Unusual presentation of more common disease/injury

Intestinal malrotation in an extremely preterm very low birthweight infant

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A case of a very low birthweight premature infant with a clinical presentation of necrotising enterocolitis that was found to have malrotation and midgut volvulus at autopsy is presented.

BACKGROUND

Volvulus is a rare finding in the premature newborn population, most commonly occurring in a term infant with bilious emesis. In this case, the clinical, laboratory and radiographic data suggested necrotising enterocolitis (NEC) – a far more common condition in premature infants.^{1–3} It is written to make the teaching point of it being among the differential diagnoses of NEC requiring immediate surgical attention.

CASE PRESENTATION

The infant was a 1080 g, 23-day-old female delivered at 26 weeks' gestation with a birth weight of 800 g. There was no perinatal asphyxia. Apgar scores were 5 and 7 at 1 and 5 min, respectively. After an uncomplicated cardiopulmonary course in the first 48 h (no intubation, assisted ventilation or nasal continuous positive airway pressure), she was advanced to full enteral breast milk feedings by orogastric tube over the next 14 days. There was no history of patent ductus arteriosus, indomethacin or ibuprofen exposure, or umbilical vessel cannulation. There was no prodrome of clinical signs such as emesis, bile-stained gastric residuals or abdominal distention. The infant was advanced to an enteral feeding volume at 150 ml/kg/day of breast milk over 14 days. She was maintained on breast milk until postnatal day 21 at which time the supply was exhausted. At that time, the feedings were provided using Prolacta ready-to-feed liquid formula (Prolacta Bioscience, Monrovia, California, USA). On the twenty-second day of postnatal life, a transfusion of packed red blood cells (PRBC) for the treatment of anaemia (haemoglobin of 8 g/dl) was provided. She was breathing room air without support and took her enteral feedings without incident. On postnatal day 23 the infant passed a grossly bloody stool and the abdomen became extremely distended, discoloured and tender and she displayed signs of clinical instability including hypotension, bradycardia and respiratory failure. Abdominal radiographs (figures 1A and 1B) revealed widespread pneumatosis intestinalis without portal venous air or intraperitoneal free air. The infant was transported from the Level III neonatal intensive care unit (NICU) where she

had been cared for since birth to the regional Level IV surgical NICU.

The infant arrived from transport bradycardic, hypotensive and in respiratory failure. The physical exam showed extremely poor perfusion throughout with thready pulses; the abdomen was severely distended and dark red-to-purple in colour. Venous and arterial access was obtained and full resuscitation was provided over the following 6 h.

INVESTIGATIONS

Laboratory data upon arrival revealed a severe metabolic acidemia (pH values ranged from 6.80–6.94; lactates >15 mmol/l; a calculated buffer base deficit 23 mmol/l). The haemoglobin fell as low as 6 g/dl during this time. The initial serum potassium was 9.8 mmol/l and rose to a maximum 10.4 mmol/l but never declined below 8.9 mmol/l. Coagulation studies demonstrated severe coagulopathy with an INR of 8.9 despite blood component therapy (fresh frozen plasma and cryoprecipitate).

Abdominal radiographs again revealed widespread pneumatosis intestinalis (figure 1B).

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for this newborn's presentation included a primary gastrointestinal infection with subsequent systemic shock, septicemia and coagulopathy or gastrointestinal ischaemia with mucosal sloughing and secondary enteric bacterial translocation into the systemic circulation and resultant septic shock and coagulopathy. These two mechanisms of disease would likely be operative in the clinical entities of NEC or malrotation with volvulus.

TREATMENT

Resuscitation included intravenous epinephrine, sodium bicarbonate, normal saline fluid boluses and blood component therapy (PRBC, platelets and fresh frozen plasma), as well as chest compressions. Antibiotics started at the referring centre were changed from ampicillin and gentamicin to vancomycin, gentamicin and meropenem. Intravenous dopamine and dobutamine were given for cardiovascular support.

Figure 1 (A) Plain abdominal flat plate radiograph afternoon of admission: diffuse pneumatosis intestinalis. (B) Plain abdominal flat plate radiograph 5 h after (A): diffuse pneumatosis intestinalis.

Consultation with the Pediatric Surgery was sought and, on arrival at the bedside, the surgeon placed a Penrose drain in the right lower quadrant of the abdomen. Foul smelling gas escaped through the surgical wound and feculent, bloody material drained from the site. Ventilation improved after the drain was placed and the abdomen decompressed, but the infant remained gravely ill.

OUTCOME AND FOLLOW-UP

Despite all efforts, the infant's acidaemia and coagulopathy continued. Florid pulmonary haemorrhage ensued and she became bradycardic. After counselling the family, life support was withdrawn. The time from initial grossly bloody stool to death was 12 h. A premortem blood culture and a peritoneal swab at the time of abdominal decompression and Penrose drain placement grew *Enterobacter sakazakii* within 24 h.

An autopsy examination using the standard Rokitansky evisceration method was performed. There was incomplete intestinal rotation with Ladd's bands at the third portion of the duodenum and caecum in midabdomen (figure 2). Histologically, there were massively dilated lymphatics, veins and arteries within the small bowel submucosa consistent with gross findings of incomplete rotation and midgut volvulus. Of a total of 161 cm of bowel, approximately 75% demonstrated necrosis with the exception of the proximal duodenum and distal rectum.

Histologically, there was extensive transmural ischaemic necrosis but no evidence of perforation. Additional findings included bilateral haemorrhagic adrenal glands

(Waterhouse–Friderichsen syndrome; figure 3). Postmortem blood cultures were positive for *E sakazakii*.

