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Intestinal Obstruction by Meconium Ileus in the Newborn Revealing Cystic Fibrosis

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Abstract **Case Report**

Objective*:** To report a case of a preterm infant with meconium ileus at birth and cystic fibrosis. ***Background*:** The meconium ileus corresponds to the obstruction of the terminal ileum by an abnormally thick meconium; it is observed most often in neonates with cystic fibrosis. ***Case description: a female infant was born by cesarean delivery section on severe pre-eclampsia with oligohydramnios and intrauterine growth restriction, weighing 1230 g at 36 weeks of gestational age, with Apgar scores of 10/10. His mother was a 26-year-old G2P2 who had attended three prenatal consultations with negative serology for vertical infections. Immediately after birth, the newborn was referred to our neonatal critical care unit due to severe abdominal distension. the Newborn had a delay in meconium emission of more than 48 hours. an abdominal X-ray showed significant intestinal distension and a barium enema was performed showing meconium plugs with a colon of normal topography and caliber, initially put on antibiotic therapy with enema by gastroraphy twice a day, an etiological assessment was carried: A genetic study was conducted, revealing that the newborn is a carrier of a variant (c.1521-1523delCTT,p.Phe508del F50 del) associated with a classic form of cystic fibrosis in a homozygous state and the first immunoreactive trypsinogen (IRT) was > 1300 ug/l (reference value >1000 ug/l), in view of the non-improvement the newborn benefited from an ileostomy**.** ***Comments:*** This case illustrates the rapid evolution of CF in a premature patient with meconium ileus as the first clinical manifestation.

Keywords: terminal ileum, oligohydramnios, vertical infections, antibiotic therapy.

BACKGROUND

Cystic fibrosis (CF) is a monogenic autosomal recessive disease that presents with wide phenotypic variability around 300 pathogenic cystic fibrosis transmembrane regulator (CFTR) variants cause the disease and it is the most prevalent fatal genetic disease in humans [1]. Distal intestinal obstruction syndrome (DIOS) is a gastrointestinal complication that is specific to CF. It was first described as syndrome of post-neonatal distal small bowel obstruction caused by meconium-like stool plugs, in 1945 [2]. It is characterized by accumulation of viscous fecal material, combined with sticky mucous secretions typically located in the distal ileum and cecum, which may adhere to the intestinal wall, causing subtotal or total obstruction. Since its initial description as ‘‘meconium-ileus equivalent,’’ not only has the denomination changed to DIOS, but also its definition has been reviewed and thus the epidemiology, recurrence, and burden of this complication have been better described and understood [2] neonatal screening may lead to early detection and allows immediate treatment of pancreatic insufficiency, nutritional deficiencies, and pulmonary involvement, improving survival and facilitating the design of treatment strategies. Less frequently, meconium ileus (MI) may be the first manifestation of CF in the neonatal period, occurring in approximately 20% of patients with pancreatic insufficiency [6]. This clinical picture is caused by obstruction of the terminal ileus with thick meconium containing high amounts of protein. About 80% of cases of MI are due to CF, and it would be ideal to perform an early sweat chloride test before 48 hours of life, although this is not always feasible [1]. Children with MI appear to have normal pulmonary function at CF diagnosis, with slower progression of lung disease than those diagnosed due to respiratory symptoms. However, it is currently believed that lung inflammation may occur early, and may even precede the onset of infection in infants with newly diagnosed cystic fibrosis [1].

CASE DESCRIPTION

A female infant was born by cesarean delivery due to severe pre-eclampsia with oligohydramnios and Intrauterine growth restriction, weighing 1230 g at 36

weeks of gestational age, with Apgar scores of 10/10. His mother was a 26-year-old G2P2 who had attended two prenatal consultations with negative serology for vertical infections. Immediately after birth, the newborn was referred to our neonatal critical care unit due to severe abdominal distension. the Newborn had a delay in meconium emission of more than 48 hours. an abdominal X-ray showed significant intestinal distension and a barium enema was performed showing meconium plugs with a colon of normal topography and caliber, initially put on antibiotic therapy with Gastrographin enema twice a day, an etiological assessment was carried: A genetic study was conducted, revealing that the newborn is a carrier of a variant (c.1521-1523delCTT,p.Phe508del F50 del) associated with a classic form of cystic fibrosis in a homozygous state, and the first immunoreactive trypsinogen (IRT) was > 1300 ug/l (reference value >1000 ug/l), in view of the non-improvement the newborn benefited from an ileostomy.

