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CASE REPORT

Heterotopic Pancreatic Tissue Obstructing the Gallbladder Neck: A Case Report

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ABSTRACT

Context Heterotopic pancreatic tissue is defined as pancreatic tissue outside the boundaries of the pancreas that has neither anatomic nor vascular continuity with the pancreas. Heterotopic pancreatic tissue in the gallbladder is uncommon and has rarely been reported to cause symptoms. We report a case of heterotopic pancreatic tissue obstructing the gallbladder neck resulting in cholecystitis. **Case report** A 26-year-old female presented with right upper quadrant abdominal pain and fever. On physical examination the right upper quadrant was tender to palpation with a positive Murphy's sign. Laboratory tests were significant for elevated aspartate aminotransferase and alanine aminotransferase. Transabdominal sonography showed gallbladder wall thickening, a positive sonographic Murphy's sign, and an apparent large non-mobile stone at the gallbladder neck. Pathologic examination revealed cholecystitis but instead of a large stone there was a tan-yellow necrotic mass at the gallbladder neck. Microscopically, the mass consisted of heterotopic pancreatic tissue containing exocrine pancreatic acini, ducts, and islets of Langerhans. The final diagnosis was acute cholecystitis secondary to obstruction by heterotopic pancreatic tissue. **Conclusion** Although heterotopic pancreatic tissue is usually an incidental finding on pathologic exam, one should not exclude it in the differential diagnosis of symptomatic gallbladder disease of indefinite etiology.

INTRODUCTION

Heterotopic pancreatic tissue is defined as pancreatic tissue outside the boundaries of the pancreas that has neither anatomic nor vascular continuity with the pancreas [1]. This tissue, rarely found in the gallbladder, is usually discovered incidentally at pathologic exam. However, there have been reports of heterotopic pancreatic tissue causing symptomatic gallbladder disease. We report a case of a 26-year-old female with heterotopic pancreatic tissue obstructing the neck of the gallbladder with clinical findings of acute cholecystitis.

CASE REPORT

A 26-year-old female presented to the Emergency Department complaining of 24 hours of progressively worsening right upper quadrant abdominal pain that radiated to her back and right scapula. On physical examination the patient was obese with a body mass index of 32 kg/m^2, febrile, and appeared to be in significant pain with a positive Murphy's sign. Laboratory results were significant for aspartate aminotransferase (AST) 57 U/L (reference range: 9-16 U/L), alanine aminotransferase (ALT) 118 U/L (reference range: 8-30 U/L), and lipase 20 U/L (reference range: 12-70 U/L). Real-time trans-abdominal sonography of the right upper quadrant was technically difficult due to the patient's obese body habitus. The ultrasound showed mild gallbladder wall thickening (2 to 4 mm), no pericholecystic fluid, a positive sonographic Murphy's sign, and a large, non-mobile, 11x8x4 mm mass with gallstone-like appearance within the gallbladder neck (Figure 1). The common bile duct measured 6 mm in diameter at the porta hepatis without evidence of choledocholithiasis. A diagnosis of acute cholecystitis was made and the pati ent underwent an uncomplicated laparoscopic cholecystectomy. Intraoperative cholangiography showed no filling defects of the common bile duct with free flow of contrast into the duodenum demonstrating no evidence of retained gallstones. The patient's right upper quadrant pain was relieved following surgery and she was discharged home on the first post-operative day. Patient remained pain free with no right upper quadrant pain at one month follow-up.

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Pathologic examination revealed an 8.5x3.5x3.0 cm gallbladder with a smooth gray-white serosal surface. The gallbladder contained a 10x6x4 mm tan-yellow necrotic mass at the gallbladder neck. Microscopic examination of the gallbladder wall showed acute cholecystitis. The tan-yellow necrotic mass was heterotopic pancreatic tissue composed of exocrine pancreatic acini, numerous ducts, and occasional islets of Langerhans (Figure 2). No direct communication with the lumen of the gallbladder was observed. We determined that the heterotopic pancreatic tissue had obstructed the gallbladder, leading to cholecystitis. This conclusion was based on the absence of large gallstones and by post-operative ultrasound review.

