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Contents

Thrice Monthly Volume 12 Number 30 October 26, 2024

EDITORIAL

MINIREVIEWS

ORIGINAL ARTICLE

Retrospective Study

Clinical and Translational Research

World Journal of Clinical Cases

Thrice Monthly Volume 12 Number 30 October 26, 2024

Contents

LETTER TO THE EDITOR

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Thrice Monthly Volume 12 Number 30 October 26, 2024

Contents

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EDITORIAL

Advances in the diagnosis and treatment of heterotopic pancreas

Li Lang, Fa-Kun Yu, Li-Min Kang

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Abstract

Heterotopic pancreas, a rare congenital malformation, manifests outside the normal pancreas. Research suggests that abnormal embryonic development is linked to the presence of heterotopic pancreas. Three prevailing theories explain its mechanism: Dislocation theory, metaplasia theory, and totipotent stem cell theory. Clinical presentations of heterotopic pancreas are often nonspecific, with most patients being asymptomatic and incidentally discovered during unrelated surgeries or examinations. Endoscopic ultrasound, computed tomography, and magnetic resonance imaging are commonly employed diagnostic tools for heterotopic pancreas. However, the accuracy of diagnosis based on these methods is not consistently high, necessitating histopathological confirmation in many cases. Treatment options for heterotopic pancreas typically involve endoscopic resection, surgical resection, or observation through follow-up.

Key Words: Heterotopic pancreas; Ultrasound endoscopy; Enhanced computed tomography; Endoscopic therapy; Surgery

Core Tip: Heterotopic pancreas is rare, and its clinical presentation lacks specificity, posing challenges in diagnosis and differential diagnosis. Endoscopic ultrasound, computed tomography, and magnetic resonance imaging are the predominant diagnostic methods used for heterotopic pancreas. However, the diagnostic accuracy based on these imaging techniques is not consistently high, often necessitating histopathological confirmation. Clinicians should possess a thorough understanding of the clinical symptoms and imaging findings associated with this condition, as well as the key factors differentiating from other gastrointestinal disorders. Treatment strategies, such as endoscopic resection, surgical resection, or close monitoring, should be tailored based on comprehensive assessments considering factors such as the location, size, and clinical manifestations of heterotopic pancreas.

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INTRODUCTION

Heterotopic pancreas is a rare congenital malformation that occurs outside the main body of the normal pancreas. The cause and mechanism of this condition remain unknown. It lacks connections with the normal pancreas in terms of anatomy, blood vessels, ducts, and innervation. It has its own independent blood supply, ducts, and innervation, and is also referred to as vagus pancreas, accessory pancreas, or pancreatic tissue heterotopia[1]. The incidence of heterotopic pancreas is low, and was historically discovered incidentally during pathological biopsies or upper abdominal surgeries. However, advancements in imaging technologies such as computed tomography (CT) scans and endoscopic procedures like gastroscopy have led to a clinical increase in the identification of heterotopic pancreas cases, particularly in the digestive tract[2]. Clinical manifestations of heterotopic pancreas are non-specific, posing challenges in diagnosis and differential diagnosis, with variations based on the site of occurrence. Heterotopic pancreas in the gallbladder is particularly rare, presenting difficulties in distinguishing it from conditions such as gallbladder adenomyosis or cancer. In this editorial, we comment on a rare case of heterotopic pancreas in the gallbladder reported by Wang *et al*[3]. The patient had no specific clinical discomfort and was initially diagnosed with adenomyomatosis by CT scan. The patient underwent a laparoscopic cholecystectomy, and pathological examination revealed heterotopic pancreatic tissue in the gallbladder. Further research is needed to deepen our understanding of the clinical characteristics and optimal treatment approaches for heterotopic pancreas. This article provides a brief overview of recent advancements in the diagnosis and management of this condition.

MECHANISM OF HETEROTOPIC PANCREAS

Current research indicates that the occurrence of heterotopic pancreas is associated with abnormal embryonic development, although the precise embryological mechanism remains unclear. Three main theories have been hypothesized: Dislocation theory, metaplasia theory, and totipotent stem cell theory[4]. The most widely accepted theory regarding the embryonic development of heterotopic pancreas is the dislocation theory, which suggests that pancreatic primordium tissue is incorrectly positioned within the developing gastrointestinal system during embryonic development, resulting in anatomical discontinuity with the native pancreas[5]. The theory regarding the pathogenesis of heterotopic pancreas is the metaplasia theory, which posits that endodermal tissue migrates to the submucosa of the gastrointestinal tract during embryonic development and transforms into heterotopic pancreas. The third theory, totipotent stem cell theory, proposes that endothelial stem cells lining the gut mistakenly differentiate into heterotopic pancreas[6].