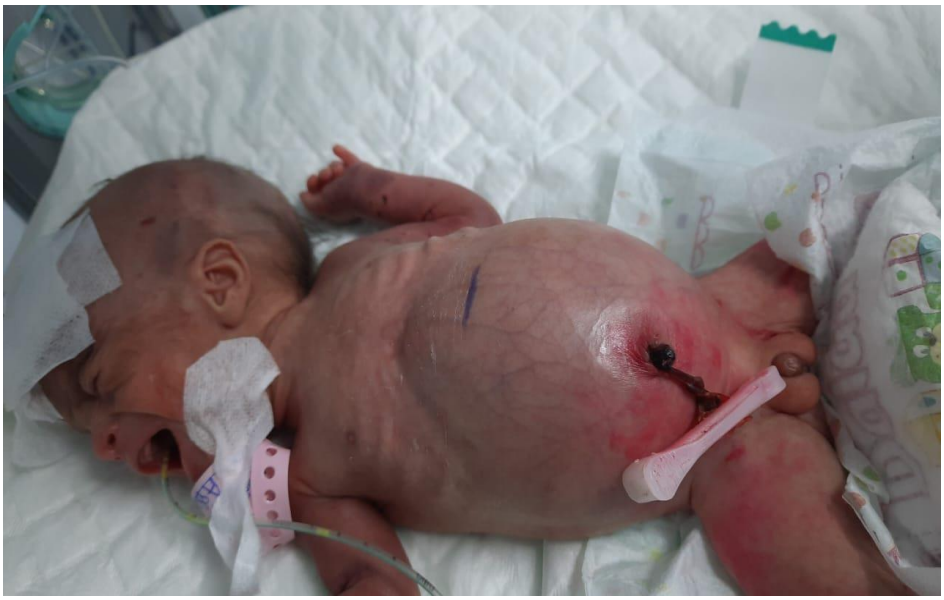

Figure 1: Newborn with abdominal distention

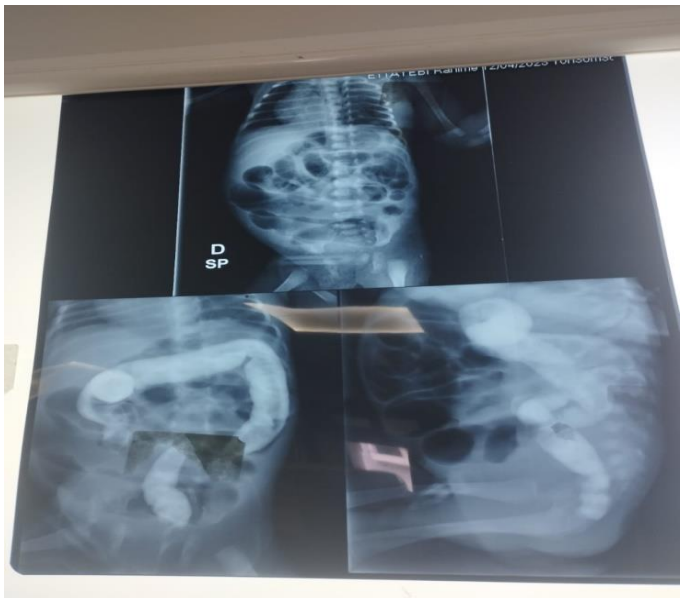

Figure 2: Plain film and contrast enema findings in MI

DISCUSSION

An intestinal ileus in the neonate may be mechanical or functional. Mechanical obstruction, for instance the impaction of the distal ileum with thick inspissated meconium, results in dilatation of the bowel upstream. Alternatively, it may be functional, as a result of temporary absence of intestinal peristalsis. This results in a lack of the normal proximal to distal movement of intraluminal contents [1]. The etiology of this type of ileus can be multifactorial and may include electrolyte derangement, sepsis, pharmacological agents, trauma, or direct handling of the bowel from surgery. The initial

management of these potentially very unstable neonates remains standard supportive measures, adequate resuscitation, and the investigation for a cause to guide definitive management [3].

Meconium plug syndrome is a condition that predominantly affects preterm neonates of low birth weight [6]. It presents with an inability to pass meconium within the first 24 to 48 hours of life, and It must be properly differentiated from similar conditions [2]. As it presents with obstruction, it must be treated accordingly and swiftly. Meconium ileus is usually caused by cystic fibrosis.

It typically causes vomiting, abdominal distension and no bowel movements during the first days after birth. Diagnosis is based on symptoms and the results of an X-ray. The obstruction is treated with enemas and/or surgery [1, 5].

Medical management of simple MI has been developed around the use of hyperosmolar enemas given under fluoroscopic guidance to ensure that the solution refluxes into/reaches the TI [14]. This technique was first described in 1969 by Noblett utilizing Gastrografin, the mechanism of action is to act as a direct solvent and shift fluid into the bowel lumen instead of competing with the intracellular space surrounding the mucosa. When utilizing such a hyperosmolar agent, adequate hydration (150 mL/kg/day minimum) via an IV line is imperative to avoid hypovolemia that can lead to shock and end-organ damage including necrotizing enterocolitis. Presence of an IV line is also essential to respond appropriately to complications such as need for emergent surgical intervention [16, 17]. Most commonly with Gastrografin, a ¼–½ dilution with water is infused under low hydrostatic pressure through a catheter under fluoroscopy through the rectum until the terminal ileum is reached. Perforation risk has been described as low as 2.7% and as high as 23%. If hyperosmolar enema is unsuccessful, then surgical intervention is pursued [12, 14].

