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Case Report

ANNULAR PANCREAS WITH DUODENAL OBSTRUCTION- CASE REPORT AND REVIEW OF LITERATURES

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Abstract:

Annular pancreas as a cause of extrinsic duodenal obstruction is rare; with estimated incidence of one in 12,000-15,000 live births. Age of presentation varies due to differences in severity of symptoms of upper gastrointestinal tract obstruction. The band of pancreatic tissue encircling the 2nd part of Duodenum is responsible for narrowing of duodenal luminal diameter which subsequently determines the degree of obstruction. Various methods of bypassing the constriction without injuring the pancreatic tissue, are available; of them diamond shaped duodeno- duodenostomy as described by Kimura is widely practiced. The surgical outcome is satisfactory if there are no associated risk factors like prematurity, low birth weight, sepsis and other congenital anomalies. We have treated only three cases in last five years from January, 2011 to December, 2015. Two of them were neonate with acute presentation and one aged five and half years with chronic symptoms. Diamond shaped duodeno-duodenostomy was done in all patients. One of the neonate expired postoperatively due to sepsis and rest of the two had uneventful surgical recovery.

Key Words: Annular Pancreas, Duodenal obstruction.

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Introduction:

Several varieties of intrinsic and extrinsic congenital lesions can cause complete or partial obstruction of duodenum¹. Annular pancreas (AP) is a rare cause of extrinsic obstruction with an incidence of 1 in every 12,000-15,000 live births². Zyromski et al found only 737 reported cases of AP in English literatures³. The pancreas begins to develop from the endodermal lining of the duodenum in the 4th week of gestation- one larger dorsal and a smaller ventral pancreatic primordium. Ventral bud then rotates dorsally to fuse with the dorsal bud during the 8th week. If the ventral bud fails to rotate completely, it remains anterior to the duodenum, and fusion with the dorsal pancreatic primordium results in a ring of pancreatic tissue encircling the duodenum, creating an annular pancreas with concomitant partial or complete obstruction^{4,5,6}. Duodenal atresia or, stenotic web may underlie the annulus; the luminal diameter at the affected site directly determines the degree of obstruction and is inversely related to symptoms⁷. We have treated only three patients of annular pancreas creating duodenal obstruction in our center in last five years and in all of them-diagnosis was confirmed during laparotomy.

Case 1:

A 5 year 6 months old boy admitted with complaints of recurrent upper abdominal pain, occasional bilious vomiting, following or just after meal for the last 3 years and fullness in upper abdomen with visible movement for 4 years. He was delivered by normal vaginal delivery at home in rural area without proper antenatal followup and screening. There was no history of consanguinity and parents observed no other symptoms except poor weight gain. Physical examination revealed failure to thrive –weighing only 11 kg, vital signs within normal limit and no features of anaemia, dehydration, edema. Upper abdomen was distended with visible peristalsis from left to right. Other features in abdomen were unremarkable. Ultrasonography (USG) showed dilated stomach, pylorus and duodenal cap; regurgitation of food materials from 1st part of duodenum to stomach was also obvious. Upper gastrointestinal tract (GIT) contrast study showed partial obstruction at 2nd part of duodenum with delay in gastric emptying. Hemoglobin, electrolytes and renal functional status were within normal limit. Exploration was planned with provisional diagnosis of malrotation with Ladd's band. On laparotomy, annular pancreas causing incomplete obstruction at 2nd part without any features of malrotation was found. Diamond shaped duodenoduodenostomy was made between proximal and distal part of obstruction without interfering the band of pancreatic tissue. No intrinsic cause of obstruction (ie septum / membrane) was detected. Postoperatively uneventful recovery observed; Sips of water followed by liquid diet was allowed from 5th postoperative date. He was discharged on 10th postoperative day with advice to take anti-ulcer and pro-kinetic agents for further 6 week. Follow up after 6 week showed weight gain of 1.5 kg with no upper GIT symptoms.