DISCUSSION

The classic description of intestinal malrotation is that of the term infant who presents with bilious emesis and then has the diagnosis confirmed with an upper gastrointestinal contrast study (UGI) that demonstrates the right-sided position of the duodenal-jejunal junction or evidence of a midgut volvulus.⁴ There exists a number of case reports of this phenomenon in term infants, but the finding has rarely been described in the very low birthweight (VLBW) preterm infant.^{5 6}

Intestinal malrotation occurs in approximately 1% of the general population. During fetal development, midgut rotation is a multistep process requiring herniation of the intestinal contents, rotation 270° around the superior mesenteric artery and return to the abdominal cavity. Non-rotation, malrotation, reverse rotation and incomplete rotation are variations of this phenomenon. With malrotation, the gut has 'floppy' attachments and can lose its blood supply through torsion or restrictive Ladd's bands. Midgut volvulus and ischaemia can occur at any age with no apparent warning or trigger, but often presents in the pediatric population. The typical early symptoms are irritability, poor feeding and bilious emesis, which are regarded as a sign of surgical emergency. Late symptoms include frankly bloody stools, abdominal distension and hypotension. Treatment requires surgical laparotomy and de-torsion or lysis of Ladd's bands (Ladd procedure). In children who

Figure 2 (A) The gastrointestinal tract (en-bloc Rokitansky evisceration method) shows a normally positioned liver (black arrows) and gallbladder (GB). Instead of its usual extraperitoneal position within the right lower quadrant, the caecum is found in the midabdomen, attached to the third portion of the duodenum by Ladd's bands (probe). This abnormal attachment led to volvulus and mechanical vascular compromise, which is grossly evidenced by discoloured small bowel. LT, ligamentum teres. (B) En-bloc genitourinary dissection shows the aorta, bilateral renal arteries, bilateral kidneys and ureters and bilateral adrenals. The adrenals show gross haemorrhage.

Figure 3 (a) Small bowel lymphovascular dilatation. A, representative photomicrograph (100 $\times$) shows marked dilatation of submucosal small muscular arteries, V, veins, L, lymphatics, as evidence of both inflow and outflow obstruction (ie, torsion). Appropriately-sized vessels are identified in the left upper corner. M, mucosa; SM, submucosa; MP, muscularis propria. (B) Waterhouse-Friderichsen syndrome. Representative photomicrograph (200 $\times$) show adrenal cortical haemorrhage extending through the ZG, zonae glomerulosa, ZF, fasciculata and ZR, reticularis.

survive, the most problematic result of midgut volvulus is short bowel syndrome, in which only a short portion of the intestine survives the ischaemic insult, but is too short to adequately absorb nutrients to meet daily needs.

Both NEC and malrotation with midgut volvulus are conditions known to have devastating results in the newborn population. The overall incidence of NEC for non-Hispanic blacks is significantly greater than for non-Hispanic whites.^{3,7} The case death rate when surgical intervention is required has been found to be as high as 50% and is highest in the smallest, least mature infants,^{8,9} thus lending further credence to NEC as a particularly devastating neonatal disease. While some plausible hypotheses for the cause of NEC include issues related to introduction or advancement of enteral feeds, alteration in normal bacterial colonisation of the gastrointestinal tract, bacterial translocation and activation of the cytokine cascade and mucosal damage from free radical production, widespread and common belief remains that NEC results primarily from ischaemic injury to the immature gut.¹⁰ In terms of phenomena that lead to intestinal ischaemia and gut hypoperfusion, malrotation with midgut volvulus remains among the rarest, yet most catastrophic of causes.

The infant in the present case had no history of bilious emesis, and advanced to full enteral feedings without complication. Accordingly, there was no index of suspicion for the presence of malrotation or midgut volvulus, and an UGI contrast study had never been performed. There have been reported cases of fetal malrotation being discovered on prenatal ultrasound by the demonstration of the 'whirlpool sign,' polyhydramnios and abnormal Doppler flow studies (among other signs).^{11–13} Some of these same studies also state, however, that the sonographic findings of volvulus are usually apparent after 27 weeks of pregnancy.¹¹ The presently described infant was born at 26 weeks gestation and prenatal ultrasounds were unremarkable.

The infectious agent found in this case is also of interest in that *E sakazakii* is typically found in cases with a history of using powdered infant formula, a risk factor known to be associated with the presence of this particular microbe.¹⁴ *E sakazakii* was identified in both the premortem blood culture and peritoneal swab and from postmortem cultures. It is a known risk factor for sepsis or NEC¹⁴ and is associated with the use of powdered formulas. However, a thorough review of this patient's NICU course revealed that she did not receive any powdered formula or human milk fortifier.

While NEC is a widely reported and well-known phenomenon in premature VLBW infants, little information regarding the presence of malrotation with midgut volvulus resulting in clinical and radiographic NEC can be found concerning these infants. The possibility of malrotation with midgut volvulus should be included in the differential diagnosis of preterm infants presenting with signs and symptoms of feeding intolerance or NEC, even in the absence of bilious emesis or other signs of gastrointestinal distress when advancing on enteral feedings. Although the present case demonstrated no gastrointestinal or other clinical symptoms of the disorder until it was essentially too late for surgical intervention, by keeping the diagnosis of malrotation with midgut volvulus in mind in preterm VLBW infants with evidence of feeding difficulties, it may be possible to avoid potentially fatal outcomes.

Learning points

- ▶ NEC is a widely reported and well-known phenomenon in premature VLBW infants and often presents with bloody stools and haemodynamic collapse.
- ▶ Less well-known considerations for this presentation, even in the VLBW infant, should include intestinal malrotation and volvulus.
- ▶ Rapid diagnostic and supportive interventions are necessary to minimise the loss of intestinal viability and save the affected infant's life.

Competing interests None.

Patient consent Obtained.

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