Meconium peritonitis is defined as a sterile chemical or foreign-body peritonitis that is caused by escape of meconium from the intestinal tract into the general peritoneal cavity during the fetal or perinatal period. Although meconium peritonitis is indicative of intrauterine perforation of the intestine, it may occur as early as the 4th to 6th month of fetal life and as late as several hours after birth. The estimated incidence is about 1 in 35,000 live births. The etiology can be categorized as perforation with obstruction, including stenosis, atresia, volvulus, extrinsic congenital band, meconium ileus, and internal hernia, or perforation without obstruction, such as appendicitis, Meckel’s diverticulum, vascular insufficiency, and ruptured ulcer. The intra-abdominal calcification is recognized as a pathognomonic presentation either by X-ray or ultrasound.

The surgical indications are intestinal obstruction or persistent leakage of the meconium into the peritoneal cavity, which may result in progressive abdominal distension and subsequent respiratory distress or sepsis. The principles of the operation are to treat the underlying pathology and restore intestinal continuity, with preservation of at least 50% of intestinal length. Previously, the surgical mortality has been high, at up to 60%. Despite improvements in antenatal diagnosis, neonatal intensive care, and postoperative management, the overall mortality is still estimated to be 11%.

Meconium Pseudocyst: A Classical and Successfully Treated Case

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Meconium peritonitis with pseudocyst formation is rare and can be lethal. We report a newborn infant with antenatal sonographic diagnosis of bowel dilatation and giant meconium pseudocyst. Postnatal presentation included a palpable abdominal mass and abdominal distention. Abdominal X-ray revealed a huge egg-shell calcified mass containing air-fluid level. Postnatal sonography revealed a cystic mass with air-fluid level and acoustic shadows. Computed tomography showed a giant communicating cyst with egg-shell calcification, which compressed the other intestinal loops to the posterior peritoneal cavity. Elective laparotomy was performed, and distal ileal atresia with sealed proximal perforation and a giant meconium pseudocyst were found. Resection of the involved small bowel, including the giant pseudocyst, followed by primary end-to-end anastomosis was performed smoothly. The postoperative course was uneventful. Advances in perinatal intensive care mean that neonates can be operated upon under stable rather than critical conditions, and elective rather than emergency laparotomy, and primary anastomosis of the intestine rather than staged enterostomy can be performed.

Key Words: calcification, computed tomography, cystic, meconium peritonitis, pseudocyst

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Here, we report a classical and comprehensive case of meconium peritonitis with pseudocyst formation, which was treated successfully with elective one-stage resection and primary anastomosis on postnatal day 3.

**Case Report**

A 39-year-old woman, gravida 3 para 2 abortion 1, was admitted to our delivery room at 38 weeks of gestation. The prenatal chromosome study performed at the 18th week of gestation revealed 46 XY chromosomes. However, at the 24th week of gestation, sonography showed dilated bowels and an echogenic cyst (Figure 1), followed by polyhydramnios at the 30th week of gestation. She received amnioreduction and 1300 mL amniotic fluid was aspirated under sonographic guidance. Finally, she underwent cesarean section because of prolonged labor and delivered a boy at 38 weeks gestation.

The baby was born smoothly without fetal distress. The birth body weight was 3700 g. The Apgar scores were 8 and 9 at 1 and 5 minutes, respectively. No congenital anomaly was found immediately after birth. Physical examination showed mild tachypnea and abdominal distention. The digital examination showed meconium in the rectum and normal anal tone. The baby showed good activity initially but progressive abdominal enlargement was noted followed by desaturation. He was transferred to our neonatal intensive care unit and supportive care was applied with a policy of nil by mouth, nasogastric tube decompression, intravenous fluid support, and prophylactic antibiotic treatment.

Abdominal plain X-ray revealed a mid-abdominal, large egg-shell calcification (Figure 2). Abdominal computed tomography (CT) showed a large calcified cyst with air and fluid content. The measurable maximal diameter revealed by CT was 8.4 cm. Communication between the cyst and adjacent dilated bowel was also found (Figure 3). After a diagnosis of meconium pseudocyst complicated with respiratory distress and intestinal obstruction, laparotomy was performed electively on postnatal day 3.