Case 2:

A 14 days old term male baby of non consanguinous parent admitted with features of acute intestinal obstruction with septicemia and jaundice. The baby was delivered by normal vaginal route at home with no antenatal checkup and weighing 2400 gm. Plain X-ray abdomen (fig. 1) revealed double bubble sign with paucity of gas distally. Finding of abdominal ultrasound (fig. 2) was also suggestive of almost complete duodenal obstruction at 2nd part. After initial assessment and attempt of resuscitation; emergency exploratory laparotomy was planned as there was no improvement. On exploration annular pancreas with a pin holed duodenal septum producing incomplete obstruction at 2nd part of duodenum was observed. The baby also had features of malrotation. Ladd's procedure and diamond shaped duodenoduodenostomy were done. Patient was expired on 2nd postoperative day due to ongoing sepsis.



Fig 1: *Plain X-ray abdomen showing double bubble sign with distal paucity of gas.*

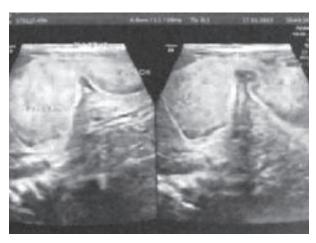


Fig 2: USG showing duodenal obstruction at second part.

Case 3:

A 10 days old female baby of non consanguinous parent with history of delivery by caesarean section for fetal distress admitted with history of bilious vomiting and passage of scanty amount of meconium since birth. On examination she was icteric, dehydrated, weighing 2300 gm with fullness in upper abdomen; and other findings on physical examination were normal. USG abdomen revealed duodenal obstruction; Barium meal X- ray and follow through examination (fig. 3) observed almost complete obstruction in the 2nd part of duodenum with abnormal dilatation of 1st part. Laparotomy with diamond shaped duodeno-duodenostomy was done to bypass the constricting band of pancreatic tissue at 2nd part. No intrinsic obstruction was noted. Postoperative period was uneventful and breast feeding was allowed from 7th postoperative day and patient was discharged from hospital on 10th postoperative day. No complain regarding feeding was observed on follow up after 2 week and 6 week with normal weight gain.



Fig 3: Barium meal examination and follow through showing almost complete obstruction at 2nd part of Duodenum with proximal dilatation.

Discussion:

The spectrum of abnormalities causing extrinsic duodenal obstruction includes annular pancreas, pre duodenal portal vein, Ladds band and volvulus^{7,8,9,10}. Tiedman first described annular pancreas in 1818 and Ecker subsequently supported it in 1862¹¹. The main characteristic feature is a band, or a ring of pancreatic tissue; which usually spans the descending duodenum close to ampulla of Vater resulting from the pathologically positioned embryonic part of the developing pancreas^{4,12}. There are three

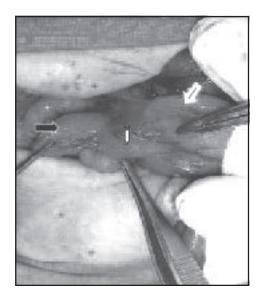


Fig 4: Peroperative picture showing proximal dilated duodenum (2) and distal narrow part () with intervening band of pancreatic tissue () creating stenosis.

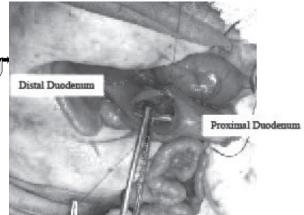


Fig 5: Diamond shaped anastomosis is going on with nasogastric tube in situ showing no distal obstruction.

