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Various phenotypes of ectopic pancreatic tissue in six children

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Abstract

Aims: Ectopic pancreas, an infrequently documented condition within the pediatric demographic, is often asymptomatic. When clinical manifestations do occur, their severity is contingent upon the location, size, and involvement of the adjacent mucosa.

Methods: Patients with heterotopic pancreas from one single institution were retrospectively analyzed, between August 2020 and January 2024.

Results: This article delineates six cases of heterotopic pancreas. Five of the six cases presented both ectopic pancreatic tissue and ectopic gastric tissue. One case exhibited duodenal localization (duodenum I), with the patient being admitted for upper gastrointestinal bleeding. An endoscopic evaluation subsequently diagnosed a perforated duodenal ulcer, and histopathological examination of the biopsy confirmed the presence of ectopic pancreatic tissue. The remaining five cases were identified at the site of Meckel's diverticulum.

Conclusion: The identification of ectopic pancreatic tissue continues to pose a diagnostic challenge, as there are currently no specific paraclinical investigations available for this purpose.

Keywords: Children; Heterotopic pancreas; Gastrointestinal bleeding; Meckel's diverticulum.

Introduction

Ectopic pancreas refers to the presence of pancreatic tissue in structures devoid of anatomical, vascular, or neuronal connections to the gland itself. This condition is infrequently reported in the pediatric population, unlike in adults [1]. It is most commonly identified in the stomach, duodenum, or jejunum, followed by locations such as Meckel's diverticulum or ileum [2]. Cases have also been documented in the gallbladder, Vater's ampulla, umbilicus, or even in the brain [3]. The incidence of ectopic pancreas is likely underestimated since most patients are asymptomatic; frequently, the detection of ectopic pancreas is incidental during histopathological examination. In symptomatic patients, the clinical presentation is dependent on the location of the ectopic pancreatic tissue. Abdominal pain is a prevalent symptom, occasionally accompanied by nausea or vomiting, hematemesis, or melena. One of the most significant complications associated with ectopic pancreatic tissue is the potential for malignancy [4]. Paraclinical investigations may reveal anemia and elevated levels of serum amylase and lipase [5]. This paper presents six cases of ectopic pancreatic tissue: one located in the duodenum and five located in Meckel's diverticulum, in association with ectopic gastric tissue.

Materials and methods

Following histopathological evaluation of intraoperative specimens sent for examination to the Pathological Anatomy Service of the Grigore Alexandrescu Emergency Hospital for Children between August 2020 and January 2024, six cases of ectopic pancreatic tissue were identified. It was recorded the patients' age, sex, presenting symptoms, diagnosis, hemoglobin on admission, and location of ectopic pancreatic tissue. Regarding the symptoms present at the time of admission, only patients whose symptoms were attributed to the presence of ectopic tissue were classified as symptomatic. Incidental identification of ectopic tissue was also noted.

Results

Among the six cases (3 boys and 3 girls) identified over a period of three years and four months, one case involved only ectopic pancreatic tissue located in the duodenum I, while five cases involved both ectopic pancreatic tissue and ectopic gastric tissue located in Meckel's diverticulum (Table 1). Four cases were symptomatic, while two cases involved incidental identification of ectopic pancreatic tissue. Only two cases were associated with anemia, requiring intravenous iron therapy and/or blood transfusion. Two cases underwent preoperative imaging (abdominal ultrasound) suggestive of Meckel's diverticulum. For all six patients, excision of ectopic pancreatic tissue and surgical resection of Meckel's diverticulum were performed (Table 1 near here). According to the Heinrich classification, modified by Gaspar-Fuentes et al., there are four histological types of ectopic pancreatic tissue: type I – presenting acini, ducts, and islets (complete heterotopia), type II – presenting only ducts (canalicular heterotopia), type III-presenting only acini (exocrine heterotopia), and type IV – presenting only islets (endocrine heterotopia) [6]. Five out of the six cases evaluated belonged to type I and one case belonged to type III (Figures 1-4).

