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Review Article

Annular pancreas and current approaches to surgical management

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ABSTRACT

Annular pancreas is an uncommon but well documented anatomical variant. Annular pancreas has a strong embryological basis, likely resulting from errors in rotation of the ventral duct of the embryological pancreas. Despite a low prevalence, annular pancreas is associated with surgical pathology that requires urgent intervention. In neonates and adults, duodenal obstruction can pose a life-threatening risk, and prolonged inflammation of related structures in adults can result in pancreatitis, cholangitis, and ultimately pancreaticobiliary malignancy. Currently, in cases of duodenal obstruction, bypass surgery has garnered favour over resection, whereas in cases of recurrent pancreatitis and cholangitis, endoscopic retrograde cholangiopancreatography (ERCP) is preferred. Further longitudinal investigation of the risk of developing foregut malignancy with bypass as opposed to resection would be beneficial in informing guidelines for surgical management.

Keywords: Embryology, Annular pancreas, Pancreatitis, Bypass surgery

INTRODUCTION

Normal anatomy and embryology of the pancreas

The pancreas is a largely secondarily retroperitoneal composite gland lying obliquely in the transpyloric plane.1 It consists of four parts; a head, neck, body, and tail. The anterior surface of the gland above the inferior border gives attachment to the transverse mesocolon. Thus, tissue below this attachment lies in the infracolic compartment, while the majority of the gland lies in the supracolic compartment to form the stomach bed. The head and tail incline towards the right and left paravertebral gutters, while the neck and body form a convexity over the aorta, inferior vena cava and the first lumbar vertebra.[1,2]

The head is encased in the C-shaped concavity of the duodenum at the level of the second lumbar vertebra, overlaying the renal veins and the inferior vena cava. Of note, the bile duct indents, or is contained within, its posterosuperior surface as it travels laterally to empty into the second part of the duodenum at the major duodenal papilla.[1,2] An inferior wedge lying over the aorta forms the uncinate process, while the head becomes the neck in front of the superior mesenteric-portal channel. Continuing from the neck, the body slopes upwards, reaching the hollow of the left kidney. The short tail passes forward from the anterior surface of the left kidney in the lienorenal ligament in contact with the hilum of the spleen. The secretions of the pancreas are drained by two ducts, the main pancreatic duct, passing from the tail to the upper portion of the head to join the bile duct at the Ampulla of Vater and the accessory pancreatic duct, which drains a lower portion of the head and uncinate process emptying to the minor papilla.[1,2]

During the 4th week of gestation, dorsal and ventral buds develop from the endoderm at the junction of the fore and midguts, growing into the ventral and dorsal

mesogastrium respectively.[3,4] The ventral bud elongates in common with the ventral outgrowth of the bile duct, as opposed to the dorsal bud which grows with an independent dorsal duct. In the 5th and 6th week of development the duodenum rotates clockwise around its long axis, shifting the entrance of the common bile duct to the dorsal side of the duodenum.[3,4] Carried by the bile duct, the ventral bud thus also rotates clockwise around the duodenum towards the dorsal bud. These buds fuse during the 7th week of gestation to form the pancreas.[3,4] The dorsal bud thus forms the upper portion of the head, body, and tail, whereas the ventral bud forms the lower head, rich in pancreatic polypeptide cells, and the uncinate process. The immature duct systems of the buds anastomose as they develop. Eventually, the duodenal end of the dorsal duct forms the accessory pancreatic duct and the remainder of the dorsal duct and ventral duct form the main pancreatic duct.[3,4]

ANATOMY AND EMBRYOLOGY OF ANNULAR PANCREAS

Annular pancreas is a variant of the normal anatomy already described. It manifests as a complete or partial band of physiologically normal pancreatic tissue surrounding the second part of the duodenum. It affects women more than men and is associated with pancreas divisum, intestinal and pancreaticobiliary malrotations and other congenital abnormalities in a third of patients.[3-7] Since being first described by Tiedermann in 1818, annular pancreas has been recognised as posing a significantly increased risk of duodenal and pancreaticobiliary pathology. Despite this, no consensus classification system is in use clinically or in the literature.

Estimations of its incidence vary. Postmortem studies conducted by Ratvitch suggest a 1 in 20,000 live birth incidence.[8] More recent studies by Baggott utilizing ERCP, albeit in a symptomatic population, have suggested a much more common incidence of 0.1%.[9] Regardless of the true incidence, the largest cohort study to date by Nagpal suggests 40% of adult patients with annular pancreas become symptomatic,6 and as such it remains a significant differential diagnosis in acute surgical pathology. Furthermore, upwards of 15% of patients with annular pancreas will develop a foregut malignancy.[3-7]

Annular pancreas has an embryological basis despite consensus on a single theory for its development lacking. One hypothesis has gained support as the most plausible explanation. Lecco's theory suggests that in annular pancreas, the distal tip of the ventral bud becomes pathologically adherent to the ventral duodenal wall during the 5th week of gestation.[3,4] Thus the ventral bud is forcibly elongated, encircling the duodenum as it completes its clockwise rotation. This theory is supported by a case series by Wellan in 93 patients, 91 demonstrated pancreatic tissue interspersed between the duodenal muscularis supporting embryonic adherence of the bud to duodenum.[14] ERCP studies by Patel showed 35 of 42 patients had continuation of the embryonic ventral duct through annular tissue draining into the main pancreatic duct, also supporting Lecco's theory.[10] The ventral bud origin of annular pancreas is further supported by histopathological studies by Suda and Sessa.[12,13] Investigation of annular pancreas tissue from 4 patients revealed a high proportion of pancreatic polypeptide cells identical to the concentration found in other ventral bud derivatives, namely, the lower portion of the head and uncinate process.

