




Incidental Finding of Annular Pancreas in a Routine Cadaveric Dissection: Case Report

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Abstract

Annular pancreas is a rare congenital anomaly that results from the malrotation of the ventral pancreatic bud. The presentation of annular pancreas varies: it can be asymptomatic or present clinical symptoms of duodenal obstruction that can affect all age groups, from newborns to adults. In the present case report, we describe a complete type of annular pancreas at the level of the second part of the duodenum, which was an incidental finding in a prosected specimen. This anomaly has significant clinical relevance to clinicians and radiologists due to its variable presentation. The embryological, clinical and radiological aspects of this congenital anomaly are discussed in detail in the present article.

Keywords

- ▶ annular pancreas
- ▶ congenital anomaly
- ▶ duodenal obstruction
- ▶ pancreatic bud

Introduction

The pancreas is a retroperitoneal organ situated within the C-shaped loop of the descending part of the duodenum across the posterior abdominal wall. It has four anatomical parts: head, neck, body and tail, as well as an uncinata process.¹ The pancreas develops from the fusion of ventral and dorsal pancreatic buds at the junction of the foregut and midgut during the fourth week of gestation. It requires a complex sequence of events, such as the fusion of the ventral bud with the dorsal pancreatic bud along with the rotation of the duodenum.² Therefore, due to its complex development, several congenital anomalies are associated with the embryonic development of the pancreas.

Annular pancreas (AP) is one such rare congenital anomaly that is due to an error in rotation, resulting in the formation of a pancreatic tissue ring that partially or completely encircles the second part of the duodenum. It is a rare type of morphological congenital anomaly, with an estimated incidence of 1 out of 12,000–15,000 newborns. Though it affects mostly newborns, constituting around 1% of all intestinal obstructions in the pediatric age group, in late adulthood its presentation mimics a wide variety of complications, such as pancreatitis, peptic ulcer, duodenal obstruction, perforation and peritonitis, which makes its diagnosis difficult yet indispensable.

The present case report deals with an incidental finding on a prosected specimen of an AP with an intact second part of the duodenum during a regular undergraduate class.

Case Report

During a regular undergraduate Bachelor of Medicine, Bachelor of Surgery (MBBS), anatomical dissection, a prosected pancreas specimen with an intact second part of the duodenum and spleen was identified. This prosected specimen had been obtained earlier from one of the cadavers used for dissection. Thus, the age, sex and medical history of the cadaver from which this specimen was procured could not be traced. These cadavers used for the purpose of research and teaching of undergraduate medical students were donated to the Department of Anatomy, Christian Medical College. Since the college is located in Vellore, India, we assumed that the cadaver was of Southern Indian origin.

This prosected specimen was identified as a case of complete AP with a ring of pancreatic tissue encircling the second part of the duodenum. This band of pancreatic tissue began as a continuation of the head of the pancreas, and traversed horizontally on the anterior aspect of the second part of the duodenum toward the right side, laterally and then posteriorly,

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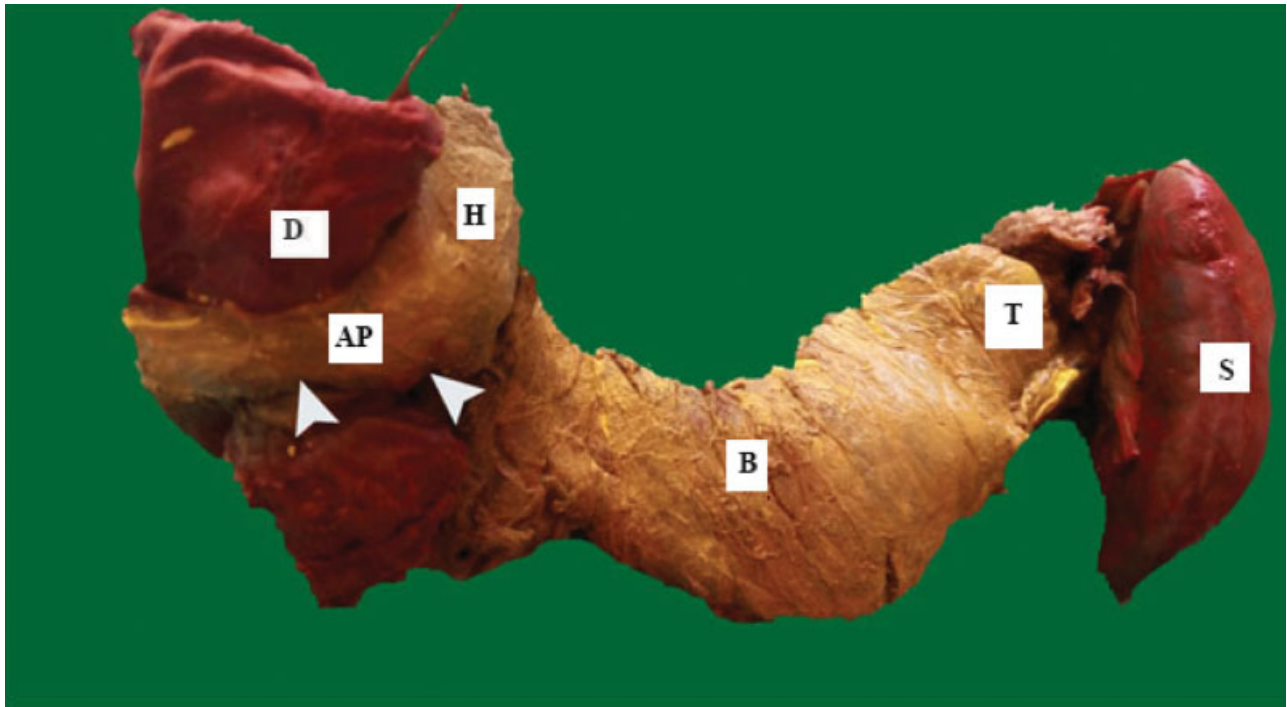