In the literature, bleeding in Meckel's diverticulum is frequently mentioned, often caused by ectopic gastric tissue. However, upper gastrointestinal bleeding caused by a perforated duodenal ulcer through ectopic pancreatic tissue is extremely rare, with only a few cases described to date. One such case involved a 13-year-old boy who presented to our clinic with a sudden onset of symptoms, including hematemesis and melena. It was noted that he had been experiencing moderate abdominal pain for approximately two weeks prior to the admission and also weight loss, approximately 10 kg due to post-prandial fulness and inability to eat. Endoscopic intervention was performed 48 hours after admission, during which a large blood clot was identified in the stomach and an adherent clot was found in the duodenum I. On the eleventh day of hospitalization, the patient experienced hemodynamic instability, expelling fresh red blood through the nasogastric tube, necessitating an immediate exploratory laparotomy. Ulceration with a detached clot and loss of wall integrity with active bleeding was observed on the posterior aspect of the duodenum I. Duodenal resection was performed 2 cm distal to the pylorus up to the level of duodenum III. Biliary leakage was observed at the level of the main bile duct and pancreatic duct, so stenting was performed for both structures. Biopsy revealed the presence of ectopic pancreas in the duodenum I. Postoperative recovery was slow but favorable. Endoscopic stents were removed after a period of five months. The patient received blood transfusions and several iron intravenous infusions. Based on the recommendation of the pediatric gastroenterologist, an enteral supplement with a hypercaloric formula was administered. Following this intervention, the patient's weight curve showed a significant upward trend, with a weight gain of approximately 10 kg over 8 months. The patient was initially hospitalized for one month and subsequently followed up on a monthly or bi-monthly basis for approximately one year. At the most recent assessment, the patient had a weight of 40 kg (Z-score-1.546), a height of 160 cm (Z-score-0.732), and a BMI of 15.6 kg/m² (Z-score-1.987).

Discussion

Although ectopic pancreatic tissue often progresses asymptomatically, clinical manifestations may occur due to inflammation, bleeding, obstruction, or malignancy of the affected structure [2]. The potential for malignant transformation of ectopic pancreatic tissue remains the most significant risk for these patients. Notably, cases of ectopic pancreatoblastoma have been reported in the pediatric population [7]. Furthermore, all pathologies affecting the pancreas, such as pancreatitis, pancreatic cysts and pancreatic cancer, can also manifest in ectopic pancreatic tissue. The etiology of heterotopic pancreas remains unclear. It is hypothesized that small portions of pancreatic tissue become separated early in fetal development during the rotation of the foregut and fusion of the dorsal and ventral parts of the pancreas, subsequently developing in ectopic locations. Another theory, pancreatic metaplasia of endodermal tissue, may explain the presence of heterotopic pancreas in organs anatomically distant from the pancreas [8]. There are no specific paraclinical investigations for identifying ectopic pancreatic tissue, rendering even the most commonly reported locations (stomach, duodenum, jejunum) a diagnostic challenge [9]. However, endoscopic examination may be useful in cases where ectopic pancreatic tissue is located in the submucosa or muscular layer of the stomach, duodenum I, or duodenum II [10]. In gastric localization, ectopic pancreatic tissue often appears as a nodule in the submucosa of the gastric antrum, frequently situated on the greater curvature of the stomach [11]. Symptoms typically occur in lesions larger than 2 cm [12]. Endoscopic Ultrasound (EUS) can also be beneficial in certain cases for differentiating ectopic pancreatic tissue from other tumor formations in the gastric submucosa [13]. The use of Video Capsule Endoscopy (VCE) may represent an imaging option for some patients, given its high sensitivity and specificity in detecting sources of gastrointestinal bleeding [14]. Thus, the diagnosis is often established following histopathological examination of intraoperative specimens when complications such as gastrointestinal bleeding, obstruction, or intussusception arise. In suspected cases of Meckel's diverticulum, a range of paraclinical investigations can be performed, including technetium-99m pertechnetate scintigraphy, abdominal Computer Tomography (CT) with contrast agent, abdominal Magnetic Resonance Imaging (MRI), abdominal ultrasound, etc. These investigations can aid in establishing a preoperative diagnosis [15]. There are a number of limitations in this paper. First, statistical elements were not presented because the analyzed time period was very short and the number of cases was very small. Additionally, five of the six cases presented both ectopic pancreatic tissue and ectopic gastric tissue. However, we consider their presentation appropriate given the low incidence of ectopic pancreas in children and the fact that

Table 1: Patients' characteristic.

