

CASE REPORT

Heterotopic Pancreas: Report of Two Cases

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ABSTRACT

Context Heterotopic pancreas is a congenital anomaly defined as pancreatic tissue occurring outside its normal anatomical location, lacking both anatomic and vascular connections. Ninety percent of heterotopic pancreata are found in the upper part of the gastrointestinal tract and clinical presentations show differences.

Case reports We present two different cases of heterotopic pancreas. One of them was a 56-year-old man noted to have a mass in the proximal duodenum during elective cholecystectomy for cholelithiasis. The mass was excised locally and primary duodenal repair was carried out. The second case was a 41-year-old man admitted to our hospital with an intestinal obstruction caused by a jejunal heterotopic pancreas; he was operated on under emergency conditions. Both patients' postoperative periods were uneventful.

Conclusion Resection of the heterotopic pancreas tissue-bearing area is advisable when the condition is encountered coincidentally at surgery, and surgical exploration should be done for all symptomatic patients, particularly in the absence of a histological diagnosis.

INTRODUCTION

Heterotopic pancreas is the presence of pancreatic tissue outside its normal location, lacking ductal or vascular continuity with the

main gland. Pancreatic heterotopia is found in 2-15% of all autopsies [1]. The majority of heterotopic pancreata are asymptomatic; the clinical prevalence is estimated as one every 500 upper abdominal operations [2].

The most common sites of a heterotopic pancreas are gastric antrum, duodenum, jejunum and Meckel diverticulum. Unusual localizations are the colon, spleen, liver biliary tract mesentery and lymph nodes. Also, heterotopic pancreas may be found in mature teratomas, particularly of mediastinal origin [3, 4]. We herein report two cases of heterotopic pancreas having different locations.

CASE REPORTS

Case 1

A 56-year-old man presented at the Gastroenterology Out-patient Clinic complaining of epigastric pain, flatulence, nausea and vomiting. His physical and laboratory findings were normal. An upper gastrointestinal endoscopy revealed superficial antral gastritis, but abdominal ultrasonography showed a 2 cm gallstone located at the fundus of the gallbladder and the patient underwent surgery. During surgery, other than cholelithiasis, an exophytic, soft, yellowish mass located on the anterior surface of the first part of duodenum, 1.5 cm in size, was detected. The cholecystectomy together with local excision of the duodenal mass with primary duodenal repair was carried out. A frozen section

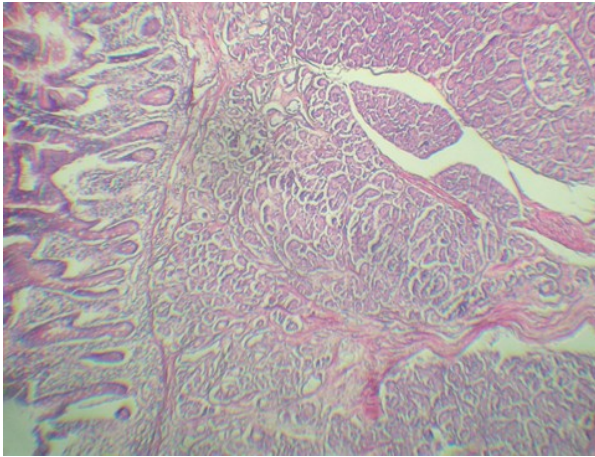


Figure 1. Case 1. Low magnification view of a heterotopic pancreas nodule in the duodenal submucosal area and an endocrine island located in the upper right corner of the image. (H&E x40)

biopsy of the duodenal lesion could not make any definitive diagnosis. The postoperative period was uneventful and pathology of the mass revealed a heterotopic pancreas (Figure 1).

Case 2

A 41-year-old man presented with a 3-day history of diffuse abdominal pain. On physical examination, there was rebound tenderness at the upper quadrant of the abdomen. Laboratory tests revealed leukocytosis and an elevated amylase level. Plain abdominal X-ray suggested intestinal obstruction. The patient was then operated on for an acute abdomen with intestinal obstruction. At laparotomy, a mass measuring 3x4 cm located in the first 20 cm of the jejunum causing lumen occlusion and dilatation of the proximal intestine, leading to partial small bowel obstruction, was found. A segmental jejunum excision with primary anastomosis was carried out. Subsequent pathology revealed a heterotopic pancreas (Figure 2). During the follow-up period, there were no sequelae from the excision of the heterotopic pancreas.

