


Annular Pancreas in a 13-Year-Old Boy: A Delayed Clinical Presentation of a Congenital Anomaly Highlighting Challenges of Diagnosis

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Introduction: The annular pancreas is a rare congenital anomaly that typically results in duodenal obstruction during the neonatal period. However, its presentation can be variable, with some cases remaining undiagnosed until adolescence and posing diagnostic challenges, especially in low-resource settings where advanced imaging may be limited.

Case Presentation: We report the case of a 13-year-old boy with a three-year history of recurrent non-bilious projectile vomiting and epigastric pain. Despite normal laboratory findings, plain computed tomography (CT) and post-intravenous (IV) contrast CT scans revealed features suggestive of gastric outlet obstruction, including significant stomach distention and abrupt tapering of the duodenum. Owing to the inconclusive imaging results, surgical exploration was pursued. Intraoperative findings confirmed the presence of a band of pancreatic tissue encircling the first part of the duodenum, thereby establishing the diagnosis of an annular pancreas. A gastroduodenostomy was successfully performed, resulting in an uneventful recovery and resolution of symptoms during subsequent follow-up.

Conclusion: This case underscores the importance of maintaining a high index of suspicion for the annular pancreas in patients presenting with chronic gastrointestinal symptoms. Although imaging modalities provide valuable clues, surgical exploration remains the gold standard for achieving a definitive diagnosis when findings are ambiguous. The successful surgical management of this patient underscores the crucial role of timely intervention, particularly in settings with limited diagnostic resources.

Keywords: annular pancreas, duodenal obstruction, a delayed clinical presentation, gastroduodenostomy

Introduction

The annular pancreas is a rare congenital malformation that causes duodenal obstruction at birth.¹ The annular pancreas is characterized by partial or total encirclement of the second segment of the duodenum by a strip of pancreatic tissue during embryonic development.² It is commonly found below the ampulla of Vater in about 85% of confirmed cases and rarely above 15%.³ Annular pancreas occurs in approximately 1 out of every 20,000 live births.⁴ Due to the infrequency of this congenital condition, the specific cause related to the formation of an annular pancreas is not well established. Still, the annular pancreas is regarded as an embryopathy.⁵ In the initial four to eight weeks of embryonic development, the pancreas typically forms as the dorsal and ventral pancreatic buds rotate and fuse, driven by the expansion of the duodenum. The ventral bud gives rise to the inferior section of the head and the uncinat process of the pancreas, while the dorsal bud develops into the body and tail of the pancreas. The formation of the annular pancreas is thought to be a migration defect occurring due to the inability of the ventral bud to rotate and grow in a way that fully or partially surrounds the second portion of the duodenum.^{6,7}

Symptoms associated with annular pancreas can vary greatly from one individual to another. In certain instances, it may lead to intense symptoms shortly after delivery, whereas in other cases, it could stay asymptomatic throughout a person's life.⁴ In neonates, typical symptoms include difficulties with feeding, vomiting, and abdominal bloating. In



adults, the symptoms often resemble those of gastric outlet obstruction, including persistent abdominal pain, nausea, a feeling of fullness after eating, and vomiting.⁸ The intensity of the symptoms is linked to the degree of pressure exerted by the annular pancreas on the duodenum.⁴ Nevertheless, the severity of duodenal obstruction and the accompanying obstructive symptoms can vary, and there have been cases of the unrecognized annular pancreas being found in teenagers or even adults.¹ Understanding the clinical features of patients with an annular pancreas is tremendously valuable in diagnosing this condition. Typically, annular pancreas is identified during routine prenatal ultrasounds through the detection of the double bubble sign in the fetal abdomen, allowing for both diagnosis and treatment to be effectively carried out shortly after delivery.⁹ Different imaging methods, such as ultrasonography, X-ray, endoscopic retrograde cholangiopancreatography (ERCP), and computed tomography (CT), can also be used to diagnose an annular pancreas. In adults who are affected, CT is more frequently used.¹⁰ A definitive diagnosis of the annular pancreas relies on imaging studies and findings observed during surgery. Surgery is the gold standard for diagnosing the annular pancreas. Imaging studies play a suggestive role in the diagnosis before surgery.¹¹ CT and MRI reveal pancreatic tissue surrounding the duodenum.¹² There are no established guidelines or protocols for managing an annular pancreas.¹³ Several surgical techniques can be employed to treat the annular pancreas, with the primary goal being to alleviate the obstructive symptoms associated with this congenital condition.¹⁰ Duodenoduodenostomy, duodenojejunostomy, or gastroduodenostomy may be carried out, and the section of the duodenum, along with a ring of pancreatic tissue, can be excised as a single unit.¹² Here, we report a rare case of annular pancreas in a 13-year-old boy with a delayed clinical presentation of a partial duodenal obstruction (obstruction of the first part of the duodenum). This case highlights the challenges of diagnosis, which was achieved through surgical exploration in a resource-limited setting, and it was treated successfully with gastroduodenostomy.

