

Case Report

Annular pancreas – a case report and review of the literature

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Abstract

Introduction: Annular pancreas is a very rare congenital problem that can manifest clinically from neonatal age to adulthood. Here we present a rare case of Annular pancreas which presented in neonatal period. The child presented with bilious vomiting and the AXR revealed classical Double bubble appearance. Laparotomy revealed the diagnosis and we did Kimura's diamond shaped Duodeno-duodenostomy. Postoperatively we gave peripheral TPN for 10 days.

Keywords: Duodenal obstruction, Malrotation, Duodenal atresia.

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INTRODUCTION

Annular pancreas (AP) is a rare congenital abnormality characterized by a complete or incomplete ring of pancreatic tissue surrounding the descending portion of the duodenum¹. It was described in 1862 by Ecker, since then only 737 cases of annular pancreas have been reported in the literature⁸. Several theories have been proposed but the etiology is still unclear. It is thought to originate from incomplete rotation of the pancreatic ventral bud and can present at any age from neonate to adulthood. The age of presentation depends upon the severity of obstruction. Up to two-thirds of cases are totally asymptomatic. ERCP studies have shown a higher prevalence of 1 in 250, or 400 cases per 100,000, adults². In this article, we report a case of annular pancreas in a neonate and review of the literature.

CASE HISTORY

A 2 days old FTND 2nd born male baby was brought to the hospital for C/O Bilious vomiting of 1 day duration. O/E: Baby was pink, active and crying normally. There was mild dehydration. The Upper abdomen was mildly distended. Bowel sounds were present. An AXR revealed double bubble appearance (Fig 1). The preoperative differential diagnosis were Duodenal Atresia, Annular Pancreas and Malrotation with Ladd's band. Without further investigations the baby was taken up for surgery. On Laparotomy the baby was found to have Annular Pancreas (Fig 2). Hence Kimura's diamond shaped Duodeno-duodenostomy was done (Fig 3). Post op baby was started on peripheral vein TPN for 6 days and then slowly oral feeds were introduced. Baby was discharged on 10th post op day when the baby was tolerating normal oral feeding.



Figure 1: X-ray abdomen showing the classical Double Bubble appearance



Figure 2: Descending portion of duodenum showing annular pancreas on laparotomy.



Figure 3: Kimura's diamond shaped Duodeno-duodenostomy was done.

Embryology

The pancreas develops from a single dorsal and two ventral buds that appear in the fifth week of gestation as outgrowths of the primitive foregut. The two ventral buds rapidly fuse. By the seventh gestational week, expansion of the duodenum causes the ventral bud to rotate and pass behind the duodenum from right to left and fuse with the dorsal bud. The ventral bud forms the inferior part of the uncinata process and the inferior head of the pancreas, and the dorsal bud gives rise to the tail and the body. Fusion of the ducts of the two buds produces the main pancreatic duct. Annular pancreas results from failure of the ventral bud to rotate with the duodenum, resulting in encircling of the duodenum. This encircling is usually by a band of pancreatic tissue that completely encircles the second portion of the duodenum. It may be even incomplete, leaving the anterior portion of the duodenum unconstricted. The pancreatic band is usually interspersed with the duodenal muscle, although it can also be free from the duodenum.

Several theories have been proposed to explain this anomaly:

- Adherence of the right ventral bud to the duodenal wall prior to rotation resulting in its persistence and encirclement of the duodenum (Lecco's theory)³.
- Persistence and enlargement of the left ventral bud (Baldwin's theory)⁴.
- Hypertrophy and fusion of the ventral and dorsal buds to form the annulus before rotation of the gut resulting in complete encirclement of the duodenum⁵.
- Another theory explained by Verga in 1972²⁵, suggests that the primary abnormality is duodenal with the pancreas "filling the space" around a narrowed duodenum. This results in a complete or in-complete stenosis of the duodenal lumen.