DISCUSSION

Autopsy series have demonstrated pancreatic heterotopia in 0.55 to 13.7% of patients; heterotopic tissue is most commonly located in the stomach, duodenum, upper jejunum, or Meckel's diverticulum [1, 2, 3, 4]. Clinically, heterotopic pancreatic tissue has been noted in one out of every 500 patients undergoing surgery of the foregut [2]. Although commonly found in the stomach, duodenum, and jejunum heterotopic pancreatic tissue is rarely found in the gallbladder. Of 683 patients with heterotopic pancreatic tissue only 1% had pancreatic tissue located in the gallbladder wall [2, 4]. In most cases the condition is an incidental pathologic finding, but there have been reports of pancreatic heterotopia causing acute and chronic cholecystopathy, gallbladder neck obstruction, and gallbladder perforation [3, 5, 6].

Heterotopic pancreatic tissue is presumed to result from an error during embryological development. Three theories have been proposed for the mechanism of this embryologic error. In the most widely accepted theory, pancreatic tissue migrates from the pancreas by longitudinal intestinal growth during the penetration of the intestinal wall by the rudimentary pancreatic duct [7]. A second theory suggests that pancreatic tissue is separated from the pancreas during embryologic rotation and dorsal and ventral anlage fusion [8]. A recent theory contends that abnormalities in the Notch signaling system give rise to heterotopic pancreatic tissue. It has been shown that Notch signaling determines the fate of pancreatic cells through local interactions and precisely coordinates cell lines in the developing pancreas [6]. Hairy and enhancer of split 1 (Hes-1), a main effector of Notch signaling, are required for region appropriate pancreatic demarcation in the developing foregut endoderm. In Hes-1 knockout mice, pancreas specific transcription factor 1a (PTF1A) is incorrectly expressed, leading to mature heterotopic pancreatic tissue formation with exocrine, endocrine, and duct cells in the stomach, duodenum, and common bile duct. These data suggest that the Hes-1 mediated Notch pathway is required for region appropriate pancreas specification and that abnormalities during embryogenesis may contribute to heterotopic pancreatic tissue in the gallbladder [9].

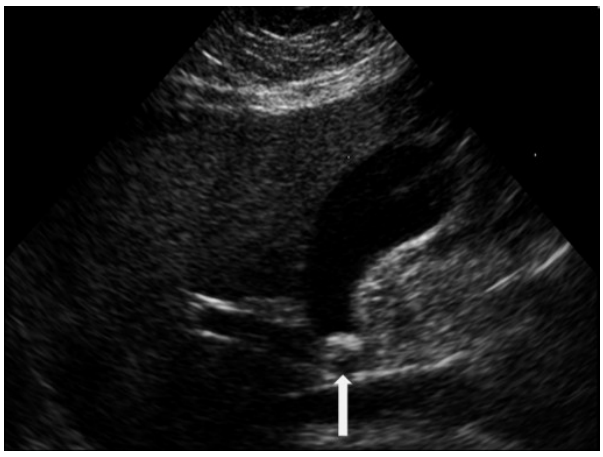

Figure 1. Right upper quadrant ultrasound showing mild gallbladder wall thickening (2 to 4 mm), no pericholecystic fluid, and a large, non-mobile, 11x8x4 mm mass with gallstone-like appearance within the gallbladder neck.

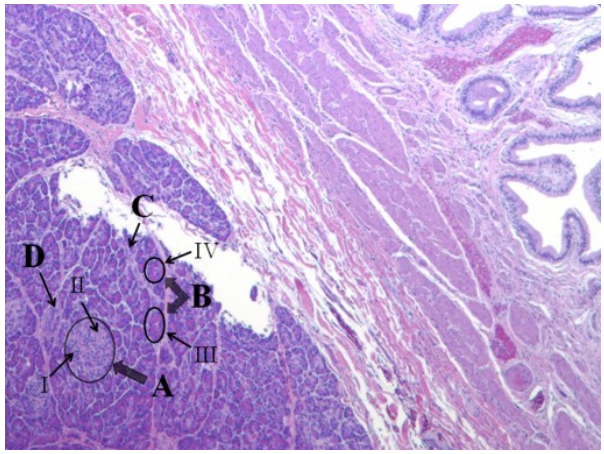

Figure 2. Hematoxylin and eosin stain of mass on gallbladder wall. A. Islet of Langerhans: I: alpha cells; II: beta cells. B. Exocrine acini: III: serous cells; IV: centroacinar cells. C. Intercalated duct. D. Interlobular duct.