Most often, meconium ileus is an early manifestation of cystic fibrosis. Cystic fibrosis is an inherited disease that makes intestinal secretions too thick and viscous. They stick to the wall of the intestine and a small bowel obstruction form. These extremely viscous secretions are the first manifestation of the disease in 10 to 20% of children with cystic fibrosis. Newborns with meconium ileus almost always develop other symptoms of cystic fibrosis later. Meconium plug syndrome is similar to meconium ileus, except that the large intestine is blocked with meconium [15].

The primary surgical intervention is decompaction of the meconium by irrigating the obstructed TI with warm saline or Gastrografin in the OR [7, 12]. The subsequent creation of a continuous enterostomy such as the bishop-Koop is preferred to allow for ongoing irrigation, if necessary, of the TI postoperatively. It also reduces the risk of postoperative complications of a primary enterostomy which can be as high as 30%. Risks of creating an enterostomy include high output losses, especially of sodium. Bowel resection is reserved for more complex cases and is dependent on the extent of bowel injury. It can vary from simple resection with primary anastomosis to simple enterostomy with anastomosis several months later requiring minimal resection of small bowel (b10 cm) to more extensive bowel resection, involving both small bowel and colon, or removal of a meconium cyst [4, 7].

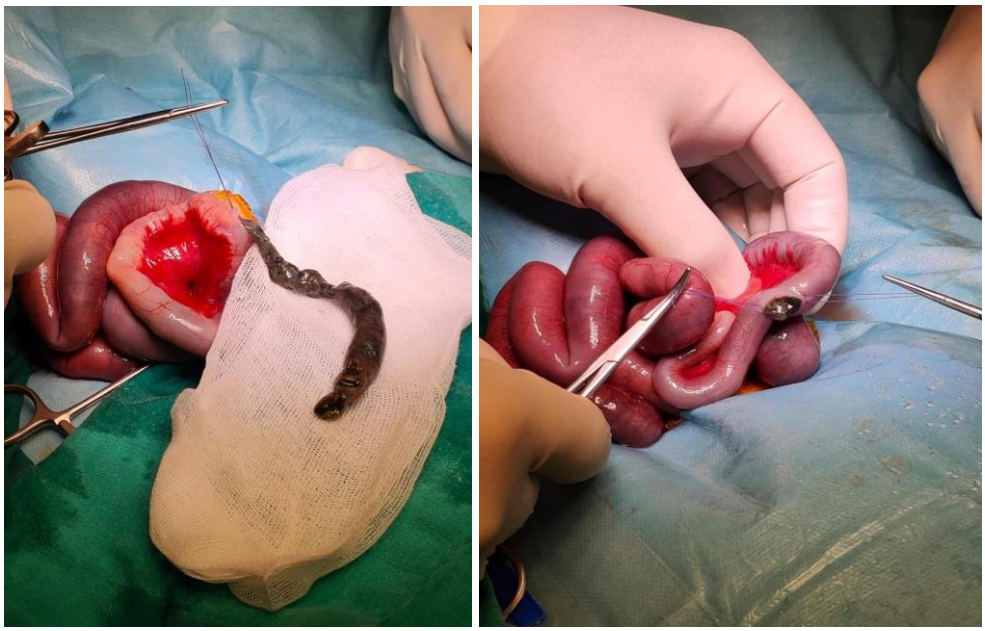

Figure 3: Operative findings the ileal meconium plug is evident with obstructed, dilated, proximal bowel on the right and collapsed distal bowel on the left

Figure 4: Meconium plug meconium ileus can sometimes lead to the following complications: perforation of the small intestine or twisting of the intestine on itself significant dilation or volvulus which cuts off the blood flow in the intestine and can lead to perforation of the small intestine. A perforation is a serious complication because it leads to leakage of meconium into the abdominal cavity. If the perforation occurs before birth, the affected part of the small intestine may atrophy and become narrower (intestinal atresia). Surgery may be needed after birth. If perforation occurs after birth, leakage of meconium leads to inflammation and peritonitis, which can lead to shock and death [4]

CONCLUSION

A combination of factors, including early diagnosis of both MI and CF, an overall improvement in the multidisciplinary approach to the care of CF patients, and specific improvements in medical and surgical management of MI (use of contrast enemas to treat MI, better surgical techniques, and early implementation of nutritional support) has resulted in a prognosis for CF patients with MI that is comparable to those CF patients without MI. The advancement of science and ongoing quality improvement in clinical care continues to make the future for our CF patients more promising by positively impacting the morbidity and mortality of CF's many manifestations.

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