CASE REPORT

Meconium Pseudocyst and Ileal Atresia Secondary to Intrauterine Intussusception

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Introduction

Major anomalies of the fetus present a critical dilemma for the pregnant mother and her immediate family regarding the prognosis and outcome of the newborn. Termination of pregnancy is not the answer in all cases of major fetal anomaly. In those cases where the anomaly is compatible with life and is surgically correctable, delivery should be conducted in a tertiary care center with facilities for pediatric surgery and neonatal intensive care. We report a case of meconium pseudocyst with ileal atresia treated successfully in our hospital.

Case Report

A 20 year old primigravida came for antenatal check up at 34 weeks of gestation (with known dates) she was married for 1 year. She was a known case of epilepsy, and was on carbamazepine 200 mg at night in the preconception period since 17 years of age and throughout her pregnancy. An

ultrasound revealed a 34 to 35 weeks gestation fetus in cephalic presentation with polyhydramnios, grossly dilated bowel loops and 7 × 6 cm cystic intraabdominal lesion with calcification within its wall (Fig. 1). The estimated fetal weight was 2.02 kg. A diagnosis of meconium ileus with intestinal obstruction and meconium pseudocyst formation was made. Other routine antenatal investigations were normal. Opinion was sought from the pediatric surgeon and neonatologist. Parents were counseled regarding prenatal diagnosis, postnatal management and prognosis. At 36 weeks and 3 days of gestation she was admitted in active labor and had a spontaneous vaginal delivery with episiotomy, of a live baby boy weighing 2 kg. The baby cried immediately after birth. Liquor was excess and clear. Apgar scores at 1 and 5 min were 7 and 8, respectively. The baby was examined at birth by the pediatric surgeon and neonatologist. All vitals, reflexes and systemic examination were normal. The abdomen was found to be distended, bowel loop peristalsis were seen, bowel sounds were present and there was no organomegaly. An ultrasound confirmed antenatal USG findings. An erect x-ray of abdomen showed a space occupying lesion in the right hypochondrium and lumbar region pushing the dilated coils of intestines medially. A barium enema showed classical microcolon suggesting intrauterine small bowel obstruction (Fig. 2). The neonate was taken up for emergency laparotomy with a diagnosis of meconium pseudocyst with ileal atresia on the first post natal day under general anesthesia. On opening the abdomen, a thick walled meconium filled cyst with a segment of devitalized bowel within it was found. There was ileal atresia in

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the terminal part of ileum and extensive intraperitoneal adhesions (Fig. 3). The devitalized bowel loop was the segment of ileum which had undergone intussusception during the intrauterine life. Excision of the meconium pseudocyst along with the devitalized bowel and excision of the blind ending ileum and end to end anastomosis was performed. During the postoperative period the baby was put on ventilator for 30 h as he was not maintaining saturation. The histopathology report showed meconium pseudocyst with small bowel intussusception and segmental transmural infarction. On subsequent follow up up to 6 months in the pediatric OPD, the baby was found to be doing well with adequate weight gain.

Discussion

The presence of fetal dilated echogenic bowel on USG after 20 weeks of gestation may suggest the possibility of meconium ileus. Meconium ileus is a manifestation of cystic fibrosis which occurs due to mutation of ΔF 508 gene over long arm of chromosome 7. Intestinal obstruction occurs in meconium ileus due to abnormalities of exocrine mucous secretion resulting in thick, tenacious and viscid intestinal mucus. Complications of such obstruction are volvulus and intestinal atresia, perforation and meconium peritonitis or meconium pseudocyst [1].

A cyst wall with calcification and dilated bowel loops indicates meconium pseudocyst which may be due to meconium ileus or other causes. Meconium pseudocyst is formed by development of fibrous wall around the meconium leaked into the peritoneal cavity due to intrauterine

Fig. 1 Antenatal ultrasound picture showing meconium pseudocyst with calcification in its wall and dilated small bowel loops

Fig. 2 Barium enema picture showing microcolon

Fig. 3 Operative photograph showing cyst wall with devitalized intussuscepted ileum and dilated small bowel loops due to ileal atresia

bowel perforation. Meconium peritonitis or meconium pseudocyst may occur from complications of intestinal obstruction not related to meconium ileus like internal hernia, jejunoileal atresia and perforation, ruptured Meckel's diverticulum and volvulus due to congenital abnormal peritoneal bands. Ileal atresia can be secondary to intrauterine/intussusception [2]. It is generally not possible to differentiate the cause of meconium peritonitis before exploration. In our case, the meconium pseudocyst formation and ileal atresia were due to intussusception, as the meconium was normal greenish black in colour and soft in consistency unlike the thick, dark and viscid consistency of meconium ileus. This fact was also confirmed by histopathology. Therefore, the screening tests for cystic fibrosis like sweat chloride test or DNA probe analysis for the ΔF 508 gene mutation on chromosome 7 was not recommended. Kubota et al. [3], have reported a case of ileal atresia with partial IKS torsion of the dilated small bowel wherein prenatal USG had shown a single cyst like

intraabdominal lesion. In another case reported by Budhiraja et al. [4], ileal atresia was associated with segmental defect of intestinal musculature. Long term outcome is good in patients with meconium peritonitis/meconium pseudocyst due to causes other than meconium ileus.

Antenatal diagnosis of life threatening fetal anomalies is not an indication for termination of pregnancy in all cases. Ming Kwang Shyu et al. [5], have correlated prenatal ultrasound findings with post natal outcome and defined persistent ascites, pseudocyst or dilated bowel loops as highly predictive of need for postnatal surgery (92%). Therefore, once diagnosed, pregnant women with fetal anomalies should deliver in a tertiary care centre where immediate neonatal and pediatric surgical facilities are available.

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