The diagnosis of heterotopic pancreatic tissue is extremely challenging because of its rarity and nonspecific symptoms. Fifty percent of heterotopic pancreatic tissues found in the gallbladder arise in the neck. The lesions are usually yellow, irregular, intramural nodules that are firm to palpation [4, 10]. Heterotopic tissue in the gallbladder neck may prevent bile flow, leading to obstruction, distention, inflammation, and edema of the gallbladder wall.

In this case, the patient's obese body habitus further complicated the diagnosis because of the technical difficulty of interpreting the ultrasound. After retrospective ultrasound review it was found that this lesion was non-mobile, had the same echogenicity as the pancreas, and did not produce a shadow. These finding are most consistent with the diagnosis of pancreatic heterotopy. Despite its high resolution, ultrasound is not specific for heterotopic pancreas and it is impossible to distinguish heterotopic pancreas from other lesions such as cholesterol polyps, adenoma, and carcinoma [1, 5].

Pathologic examination in this case revealed submucosal pancreatic tissue composed of exocrine pancreatic acini, numerous ducts, and occasional islets of Langerhans. This submucosal presentation is the most common and is found in 73% of cases. Less common presentations include intramuscular (17%), and subserosal (10%) [11]. Histologically, acinar and ductal tissues are always present, whereas islet tissue is found in only one-third of cases [12]. Pancreatitis may also occur in heterotopic pancreatic tissue [13]; however, this did not occur in our patient.

Although most cases of pancreatic heterotopia are asymptomatic and remain undiagnosed until incidental discovery on pathologic examination, clinically important conditions such as acute or chronic cholecystopathies can also occur. Even when heterotopic pancreatic tissue does cause symptoms, the preoperative diagnosis is difficult primarily due to nonspecific clinical presentations. When operating in the abdominal cavity, it is important to be aware of pancreatic heterotopia and excision is indicated when lesions are encountered. Although heterotopic pancreatic tissue is usually an incidental finding, one should not exclude it in the differential diagnosis of symptomatic gallbladder disease without definitive clinical findings.

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Conflict of interest The authors have no potential conflicts of interest

References

1. Kondi-Paphiti A, Antoniou AG, Kotsis T, Polimeneas G. Aberrant pancreas in the gallbladder wall. Eur Radiol 1997;7:1064-1066. [PMID 9265676]

2. Barbosa J, Dockerty M, Waugh J. Aberrant pancreas in the gallbladder wall. Surgery, Gynecology, and Obstetrics 1946;82:527-542.

3. Ben-Baruch D, Sandbank Y, Wolloch Y. Heterotopic pancreatic tissue in the gallbladder. Acta Chir Scand 1986;152:557-558. [PMID 3788403]

4. Dolan RV, ReMine WH, Dockerty MB. The fate of heterotopic pancreatic tissue. A study of 212 cases. Arch Surg 1974;109:762-765. [PMID 4420439]

5. Monig SP, Selzner M, Raab M, Eidt S. Heterotopic pancreas. A difficult diagnosis. Dig Dis Sci 1996;41:1238-1240. [PMID 8654158]

6. Shiwani MH, Gosling J. Heterotopic pancreas of the gallbladder associated with chronic cholecystitis. Jop 2008;9:30-32. [PMID 18182740]

7. Pang LC. Pancreatic heterotopia: a reappraisal and clinicopathologic analysis of 32 cases. South Med J 1988;81:1264-1275. [PMID 3051429]

8. Bhana BD, Chetty R. Heterotopic pancreas--an unusual cause of cholecystitis. S Afr J Surg 1999;37:105-107. [PMID 10701341]

9. Artavanis-Tsakonas S, Rand MD, Lake RJ. Notch signaling: cell fate control and signal integration in development. Science 1999;284:770-776. [PMID 10221902]

10. Palmer ED. Benign intramural tumors of the stomach: a review with special reference to gross pathology. Medicine (Baltimore) 1951;30:81-181. [PMID 14826371]

11. Jeng KS, Yang KC, Kuo SH. Malignant degeneration of heterotopic pancreas. Gastrointest Endosc 1991;37:196-198. [PMID 2032610]

12. Hara M, Tsutsumi Y. Immunohistochemical studies of endocrine cells in heterotopic pancreas. Virchows Arch A Pathol Anat Histopathol 1986;408:385-394. [PMID 2418578]

13. Qizilbash AH. Acute pancreatitis occurring in heterotopic pancreatic tissue in the gallbladder. Can J Surg 1976;19:413-414. [PMID 974927]