Fig. 1 Anterolateral view of the annular band of pancreatic tissue completely encircling the second part of the duodenum. Abbreviations: AP, annular pancreas; B, body of the pancreas; D, duodenum; H, head of the pancreas; S, spleen; T, tail of the pancreas.

thus completely surrounding the second part of the duodenum. It became continuous with the head of the pancreas on the posterior aspect (► **Fig. 1**). The color and texture of the pancreatic tissue ring was similar to that of the rest of the pancreas. The width of the AP was not uniform throughout the duodenum. We measured the width at three sites: anteriorly, it had 1.5 cm, laterally, it had 1.8 cm, and posteriorly, it had 2 cm. The margins of this band were well-defined and showed no adhesion to the duodenal wall. However, a constricted lumen of the second part of the duodenum was observed encircled by the pancreatic tissue ring. In addition, the duodenal luminal diameter showed an appreciable dilation above the level of the annular band, as compared with the distal part. Upon examination, the neck, body and tail of the pancreas, along with spleen, were found to be normal. As this was a prosected specimen, we were unable to observe and comment on the relevant anatomic findings of the surrounding area in the cadaver.

Discussion

The AP was first reported by Tiedemann in 1818, but it was spotted in autopsy material in 1862 by Ecker, who named it after the ring of pancreatic tissue as “annular pancreas”.^{3,4} It is a rare variety of congenital anomaly, occurring in every 12,000–15,000 births.⁵ The true prevalence of AP in a specific population has not yet been reported. However, a few studies on endoscopic retrograde cholangiopancreatography (ERCP) have shown a prevalence of 1 in 250 or 400 cases per 100,000 adults.⁶ In an autopsy series, the prevalence of AP ranged from 5–15 per 100,000 adults.⁷

The development of the pancreas begins with the formation of a dorsal and ventral pancreatic bud in the corresponding mesenteries of the duodenum during the fourth week of gestation. By the seventh week, the rotation of the duodenum causes the ventral bud to also rotate and course behind the duodenum to fuse with the dorsal pancreatic bud. The dorsal pancreatic bud forms the upper part of the head, neck, body and tail of the pancreas, whereas the ventral bud forms the lower part of the head and the uncinete process. Sometimes, the ventral bud splits into right and left parts. Later, a faulty migration and fusion of these two parts of the ventral pancreatic bud along with the dorsal pancreatic bud result in a band of pancreatic tissue surrounding the descending part of the duodenum, constricting the duodenal lumen at the site. This annular mass of pancreatic tissue maintains its continuity with the head tissue. Numerous theories have been put forward to explain the development of AP, and, among them those by Lecco⁸ and Baldwin⁹ are widely accepted. These authors postulate that the faulty migration of the ventral pancreatic bud results in a variable band of pancreatic tissue around the second part of the duodenum.^{8,9} Further studies in this line have highlighted the role of the hedgehog signaling pathway in the development of this anomaly. One study¹⁰ hypothesized the overexpression of the ventral-specific gene *transmembrane 4 superfamily member 3 (tm4sf3)*, which plays a role in the development of this anomaly.¹⁰ On the other hand other theories postulate that the primary duodenal stenosis results in a band of pancreatic tissue around it.¹¹ Alternatively, Glazer and Margulis¹² proposed that hypertrophy or atrophy of the ventral and dorsal pancreatic buds might be associated with the development of AP.¹² Apparently, the AP is associated with variable ductal abnormalities, and, based on that, it has been

classified into six types, taking into account the drainage site of the main pancreatic duct.¹³

Children usually present with symptoms of gastrointestinal (GI) obstruction, such as poor feeding, vomiting and abdominal distention in the first weeks of life. A few cases of the association of the AP with duodenal ulcer in childhood have also been reported by Fu et al.¹⁴ Previous studies have reported that the occurrence of AP is as common in adults as it is in children.^{15,16}

