International Journal of Radiology Research
www.radiologyjournal.in
E-ISSN: 2663-4562, P-ISSN: 2663-4554
Received: 12-10-2020; Accepted: 27-10-2020; Published: 17-11-2020
Volume 2; Issue 2; 2020; Page No. 42-45

Annular pancreas combined with intestinal malrotation: A rare case of multiple alimentary tract anomalies in a neonate

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Abstract
Malrotation of the mid gut is the most common congenital anomaly which might be present in infants, children or adults at various point of time in the form of acute intestinal obstruction or intestinal ischaemia with caecal or midgut volvulus or with chronic abdominal pain. Annular pancreas is a rare congenital anomaly which is consists of a rim of pancreatic tissue which encircles the second part of duodenum either partially or completely. Annular pancreas may be asymptomatic for a very long period of time before presenting with vomiting or features of intestinal obstruction. Here, we are presenting two female neonatal patient presented to us with bilious vomiting, weight loss, and irritability, abdominal distention who was diagnosed as mid gut volvulus which eventually turned out to be malrotation of mid gut and with incidental finding of annular pancreas.

Keywords: Annular pancreas, intestinal malrotation

Introduction

Annular pancreas (AP) is a rare congenital anomaly characterized by the presence of pancreatic tissue of variable width, completely or partially obstructing the second part of the duodenum. It forms due to the failure of normal migration of the ventral pancreatic bud, which results in some of the pancreatic tissue encircling the duodenum. It is estimated that it occurs in one of every 12,000–15,000 live births[2]. The incidence of annular pancreas has been reported to be 0.005%–0.015% in autopsy cases in adults. It is frequently associated with other congenital abnormalities, such as: esophageal atresia, imperforate anus, congenital heart disease, malrotation of the midgut and Down's syndrome. In contrast to adults, children with AP always present with signs and symptoms of gastrointestinal obstruction and AP in children is frequently associated with other congenital anomalies. Only 737 cases have been reported in the English literature to date, with a slight female preponderance. In newborns the main mode of presentation is that of duodenal obstruction. We report a rare case of a combination of annular pancreas with malrotation in a 14 old girl and 4 day old female infant.

Case reports

Two female infant was born at term without complications. At 14 & 4 days of old, both presented to a pediatric emergency department for investigation of frequent bilious vomiting, weight loss, and irritability, abdominal distention. An X-ray of the abdomen showed gross gaseous distention of the stomach, with minimal gas in the distal portion fig 1. The findings were suggestive of duodenal obstruction. Abdominal ultrasound confirmed the X-ray findings, revealing distention of the stomach. An upper gastrointestinal water soluble contrast study revealed that the duodenum and jejunum were distributed abnormally on right side with gross distention of the stomach fig 2. Based on the radiography findings, exploratory laparotomy was performed, which revealed a markedly distended stomach and proximal duodenum. Following mobilization of the duodenum, an annular pancreas, with complete duodenal obstruction, was found. Distally, the small bowel was normal fig 3. The annular pancreas was treated by Kimura's diamond shaped duodenoduodenostomy and the intestinal malrotation with Ladd's procedure.