Patient	Sex	Age	Admission Symptoms	Initial diagnosis	Final diagnosis	Hemoglobin on admission	Localization of heterotopic tissue	Histopathological finding
1	M	13 years	Abdominal pain, haematemesis, Melena	Upper gastrointestinal bleeding	Perforated duodenal ulcer	HGB 7.1 g/dl	Duodenum	Pancreatic heterotopic tissue
2	F	11 years	Abdominal pain, vomiting	Acute surgical abdomen	Perforated Meckel's diverticulum	HGB 13.6 g/dl	Meckel's Diverticulum	Pancreatic and gastric heterotopic tissue
3	F	11 years	Abdominal pain, hematochezia	Lower gastrointestinal bleeding	Ulcerated Meckel's Diverticulum	HGB 7.5 g/dl	Meckel's Diverticulum	Pancreatic and gastric heterotopic tissue
4	F	5 years	Abdominal pain	Meckel diverticulitis (ultrasound finding, with normal size of the appendix)	Meckel diverticulitis	HGB 11.8 g/dl	Meckel's Diverticulum	Pancreatic and gastric heterotopic tissue
5	M	15 days	! Incidental finding of ectopic tissue Right scrotal swelling, vomiting	Right inguinal hernia	Right inguinal hernia; Intraoperative finding of Meckel's diverticulum; Incidental finding of ectopic tissue	HGB 14.2 g/dl	Meckel's Diverticulum	Pancreatic and gastric heterotopic tissue
6	M	9 years	! Incidental finding of ectopic tissue Asymptomatic	Meckel's Diverticulum (incidental finding during appendicectomy performed before)	Meckel's Diverticulum; Incidental finding of ectopic tissue	HGB 13 g/dl	Meckel's Diverticulum	Pancreatic and gastric heterotopic tissue

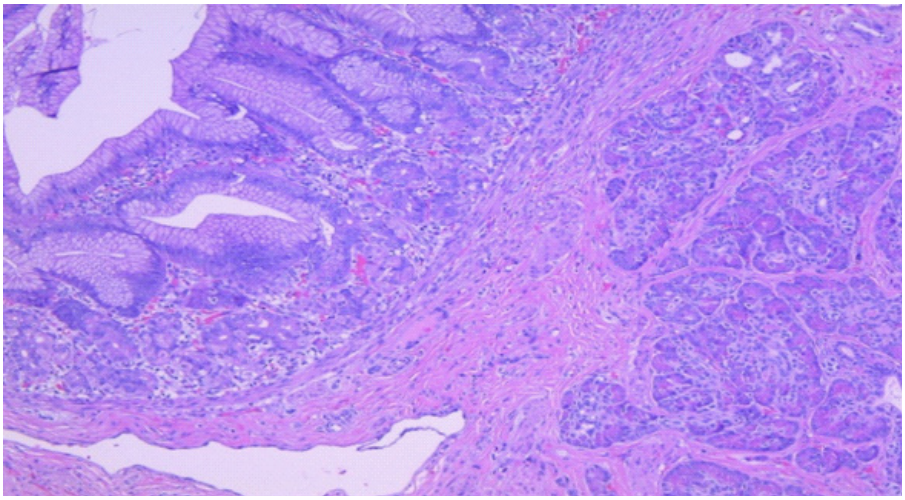

Figure 1: Detail with exocrine serous acini in the ectopic pancreatic tissue, HE, 200x.