Case Presentation

A 13-year-old boy came to the emergency department complaining of recurrent episodes of vomiting and abdominal pain over the past 3 years. The vomiting was non-bilious, projectile, and occurred after meals. Its frequency and severity have been gradually increasing. The abdominal pain was primarily in the epigastric region and was relieved after vomiting. On physical examination, the patient was in fair general condition, alert, and slightly dehydrated with normal vital parameters. The patient's systemic examination was normal. On laboratory examination, the white blood cell count, hemoglobin, electrolytes, urea, creatinine, and albumin were all within normal limits. On imaging, plain CT and post-IV contrast CT of the abdomen and pelvis revealed marked distention of the stomach with abrupt tapering of the first part of the duodenum and collapse of the rest of the duodenum. The pancreas was normal in size and showed homogeneous enhancement on post-contrast scans, with no evidence of calcifications. The rest of the small bowel loops, liver, gall bladder, spleen, kidneys, urinary bladder, and abdominal aorta were normal (Figure 1). The patient underwent a surgical exploration. Intraoperatively, a band of pancreatic tissue is found encircling the first part of the duodenum, causing an obstruction (Figure 2), with marked dilatation of the stomach (Figure 3), without any additional abnormal findings. A gastroduodenostomy was performed. The patient experienced an uneventful postoperative recovery, during which he received intravenous fluids, analgesics, and antibiotics. On the third day after surgery, oral sips were introduced, gradually increasing the diet from liquids to semi-liquids and then to full solid foods over three weeks. After discharge from the hospital, the patients attended follow-up appointments and showed improvement with no further vomiting, abdominal pain, or negative complications.

Discussion

The annular pancreas is a rare congenital anomaly characterized by the abnormal rotation of the pancreas. This condition infrequently occurs in adults.¹¹ The differential diagnosis can be divided into intrinsic and extrinsic. Notable intrinsic causes to consider include duodenal atresia, duodenal stenosis, paraduodenal hernias, Meckel diverticulum, and duodenal webs. At the same time, significant extrinsic factors to consider are gut malrotation and midgut volvulus. In adults suspected of having annular pancreas, peptic ulcer disease, pancreatic divisum, and primary duodenal and pancreatic cancers should also be included in the differential diagnosis.^{14,15} Various imaging modalities are being actively considered for diagnosing annular pancreas. CT is used to diagnose and analyze the annular pancreas.¹⁰ CT results

