Appendiceal Necrosis and Pneumatosis as Part of Necrotizing Enterocolitis

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Key words: necrotizing enterocolitis, pneumatosis, appendix

IMAJ 2008;10:479–480

Necrotizing enterocolitis is a disease of multifactorial origin that almost exclusively affects stressed premature infants [1]. Common factors in most infants with NEC include intestinal ischemia, bacterial colonization of the gut, and enteral feedings of synthetic formulas [2,3]. Free intraperitoneal air and signs of diffuse peritonitis are obvious indications for surgery. Surgical findings of intestinal wall necrosis are most common in the ileum and to a lesser degree in the colon [2,4]. We present a rare case of a neonate with NEC and ileal perforation. Upon surgery, pneomatosis and necrosis of the terminal ileum, cecum and the appendix were evident. To the best of our knowledge this is the first report of pneumatosis of the appendix.

Patient Description

A 1355 g, first of twins, baby girl was delivered vaginally at 31 weeks of gestation because of placenta abruption of the second twin. Apgar scores were 9 and 10 at 1 and 5 minutes, respectively. Her physical examination was normal for gestational age and the first days of her hospitalization were uneventful. Enteral feeding was instituted on her second day of life.

At age 10 days, necrotizing enterocolitis was diagnosed based on symptoms of distended abdomen and pneumatosis intestinalis mainly in the right lower and right upper quadrants on serial abdominal X-rays. Conservative treatment was begun. Three days later, a pneumoperitoneum was documented in a routine follow-up abdominal X-ray.

NEC = necrotizing enterocolitis

Explorative laparotomy revealed intestinal content and peritoneal fluid. The small intestine was normal up to 12 cm proximal to the cecum. This segment of the terminal ileum, the cecum and the appendix [Figure] all had wall necrosis and pneumatosis. Histopathological examination confirmed wall necrosis and pneumatosis; appendicitis was ruled out. A clear perforation was identified at the involved segment of the terminal ileum. A segmentectomy of the ileum and appendectomy were undertaken. The remaining ileum and the cecum were brought to the abdominal wall as ileostomy and mucous fistula, respectively. The baby recovered from surgery and enteral feeding was resumed on postoperative day 9 with satisfactory weight gain.

A 2 cm-long appendix, with necrosis and pneumatosis at the distal end wall.

Comment

Necrotizing enterocolitis is the most common gastrointestinal medical and/or surgical emergency occurring in neonates. With mortality rates approaching 50% in infants who weigh less than 1500 g, NEC represents a significant clinical problem. Although it is more common in premature infants, it can also be observed in term babies. Despite intensive study over the past 30 years, its etiology remains elusive. Research suggests that it is multifactorial; ischemia and/or reperfusion injury may play a significant role. Cases that cluster in epidemics suggest an infectious etiology; however, a single causative organism has not been identified [1].

The mainstay of diagnostic imaging is abdominal radiography. An anteroposterior abdominal radiograph and a left lateral decubitus radiograph (left side down) are essential for initially evaluating any baby suspected of NEC. Characteristic findings on an anteroposterior abdominal radiograph include an abnormal gas pattern, dilated loops, and thickened bowel walls (suggesting edema/inflammation). Pneumatosis intestinalis is a radiological sign pathognomonic of NEC. Intramural air bubbles represent extravasated air from within the intestinal lumen caused by invasion of the ischemic mucosa by gas-forming organisms [2].

Obvious indications for surgery include free intraperitoneal air and diffuse peritonitis. Children with significant acidosis despite resuscitation and those with hepatoportal venous gas who do not im-

prove promptly with medical therapy are considered surgical candidates. Surgical findings of intestinal wall necrosis and pneumatosis are most common in the ileum and to a lesser degree in the colon [3]. However, we were unable to find any reports of pneumatosis and wall necrosis of the appendix. To the best of our knowledge this is the first report and image of NEC-associated pneumatosis and wall necrosis of the appendix.

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