None of the above theories explain the variations in position of the annular ducts⁶. Kamisawa *et al* proposed

that the tip of the left ventral bud adheres to the bud and stretches to form a ring⁴¹. There are some reports of familial annular pancreas supporting a genetic basis for the disease, as the same has been described in siblings³⁷⁻³⁹. In a recent study on *Xenopus* embryos it has been shown that the cells that form the annulus are derived entirely from the ventral pancreas²⁶. It has also been demonstrated that the tetraspanin Tm4sf3 which is localized to the ventral pancreas regulates fusion of the dorsal and ventral pancreatic buds. The inactivation of trans-membrane 4 super family member 3 (TM4SF3) inhibited fusion of the dorsal and ventral pancreatic buds. Over expression of the same gene promote development of annular pancreas.

In another study 42% of mice embryos with a targeted inactivation of the Indian Hedgehog (IHH) gene, a member of the mammalian Hedgehog family, changes in the morphology of the ventral pancreatic bud similar to annular pancreas were seen⁴⁰. Johnston *et al* in 1978 classified annular pancreas into two distinct subtypes namely the extramural and intramural type. In the Extramural type, the presenting symptoms are those of high gastrointestinal obstruction. In the Intramural type, the symptoms are those of duodenal ulceration. Annular pancreas is also frequently seen after inactivation of Sonic Hedgehog (SHH) SHH is not expressed in pancreatic tissue; therefore it has been suggested that a defect in duodenal Hedgehog signaling may lead to the anomaly because both SHH and IHH are expressed in the developing gut^{27, 28}. SHH knockout mice embryos also demonstrate a number of gastrointestinal abnormalities, strikingly similar to that seen in patients with annular pancreas.

CLINICAL FEATURES

The age at presentation depends upon the severity of duodenal obstruction. AP can become symptomatic only at adult hood, usually being in the third to sixth decade of life. About one half and two thirds of cases of annular

pancreas in adults remain asymptomatic²⁴. Congenital anomalies and duodenal obstruction are the predominant features in children, and pancreatitis is the main presentation in adults⁸. More than two-thirds of children present during the neonatal period especially during the first week, typically with features of gastric outlet obstruction including feeding intolerance, bilious vomiting and abdominal distension⁹. In infants, the presence of AP has been most commonly associated with Down's syndrome, the rest being maternal polyhydramnios, esophageal atresia, duodenal atresia, intestinal malrotation, situs inversus, tracheoesophageal fistula, omphalocele, imperforate anus, Meckel's diverticulum and cardiac abnormalities¹⁰⁻¹². The presentation in adults differs from that in children. Adults may present with gastric outlet obstruction, upper GI bleeding (from peptic ulceration), acute or chronic pancreatitis and rarely biliary obstruction¹³⁻¹⁶. Obstructive symptoms presenting in adults may be due to repeated inflammation, edema leading to fibrosis and scarring. Impaired pancreatic flow through the annular duct causes edema and fibrosis which is limited to the annular duct and adjoining duct in the pancreatic head. It can cause partial obstruction of these ducts leading to jaundice and pancreatitis¹⁷. In addition, the incidence of pancreatic and periampullary neoplasia in adults with annular pancreas is substantial and should be considered when the diagnosis of annular pancreas is established in adults^{19, 20, 42-45}.

DIAGNOSIS

In symptomatic neonates a plain abdominal X-ray or ultrasound will show the classic 'double bubble' sign- air in the stomach and first part of duodenum suggestive of duodenal obstruction. A double bubble sign is not specific for annular pancreas since it can also be seen in other conditions like duodenal atresia, and intestinal malrotation²¹⁻²⁴. However, no further testing is usually required because all patients in this age group with complete or partial duodenal obstruction require surgery. In older children and adults; the diagnosis is usually made with an upper GI series or an abdominal CT scan, although surgery remains the diagnostic gold standard. AP should be considered if the UGI series or abdominal CT scan shows descending duodenal narrowing. The upper GI series may show an annular filling defect across the second portion of the duodenum, symmetrical dilatation of the proximal duodenum, or reverse peristalsis of the duodenal segment proximal to the annulus^{16, 19}. In an abdominal CT scan a complete ring of pancreatic tissue around the duodenum is not required for a diagnosis of annular pancreas. An incomplete annulus most likely will have only a thin band of pancreatic tissue