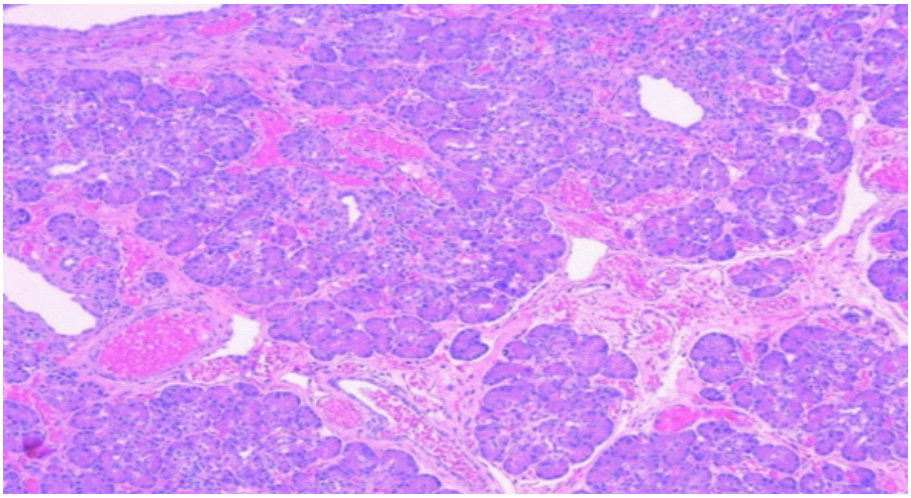

Figure 2: Ectopic pancreatic tissue within the submucosa of a Meckel diverticulum. In the left upper half of the image, the lining of the diverticulum is formed by oxyntic glands and gastric foveolar epithelium, HE, 100x.

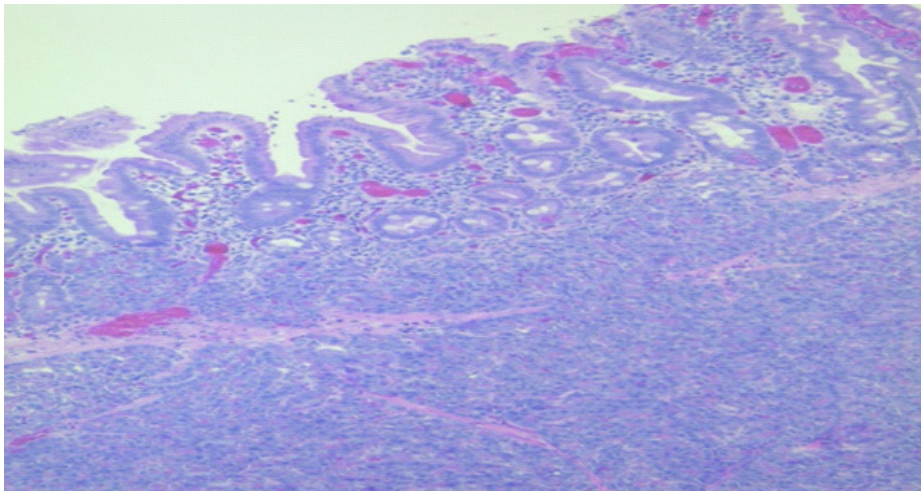

Figure 3: Duodenal wall with flattened mucosa and pancreatic tissue within the basal part of the mucosa and submucosa, HE, 100x.

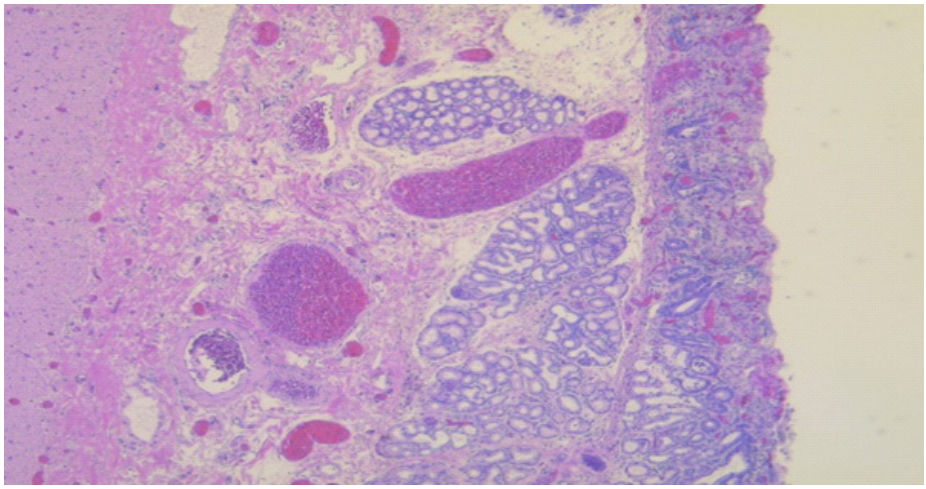

Figure 4: Duodenal wall with erosions and regenerative epithelium in the adjacent sections with ectopic pancreas, HE, 50x.

establishing a diagnosis of ectopic pancreas remains extremely challenging. In conclusion, ectopic pancreatic tissue should be considered in patients presenting with severe symptoms, such as gastrointestinal bleeding, as well as in those with milder manifestations, such as dyspeptic syndrome. Ectopic pancreatic tissue in the pediatric population remains a rare condition, and its growth with age may explain why symptoms typically appear in adulthood. The recommended therapeutic intervention for symptomatic patients remains surgical resection.

