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Case Report

Pancreatic Heterotopia

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Keywords
Ectopic Pancreas
Heterotopia
Intestinal obstruction
Intussusception
Pain abdomen

Abbreviations
GI - Gastro intestinal
PH - Pancreatic heterotopia
USG - Ultrasonography

Abstract

Pancreatic heterotopia (PH) is a rare congenital anomaly that is also known as ectopic pancreas or pancreatic choristoma. It is usually asymptomatic but rarely may cause abdominal pain, lower gastrointestinal bleeding or intussusception. This report describes two cases of PH: a 9-month-old boy presented with intussusception and a 6-year-old girl presented with a jejuno-colic inflammatory mass. Surgical excision of PH cured both of them.

INTRODUCTION

Pancreatic heterotopia (PH) is a rare congenital anomaly that mostly remains silent for several years. It is also known as ectopic pancreas or pancreatic choristoma. Rarely PH may become symptomatic, presenting with abdominal pain. The common age of presentation is usually the third decade. In the pediatric age group, it forms the pathological lead point of intussusception.(1)

The pathogenesis of PH is explained by the misplacement theory. During the embryonic rotation and fusion of the ventral and dorsal pancreatic buds, some of the pancreatic tissue may get separated from the main gland, and gets attached to the adjoining structures such as the stomach, small bowel, colon and Meckel's diverticulum. This ectopic tissue has its own independent vascularity and ductal com-ponents. The degree of maturation of the ectopic tissue is variable.(2,3) In PH, the pancreatic tissue is noted within the submucosa of the gastrointestinal (GI) tract, making its endoscopic diagnosis difficult. Gastrointestinal stromal tumor (GIST), leiomyoma and submucosal lymphoid hyperplasia or tumors are the closest differential diagnosis of PH.(2,3) Endoscopy or endoscopic ultra sound (EUS) combined with fine-needle aspiration cytology may be helpful in the diagnosis. Surgical excision is the only effective treatment of PH. In this report we share our experience with 2 such cases of PH.

CASE REPORTS

Case 1

A 9-month-old male child presented with non-bilious vomiting and excessive crying. On clinical examination, the abdomen was soft and non-tender without any palpable mass. Plain x-ray of

the abdomen was non-contributory. An ultrasonography (USG) revealed ileo-colic intussusception. After a failed attempt of USG-guided hydrostatic reduction, it was manually reduced at laparotomy (Fig. 1A), where upon a palpable nodule of 2 x 1.5 cm was noted at its apex. The lead-point was located at the distal ileum at about 10 cm proximal to the ileo-cecal junction. (Fig. 1B) The involved segment of the ileum was resected and primary end-to-end bowel anastomosis was done. Post-operative recovery was uneventful.

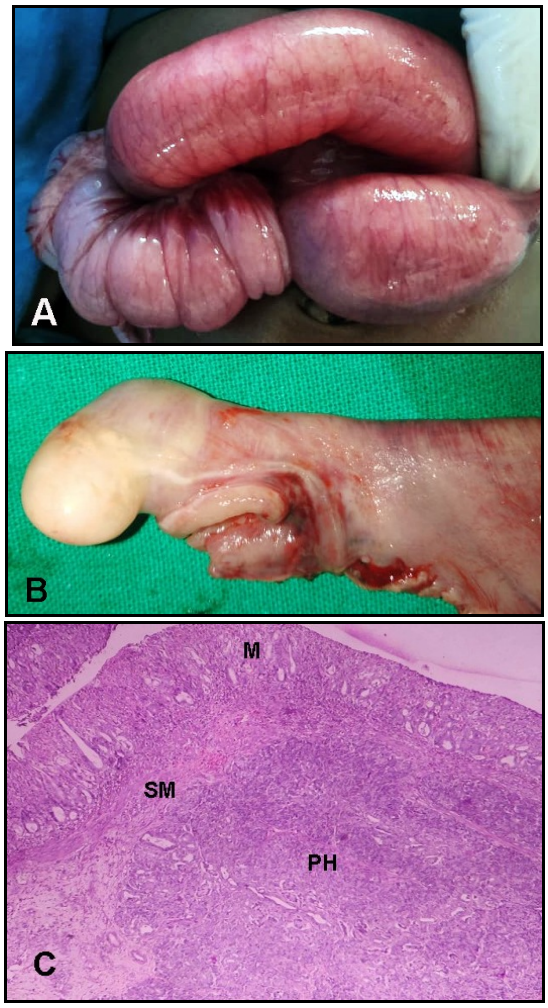

Fig 1. *Pancreatic heterotopia (case 1): (A) Intra-operative photograph showing ileo-colic intussusception, (B) pathological lead point at the apex of the reduced intussusception, (C) Histopathology of the apical nodule showing pancreatic heterotopia (PH) deeper to the mucosa (M) and submucosa (SM) Magnification 20X, Hematoxylin-Eosin staining*

Histopathology of the resected nodule (Fig. 1C) revealed presence of heterotopic pancreatic tissue in the submucosa along with focal ulceration of the overlying mucosa. The child was well on follow-up at 8 months.