CLINICAL MANIFESTATIONS OF HETEROTOPIC PANCREAS

The clinical manifestations of heterotopic pancreas are often nonspecific, with most patients being asymptomatic and typically discovered incidentally during unrelated surgeries or examinations. However, a small percentage of cases can develop complications such as pancreatitis, pseudocyst, intussusception, insulinoma, and even pancreatic cancer[7]. These complications may present with non-specific symptoms including abdominal pain, bloating, nausea, vomiting, bleeding, and obstruction. Among these, abdominal pain and bloating are the most commonly reported symptoms, although precise epidemiological data are currently lacking[8]. Some studies[9] have indicated that the incidence rate of abdominal pain and bloating in patients with gastrointestinal heterotopic pancreas is 21.4%. Despite the non-specific nature of these symptoms, they should raise suspicion when patients present with unexplained abdominal pain and bloating, prompting the use of advanced imaging techniques to investigate the possibility of heterotopic pancreas and prevent potential misdiagnosis[10].

IMAGING FEATURES OF HETEROTOPIC PANCREAS

Endoscopic ultrasonography, CT, and magnetic resonance imaging (MRI) are the most commonly used methods for diagnosing heterotopic pancreas[11]. While gastroenteroscopy and endoscopic ultrasound alone may not have a high diagnostic rate, most cases can still be diagnosed based on histopathological reports[12]. The ultrasonic echo of heterotopic pancreas is closely related to its histopathological components, with uneven echoes mainly located in the submucosa and/or muscularis propria. Additionally, the lesions may present with anechoic tubular or cystic areas, aiding in the preoperative diagnosis[13]. Enhanced CT is the preferred imaging technique for heterotopic pancreas, where it appears as oval soft tissue density nodules with clear edges on plain images. Characteristic manifestations include an umbilical-like depression on the lesion's surface[14]. Heterotopic pancreas typically shows continuous enhancement during CT scans, with varying degrees of enhancement based on pathological components, blood supply, and other factors[15]. Acinar components exhibit significant enhancement, while duct-dominated types show less enhancement compared to normal pancreas. Heterotopic pancreatic pseudocysts and cystic atrophy may present as anechoic or occasionally with a reinforced septal component[16]. Heterotopic pancreas exhibits a similar appearance to the native pancreas on MRI images. It typically demonstrates a slightly high signal on T1-weighted images, along with notable enhancement in the late arterial phase. In some cases, the enhancement may even appear more pronounced than that of the native pancreas[17].

TREATMENT OF HETEROTOPIC PANCREAS

There is currently no consensus on the treatment of heterotopic pancreas. Options include endoscopic resection, surgical resection, or observation. Some studies suggest that surgical intervention is necessary due to the potential risks of cystic degeneration, necrosis, and malignant transformation, regardless of symptoms[18]. However, other studies indicate that malignant transformation is rare and most cases are asymptomatic, recommending follow-up observation instead[19]. Endoscopic treatments are typically suitable for lesions originating from the mucosa and submucosa that do not involve deeper layers. Advancements in endoscopic technology have led to the popularity of minimally invasive techniques such as endoscopic mucosal resection and endoscopic submucosal dissection, showing advantages in safety, effectiveness, quick recovery, and low complication rates[20]. Surgical treatment such as subtotal gastrectomy or intestinal resection is recommended when heterotopic pancreas undergoes secondary pathological changes and causes significant symptoms [21]. If a heterotopic pancreas is incidentally found during other surgeries and the patient is asymptomatic, it should be removed if feasible without impacting the original surgery. Frozen sections should be taken during the operation to assess for cancer, with the possibility of expanding the resection or performing radical surgery if needed[22].

CONCLUSION

Heterotopic pancreas is a congenital developmental variant characterized by mostly solitary lesions found in the stomach, duodenum, or proximal jejunum. Diagnosis and differential diagnosis of heterotopic pancreas can be challenging due to atypical clinical symptoms and imaging findings. Clinicians should be well-versed in the clinical symptoms and imaging manifestations of this disease, and understand the key points for differentiating it from other gastrointestinal diseases. This knowledge can help prevent missed diagnoses and misdiagnoses, ultimately reducing the need for unnecessary surgeries.

FOOTNOTES

Author contributions: Lang L, Yu FK, Kang LM designed the study, performed the study, analyzed the data, and wrote the manuscript; Kang LM has read and approve the final manuscript.

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