

Meconium peritonitis and pseudocyst in a preterm infant with trisomy 21

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Abstract

Presentation

A preterm neonate born at 36 weeks of gestation and trisomy 21 presented with abdominal distension at birth. Antenatal scans showed dilated bowel loops, moderate hyperechogenic bowel and polyhydramnios.

Diagnosis

Abdominal x-ray and upper gastrointestinal contrast study showed a large gas bubble and a calcified rim in the right upper quadrant. Laparotomy confirmed meconium peritonitis and pseudocyst due to two small internal hernias through a mesenteric defect.

Treatment

The cyst was partially resected and jejunostomy with mucus fistula was fashioned. Reversal of stoma was uncomplicated at second laparotomy.

Discussion

Although gastrointestinal malformations are common in infants with trisomy 21, meconium peritonitis (MP) pseudocyst due to internal hernias is extremely rare. This aetiology should be included in the differential diagnosis of neonatal gastrointestinal obstruction, even in infants who do not fulfil the prenatal classical diagnostic criteria.

Introduction

Meconium peritonitis (MP) pseudo-cyst is very rare and is defined as an extra luminal collection of meconium surrounded by dense calcified mass of fibrous tissue sealing off the perforation. A small muscle sheath usually connects the cyst to the rest of the bowel¹. The typical prenatal ultrasound

features are calcifications, polyhydramnios and ascites². Although gastrointestinal malformations are observed in 6 % of infants with trisomy 21³, to our knowledge, this is the first report on an internal hernia in a mesenteric defect that caused meconium peritonitis pseudocyst.

Case Report

A 36 week gestation preterm neonate was referred from a local unit with abdominal distension since birth. The pregnancy was spontaneous, in a healthy mother and uneventful until 33 weeks of gestation. Antenatal ultrasound then noted dilated bowel loops, moderate hyper echogenic bowel, and polyhydramnios (suspected trachea-oesophageal fistula). Non-Invasive Pre-Natal Screening showed high risk for trisomy 21 and postnatal karyotype confirmed 47 XX+21. The patient was born by emergency caesarean section for concerns regarding foetal position and previous caesarean section. Apgar scores were 7 and 9 at 1 and at 5 minutes of life respectively. Abdominal distension was noted immediately after birth and a nasogastric tube was placed easily even though the possibility of tracheoesophageal fistula had been raised on antenatal scans. At 9 hours of life, an abdominal x-ray was done [Figure (1)] which showed distended loops, with gas bubble and a calcified rim in the right upper quadrant.

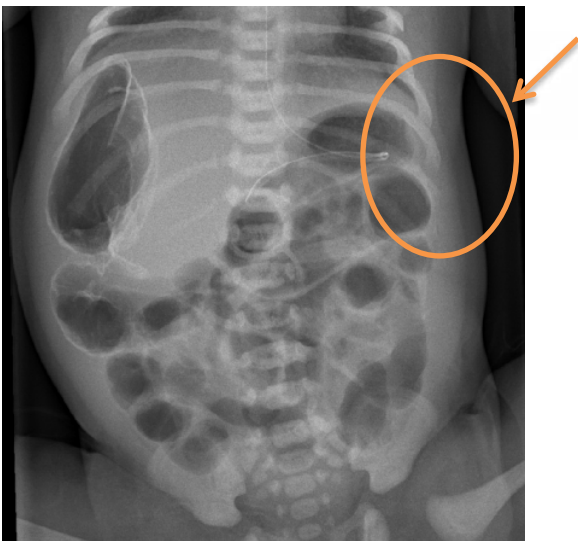


Figure 1.

The decision to perform an upper gastro-intestinal study was taken because of the unusual appearance of the abdominal X-Ray and to assess if the calcified area in the right upper quadrant had any connection with the bowel. The contrast study showed normal position of the pylorus, duodenum and duodeno-jejunal flexure with no evidence of malrotation or volvulus. It also suggested that the right upper quadrant gas bubble represents a contained perforation, probably of the hepatic flexure. Another image was taken 3 hours post contrast ingestion, showing contrast had passed all the way through to the colon. There was also an impression that contrast had got into the cyst, thus communication existed between bowel and cyst [Figure (3)]