After laparotomy, severe adhesive bands between the cyst and the liver, spleen, mesentery, omentum, bladder and abdominal wall were found. Therefore, meticulous adhesiolysis was performed. After mobilization of the cyst and the intestine, we found the communicating point between the cyst and the proximal small bowel, as indicated by CT. The distal small bowel showed no communication with the cyst (Figure 4A).
The junction between the proximal small bowel and the cyst was 80 cm from the ligament of Treitz, and the junction between the cyst and the distal small bowel was 45 cm from the ileocecal valve. The proximal small bowel showed two variable sizes, its proximal section was about 1.5 cm in diameter and 50 cm in length, followed by the distal section, which was 2.5–3.0 cm in diameter and 30 cm in length. The distal small bowel was 0.4–0.6 cm in diameter and 45 cm in length (Figure 4B). Resection of the cyst and dilated malfunctioning small intestine was performed, followed by primary end-to-end anastomosis. Importantly, distal gastrointestinal tract patency was confirmed before the anastomosis by instilling warm saline via a Nelaton tube placed in the distal bowel. The preserved small intestine was 95 cm in length.

Pathologic examination showed an 8 × 7 × 6 cm, meconium-containing ovoid cyst, which was consistent with meconium pseudocyst secondary to intestinal atresia (Figure 5A). Microscopically, the intestinal wall around the perforation site showed discontinuity of the muscle layers replaced by fibrous tissue that indicated the perforation site. Many calcification foci were visible in the cyst wall (Figure 5B). Ganglion cells were found over the proximal and distal intestinal wall.

Flatus and stool passage were noted on postoperative day 2. Feeding with 15 mL regular formula every 4 hours was tolerated well, and the nasogastric tube was removed. The feeding was increased gradually up to 70 mL every 4 hours on postoperative day 4. As a result of adequate bowel movement, the patient was discharged on postoperative day 4. The patient’s body weight had increased to 9 kg at the last clinic follow-up at the age of 5 months.
Meconium peritonitis was first described by Morgagni in 1761 and more comprehensively by Simpson in 1838. The clinical course ranges from spontaneous healing to rapid fatality, depending on the timing of perforation and if the perforation persists after birth. Postnatal surgical intervention to relieve the intestinal obstruction or close the perforation and restore the intestinal continuity is usually necessary.

Since the 1980s, antenatal diagnosis by maternal sonography has become possible. Antenatal diagnosis of meconium peritonitis is believed to reduce the mortality rate from 50% to 11%, and predict postnatal surgery in 50% of those with meconium pseudocyst, persistent ascites, and dilated bowel loops. Postnatal contrast abdominal CT can identify the finely distributed calcification and the persistent intestinal perforation invisible with prenatal ultrasound. A fetal intraperitoneal cystic mass that shows high T1 and low T2 signals in prenatal magnetic resonance imaging is considered pathognomonic of meconium pseudocyst. In our case, the findings of antenatal sonography, postnatal plain X-ray, and postnatal abdominal CT all suggested a large meconium pseudocyst with calcification. Furthermore, CT revealed that the cyst had a communication channel with the nearby dilated bowel. To the best of our knowledge, this is the first report to demonstrate communication between the meconium pseudocyst and dilated bowel loop, by postnatal abdominal CT. In addition, we presented clear intraoperative pictures and pathologic findings exactly consistent with the preoperative images.

Our intraoperative findings and formal pathologic reports demonstrated the diagnosis of meconium peritonitis with pseudocyst formation secondary to ileal atresia. However, some may doubt why digital examination still showed meconium in the rectum at delivery in this case of ileal atresia at 24 weeks of gestation. From the embryologic point of view, meconium formation begins about the 3rd month of gestation. Intestinal peristalsis begins about the 5th month of gestation and the meconium fills the entire intestine. If this vascular event occurs later than meconium passage, one could still find meconium in cases of ileal atresia at 24 weeks of gestation, as in our case. Therefore, the appearance of meconium at 24 weeks of gestation does not rule out the possibility of intestinal atresia.

