Annular Pancreas

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Continuing Education Activity

Annular pancreas (AP) is a rare congenital anomaly characterized by encasement of the duodenum by a band of pancreatic tissue. AP is not commonly encountered in clinical practice and affects both children and adults. Prompt diagnosis and treatment of this condition can improve the morbidity and mortality associated with this condition. This activity reviews the evaluation and treatment of AP and highlights the role of the interprofessional team in the care of patients with this condition.

Objectives:

- Describe the etiology of annular pancreas.
- Summarize the diagnostic evaluation of annular pancreas.
- Outline the currently available treatment options in the management of annular pancreas.
- Summarize the interprofessional team strategies for improving care coordination and communication to advance and improve outcomes in patients diagnosed with annular pancreas.

Access free multiple choice questions on this topic.

Introduction

Annular pancreas (AP) is a rare congenital anomaly characterized by partial or complete circumferential encasement of the second part of the duodenum by a band of pancreatic tissue during embryogenesis.[1] It is usually located above the papilla of Vater in approximately 85% of diagnosed cases.[2] Older studies have reported the prevalence of AP in three of 20,000 autopsies and three of 24,519 surgical cases.[3][4] With the advent of newer and improved diagnostic modalities, this condition is being recognized more frequently. Many individuals with this anomaly remain asymptomatic throughout their lifetime and are often diagnosed incidentally on imaging or during autopsies. However, a fraction of patients with AP tends to present with clinical manifestations either early in life or during adulthood usually between 20 and 50 years of age.[5][6][7]

Based on the morphologic distribution of pancreatic tissue, AP has been classified into a complete or incomplete type. Complete type AP shows pancreatic parenchyma or annular duct completely encircling the second part of the duodenum confirmed by macroscopic inspection, and incomplete type AP demonstrates partial circumferential encasement of the duodenum by pancreatic tissue confirmed by endoscopic retrograde cholangiopancreatography (ERCP) or surgical evaluation.[8] The annular pancreatic duct (APD) commonly drains in the main pancreatic duct (MPD) but can communicate with the intrapancreatic common bile duct, the duct of Santorini, or the duct of Wirsung.[9] Yogi et al. classified AP into six subtypes with type I type demonstrating communication of APD with the duct of Wirsung and type II showing the duodenum encircled by MPD. Types I and II are the most common subtypes with the other 4 subtypes being less frequently encountered and correspond to the communication of the APD with the duct of Santorini or the common bile duct (CBD).[10]

Etiology

Given the rarity of this congenital condition, a precise etiology implicating the development of annular pancreas is not well defined, but AP is considered to be an embryopathy.[5] During the first four to eight weeks of embryonic development, the pancreas normally develops following rotation and fusion of the dorsal and ventral pancreatic buds as a result of the expansion of the duodenum. The ventral bud develops into the inferior part of the head and uncinate process of the pancreas with the dorsal bud developing into the body and tail of the pancreas.[8][11] The development of AP is proposed to be a migration anomaly resulting from the failure of the ventral bud to rotate and extend to encase the second portion of the duodenum either partially or completely.[8][11]

Several theories, some dating back to the 1900s, have been proposed to explain the formation of AP, but none of them have convincingly been able to explain the same in all cases of AP. However, immunohistochemistry studies have shown the origin of AP to be of the ventral pancreatic anlage.[7] Although not well studied, the role of genetic factors has been linked to this anomaly based on isolated cases reports of familial annular pancreas and overexpression of the ventral-specific gene transmembrane 4 superfamily member 3 (tm4sf3).[12]

Epidemiology

The true prevalence of annular pancreas is currently unknown owing to the rarity and infrequent reporting of this congenital anomaly. Before the availability of contemporary imaging, this anomaly was noted incidentally in 3 of 20,000 autopsies and in 3 of 24,519 individuals who underwent abdominal surgeries.[3][4] With the availability of novel imaging modalities, the prevalence of AP has marginally increased and is estimated to be approximately 1 in 1000 cases.[13][14][15] AP affects both sexes with a slight male preponderance, which has been a contentious issue.[16][17]