which may not be seen at CT or MRI, or it may be incorporated in the duodenal wall^{29,30}. Nevertheless, an imaging finding of pancreatic tissue extending in a postero-lateral direction to the second part of the duodenum in the appropriate clinical setting, such as unexplained chronic pancreatic or gastric outlet obstruction, should raise the suspicion of annular pancreas. The antero-lateral extension of pancreatic tissue to the second part of the duodenum is less specific for annular pancreas. ERCP is helpful in the diagnosis of AP, especially when the results from barium studies or CT are equivocal. It allows for the exact delineation of the anatomic structure of the accessory pancreatic duct and the C-loop duct encircling the duodenum^{13, 29}. AP has been classified into six types based upon the drainage site of the annular duct:

- Type I (the most common form) in which the annular duct flows directly into the main pancreatic duct
- Type II (the second most common variant) in which the duct of Wirsung encircles the duodenum but still drains at the major papilla
- The other four subtypes are much less common²⁰.

The best non invasive method of ascertaining the ductal configuration and the presence of disease in annular pancreas appears to be secretin-enhanced MRCP. In up to 40% of cases, a definitive diagnosis is made at the time of laparotomy only⁷.

MANAGEMENT

Surgery remains the procedure of choice in patients with symptomatic Annular pancreas. The optimal operation has been a matter of debate. The goal of surgery is to relieve duodenal or gastric outlet obstruction^{7,19} by a bypass surgery of the annulus, which can be achieved via a duodenoduodenostomy, gastro jejunostomy, or a duodenojejunostomy. Resection of the annulus should be avoided since it is associated with serious complications such as pancreatitis, pancreatic fistula formation, and incomplete relief of obstruction leading to unacceptably high morbidity^{9,14,33}. In neonates, duodenoduodenostomy has replaced duodenojejunostomy as the treatment of choice because it has a lower incidence of postoperative complications, particularly obstruction and blind-loop syndromes³⁰. An end-to-end or side-to-side duodenoduodenostomy should be performed, ensuring adequate mobilization of proximal and distal ends. When the first part of the duodenum is distended, a tapering duodenoplasty or plication can be performed. A gastrostomy tube may be placed in those patients with other complex gastrointestinal malformations or chromosomal abnormalities. In early pediatric series when duodenoduodenostomy was done, mortality rates

remained high, likely due to the presence of other congenital malformations and the lack of proper perioperative care³¹⁻³³. Outcome after surgery has improved markedly, with early mortality rates decreasing from 83% in the 1950s³² to less than 10% in most recent series^{30,8} due to improvements in surgical techniques and advances in neonatal intensive care and anesthesia. Deaths in contemporary series are usually attributed to severe associated congenital anomalies. In adults (in whom the duodenum is less mobile) a duodenojejunostomy or gastrojejunostomy is recommended. The extramural obstructing type should be treated by a bypass procedure mentioned above and the intramural type with duodenal ulcer should be treated by subtotal gastrectomy with or without vagotomy. In the presence of AP associated with obstructive jaundice, biliary bypass with choledochenterostomy or placement of an internal stent may be necessary to relieve the obstruction^{3,21}. In adults even Pancreaticoduodenectomy has been recommended when AP is associated with pancreatolithiasis or if associated with periampullary lesions and suspected coexisting malignancy^{22,36}.

CONCLUSION

Annular Pancreas is a rare disorder which is not difficult to diagnose. We treat this condition without touching the actual organ involved. The treatment modalities have evolved and are fairly well defined today.

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