CLINICAL PRESENTATIONS OF ANNULAR PANCREAS

A rare variant, symptomatic annular pancreas may manifest variably based on its complex anatomical relationships to the surrounding structures of the foregut. In Nagpal's cohort study of 198 patients with annular pancreas, 80 were symptomatic. 48.8% of those patients had severe abdominal pain, 31% had duodenal obstruction and 16% had acute pancreatitis.[6] The prevalence of these presenting complaints are supported by other smaller cohort studies. Zyromski, Sandrasegaran and Cai investigated 55, 24 and 11 symptomatic annular pancreas patients respectively, and reported the prevalence of abdominal pain between 70 and 100%, duodenal obstruction in 35 to 55%, and acute pancreatitis in 20 to 25%.[7,11,15] These populations showed a consistent prevalence between 8 and 15% of pancreaticobiliary or duodenal malignancy.

The mechanisms of these presenting complaints are complex. Duodenal obstruction occurs by both the mechanical constriction, but also due to duodenal stenosis occurring secondary to the chronic inflammatory response induced by ectopic pancreatic tissue within the duodenal muscularis.[3-7] This inflammatory environment also may induce concurrent fibrosis of the ventral annulus duct, as well as increasing the intrinsic susceptibility of annulus tissue to damage. Thus, pancreatic secretions from the histologically normal annulus tissue are unable to drain to the main pancreatic duct, resulting in localised pancreatitis of the annulus and eventually pancreatic malignancy.[3-7]

APPROACHES TO MANAGEMENT OF ANNULAR PANCREAS

There are no clinical guidelines for the management of annular pancreas. In neonates who almost exclusively present in duodenal obstruction, duodenal bypass is necessary. In adults who have a more varied presentation, options are numerous.

For patients presenting with duodenal obstruction, bypass surgery is almost always required, with the age of the patient influencing the location of the anastomosis.[6,7,10,16-19] In younger patients, who generally have a high level of

gastric acid secretion, duodenoduodenostomy or duodenojejunostomy is preferred to gastrojejunostomy to avoid ulceration of anastomosis sites. If gastrojejunostomy is performed concurrent truncal vagotomy may also be performed to counteract this. Conversely, in older patients, gastrojejunostomy without vagotomy can be considered. Of Nagpal's cohort of 25 adult patients with duodenal obstruction, 20 underwent a laparoscopic bypass surgery.[6] At 24 months postoperatively no patient showed recurrence of symptoms. Chittawadagi reported successful treatment of duodenal obstruction by laparoscopic duodenojejunostomy in six patients with no perioperative complications and no symptom recurrence at 24 months.[17]

For neonatal patients with annular pancreas, duodenoduodenostomy is the preferred bypass operation. The type of anastomosis utilised is the main point of contention in this population. In a retrospective review by Liang, 52 patients aged between 2 and 10-days-old with annular pancreas underwent laparoscopic duodenoduodenostomy with either a diamond-shaped (n=44) or side-to-side (n=8) anastomosis.[19] A majority of these neonatal patients presented with bilious vomiting or other features suggesting duodenal obstruction. Liang reported no clinically significant difference in operative time, rate of bleeding, time of initial oral feeding, time to full oral feeding, feeding intolerance, or length of admission between these anastomosis types. Of note, at a follow-up period between 19 to 85 months, no severe acute or chronic complications or recurrence in either group was demonstrated.

Although previously preferred, resection of the annular pancreas has fallen out of favour in cases of duodenal obstruction without malignancy. The reasons for this are twofold. Prospective studies of patients who received annulus pancreatectomy showed significant rates of post operative pancreatitis and pancreatic fistula.[6,7,20] Secondly, the persistence of pancreatic tissue within the duodenal muscularis resulted in symptom recurrence and the need for further intervention. Malignancy is normally treated as per other pancreaticobiliary malignancies. It is unclear whether bypass surgery increases the long-term risk of malignancy arising from remnant annulus or bypassed duodenum as opposed to annulus pancreatectomy. Lim recently described an approach of laparoscopic primary division of annular pancreas in an adult male. This patient required endoscopic duodenal dilation at 1-week post-operatively due to residual symptoms, although at 1 year follow-up was asymptomatic without operative complication.[21]

In patients presenting with recurrent pancreatitis, or cholangitis, ERCP is preferred. Fogel performed ERCP with sphincterotomy in 32 patients 26 of 27 patients had successful symptom relief after one attempt.[22] Unfortunately, no long term follow up was performed in this group and as such, rates of symptom recurrence and need for definitive management is unclear. Nagpal reported successful conservative management of acute pancreatitis in eight of 13 patients, with the remainder requiring endoscopic therapy.[6]

CONCLUSION

Although uncommon annular pancreas remains an important differential for foregut pathology in both neonates and adults. The embryonic basis of annular pancreas and its relationship to the development of its surrounding foregut structures allows for the consideration of the possible presenting complaints in annular pancreas. Short and medium-term outcomes concerning morbidity, symptom relief and recurrence have shown bypass surgery and endoscopic therapy are appropriate for duodenal obstruction or biliary tree pathology in annular pancreas. Acute pancreatitis can be managed conservatively. However, the risk of recurrence and malignancy should always be considered in patients with annular pancreas. Further research should be conducted to establish the long-term outcomes of surgical interventions for annular pancreas to allow for the development of management guidelines.

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