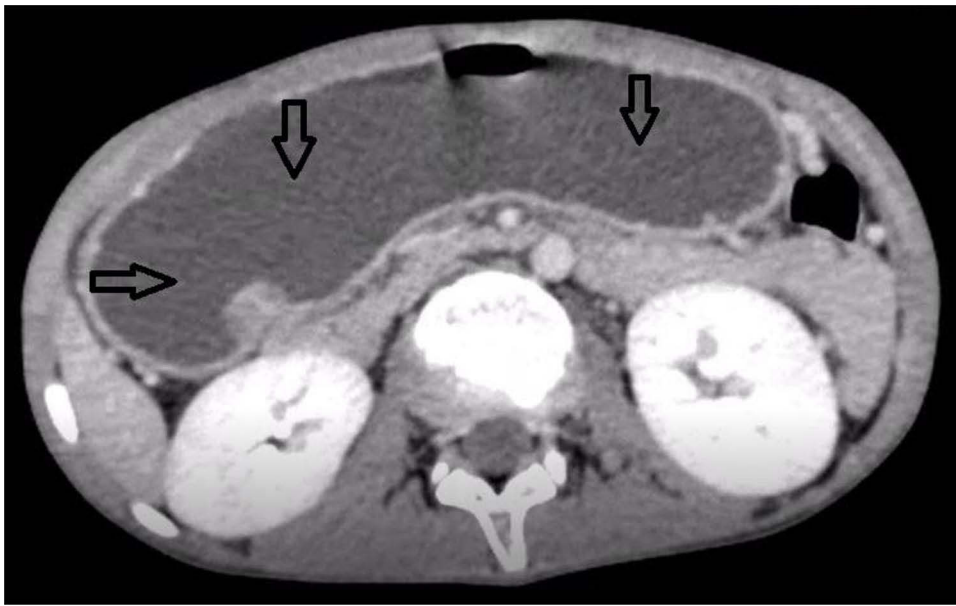


Figure 1 Abdominal CT shows marked distension of the stomach (thick arrows) with abrupt tapering of the first part of the duodenum and collapse of the rest of the duodenum.

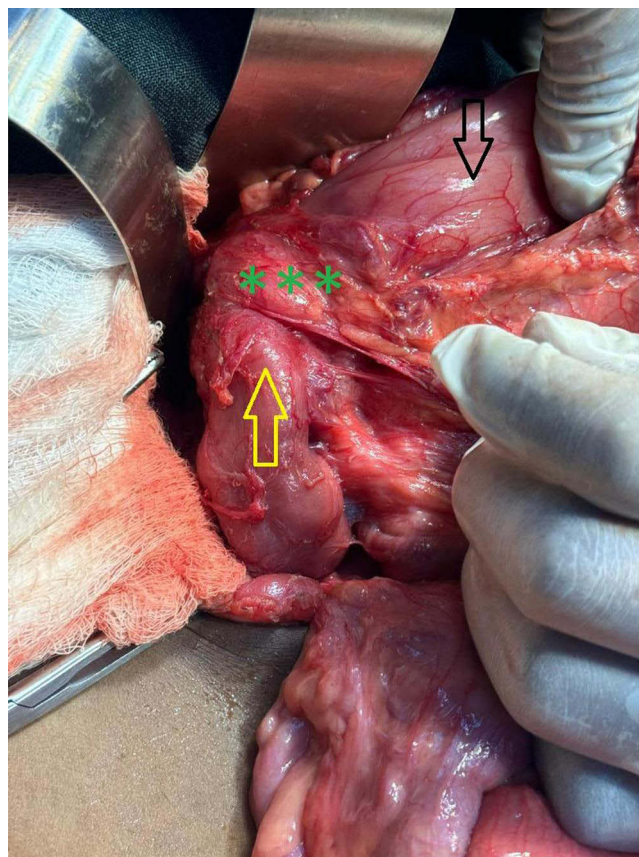


Figure 2 Intraoperative image showing the pyloric region of the stomach (black arrow), a band of pancreatic tissue encircling the first part of the duodenum, causing obstruction (green asterisks). The second part of the duodenum is normal (yellow arrow).

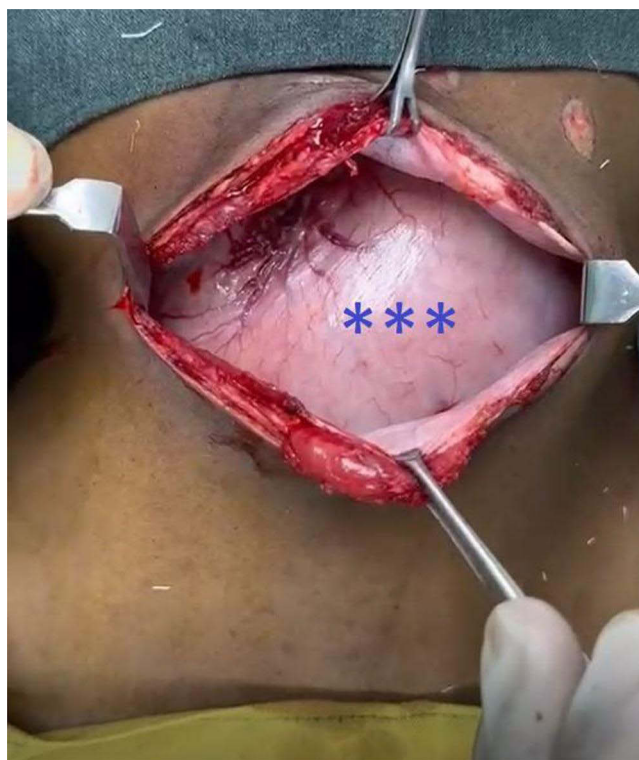


Figure 3 Intraoperative image showing a dilatation of the stomach (blue asterisks).

