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Heterotopic Pancreas: Report of Two Cases with unique presentation and review of literature.

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ABSTRACT

Heterotopic pancreas (HP) is defined as the presence of pancreatic tissue outside its normal location, lacking ductal or vascular continuity with the main gland. Though pancreatic heterotopia is usually incidental, it may become clinically evident when complicated by pathological changes such as inflammation, bleeding, obstruction. In this report of two cases, a 42-year female with chronic cholecystitis with mural heterotopic pancreatic tissue is described, along with a 35-year female with abdominal pain caused by heterotopic pancreatic tissue located in proximal jejunum. In majority of reported cases, HP has been found in stomach, duodenum, upper jejunum, whereas its presence in the gallbladder is very rare. Awareness of this condition may provide insights on its clinical significance

INTRODUCTION

Pancreatic heterotopia is defined as pancreatic tissue located outside the gland without any anatomical or vascular connection to the pancreas. [1] Ectopic pancreatic tissue in gastrointestinal tract is found in 0.6-13.7% of autopsy series and the most common localizations are stomach (25-60%) and duodenum (25-35%). [2] It is often found incidentally, while it may sometimes present symptomatically, and the manifestations vary depending on the location of the lesion. In 85% to 90% of reported cases, HP has been found in stomach, duodenum, upper jejunum, whereas its presence in the gallbladder is very rare. [3] We report two cases of heterotopic pancreatic lesions with unusual presentations - one in the jejunum and the other in the gall bladder with accompanying review of the current literature.

CASE REPORT

Case 1

A 42-year female presented to the surgical outpatient clinic with a history of right upper quadrant abdominal pain for three months, associated with vomiting off and on. Upon general examination, she was afebrile, normotensive and had no jaundice. Per abdominal examination revealed tenderness in the right hypochondrium. Routine blood investigations and Liver Function Tests were normal. Ultrasonographic examination of the whole abdomen showed no abnormality, except for cholelithiasis [Figure 1].

Laparoscopic cholecystectomy was performed. On gross examination, the gallbladder measured 6 cm in length and 2.3 cm in circumference, with a wall thickness ranging from 0.2 to 0.4 cm. The serosa was unremarkable. On cutting open, the mucosa was velvety, flattened and focally ulcerated. A single yellowish black stone was noted.

Microscopic examination revealed chronic cholecystitis and a 7 mm-wide area of heterotopic pancreatic tissue within the gallbladder wall in the fundus area, composed of lobules of exocrine pancreatic acini and an occasional duct [Figure 2]. Islets of Langerhans were not seen. The remaining sections showed features of chronic cholecystitis.

The diagnosis was thereby established as chronic cholecystitis with HP. All symptoms disappeared following cholecystectomy and the patient recovered completely.

Case 2

A 35-year female presented with a history of constant abdominal pain and episodes of hematemesis for last one month. Physical examination showed tenderness in the left upper