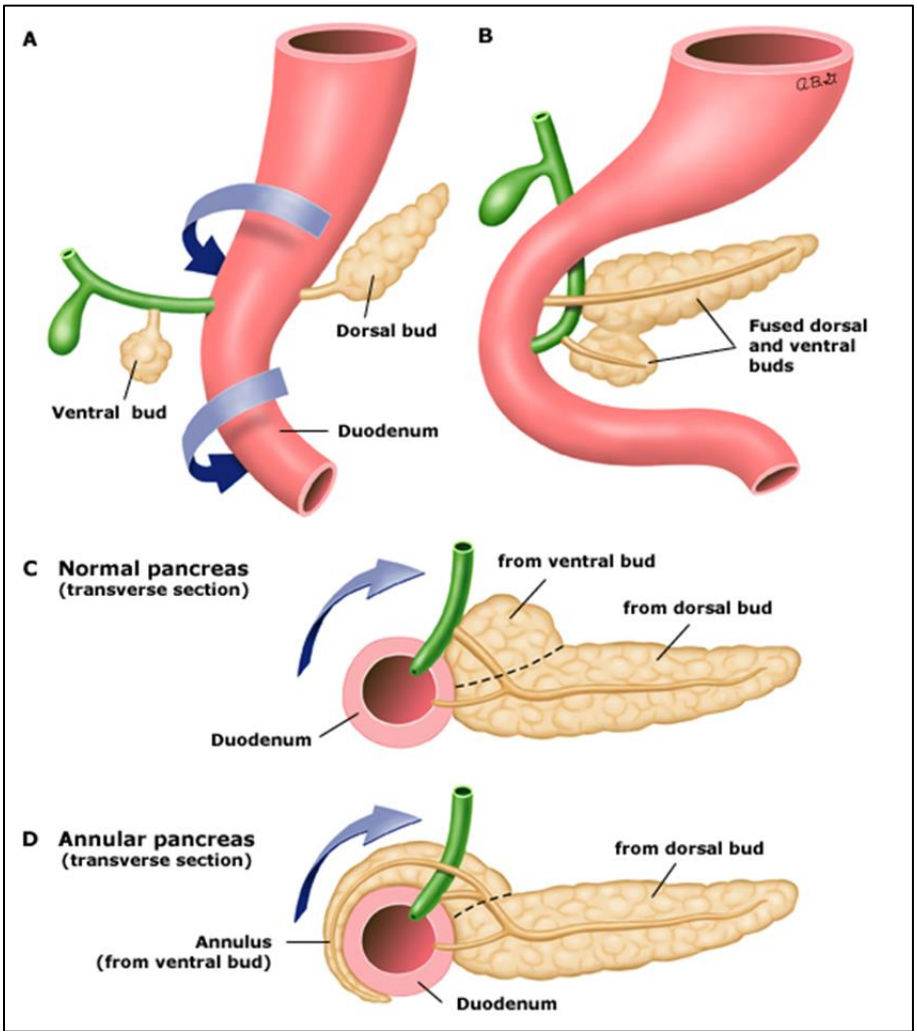

Fig 1

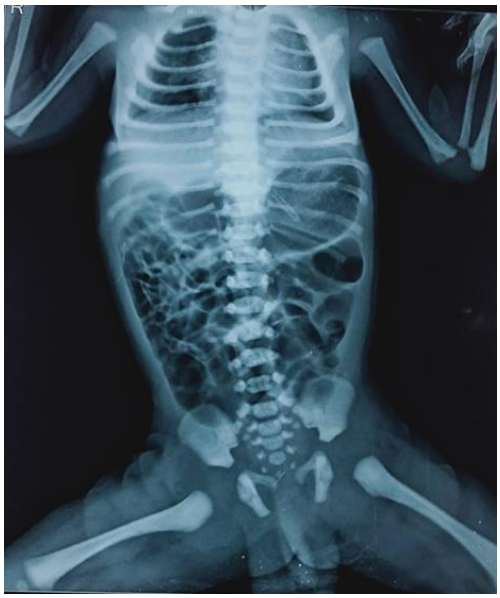

Fig 2: Plainradiogram abdomen, showing clumping of small bowel loops on the right side of the abdomen

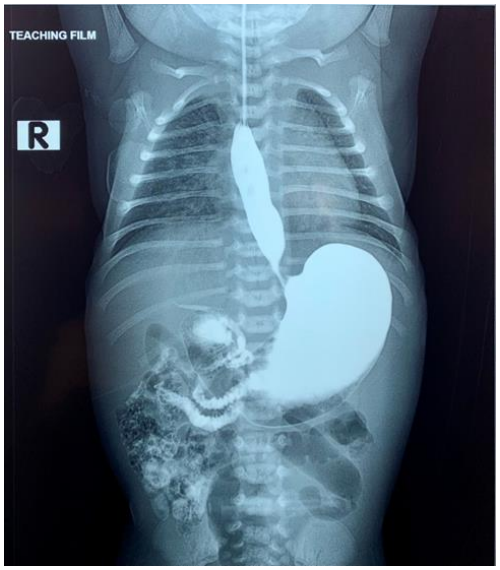

Fig 3: Upper GI contrast study through showing gross dilatation of stomach, narrowing of duodenum and third and fourth part of duodenum on the right side.

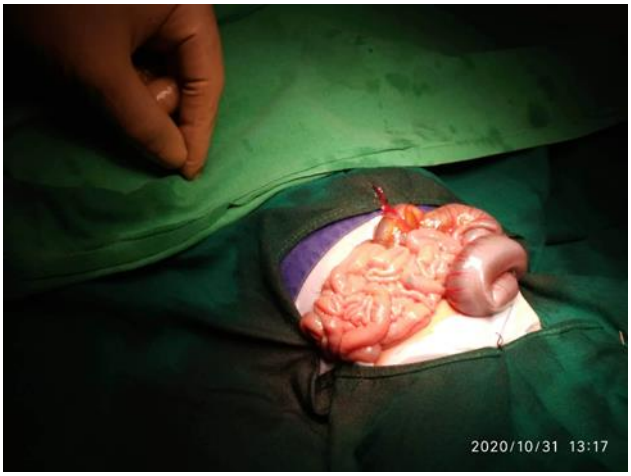

Fig 4: Operative photo showing clumping of small bowel loops.