indicate an enlargement of the pancreatic head with enhanced visibility of the second portion of the duodenum.¹³ In a case of annular pancreas reported by Moon, an abdominal CT scan revealed pancreatic tissue surrounding the second part of the duodenum, leading to the diagnosis of an annular pancreas.¹ In this patient, plain CT and post-IV contrast CT of the abdomen and pelvis revealed marked distention of the stomach with abrupt tapering of the first part of the duodenum and collapse of the rest of the duodenum. The pancreas was normal in size and showed homogeneous enhancement on post-contrast scans, with no evidence of calcifications (Figure 1). In this case, the preoperative imaging with plain CT provided features of gastric outlet obstruction, which made the diagnosis challenging. This challenge necessitated reliance on surgical exploration for accurate diagnosis. Although the patient's clinical characteristics are not unique to the annular pancreas, they provided some clues to the diagnosis, which necessitated the decision to perform surgical exploration. The definitive diagnosis was confirmed by surgical exploration, which revealed a band of pancreatic tissue encircling the first part of the duodenum, causing an obstruction. In contrast, the second part of the duodenum was normal (Figure 2). This finding is consistent with a case reported by Jha et al.⁸ This decision aligns with the existing literature, which emphasizes the importance of having a thorough understanding of the clinical characteristics of the annular pancreas and its associated anomalies when managing this uncommon congenital anomaly.¹⁰ A significant number of individuals with this anomaly stay asymptomatic for their entire lives and are frequently identified incidentally through imaging studies or during autopsies. Nevertheless, a small percentage of patients with an annular pancreas may show clinical symptoms either in childhood or later in life, typically between the ages of 20 and 50.¹⁶ The clinical presentation of the annular pancreas varies by age, resulting in differences in diagnosis.¹³ The emergence of advanced diagnostic techniques has led to an increased recognition of this condition. However, despite radiological advancements, surgical confirmation is required in approximately 40% of annular pancreas cases, establishing it as the gold standard.^{13,16} The clinical presentation of the annular pancreas varies by age, resulting in age-related variations in management. The approach to treating an annular pancreas varies based on its clinical presentation. Surgical procedures using different bypass methods, such as duodenoduodenostomy, gastrojejunostomy, gastroduodenostomy, and duodenojejunostomy, are necessary.¹³ However, the decision should be customized for the individual patient.¹⁶ In our patient, we

successfully performed a gastroduodenostomy with a transverse incision of the pylorus and a longitudinal incision of the duodenum, resulting in a diamond-shaped anastomosis. After more than six months of follow-up, the patient was feeling well and started to gain weight. The annular pancreas is a rare condition that warrants attention in the differential diagnosis of recurrent vomiting and abdominal pain. This unusual formation of pancreatic tissue encircling the duodenum can lead to gastrointestinal obstruction, which may manifest as persistent nausea, abdominal pain, and vomiting. We recommend that clinicians, especially surgical pediatricians, be vigilant in considering this possibility when evaluating patients with these symptoms.

Conclusion

This case underscores that the annular pancreas, although a congenital condition typically detected in the neonatal period, can manifest later in childhood with insidious and non-specific symptoms such as recurrent non-bilious vomiting and epigastric pain. In our 13-year-old patient, preoperative imaging provided features of gastric outlet obstruction, necessitating surgical exploration for confirmation. The subsequent intraoperative identification of a pancreatic tissue band encircling the duodenum, along with the successful execution of a gastroduodenostomy, highlights not only the diagnostic challenges but also the efficacy of tailored surgical management in resource-limited settings. The favorable postoperative recovery, marked by the resolution of symptoms and gradual weight gain, reinforces the importance of maintaining a high index of suspicion for the annular pancreas in adolescents presenting with chronic gastrointestinal symptoms. This report serves as a reminder that even in the absence of classical radiological findings, meticulous clinical evaluation coupled with timely surgical intervention can markedly improve patient outcomes. Furthermore, documenting such cases is crucial for deepening our understanding of the variable presentations of the annular pancreas. It may pave the way for the development of more standardized diagnostic and therapeutic protocols in the future.

Ethics Statement

An institution's ethics committee approval is not required for the case reports.

Informed Consent Statement

Written informed consent was obtained from the patient's parent for the publication of this case report and any accompanying images. The patient's parent was told about the purpose of this publication and that his identity would be protected.

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Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising, or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

Disclosure

The authors declare no conflicts of interest in this work.

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