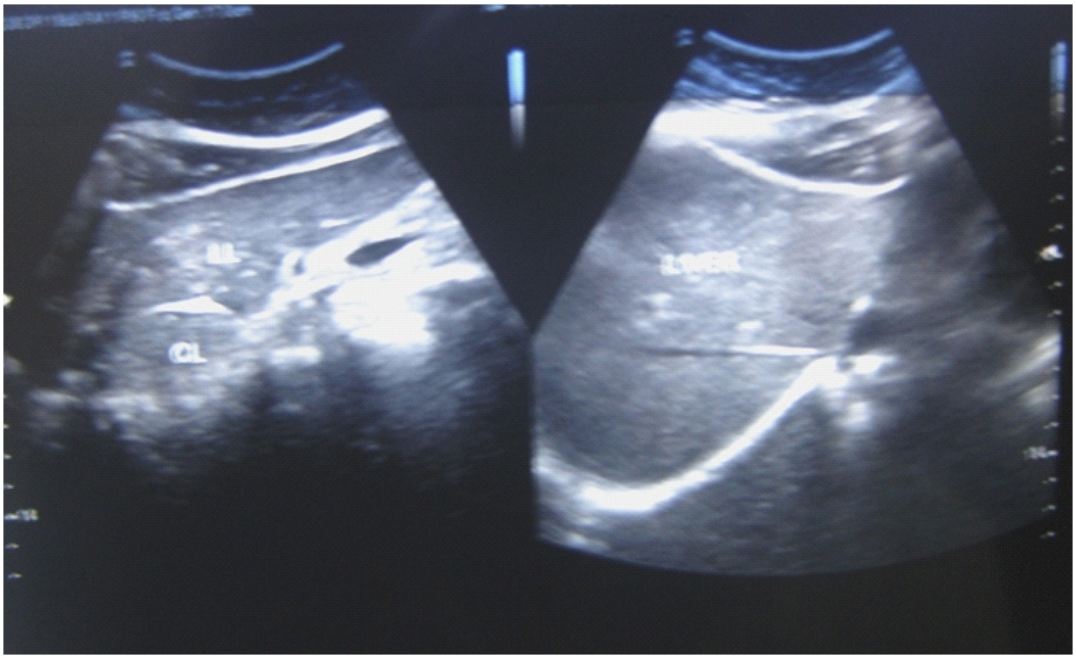

Figure 1

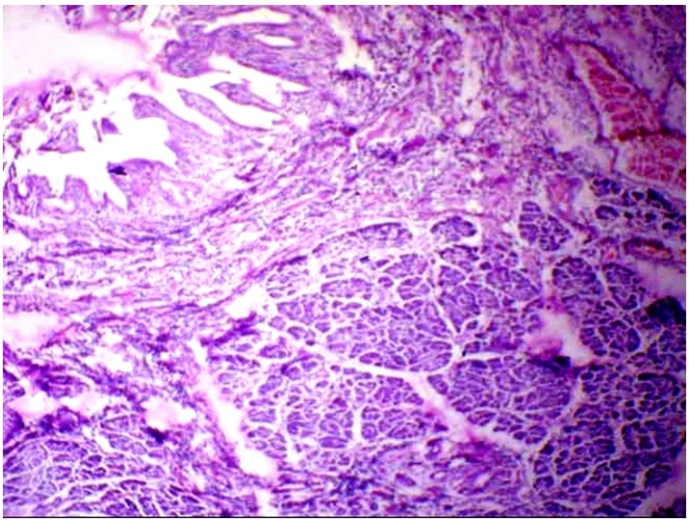

Figure 2

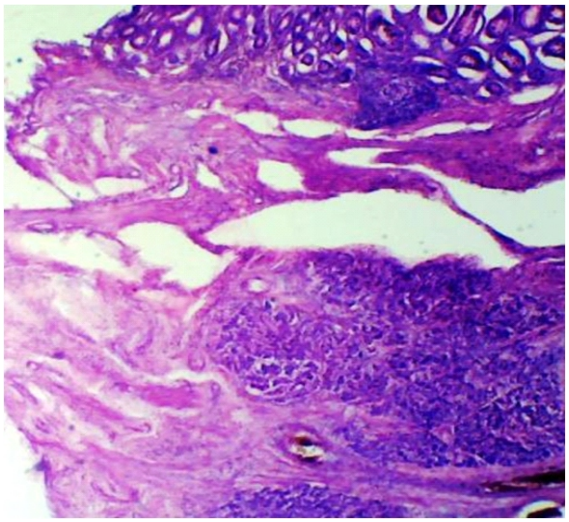

Figure 3

Figure 4

quadrant and epigastrium, without any rebound tenderness. Laboratory tests revealed leukocytosis and anemia. Abdominal ultrasound was unrevealing. Upper GI endoscopy revealed a sessile polypoid mass in proximal jejunum, partially obstructing the lumen. Then the patient underwent polypectomy and segmental resection and sent for histopathological examination.On gross examination a loop of jejunum measuring 12 cm. in length was present, cut surface of which showed a sessile polyp measuring 4 X 2.5 cm, surface smooth, no ulceration present. Mucosal rugosity proximal and distal to the polyp was normal. Subsequent histopathology of the polyp revealed a heterotopic pancreatic tissue located predominantly at submucosa without extension into the mucosa [Figure 3]. The follow-up upto 6 months was uneventful.

DISCUSSION

Heterotopic rests of pancreatic tissue are frequently identified in the upper GI tract and, despite earlier speculation that these

lesions resulted from metaplasia in response to chronic injury, they likely represent developmental abnormalities. [4]

Embryologically, the pancreas develops from invaginations of endodermal tissue in the primitive duodenum. The ventral aspect forms the head and the dorsal aspect forms the body and tail of the pancreas. The misplacement theory proposes that, during rotation of the foregut, several elements of the primitive pancreas become separated and eventually form mature pancreatic tissue along the length of the gastrointestinal tract. [5] Despite its congenital origin, an HP clinically manifests itself in older adults.Mural heterotopias are more likely to produce abdominal pain, gastric outlet obstruction, intussusception, stricture, or bleeding than those confined to the mucosa. [6]

Pancreatic heterotopias are typically composed of lobules or nests of pancreatic acini and ductules. Pancreatic islets are seen in one third of cases; however, polyps composed entirely of endocrine cells are uncommon. The acini within pancreatic heterotopias contain polarized cells with basally oriented nuclei and amphophilic or basophilic granular cytoplasm. Single lobules of acini are more commonly seen in mucosa-based heterotopias, whereas mural lesions may be grossly and histologically indistinguishable from the normal pancreas. [4]Microscopically, HP has been classified into three types by von Heinrich Type 1: Ectopic tissue with acini, ducts, and islets of Langerhans - Type 2: Ectopic tissue containing only a few acini and ducts, with absent endocrine elements in complete arrangement; Type 3: Ectopic tissue with only proliferating excretory ducts and absent exocrine acini and endocrine elements. [7] Patients with HP can be asymptomatic, or present with abdominal pain and distension. In addition, it can manifest clinically in some rare diseases of the pancreas including pancreatitis, islet cell tumor, pancreatic carcinoma, and pancreatic cyst. [8]

There is no specific examination and diagnostic method at present, and it is difficult to diagnose this disease definitely before laparotomy. [6] But the complications related to HP, such as small bowel obstruction, pancreatitis or GI bleeding, may also occur as presenting symptom as in case 2.

Despite the frequent occurrence of HP in the stomach, duodenum and upper jejunum, occurrence in the gallbladder is extremely rare. In HP of gall bladder there is a higher incidence of female patients between 40 and 50 years of age, [9] similar to our case (case 1).

HP in the gallbladder is very rarely symptomatic. In most reported cases, it is an incidental pathological finding and coexists with gallstones [9] as in our case 1. Symptomatology and clinical findings in most cases suggest gallbladder disease, mainly calculus cholecystitis. [9] As there is no submucosal layer in the gall bladder, HP is usually seen in the muscularis similar to our case. According to recent studies, abnormalities in the Notch signaling system, especially in Hes-1 expression during embryogenesis may also contribute to the formation of HP of the gallbladder. [10]

CONCLUSION

In conclusion, the incidence of HP is low, more so in gallbladder and preoperative diagnosis is difficult. Awareness of this under-reported condition may facilitate its recognition and this, in turn, may shed more light on its clinical significance.

ABREVIATIONS

HP- Heterotopic pancreas

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