Case 2

A 6-year-old girl presented with insidious onset of abdominal pain and vomiting. On clinical examination, a vague mass was felt in the upper abdomen. Diagnostic USG was not helpful due to interference of bowel gas. Plain radiograph was also non-contri butory. Persistent pain and the palpable mass prompted an exploratory laparotomy.

Intraoperatively, the proximal jejunum was found to be adherent with the transverse colon and the omentum. (Fig 2A) Further dissection revealed a nodule of 2 x 1.5 cm in the ante-mesenteric border of the jejunum. The nodule was excised by wedge resection of bowel and the rent was repaired. Histology confirmed the nodule as PH. (Fig 1C) Retrospectively, a diagnosis of ectopic pancreas causing inflammatory mass was made. The child was doing well on follow-up at 6 months.

DISCUSSION

PH is usually present in the submucosa of the upper GI tract, including the stomach, duodenum and less commonly in the jejunum, ileum and Meckel's diverticulum. It is rarely found in the esophagus, liver, gallbladder, biliary tree, spleen, omentum, lungs, mediastinum, Fallopian tubes and the umbilicus.[4] Comparatively PH is rarer in the lower GI tract than the upper tract. PH may also be complicated by inflammation and stricture. Ductal adenocarcinoma arising from rectal PH has been described.[1] Incidental diagnosis of PH during laparotomy is well known. In our cases, PH was identified in the ileum and the jejunum in one case each. These are less common sites according to the literature.

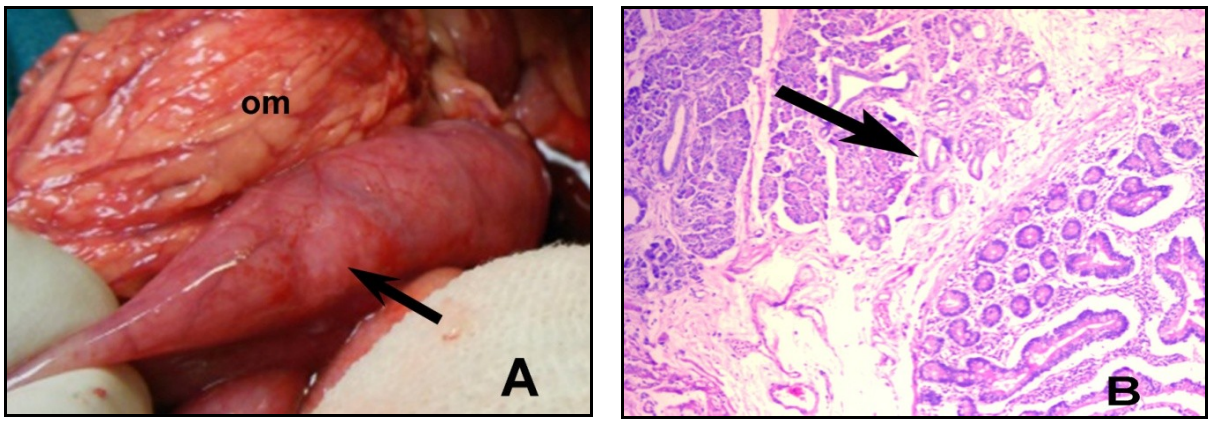

Fig 2. *Pancreatic heterotopia (case 2): (A) Intra-operative photograph showing the jejunal nodule (arrow) and the adherent omentum (om); (B) Histopathology of the nodule showing typical pancreatic acini (arrow) in the submucosa of the Jejunum (Magnification 100X, Hematoxylin-Eosin staining)*

PE may cause symptoms related to mechanical complications (e.g. gastric outlet obstruction, intus susception), bleeding from the ulcerated mucosa or metaplasia leading to malignancy in the late adulthood. Even though exceedingly rare, neoplasia arising from PH have been described even in children.[1] In our patients, one presented with intussusception and another with inflammatory bowel obstruction.