Pathophysiology

The pathophysiology of annular pancreas is explained by the direct impact of the migratory defect during embryogenesis. Duodenal obstruction is secondary to a mechanical defect, which can be extrinsic from the encasement of the duodenum or intrinsic from scarring, stenosis, or the presence of duodenal webs. The pathogenesis of pancreatitis in AP is unclear. Pancreatitis, when it occurs, is usually confined to the annulus and the adjoining pancreatic head sparing the body and tail of the gland which is likely secondary to fibrosis resulting in the inability of the pancreatic secretions to pass through the annular duct and increased intrinsic susceptibility of the annular pancreatic tissue to damage.[18][19][20] Peptic ulcer disease may be related to gastric stasis and antral overdistension with resulting hypergastrinemia.[7][21] Biliary ductal dilation is attributed likely due to the progression of chronic pancreatitis resulting in fibrosis or ductal compression by the annular gland.[8]

Histopathology

Microscopic examination of the annular pancreatic tissue demonstrates the presence of many pancreatic polypeptide (PP) cells of the ventral pancreatic anlage arranged in irregularly shaped islets.[22] Histologically, AP has been classified into two types: intramural and extramural type based on the presence of pancreatic tissue in the duodenal wall. The extramural type exhibits the presence of ventral pancreatic duct encircling the duodenum and the intramural type reveals the presence of pancreatic tissue woven into the duodenal wall with evidence of small ducts draining into the duodenum.[21]

History and Physical

Clinical manifestations of annular pancreas can occur at any age, from infancy to adulthood, and considering its bimodal age distribution, it precisely depends on the age of presentation and the degree of duodenal constriction.[8] Infants with AP associated with significant duodenal constriction present with non-bilious vomiting, bloating, and feeding intolerance. Other infants with minimal constriction or non-existent constriction can remain asymptomatic lifelong and a small fraction can become symptomatic usually between the third and sixth decade of life presenting with symptoms of abdominal pain, duodenal obstruction, and pancreatitis as evidenced by the review of a large case series cohort by S.J.S. Nagpal et al.[1] The other less common manifestations of AP in adulthood are peptic ulcer disease (PUD) and biliary obstruction.[23][24] The association of chromosomal abnormalities in AP is well known, with trisomy 21 being the most frequently detected anomaly.[25]

Many infants with AP also have various other associated congenital anomalies such as malrotation, tracheoesophageal fistula, esophageal atresia, duodenal atresia, renal anomalies, duodenal diverticulum, pancreas divisum, biliary atresia, anorectal malformations, and congenital heart disease.[5][24][25] In adults with AP, the most common associated anomalies were pancreatic divisum, malrotation, duodenal webs, and Schatzki ring.[25]

Evaluation

There is no biochemical test or genetic test available specific for annular pancreas (AP). The diagnosis of AP is radiologic and usually made incidentally or on the evaluation of clinical manifestations of AP. The diagnosis of AP can be made prenatally, preoperatively, or intraoperatively by many distinct non-invasive and invasive diagnostic techniques. Prenatally, AP can be diagnosed by prenatal ultrasonography.[25][26] In infants presenting with signs of intestinal obstruction, the diagnosis can be made by ultrasonography or plain abdominal radiographs, which demonstrates the classic “double bubble sign,” which although is nonspecific for AP.[27][28]

Newer techniques of ultrasonography incorporating upper gastrointestinal (GI) saline-contrast have been studied to reduce the possibility of misdiagnosis of neonatal AP.[29] In adults, the diagnosis is usually made by computerized tomography (CT) imaging or magnetic resonance imaging (MRI). A large retrospective case series by S.J.S. Nagpal et al. showed a significant number of patients diagnosed by CT imaging alone.[1]

Other available imaging techniques to aid in the diagnosis of AP are magnetic resonance cholangiopancreatography (MRCP), endoscopic retrograde cholangiopancreatography (ERCP), or endoscopic ultrasonography (EUS).[30][31] Sandrasegaran et al. showed that complete encasement of the duodenum with pancreatic tissue is not essential for the diagnosis of AP.[8] Newer imaging techniques with positron emission tomography (PET)/CT incorporating radiotracers C-11 choline and F-18 fluciclovine have demonstrated the diagnosis of AP.[32] Despite many advances in diagnostic techniques, the gold standard test for diagnosing AP remains to be laparotomy with a thorough gross examination of the duodenum and the head of the pancreas.[25]