In Western countries, 15–40% of infants with a diagnosis of meconium peritonitis are reported to have meconium ileus and cystic fibrosis. Foster et al suggested that the presence of abdominal

Figure 5. (A) Gross pathology showed a cystic mass that contained greenish meconium. Communication between the cyst and proximal small intestine was found (large arrow). The connection between the cyst and distal small intestine was obstructed (small arrow). (B) Microscopic pathology showed discontinuity of the muscle layer (large arrow) and calcification spots (small arrow) in the inner aspect of the pseudocyst wall.
calcification is associated with meconium peritonitis rather than cystic fibrosis. This may be because pancreatic enzymes, which are deficient in 80% of patients with cystic fibrosis, are necessary for calcification to occur. In particular, cystic fibrosis is considered non-existent in oriental people, except in a few cases of gene mutation. Based on the findings mentioned above, we did not routinely perform a sweat test to determine if cystic fibrosis was present in our case of meconium peritonitis.

In 1943, Agerty and associates were the first to record survival of a patient with meconium peritonitis treated surgically. Since then, several reports of survival after surgery for meconium peritonitis have been published. However, there is no standardized operative procedure for meconium peritonitis. The purpose of surgery is to establish intestinal continuity and to preserve at least 50% of the intestinal length. Underlying pathologic processes should also be corrected. In 1966, Lorimer and Ellis reported postoperative survival in five patients with meconium peritonitis, and tried to summarize a concept of surgical management based on their pathologic classification. In general, meconium peritonitis can be classified into three pathologic variations: fibroadhesive; cystic; and generalized. In the fibroadhesive type, the perforation usually seals off before birth as a result of an intensive fibroplastic reaction. Subsequent obstruction is caused by adhesions. Surgery aims to achieve adhesiolysis and resection of the non-viable intestine, but dissection is usually difficult. Therefore, it has been suggested that if > 50% of the intestinal length was preserved, en masse resection of the severely adhesive bowel loop should be considered. In the cystic type, the perforation generally remains open. By opening the cyst and finding the perforation, the cause of the obstruction can be determined. Decortication must be done to dissect the cyst from the entrapped intestine. After an adequate length of intestine is freed, the non-viable or severely entrapped intestine can be resected, followed by bowel exteriorization. In the generalized type, the perforation occurs in the perinatal period and results in wide dissemination of meconium in the peritoneal cavity. Resection of the pathologic segment of the intestine, followed by enterostomy, has been suggested because of the questionable bowel viability and severe peritonitis.

Our patient had an unusual form of meconium peritonitis with a high mortality rate. Early recognition, pseudocyst resection, decortication, temporary enterostomy, antibiotics and meticulous postoperative care offer the best opportunity for survival. Mikulicz enterostomy with early crushing of the spur appears to be the best means for re-establishing intestinal continuity. The moderate disparity in the size of proximal and distal intestinal segments, and the thickened nature of both segments after chronic antenatal inflammatory changes, makes primary anastomosis unduly difficult. Some authors have reported successful primary anastomosis after bowel resection in patients with cystic meconium peritonitis. Tanaka et al recommended cyst drainage after birth and elective surgery later. They consider that dissection, cyst resection and primary anastomosis become easier during later surgery.

In our case, we performed an elective operation. After exploratory laparotomy, we performed meticulous dissection to mobilize the pseudocyst rather than opening the cyst. We did our best to resect the pseudocyst en bloc so that no meconium spilled into the peritoneal cavity. This was important because we performed the operation electively on postnatal day 3. Theoretically, meconium is sterile during fetal life and probably does not become contaminated by bacterial invasion until about the 3rd day of extrauterine life. After adequate mobilization of the pseudocyst, we had the chance to identify the primary obstructive lesion, which turned out to be type I intestinal atresia. The pathologically dilated small bowel, atresia segment, and the meconium pseudocyst were excised smoothly in an en bloc fashion. After confirming the patency of the distal bowel, we performed primary end-to-oblique anastomosis without difficulty. The postoperative course was uneventful.

In conclusion, we believe that the successful surgical treatment in our case was attributed to
antenatal sonographic diagnosis, antenatal counseling, postnatal fluid and electrolyte correction, prophylactic antibiotics, postnatal CT that demonstrated communication between the cyst and the bowel loop, meticulous surgical technique, and postnatal care. Advances in perinatal intensive care mean that surgery can be performed under stable rather than critical conditions, and elective rather than emergency laparotomy, and primary anastomosis of the intestine rather than staged enterostomy can be carried out. In addition, this is believed to be the first report of meconium peritonitis with pseudocyst formation that demonstrated clear communication with a dilated bowel loop by CT.

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