It is not possible to differentiate between normal and ectopic pancreatic tissue in histology as PH may have all the normal components (acini, ducts, islet cells) of pancreas. PH is also prone for common diseases of the orthotopic pancreas such as acute pancreatitis and pseudocysts.[5]

Persano et al. reported 14 pediatric patients with PH, of whom 50% were asymptomatic. Older children with PH were more often symptomatic than infants and toddlers. The most common presenting symptoms were melena and recurrent abdominal pain. PH was more frequently recognized during emergency surgery than with elective operations.[6]

Preoperative diagnosis of PH is often difficult. If identified before surgical management, it will facilitate planning of operative procedures including minimal access or endoscopic procedures. When located in the stomach or duodenum, PH can be suspected based on the typical findings of upper GI series. In contrast imaging PH is seen as a mass with broad base and smooth surface that is characteristic of extra-mucosal intramural tumors. Umbilication (visualization of a contrast-filled pit at the center of the lesion) is thought to represent the ductal remnant of PH. In magnetic resonance imaging (MRI) PH is seen as hyper- or iso-intense lesion in comparison with the orthotopic pancreas in unenhanced T1-weighted images and is seen as iso- or hypo-intense lesions in the T2-weighted images. On dynamic MRI, PH appears as an iso-intense lesion in comparison to the orthotopic pancreas in the arterial phase image. Radiologic findings of PH distal to the duodeno-jejunal junction are non-specific.[7]

In children with dyspepsia, upper GI endoscopy may reveal PH. As the lesion is located in the submucosal layer, endoscopy can be useful in the diagnosis as well as therapy (endoscopic resection whenever feasible).[8]

Another rare presentation of PH is an antenatally detected intra-abdominal cyst, postnatally complicated with unexplained of neonatal hypoglycemia. Hypoglycemia unresponsive to medical treatment

will improve on excision of the cyst containing PH.[9] It could be a differential diagnosis of acute cholangitis or obstructive jaundice when the ectopic tissue is located at the ampulla of Vater.[10]

Heinrich's histological classification of PH has no direct implications in the clinical management. It is very difficult to diagnose PH purely based on the macroscopic appearance during surgery, and hence histological confirmation is mandatory.

Table 1: Heinrich's classification of Pancreatic Heterotopia*

Type	Histological Description
1 (Complete)	Presence of typical pancreatic tissue comprising of acini, ducts, and islet cells
2 (Canalicular)	Presence of only ducts
3 (Exocrine)	Presence of only acinar tissue
4 (Endocrine)	Presence of only islet cells

* *Ref. Heinrich[11]*

CONCLUSION

PH is an uncommon entity, especially as a symptomatic lesion causing pediatric acute abdomen. It should be considered in the differential diagnosis of recurrent abdominal pain, lower GI bleed or intussusception. Diagnosis of PH would require histopathological confirmation after excision.

REFERENCES

[1] Yang ZH, Gao JB, Yue SW, Yang XH, Guo H. Synchronous ectopic pancreatoblastoma in a child: a case report. J Korean Med Sci. 2011 Jun; 26(6): 832-5.

[2] Chin NH, Wu JM, Chen KC, Lee TH, Lin CK, Chung CS. Pancreatic heterotopia in the small bowel: a case report and literature review. Pancreas. 2022 Jul 1; 51(6):700-4.

[3] Nguyen TL, Kapur S, Haerer SC, Gurda GT, Folkers ME. Unusual tissue - Unusual issue: Pancreatic heterotopia presenting as gastric outlet obstruction. Case Rep Gastro enterol. 2021 Mar 11; 15(1): 338-343.

[4] Zhou Y, Zhou S, Shi Y, Zheng S, Liu B. Endoscopic submucosal dissection for gastric ectopic pancreas: a single-center experience. World J Surg Oncol. 2019 Apr 16; 17 (1): 69.

[5] Tang XB, Liao MY, Wang WL, Bai YZ. Mesenteric heterotopic pancreas in a pediatric patient: A case report and review of literature. World J Clin Cases. 2018 Nov 26; 6 (14): 847-853.

[6] Persano G, Cantone N, Pani E, Ciardini E, Noccioli B. Heterotopic pancreas in the gastrointestinal tract in children: a single-center experience and a review of the literature. Ital J Pediatr. 2019 Nov 9; 45(1): 142.

[7] Kung JW, Brown A, Kruskal JB, Goldsmith JD, Pedrosa I. Heterotopic pancreas: typical and atypical imaging findings. Clin Radiol. 2010 May; 65(5): 403-7.

[8] Zhong YS, Shi Q, Yao LQ, Zhou PH, Xu MD, Wang P. Endoscopic mucosal resection/endoscopic submucosal dissection for gastric heterotopic pancreas. Turk J Gastro enterol. 2013; 24(4): 322-9.

[9] Seymore NM, Zoghbi B, Sotelo-Avila C, Farmakis SG. Pancreatic heterotopia in a neonatal abdomino-pelvic cyst. Pediatr Radiol. 2019 Mar; 49(3): 415-418.

[10] Christodoulidis G, Zacharoulis D, Barbanis S, Katsogridakis E, Hatzitheofilou K. Heterotopic pancreas in the stomach: a case report and literature review. World J Gastroenterol. 2007 Dec 7; 13(45): 6098-100.

[11] Heinrich H. Ein Beitrag zur Histologie des sogenannten akzessorischen Pankreas. Virchows Arch A Pathol Anat Histopathol. 1909; 198: 392–440.

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