Treatment / Management

Best practice guidelines for the management of annular pancreas are lacking, and the treatment of AP is individualized based on presenting symptoms.[28] Asymptomatic patients who do not demonstrate signs and symptoms of AP can be followed up closely without any intervention.[5] Patients presenting with symptoms of acute pancreatitis should be managed with supportive care. Recurrent disabling attacks of acute on chronic pancreatitis can be considered for a pylorus-preserving Whipple's procedure on a case by case basis.[1]

Division of the pancreatic annulus is not advised due to the increased incidence of postoperative complications like pancreatitis, pancreatic fistulas, and duodenal stenosis.[28] The management for symptomatic patients from an obstructing AP is purely surgical with duodenal bypass procedures such as gastrojejunostomy, duodenoduodenostomy, or duodenojejunostomy in both adults and children.[17][28]

In contrast to children undergoing duodenal bypass procedures, Zyromski et al. highlighted the complex pancreaticobiliary pathology associated with AP in the adult population by describing that 20% of adult patients in their study group requiring surgical intervention underwent complex pancreaticobiliary surgical procedures which included pancreaticoduodenectomy, lateral pancreatojejunostomy, biliary/pancreatic sphincteroplasty, and bypass of the biliary system.[25]

Differential Diagnosis

As the clinical manifestations of annular pancreas exhibit a bimodal age distribution presenting either early during infancy or adulthood, the differential diagnosis of this anomaly is also based on the age of presentation. Infants commonly present with duodenal obstruction with the classic "double bubble sign" on imaging, and the differential diagnosis can be classified into either intrinsic or extrinsic causes.[33] Duodenal atresia, duodenal stenosis, paraduodenal hernias, Meckel diverticulum, and duodenal webs are some important intrinsic causes that need to be considered.[27]

Besides AP, the other notable extrinsic causes that need to be considered are malrotation of the gut or midgut volvulus. Peptic ulcer disease, pancreatic divisum, primary duodenal and pancreatic malignancies should be considered in the differential diagnosis of an adult patient suspected to have AP.[34] Gastroduodenal tuberculosis should be considered in patients from geographical areas endemic for tuberculosis.[35]

Prognosis

The prognosis of annular pancreas in children is excellent with increased overall survival rates, despite the presence of associated congenital malformations and chromosomal anomalies.[36] This has been attributed to improved neonatal care, nutritional management, early detection, and management of associated anomalies.[24] The prognosis of AP in adults is favorable if it is not complicated by underlying malignancy.

Complications

Complications of annular pancreas are secondary to the mechanical migratory effect of the pancreatic anlage and include acute and chronic pancreatitis, common bile duct obstruction, peptic ulcer disease, cholecystolithiasis, and pancreatitis.[24] Emerging data from large case series studies report the increased incidence of neoplastic processes in adult patients with AP.

Deterrence and Patient Education

Annular pancreas is a rare congenital abnormality that causes the encasement of the duodenum by a band of pancreatic tissue. Previously, this anomaly was noted on autopsies and abdominal surgeries, but with the emergence of newer diagnostic modalities, it has been recognized more regularly. Most individuals with this abnormality are asymptomatic but can present with features of intestinal obstruction in infancy or abdominal pain, peptic ulcer disease, and pancreatitis during adulthood. This condition is often associated with chromosomal abnormalities, with the most common being trisomy 21. The management for symptomatic patients from AP is purely surgical with duodenal bypass procedures such as gastrojejunostomy, duodenoduodenostomy, or duodenojejunostomy in both adults and children.

Enhancing Healthcare Team Outcomes

Annular pancreas is not commonly encountered in clinical practice and is primarily considered to be a disease of infancy, which is untrue as the diagnosis of AP is made with nearly equal frequency in children and adults. However, specific guidelines outlining the management of AP are lacking; therefore, the management must be individualized.[28] A holistic approach is needed for the care of these patients, which can help achieve the best possible outcomes. An interprofessional team that includes neonatologists, surgeons, gastroenterologists, and radiologists needs to be familiar with this condition and the methods available for making a diagnosis and its management.[25][37]

Timely identification and management of chromosomal anomalies and malformations associated with this condition are important to reduce morbidity and mortality. Prenatal diagnosis of this anomaly helps in preplanning and management at a center equipped with advanced neonatal care. [38] Assessment of nutritional status and optimizing nutrition are important in the management of AP. Adult patients with a diagnosis of AP should be followed up closely, considering the incidence of neoplasia associated with this anomaly is significant.[25]

Review Questions

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