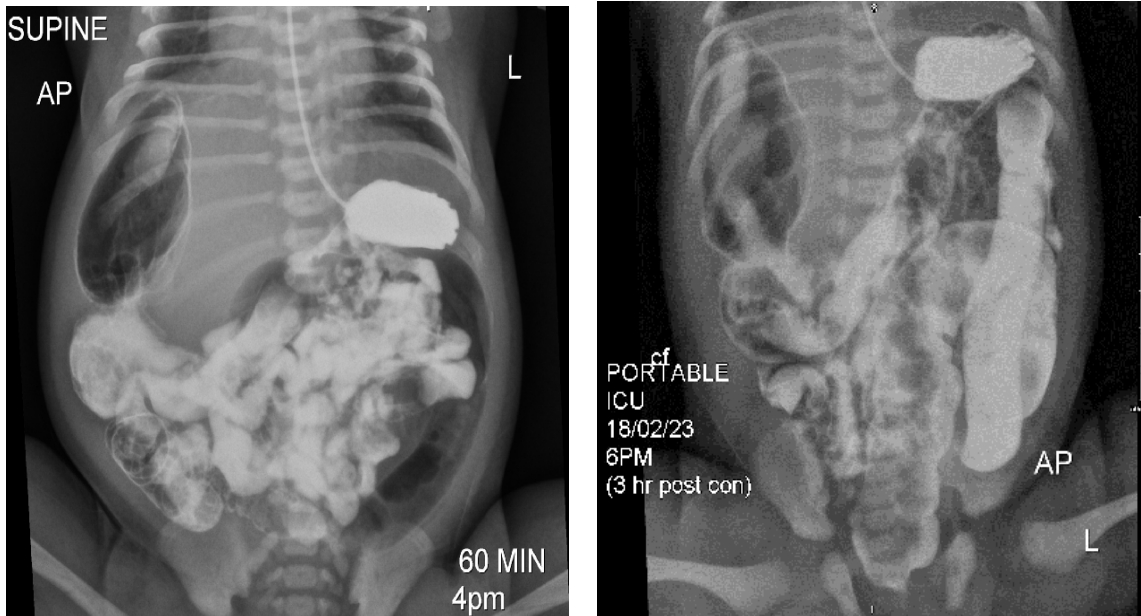


Figure (2) and (3): Those pictures were taken 60 minutes and 90 minutes after 10 cc of contrast was administered

Based on the imaging and clinical findings; the decision for exploratory laparotomy was made on day life 2. Through a transverse abdominal incision, the distal ileum was found to be attached to the under surface of the liver and the lateral abdominal wall. Two small internal hernias through a mesenteric defect were found surrounded by areas of calcification. The cyst was partially resected as liver bleeding concerns occurred during the surgery. Proximal jejunostomy and mucus fistula were fashioned, taking into consideration the jejunal perforation, the unhealthy condition of the jejunum and the constant ooze from the under surface of the liver, it was felt that a formation of a jejunostomy, proximal to the perforation and mucous fistula with resection of the meconium pseudocyst, was the best surgical option and future plan for reversal was made. Reversal surgery was done on 54 day of life. End to end jejuno-jejuno anastomosis was done with no intra or post-operative complications.



Figure 4: Intraoperative picture of the cyst before excision

Enteral feeding was started on day 2 post initial surgery and advanced slowly with parenteral nutrition support. Cystic fibrosis testing was negative and rectal suction biopsy ruled out Hirschsprung disease at 46 day of life. Congenital hypothyroidism was diagnosed at 11 day of life and thyroxin treatment was started. Stoma reversal was successfully performed at 54 day of life due to high stoma output when the infant reached full enteral nutrition. Full oral feeding was achieved at 64 days after the second surgery with discharge home at day 81 of life.

Discussion

Internal hernia may cause bowel obstruction and secondary meconium peritonitis. The surrounding intestinal loops adhere to the meconium leak, resulting in a meconium pseudocyst⁴. The extravasated meconium's extracellular calcifications are the result of a chemical reaction between pancreatic enzymes⁵. Intraabdominal calcifications in the neonatal period have a wide range of differential diagnoses⁶. Pseudocyst treatment involves resection of the small bowel segment and the pseudocyst, followed by an enteral anastomosis⁷.

This case highlights that meconium peritonitis pseudocyst should be included in the differential diagnosis of prenatal hyperechogenic gut as it does not always fulfil the classical pattern of eggshell calcifications, polyhydramnios and ascites; especially in infants with trisomy 21 where abdominal malformations are more common than in the general population.

Declarations of Conflicts of Interest:

None declared.

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