developmental theories behind the formation of an annular pancreas: i) Baldwin in 1910: failure of atrophy of left component of ventral pancreatic anlage; ii) Lecco's theory: ventral pancreas adhered to surrounding tissues at its site of origin in the ventral mesogastrium; iii) Verga in 1972: the pancreas is filling the space around the abnormally narrowed duodenum where the primary abnormality is in duodenum¹. In 75%; the duodenum is circled only partially and in rest totally¹³. This condition may be completely asymptomatic and may be diagnosed in adulthood or even at necropsy. Annular pancreas frequently coexists with intrinsic obstruction and 40% of annular pancreas cases are associated with duodenal atresia or obstruction that needs immediate surgical attention¹⁴. One of the neonates in our report had additional duodenal web creating intrinsic obstruction. Majority of duodenal obstructions either complete or incomplete could be detected before birth due to advance in prenatal care^{12,15,16}. Maternal polyhydroamnios noted in 30-65% cases^{1,17}. Classic double bubble obstructive pattern is usually identifiable on fetal ultrasonography, an annular pancreas may also be recognizable^{2,10,12,16,18,19,20}. None of the mothers of the affected babies in our report received proper antenatal follow-up and so screening was not possible. Repeated bilious vomiting with upper abdominal fullness is the characteristic clinical feature in newborn. The recognition of partial obstruction may be considerably delayed if the obstruction is minor as we have observed in our case 1; who was a 5 year 6 months old boy⁷. In this situation, symptoms may first occur when the infant starts taking solid food from formula or, may only be unmasked much later in infancy, childhood or, rarely in adulthood due to progressive decrease in motility or impaction of food^{7,21,22}.

USG abdomen was done in all three patients but duodenal obstruction was suspected in only one (Fig. 2). Plain X-ray abdomen is also helpful to show typical double bubble shadows with or without distal gas (Fig. 1). The upper gastrointestinal contrast study is the most useful for evaluation of older infants and children with symptoms of chronic partial obstruction (Fig. 3); gastroduodenoscopy may also be done^{23,24}.

Immediate surgical correction is not necessary. Surgery should be done once the infant is optimized and associated anomalies have been appropriately studied. Common associated anomalies with annular pancreas were Trisomy 21, congenital heart disease, malrotation, duodenal atresia and stenosis- as observed by Yigiter⁸. We have not diagnosed any associated anomalies in our patients.

It is only in the last half of 20th century when effective treatment of annular pancreas was first accomplished. Gross and Chisholm in 1944 reported first successful operation of duodenal bypass instead of releasing the band of pancreatic tissue around the duodenum²⁵. Morbidity and mortality from bypass was understood in later reports explaining that annular pancreas may not only cause extrinsic obstruction but may lead to or be associated with intrinsic obstruction also²⁶. The operation is best accomplished by right upper quadrant

transverse incision and entire duodenum should be inspected to locate the probable site and type of obstruction by mobilizing right colon along with hepatic flexure. When there is continuity of proximal and distal duodenum a duodeno-duodenostomy joining the bowel just proximal and distal to obstruction is the best corrective option⁷. In patients with annular pancreas, pancreatic tissue will be seen extending circumferentially around the 2nd part of duodenum (Fig. 4) and the ring of pancreatic tissue should never be transected because of major ductal structures that traverse it. Damage to these structures will lead to leakage of pancreatic fluid and or, pancreatitis^{7, 27}. A diamond anastomosis (Fig. 5) first described by Kimura is preferred over the simple anastomosis ²⁸ ^{,29,30}. This asymmetric anastomosis will maintain itself in a more wide open position, and permit earlier transit of duodenal contents. Proper positioning of nasogastric tube and presence of any intrinsic obstruction must be checked. Postoperatively, total parenteral nutrition (TPN) should be maintained along with monitoring of nasogastric tube output. But in our centre TPN facility is not available and we just put intravenous fluid with glucose, electrolytes and fresh blood/fresh frozen plasma in selective patients. Oral feeding was possible within 5-7 postoperative day in both of our survived patients and discharged within 8-12 days after operation. Mean time till onset of oral feeding in different studies ranged 4.1 to 15.9 days^{3,8,26,30,31} and mean hospital stay ranged 9 to 24 days^{3,26,30,31}. Zyromski³ observed 6% mortality and Yigiter⁸ -36.3% mortality in there series respectively. Gastroduodenal dysfunctions like gastric stasis, gastritis, gastroesophageal reflux, duodeno-gastric reflux, peptic ulcer disease and delayed transit are observed late complications following surgical correction ^{8,26,29,32}.

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