Discussion

Annular pancreas (AP) is a rare congenital anomaly that is estimated to occur in 1 of 12000–15000 newborns [12]. Only 737 cases have been reported in the English literature to date, with a slight female preponderance. Annular pancreas, as first described by Tiedemann in 1818, is a rare congenital abnormality that accounts for 1% of all intestinal obstructions in the paediatric population [13]. The majority of the reported cases have been reported in the newborn population. It is thought to represent an aberration in the development of the ventral pancreatic bud and is characterized by an extension of pancreatic tissue around the second part of the duodenum fig 1. Malrotation of the intestinal tract is a congenital anomaly that refers to either lack of or incomplete rotation of the fetal intestines around the axis of the superior mesenteric artery during fetal development. Incidence is 3 of 2000 autopsies and in 3 of 24, 519 individuals who underwent abdominal surgeries.

Plain abdominal radiography and upper gastrointestinal contrast-enhanced radiography will confirm duodenal obstruction. Diagnosis of annular pancreas is made on the basis of one of the imaging modalities, namely, sonography and computed tomography, or by endoscopic retrograde cholangiopancreatography [14]. Duodenal stenosis was found. Based on these findings, the boy was diagnosed as having AP and intestinal anomaly. Based on these findings, the boy was diagnosed as having? AP and midgut malrotation, and exploratory operation was performed. The ectopic pancreatic tissue was soft and partly encircling the duodenum at the second part of the duodenum.

Congenital duodenal obstruction is relatively common during the neonatal period. It can be categorized as complete or partial and as intrinsic or extrinsic. Extrinsic duodenal obstruction has many causes, including annular pancreas, malrotation, and anterior portal vein. Its embryological origin begins between the fifth and seventh gestational weeks, when the two pancreatic buds (dorsal and ventral) rotate as part of the process of intestinal rotation. During that period, the duodenum rotates from left to right, the ventral pancreatic bud typically migrates posteriorly and inferiorly, merging with the more caudal portion of the pancreatic head and the uncinate process, and the dorsal bud develops into the body and tail of the pancreas. An annular pancreas is due to failure of the ventral bud to rotate, resulting in incarceration of the duodenum [6, 7].

Annular pancreas presentation has a bimodal distribution; the first peak is in infancy and the second one in the fourth decade of life. Pediatric cases usually present with intestinal obstruction or associated anomalies. Associated conditions include mongolism, intestinal malrotations, duodenal atresia or webs, tracheoesophageal fistulas and cardiac defects. Between one half to two thirds of cases of annular pancreas in adults remain asymptomatic. Adult annular pancreas is associated with duodenal obstruction (60%), pancreatitis (15-50%) and peptic ulceration (26-48%). Malrotation of mid gut with annular pancreas is very rare entity. Hyperechoic band completely surrounding the duodenum on antenatal as well as postnatal ultrasound feature [6, 9, 11] down syndrome, intestinal malrotation, gallbladder agenesis and CHARGE syndrome. Single umbilical artery, preaxial polydactyly, and annular pancreas are, associated anomalies of the annular pancreas. In general, an annular pancreas is symptomatic in children, especially in the neonatal period. It should be borne in mind that even if the radiological and endoscopic findings both suggest an annular pancreas, the definitive diagnosis is established only during surgery.

Duodenal atresia, duodenal stenosis, paraduodenal hernias, Meckel's diverticulum, and duodenal webs are some important intrinsic causes that need to be considered. In patients with symptoms of obstruction, laparotomy can reveal a band of pancreatic tissue surrounding the second portion of the duodenum, supporting the diagnostic hypothesis, which can be confirmed by examining the resected specimen.

Association of AP, intestinal malrotation in preadolescents is rare [1, 2]. Pre-operative radiological evaluation, careful evaluation of the gastrointestinal tract and a delicate operative technique are necessary to avoid misdiagnosis, missed diagnosis or repeated operations [3, 4]

Conclusion

Due to rarity, this combination of congenital duodenal obstruction must always be kept in mind during diagnosis of congenital duodenal obstruction we presented a case of a rare congenital developmental abnormality of the pancreas combined with intestinal malrotation. Duodenoduodenostomy is the appropriate treatment for annular pancreas and Ladd's procedure for malrotation

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Limitation of study

The present study suffered from the following limitation:

Small sample size

We are thankful to Dr Jayanthi, Principal, Mr. Manoj, Director and Dr Venkatachalapathy, Paediatric surgeon for their support

in academics.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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