DISCUSSION

The pancreas is derived from several endodermal invaginations of the primitive duodenal wall. The dorsal diverticulum becomes the body and tail and the ventral

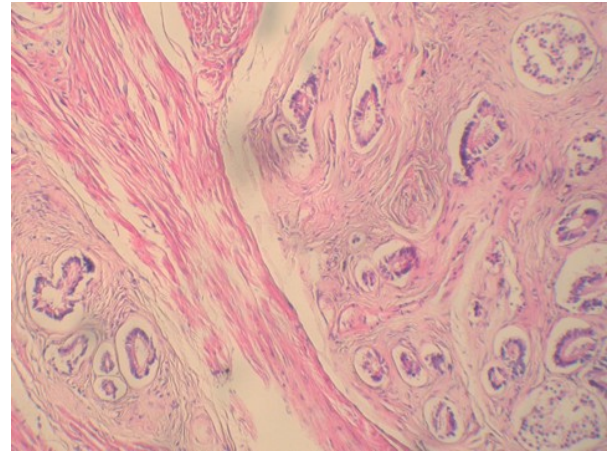


Figure 2. Case 2. A heterotopic pancreas nodule showing variably dilated ducts, acini and two islands of endocrine cells present in the upper and lower right corners. (H&E x100)

portion becomes the head of the pancreas. The pathogenesis of these lesions is unknown; they are thought to arise during rotation of the foregut when fragments of the pancreas become separated and develop into mature elements (“misplacement theory”) or from pancreatic metaplasia of endodermal tissue which migrates to the submucosa during embryonic life [2, 5].

In the gastrointestinal tract, pancreatic heterotopia forms round or lobulated white or yellowish lumps of tissue ranging from a few millimeters up to a few centimeters; larger tumors tend to have central umbilication which represents the site of the draining duct. The mass lies most commonly in the submucosa but can present as a subserosal nodule [3, 6].

Despite its congenital origin, a heterotopic pancreas clinically manifests itself in older adults. Heterotopic pancreas is usually an incidental finding, either at the time of laparotomy for another disease or during radiographic or endoscopic examination of the upper gut. We recommend a frozen section in cases found incidentally during surgery in order to identify these lesions especially if the upper GI endoscopy was done and did not show any mucosal lesions preoperatively. Moreover, if there is the feasibility of carrying out intraoperative ultrasonography or endoscopic ultrasonography, it also helps the surgeon to make a

decision intraoperatively. However, a heterotopic pancreas may be present with gastric outlet obstruction and upper gastrointestinal bleeding [2, 4, 6]. Small bowel obstruction and obstructive jaundice from a heterotopic pancreas are rare [7, 8]. Adenocarcinoma, islet cell tumors and cystic tumors are reported in a heterotopic pancreas [9, 10].

The preoperative diagnosis of a heterotopic pancreas is still difficult regardless of the recent advance in diagnostic tools and techniques. There is no specific indicator or biochemical marker to diagnose a heterotopic pancreas. But the complications related to heterotopic pancreas, such as small bowel obstruction, pancreatitis or GI bleeding, might lead to increased amylase levels as in our case report. A heterotopic pancreas typically presents in the upper gastrointestinal series as a well-delineated submucosal filling defect with a characteristic central indentation [3, 4]. Endoscopically, the lesion is seen as a submucosal tumor with a central compression or umbilication. The CT appearance of most heterotopic pancreata enhance brightly, similar to an orthotopic pancreas. Reduced enhancement may be seen with inflammation of a heterotopic pancreas [6].

Klob first described the histological appearance of a heterotopic pancreas in 1859 [11]. Histopathologically, a heterotopic pancreas contains a mixture of tissues which may be found in the pancreas. It usually consists primarily of ducts and the surrounding mucin-producing glands. Occasionally, only ducts or islets are present. Heinrich classified heterotopic pancreata into 3 types: a) Type 1: ducts plus acini plus endocrine islets; b) Type 2: ducts plus acini and c) Type 3: ducts with few acini or dilated ducts only, so-called adenomyoma [12].

In the treatment of heterotopic pancreas, where feasible, local excision rather than radical surgery is the preferred way of treatment of heterotopic pancreata [4, 7]. However, given the difficulties with accurate preoperative diagnosis and the suspicion of an underlying malignancy, radical surgical treatment might be chosen in order to prevent

re-operation and diagnostic difficulties [2, 5, 8].

In a review of the literature, and also in our experience, resection of the heterotopic pancreas tissue-bearing area is advisable when the condition is encountered coincidentally at surgery, and surgical exploration should be done for all symptomatic patients, particularly in the absence of histological diagnosis.

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