Case Report

Duodenal atresia with bile-duct anomaly

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ABSTRACT

A full-term female infant is born via vaginal delivery to a 26-year-old primigravida with unexplained polyhydramnios and antenatal diagnosis of duodenal atresia. After birth, bilious fluid was aspirated through the orogastric tube and the initial abdominal radiograph was suggestive of double bubble sign with the absence of air in the distal bowel. Baby passed meconium at 4 h of life and repeat abdominal radiograph showed double bubble sign with the presence of air in distal bowel. Upper gastrointestinal contrast study done revealed distension of the stomach and duodenal bulb with an abrupt narrowing. It also revealed flow of contrast into the jejunal loops on the right side of the spine which was suggestive of intestinal malrotation as the leading diagnosis. However, intra-operative findings confirmed a Type III Duodenal atresia with bile proximal and distal to atretic. Occasionally, distal bowel gases can be seen in radiographs of infants with duodenal atresia due to abnormal development of the biliary system that can allow passage of air from the proximal duodenal segment to the distal. Here, we report a case and discuss the embryology of this rare entity.

Keywords: Anomalous duct, double bubble, duodenal atresia

INTRODUCTION

Failure of duodenal recanalisation results in a range of upper gastrointestinal (UGI) obstruction from stenosis to atresia. The incidence of duodenal atresia is 0.9 per 10,000 live births.[1] The double-bubble sign in the abdominal X-ray with the absence of distal bowel gases strongly suggests the presence of either duodenal atresia or severe stenosis. An antenatal ultrasound can also demonstrate a similar picture along with polyhydramnios. The proximal gas shadow represents the dilated stomach and the distal gas shadow represents the dilated duodenal segment proximal to the obstruction.[2] If bowel gases are observed distal to the double bubble then the usual possible diagnosis are duodenal stenosis, duodenal web and intestinal malrotation with midgut volvulus. Occasionally, distal bowel gases can be seen in radiographs of infants with duodenal atresia due to abnormal development of the biliary system. An abnormal bile-duct channel is formed between the two duodenal segments that can allow passage of air from the proximal duodenal segment to the distal.[3-5] Here, we report a case of an infant with duodenal atresia with distal gas shadows due to anomalous biliary channel.

CASE REPORT

A female infant was born by vaginal delivery at 40 weeks to a 26-year-old primigravida woman who had polyhydramnios with a double bubble sign on antenatal scan suggestive of duodenal atresia. The baby was born vigorous with Apgar scores of 8 and 9 at 1 and 5 min, respectively. There were no apparent congenital anomalies or facial dysmorphisms related to aneuploidy present. Her birth weight, length and head circumference were 2400 g (2nd percentile),

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48 cm (27th percentile) and 33 cm (23rd percentile), respectively, as per the WHO growth chart. The baby was shifted to the neonatal care unit and abdominal X-ray done revealed a double-bubble sign with the absence of distal gas [Figure 1]. She had continuous bilious aspirates through the orogastric tube. These radiographic and clinical findings were consistent with the diagnosis of duodenal atresia, but she passed meconium 4 h after birth which was not expected in a case of duodenal atresia. A repeat X-ray abdomen done showed a double-bubble sign but this time with the presence of distal gas shadows [Figure 2]. To exclude the possibility of other differentials like malrotation, an UGI contrast study was done that showed dilated stomach and proximal duodenum with the passage of small amount of contrast into the distal small bowel. With these findings, possible pre-operative diagnosis of duodenal stenosis or duodenal web or intestinal malrotation was made and the baby was taken up for laparotomy. Ultrasound kidney, ureter and bladder and echocardiogram were normal. Intraoperatively, duodenoduodenostomy was performed as Type III duodenal atresia was found with the presence of bile in the distal duodenum. Post-operative course remained uneventful and feeds could be started by post-operative day 5 and she reached full feeds by post-operative day 14.

DISCUSSION

During embryogenesis, various events such as folding, lengthening and luminal dilatation result in the formation of the foregut, midgut and hindgut. Formation of the duodenum starts at around the 4th week of gestation with the active proliferation of duodenal epithelium that completely obstructs its lumen converting it into a solid cord. Later, this solid cord recanalises by a process called coalescence of vacuoles into two parallel channels and each channel receives its own hepatopancreatic duct and accessory pancreatic duct. Later, these two duodenal channels fuse and only one hepatopancreatic duct and one accessory pancreatic are left behind to serve the new duodenum as the other two regress.[3-5]

Delayed recanalisation of the bile duct supersedes and restricts the duodenal recanalisation causing duodenal atresia that would most of the time appear near the periampullary region. An anomalous biliary system may coexists where the hepatopancreatic duct bifurcates into two branches and enters both proximal and distal to the duodenum's atretic segment.[4,5] Failure to pass meconium and bilious vomiting without abdominal distension strongly indicate a proximal intestinal obstruction. However, in this case, the passage of meconium and the presence of the distal bowel gas on subsequent radiographs did not suggest classical duodenal atresia. Various differentials in such a scenario would be a duodenal web or stenosis or intestinal malrotation.[3] In this case, UGI contrast study revealed distension of the stomach and duodenal bulb with an abrupt narrowing. There was also flow of contrast seen in the jejunal loops that were present on the right side of the spine. These findings were consistent with intestinal malrotation with midgut volvulus rather than a duodenal web [Figure 3]. However, none of these conditions were present in this case as during laparotomy a complete duodenal obstruction was found in the form of