Declarations

Ethical consideration: The study commenced after obtaining approval from the Institutional Ethics Committee (IEC 21585/15.07.2024). Informed written consent was obtained from the guardian/parents.

Conflict of interest: The authors declare no conflicts of interest.

References

1. Xiao WD, Chen W, Yang H. Heterotopic Pancreas within Meckel's diverticulum with Obscure then Massive gastrointestinal Bleeding in a 12-year-old Child: case Report and Review of the Literature. J Int Med Res. 2009; 37: 967-972. doi: 10.1177/147323000903700343.

2. Trifan A, Târcoveanu E, Danciu M, Huţanaşu C, Cojocariu C, et al. Gastric heterotopic pancreas: An unusual case and review of the literature. J Gastrointestin Liver Dis. 2012; 21: 209-212.

3. Abdelgabar A, Wojciechowski M, Van de Vijver E, Ruppert M, Hubens G, et al. Ectopic pancreatic tissue at the umbilicus in childhood: a case report and review of the literature. Acta Chir Belg. 2013; 113: 452-454. doi: 10.1080/00015458.2013.11680963.

4. Henderson L, Nour S, Dagash H. Heterotopic pancreas: A rare cause of gastrointestinal bleeding in children. Dig Dis Sci. 2018; 63: 1363-1365. doi: 10.1007/s10620-018-4981-z.

5. Kung JW, Brown A, Kruskal JB, Goldsmith JD, Pedrosa I. Heterotopic pancreas: typical and atypical imaging findings. Clin Radiol. 2010; 65: 403-407. doi: 10.1016/j.crad.2010.01.005.

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; 45: 142. doi: 10.1186/s13052-019-0738-3.

7. Yang X, Wang X. Imaging findings of pancreatoblastoma in 4 children including a case of ectopic pancreatoblastoma. Pediatr Radiol. 2010; 40: 1609-1614. doi: 10.1007/s00247-010-1776-6.

8. Ormarsson OT, Gudmundsdottir I, Mårvik R. Diagnosis and treatment of gastric heterotopic pancreas. World J Surg. 2006; 30(9): 1682-1689. doi: 10.1007/s00268-005-0669-6.

9. Juricic M, Djagbare DY, Carmassi M, Panait N, Faure A, et al. Heterotopic pancreas without Meckel's diverticulum in children as unique cause of gastrointestinal bleeding: think about it! Surg Radiol Anat. 2018; 40: 963-965. doi: 10.1007/s00276-018-2042-0.

10. Zheng LD, Tong QS, Tang ST, Du ZY, Ruan QL. Duodenal heterotopic pancreas in a child. World J Pediatr. 2009; 5: 146-148. doi: 10.1007/s12519-009-0029-y.

11. Seddon K, Stringer MD. Gastric heterotopic pancreas in children: A prospective endoscopic study. J Pediatr Surg. 2020; 55: 2154-2158. doi: 10.1016/j.jpedsurg.2019.10.053.

12. Betzler A, Mees ST, Pump J, Schölch S, Zimmermann C, et al. Clinical impact of duodenal pancreatic heterotopia – is there a need for surgical treatment? BMC Surg. 2017; 17: 53. doi: 10.1186/s12893-017-0250-x.

13. Kim JH, Lim JS, Lee YC, Hyung WJ, Lee JH, et al. Endosonographic features of gastric ectopic pancreases distinguishable from mesenchymal tumors. J Gastroenterol Hepatol. 2008; 23: 301-307. doi: 10.1111/j.1440-1746.2008.05351.

14. De Angelis GL, Fornaroli F, De Angelis N, Magiteri B, Bizzarri B. Wireless capsule endoscopy for pediatric small-bowel diseases. Am J Gastroenterol. 1758; 102: 1749-1757. doi: 10.1111/j.1572-0241.2007.01209.

15. Kopácová M, Vykouril L, Vacek Z, Tycová V, Bártová J, et al. Inverted Meckel's diverticulum with ectopic pancreatic tissue as a source of severe gastrointestinal bleeding. J Gastrointest Surg. 2010; 14: 578-581. doi: 10.1007/s11605-009-0838-2.