In adults, it can remain asymptomatic until diagnosed as an incidental finding during routine radiological investigations, or present with secondary clinical symptoms of GI obstruction like nausea, vomiting, abdominal pain etc. In adults, the AP usually presents around the second to the fifth decades of life.¹⁵ The severity of this condition depends on its type, whether complete or incomplete, and this diagnosis is considered an important factor from the clinical and management points of view. In around 75% of the cases, the ring of tissues is incomplete, while in 25% it completely encircles the duodenum.¹⁷ The severe form, which compromises the duodenal lumen, mandates immediate surgical intervention.^{18,19} Other clinical complications, like obstructive jaundice, pancreatitis, peptic ulcer, and peritonitis secondary to perforation of the duodenum, have been reported.¹⁹ A strong association with several other congenital anomalies, such as esophageal atresia, tracheoesophageal fistula, imperforate anus, congenital heart disease, Hirschsprung disease, malrotation of the midgut, and Down syndrome have been reported in conjunction with AP.²⁰ Although this condition does not have a well-established genetic basis, around 42% of Indian hedgehog (IHH) mutant mice developed AP.²¹ However, how IHH gene loss is associated with development of AP is not yet established. Moreover, isolated case reports of familial AP have also been documented, suggesting a genetic basis for the development of this anomaly.²² A rare presentation of AP has been documented by Li et al²³ in an 8-year-old girl with sparse scalp hair, bulbous nose, thin upper lip, broad eyebrows, phalangeal abnormalities in both hands and toes, multiple exostoses, mild intellectual impairment and severe malnutrition, presumably suffering from trichorhinophalangeal syndrome type II, a rare autosomal dominant genetic disorder affecting the craniofacial and skeletal development, which is associated with loss of functional copies of the *TRPS1* gene at 8q23.3 and the *EXT1* gene at 8q24.11.²³

Glazer and Margulis,¹² Sandrasegaran et al¹⁵, Maker et al,²⁴ and Jarry et al²⁵ have reported that the AP affects the second part of the duodenum in 74% of cases. This is supported by cadaveric case reports by Russo and Ugon,²⁶ and Vinoth et al.¹⁹ Nayak et al²⁷ reported that in 21% of the cases the AP has also been found around the first and third parts of the duodenum.²⁷

The AP can remain asymptomatic in adults and go unnoticed throughout life, or it can present during the second to fifth decades of life with clinical symptoms like abdominal pain, vomiting, peptic ulcer, pancreatitis (acute or chronic) and biliary obstruction.¹⁵ The late onset of the obstructive symptoms in the elderly has been attributed to the development of pancreatitis (13–22%) due to insufficient drainage of

the pancreatic juice through the annular duct, leading to stagnation and initiation of an inflammatory cascade.^{16,28}

With the advent of different types of diagnostic and therapeutic imaging, such as ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI), ERCP, and magnetic resonance cholangiopancreatography (MRCP), the awareness regarding this kind of incidental finding of APs gained paramount clinical significance, and became important for clinicians and radiologists.¹⁵ When symptomatic, the aforementioned imaging modalities can aid in the diagnosis. The MRCP appears to be the best non-invasive alternative method for the diagnosis of AP, since the ERCP may not be feasible at times due to the variable amount of duodenal obstruction by the pancreatic ring.²⁹

The management of AP is usually primarily aimed at relieving the obstruction. Surgical interventions like enterenterostomy are being commonly performed as a treatment modality.^{24,30} Duodenoduodenostomy or duodenojejunostomy are the safest and most successful way of bypassing the annular constriction.³¹ In cases of AP associated with suspicion of periampullary malignancy, pancreatolithiasis and localized chronic pancreatitis, duodenopancreatectomy might be the treatment option.^{12,32} However, resection of the AP band is usually avoided because of severe postoperative complications, including fistula formation and pancreatitis, with incomplete permanent cure rate.³³

In conclusion, any symptoms of intestinal obstruction, including mild ones, such as vomiting and abdominal pain, at any age, can raise the suspicion of AP, though it is rare. Thus, this should be kept in mind during the investigation, and the AP must be ruled out. Asymptomatic cases can pose a threat and cause complications during various abdominal surgeries, such as kidney transplants and liver surgeries. Any inadvertent injury might lead to a leak of the active pancreatic enzymes in the pancreatic juice into the surrounding area.

Hence, this rare congenital anomaly, a complete AP around the second part of the duodenum in an adult (that can remain asymptomatic or symptomatic) has significant clinical relevance to clinicians and radiologists. But the importance and effect of this anomaly in the present case report could not be appreciated, since it was an incidental finding in a prosected specimen with unknown cause of death.

Conflicts of Interest

The authors have none to declare.

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