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INTESTINAL DUPLICATION CYST MIMICKING NECROTISING ENTEROCOLITIS IN AN EXTREME PREMATURITY INFANT.

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ABSTRACT

Intestinal duplication cyst is a rare congenital anomaly that mimics presentation of an intestinal obstruction in neonates. It is a congenital malformation formed during the abnormal development of the Gastro-Intestinal tract during embryonic development. Ante-natal ultrasound is the imaging modality of choice for early detection. Early detection and prompt referral is of importance for pediatric surgeons, as this reduces morbidity and mortality risks, and offers a more desirable outcome for patient.

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Intestinal duplication cyst is a rare congenital anomaly that mimics presentation of an intestinal obstruction in neonates. It is a congenital malformation formed during the abnormal development of the Gastro-Intestinal tract during embryonic development. Ante-natal ultrasound is the imaging modality of choice for early detection. Early detection and prompt referral is of importance for pediatric surgeons, as this reduces morbidity and mortality risks, and offers a more desirable outcome for patient.

Keywords: Antenatal; Cyst, Gastrointestinal Duplication; Infant, Extreme Prematurity; Necrotising enterocolitis.

INTRODUCTION
Intestinal duplication cyst is a rare congenital anomaly that mimics presentation of an intestinal obstruction in neonates. In 1930s the term "duplication" was popularized by William Ladd.[1] Bentley proposed the "Split notochord theory", an embryologic error may result in abnormal diverticularization of the GI endoderm through the developing notochord during the fourth week of gestation.[1] The incidence of gastrointestinal duplication is about 1 in 4500 live births and is found in 0.2% of the children.[2] It is a rare condition with prevalence observed 1 in 4500 autopsies done.[3] It is most commonly found in ileum (33%), followed by esophagus (20%), colon (13%), jejunum (10%), stomach (7%) and duodenum (5%).[4-5] The cyst or duplicated bowel is typically lined with intestinal mucosa, but heterotopic tissue from other areas of the body may be found within it. Two-thirds of all intestinal duplications are identified within the first two years of life while the remaining one-third are discovered in the newborn period.[6-7] We report a rare case of intestinal duplication cyst in an infant born with extreme prematurity with its associated management challenges.

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CASE REPORT
A baby boy was born with extreme prematurity at 24w2d POA via spontaneous vaginal de-

livery as mother went into active labor following spontaneous rupture of membrane (SROM). This infant weighed 710g, with length of 31cm and head circumference of 23cm.

Ante-natal follow up scans were noted to be normal, and no advance detailed feto-maternal specialist scan was done prior to delivery. Mother was 36 years old during pregnancy and hepatitis B positive of unsure cause. She had defaulted follow-up for this but had no history of Diabetes Mellitus or hypertension during this pregnancy.

At birth, the baby had an APGAR score of 6 at 1st minute and 8 at 5th minute. Ballard score was corresponding to 28 weeks. The neonate was noted to be tachypneic with poor respiratory effort, with SpO2 <93%. Attending pediatric team decided to intubate the baby. Abdomen at birth was noted to be normal with no organomegaly, and bowel sounds were present. The baby was admitted to NICU (Neonatal Intensive Care Unit) for severe prematurity with meconium aspiration syndrome for close monitoring.

At day 10 of life, the neonate was noted to have bilious vomiting and abdominal distension. He was hemodynamically stable, however blood smear showed leukocytosis with a count of 36 x 10^{9}/L. Abdominal radiograph revealed soap-bubble appearance. A diagnosis of Necrotizing enterocolitis stage II was made and he was started on intravenous antibiotics. Serial abdominal radiographs following treatment noted improving signs with presence of rectal gas seen. Lateral decubitus abdominal x-ray showed no pneumoperitoneum; hence child was treated conservatively.

At day 37 of life, despite supportive therapy, the infant's condition continued to be progressively unwell, with reduced feeding tolerance and further billous vomiting. Septic parameters continued to worsen and ventilatory support was increased with escalation to HFOV (High Frequency Oscillatory Ventilation). Abdominal findings then were soft, distended, no skin discoloration, tender over the right abdomen and liver was 2cm palpable below the costal margin. Abdominal radiograph noted persistent abnormal multi-cystic bowel shadow over the right side. Chest radiograph noted having broncho-pulmonary dysplasia with pneumonia changes. Urgent ultrasound noted prominent fluid filled small bowel on right side, with thickened bowel wall. No abdominal mass was seen. Based on the findings above, a decision to perform emergency laparotomy was made.

