Necrotizing enterocolitis manifesting with pneumatosis ani in a patient with gastroschisis

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\textbf{Abstract}

We present the case of a 1-month-old female with repaired gastroschisis who presented with pneumatosis ani as the initial manifestation of necrotizing enterocolitis (NEC). NEC may present in as much as 20% of patients with gastroschisis. This relationship should prompt urgent evaluation and management as soon as any sign or symptom of the disease appears. Even though pneumatosis ani is a rare clinical finding, the anal region should always be examined in all patients where NEC is suspected.

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Pneumatosis intestinalis, which is also known as pneumatosis cystoides intestinalis, intramural gas, pneumatosis coli, pseudolipomatosis, intestinal emphysema, bullous emphysema of the intestine or lymphopneumatosis [1,2], has been associated with several diseases, including necrotizing enterocolitis (NEC). We report an unusual presentation of NEC in a patient with gastroschisis.

\section{Case report}

We present a case with an unusual manifestation of pneumatosis intestinalis secondary to NEC in a 1-month-old female preterm infant. As part of the prenatal diagnosis, an ultrasound was performed at 28 gestational weeks, in which gastroschisis was diagnosed; maternal age of 18 years and paternal age of 23 years were the only prenatal risk factors identified. The delivery took place in an operating room by elective c-section at 35 gestational weeks with a birth weight of 1850 g, after which the patient was immediately transferred to our Neonatal Intensive Care Unit (NICU). The patient’s defect was 4 cm diameter, with the umbilical cord to the left; both the small and large bowel were externally exposed. The same day of birth the baby was taken to the operating room and a Silastic\textsuperscript{g} mesh silo was constructed and sutured to the abdominal aponeurosis to cover the exposed bowel. A central venous catheter was placed in the right internal jugular vein using the modified Seldinger technique under fluoroscopic control. Total parenteral nutrition was started on the following day. The intestines were slowly pushed 1–2 cm daily during the 5 following days until they were at the same level of the abdominal wall. On day 6, the patient was taken again to the operating room for silo removal and abdominal wall closure using 3-0 Vicryl\textsuperscript{h} interrupted stitches on the fascia and a 4-0 nylon running suture on the skin; there were no complications. The following days intravesical pressure was measured, with values between 5 and 15 cm H\textsubscript{2}O (average of 10 cm H\textsubscript{2}O). There were no ventilatory complications. The patient was slowly weaned from mechanical ventilation and extubated on day 9.

On day 21, after normal bowel function was noticed, oral feeds with breast milk were initiated, slowly weaning from parenteral nutrition as tolerance was improving. On day 28 the patient suddenly presented intestinal ileus, abdominal distention and tenderness, bloody stools and gastric vomits. NEC was suspected and abdominal X-rays were...
ordered. Meanwhile, the mother noticed something “strange” in the baby’s anus. Exploration showed gas bubbles in the anal mucosa and perianal tissue (pneumatosis ani), confirming the diagnosis of NEC (Fig. 1). Abdominal X-rays showed pneumatosis intestinalis in the right and left lower quadrants (image not available).

The patient underwent non-surgical medical treatment with two weeks NPO and TPN, orogastric suction and IV antibiotics (cefotaxime and metronidazole). Evolution was favorable and she was discharged after tolerating full enteral feeds, on day 50. She is currently being followed in clinic without complications.

2. Discussion

The purpose of reporting this case of NEC in a patient with repaired gastroschisis, is to acknowledge pneumatosis ani as a rare clinical manifestation of NEC. To our knowledge, this is the first report of such a rare clinical finding. The relationship between NEC and gastroschisis has been reported since 1988 by Oldham et al. [1]. NEC has been documented in up to 20% of infants after repair of gastroschisis and is responsible for significant morbidity [3]. Since the disease was named in 1953, the incidence has greatly increased due to the advances in intensive neonatal care technology, which allow the viability of increasingly premature babies [4]. NEC is one of the most serious gastrointestinal disorders and the most common intra-abdominal emergency in neonates, characterized by bowel necrosis and multisystem organ failure. It is also the leading cause of the short bowel syndrome in this age group [4–7]. The incidence of necrotizing enterocolitis in the general population decreases with increasing gestational age and birth weight, and is about 6–7 percent in very low birth weight infants less than 1500 g [8–10].

Mortality is around 10% overall and 25% in cases where surgical intervention is required [4–7]. Some authors report the mortality as high as 50% when surgical treatment is required and 20–40% overall in affected children [6]. The consistent findings between clinical manifestations and radiological signs of pneumatosis intestinalis makes the diagnosis of necrotizing enterocolitis without the need of any further studies.

Lee and colleagues concluded that the mode of delivery, method of closure, birth weight and gestational age, and the presence of intestinal atresia do not appear to correlate with survival in infants with gastroschisis. Only the presence of another major anomaly, the era of repair, and the development of NEC were associated with increased mortality. Degree of prematurity and development of NEC were associated with an increased incidence of septic complications. Low birth weight was a marker for closure complications [11]. Prenatal diagnosis of gastroschisis has been associated with better outcomes including earlier enteral feedings and shorter hospital stays [12].

An 11-year follow-up study of patients with gastroschisis showed that the most important causes of mortality in the earlier years were abdominal compartment syndrome and multi-organ failure, while in recent years the most important predictors of mortality are long hospital stay and sepsis [13].

3. Conclusion

NEC after gastroschisis repair is more likely multi-factorial in nature. It occurs more often in children with more severe gut dysfunction, and in children with delayed feeding, as well as low birth weight infants who had perinatal stress. Conservative measures are the mainstay of treatment in non-complicated cases [14–18]. The intimate relationship between gastroschisis and NEC should never be forgotten and preventive maneuvers should be performed. This case report is relevant because to our knowledge pneumatosis ani has never been described as the initial clinical sign of NEC. Even when pneumatosis ani is such a rare clinical finding, the anal region should always be examined in patients where NEC is suspected.