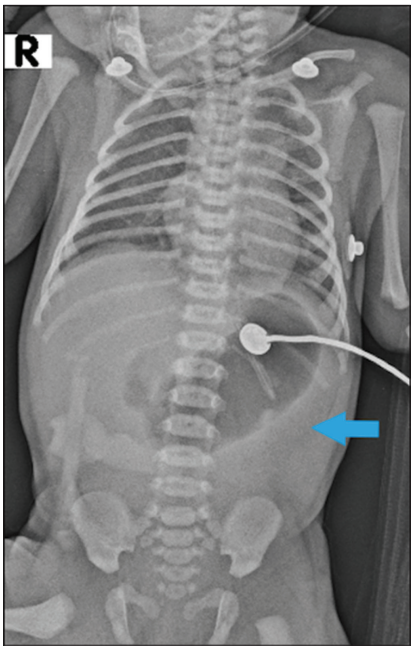

Figure 1: X-ray abdomen with double bubble sign

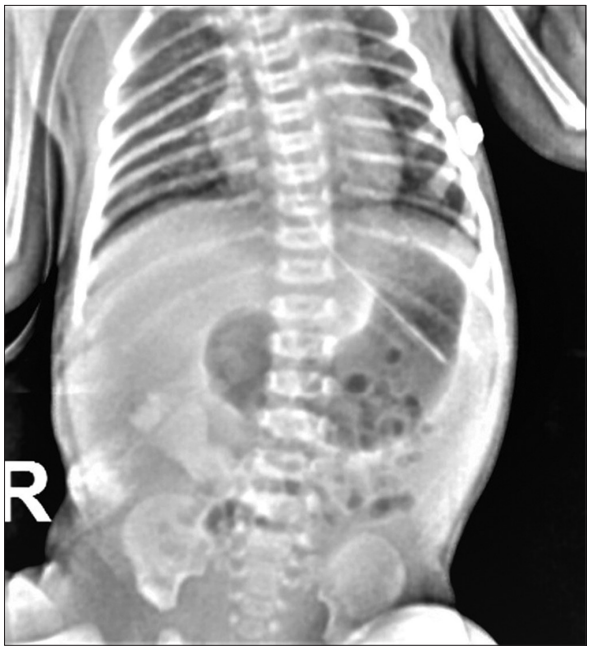

Figure 2: X-ray abdomen showing distal gas in the small bowel

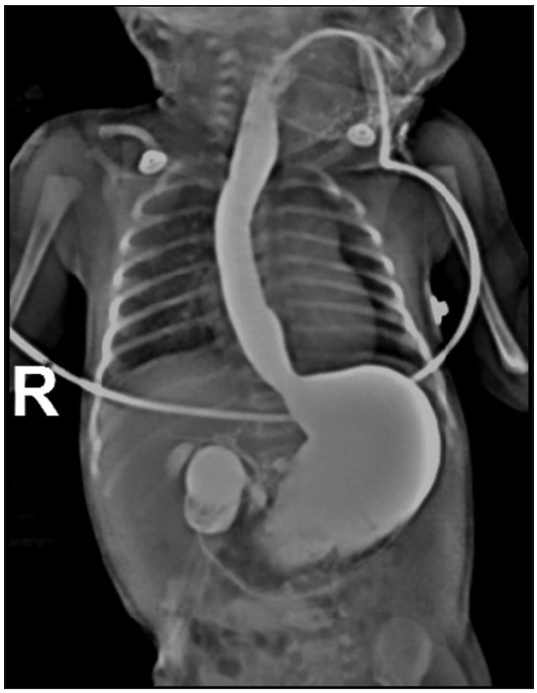

Figure 3: Upper gastrointestinal contrast study showing distension of the stomach, duodenal bulb with an abrupt narrowing, and jejunal loops on the right side of the spine with anomalous biliary connection with the duodenum

Type III duodenal atresia with the presence of bile in the jejunum. Occasionally, duodenal atresia with anomalous biliary ducts can allow passage of air from the proximal duodenal segment to the distal that is sufficient enough to give a wrong impression of partial obstruction.[3,5]

Duodenal atresia with anomalous biliary system should also be suspected when a double bubble sign with distal gas is present in an abdominal X-ray. UGI contrast study should be performed to exclude other possible diagnosis like intestinal malrotation as they are more common and can be potentially more dangerous.

CONCLUSION

The presence of gas in the small bowel distal to the double bubble suggests the possibility of intestinal malrotation, duodenal web or stenosis and duodenal atresia with anomalous biliary communication.

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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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