The infant underwent emergency laparotomy at day 38 of life. Intra-operatively a duplicated jejunal cyst at 15cm from duodeno-jejunal junction was noted. It and was connected to proximal small bowel as shown in figure 1. At distal part of normal small bowel noted another 15cm blind ending bowel in continuum with the cyst was seen. This part of the bowel had no luminal connection with the cyst. Resection of the duplicated small bowel cyst was performed and the blind end of the bowel incorporated into distal bowel to make 35cm of distal limb to be brought out as stoma. Total small bowel length left up to ileo-caecal valve was 50 cm. Duodeno-jejunal junction was seen to be located on the left side of the spine. The decision to bring out

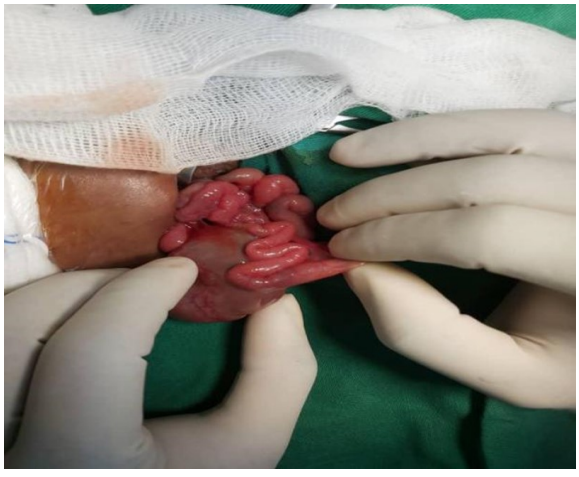

Figure 1: Intra-operative image showing the jejunal duplication cyst.

a stoma was part of the damage control surgery strategy, as child was having severe broncho-pulmonary dysplasia (BPD) with intermittent haemodynamic instability intra-operatively.

Post-operatively, child was escalated back to HFOV based on the worsening oxygenation demand and deteriorating clinical condition. Stoma was healthy and functioning by day 2 post-op. However, at day 40 of life, child succumbed to death due to haemodynamic instability and poor lung function. This child also suffered severe BPD, owing to its extreme prematurity and immature lungs. Death was attributed to difficulty in ventilation due to severe BPD.

DISCUSSION

Intestinal duplication cyst is a rare congenital anomaly occurring during embryogenesis.[4-5] Associated anomalies such as spinal defects, cardiac or urinary malformations, are reported with an incidence rate of 16–26%.[5] Other digestive anomalies are present in about 10% of cases.[5] Criteria for diagnosis are intimate attachment to the native GI tract, smooth muscle coat and alimentary mucosal lining.[6] Before the availability of prenatal US, enteric duplications were likely to remain undetected unless the patient had signs and symptoms of bowel obstruction.[7]

Abdominal radio-graph is non-specific and less sensitive to detect intestinal duplication cyst, but useful for showing intestinal obstruction.[1,7-8] Variation in the ultrasonographic features of gastrointestinal duplications have been described well in literatures.[5,7-8] Although it is difficult to distinguish between intestinal duplication cyst to other similar GIT diseases of newborn by ultrasound, a more characteristic description should be made available for the radiology team to conclude.[7-8]

Extreme pre-maturity also further added difficulty in ante-natal management by obstetricians and primary care physicians to schedule a detailed scan assessment in our case. In our opinion, ante-natal detection gives a better preparation in planning management of the infant and mother by a multi-disciplinary team discussion involving obstetricians, paediatricians and paediatric surgeons. There should be a local database made available on reporting such cases for better planning of future similar encounters. This database will allow detailed study to identify risk factors, common presentation in relation to age of gestation, infant weight and many other important factors.

A high index of suspicion for diagnosis of intestinal duplication cyst should be kept in the mind for neonates presenting with intestinal obstruction, provided common causes have been ruled out. Early detection offers more appropriate management and reduces the risk of morbidity and mortality.[2] In asymptomatic patients, a delayed diagnosis of intestinal duplication cyst may not cause any significant morbidity.[1] However, in symptomatic patients, early detection reduces risk of intestinal obstruction, ischemia and perforation. This reduces the risk of sepsis and overall mortality rate.

In such cases, timing of surgery need to be considered on the infant's general well-being. Risk versus benefit assessment should be made between bowel saving versus life-saving factors. In unstable symptomatic cases, a damage control surgery approach should be considered to offer the best solution at the shortest time. In our case, the duplication cyst excision and diverting stoma decision was considered an appropriate approach, as the surgery was completed in less than 2 hours.

CONCLUSION

Neonatal small bowel duplication cyst is a rare congenital anomaly. Although it may manifest asymptomatically in early life, it can also present as intestinal obstruction during infancy. An ante-natal scan or high index of suspicion based on symptoms is essential for early diagnosis and referral to paediatric surgery team, as this reduces morbidity and mortality risks, and offers a more desirable outcome for patient.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

DECLARATION

Written consent was obtained from the patient's guardian.

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