gland (normal pancreatic tissue)
2. Type two: canalicular variant pancreatic duct
3. Type three: exocrine pancreas with acinar tissue
4. Type four: endocrine pancreas with cellular islets

According to what is mentioned above, this case is compatible with type three (figure1-3).

These ectopic pieces of tissue macroscopically can appear as an exophytic mass, similar to polypoid lesions, or as a nodule with a yellow-colored appearance with sizes varying from a few millimeters to even 4cm (13). Still, they are generally asymptomatic and only discovered histopathologically. However, in some cases, it can cause various nonspecific symptoms, depending on the location. Symptoms can include jaundice if obstructing bile ducts or biliary colic-like symptoms (e.g., right upper quadrant pain, anorexia, nausea, and vomiting after meals); nevertheless, such symptoms presumably result from simultaneous lithiasic cholecystitis. Other conditions can be derived from EPT, such as cholelithiasis, acute or chronic cholecystitis, or carcinoma (2, 10).

As Sato et al. reported that pancreatic enzymes (amylase and lipase) secreted from EPT in the gallbladder could impact its mucosa leading to gallbladder dysplasia and carcinoma. Therefore, cases with EPT in the gallbladder must undergo cholecystectomy as a definite treatment preventing any potential malignant transformation. Moreover, EPT can potentially cause the same pathologies as typical pancreatic tissue, which includes cysts, pseudocysts formation, abscess, and acute or chronic pancreatitis (14, 15).

Diagnosing EPT in the gallbladder before and during an operation is impossible. Preoperative radiologic evaluation (ultrasound or computed tomography scan) usually cannot detect EPT in the gallbladder; neither this case did (16). Therefore, only a precise histopathologic examination can provide a definite diagnosis; consequently, it’s essential for anatomical pathologists to be aware of this uncommon presentation of EPT in gallbladder to discriminate it from a masquerading malignancy.

References


Figures’ legend:

Figure1. (x10, a) and (x40, b) Heterotopic pancreatic islands consists of exocrine pancreas with acinar glands arrow heads and pancreatic ducts arrows.

Figure2. Small pancreatic ducts arrows with adjacent atrophic gallbladder mucosa asterisk (x10)

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