Spontaneous intestinal perforation in a preterm neonate

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DESCRIPTION

A female second twin infant was born to a 39-year-old primigravida via elective caesarean section at 28 weeks of gestation due to intra-uterine growth restriction of the second fetus. A full course of antenatal corticosteroid therapy was administered. The birth weight was 645 g, and the Apgar score was 9 at both 1 and 5 min. The infant was admitted to the neonatal intensive care unit, with respiratory support by nasal intermittent mandatory ventilation. A venous umbilical catheter was placed, and she was started on ampicillin and gentamicin. Parenteral nutrition was initiated, and trophic enteral feeding with breast milk via gastric tube feeding began at the 10th hour of life. The first passage of meconium occurred at 40 hours of life. Abdominal distension was noted 8 hours later, despite the absence of feeding intolerance or haemodynamic instability. Necrotising enterocolitis (NEC) was suspected, and enteral feeding was discontinued. Appropriate empirical antibiotic treatment with vancomycin, cefotaxime and metronidazole was initiated. Endotracheal intubation was electively performed, and ventilation support was given using synchronised intermittent positive ventilation. Serial abdominal radiographs were obtained and revealed pneumoperitoneum (figure 1). This finding led to an urgent

abdominal laparotomy that revealed a small focal perforation in the terminal ileum, 7 cm above the ileocaecal valve. A 2.5 cm long segment of intestine was resected, and a temporary ileostomy was performed. Histopathology revealed a thinning mucosa and a transmural perforation with ischaemic necrosis and haemorrhage, with adjacent viable bowel. The infant had an uneventful postoperative recovery, and enteric feeding was restarted 4 days after surgery.

Intestinal perforation is a major life-threatening complication in preterm infants, with high morbidity and mortality.¹ NEC is the most common aetiology, but alternate causes, such as intestinal obstruction, gavage-related mechanical injury and spontaneous intestinal perforation (SIP), must be considered.²

SIP is an acquired neonatal intestinal disease, and it is defined as a single or, less frequently, multiple perforation, typically in the terminal ileum, without evident cause,² as in the present case. Current evidence on the pathogenesis and potential risk factors, except prematurity and extremely low birth weight, remains controversial.³

SIP presents earlier in life, at a mean age of 7 days (vs 15 days for NEC), often with a typical black-bluish discoloration of the

Figure 1 Plain radiographs confirming pneumoperitoneum. (A) Chest and abdominal radiography—image acquired in supine position, with vertical X-rays, revealing subdiaphragmatic free gas (blue arrows) which makes visible the falciform ligament (red arrow), known as Silver sign. (B) Abdominal radiography—image acquired in supine position, with horizontal X-rays, revealing marked abdominal distension and free gas between the anterior abdominal wall and the bowel loops.

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Images in...

abdomen,^{2,3} which was absent in our case. Plain radiographs of the abdomen reveal the pneumoperitoneum and sometimes a gasless abdomen, generally without the imaging hallmarks of NEC (ie, pneumatosis intestinalis or portal venous gas).² SIP is characterised histopathologically by a small perforation and mucosal thinning, without ischaemic necrosis or neutrophil infiltrates, which are typically found in NEC. However, SIP cases with anatomopathological features of necrosis have been described.^{2,3}

Primary peritoneal drainage has been proposed as a treatment option, but surgery remains the current treatment of choice for SIP.²

Learning points

- Spontaneous intestinal perforation (SIP) is an idiopathic perforation with potential life-threatening complications. It mainly occurs in the context of prematurity, and the terminal ileum is the most frequently affected area.
- SIP typically presents in the first days of life and may cause the abdomen to appear blue or discoloured. The abdominal radiograph usually shows a pneumoperitoneum without the imaging hallmarks of necrotising enterocolitis.

Infants with SIP generally exhibit better survival rates than NEC infants, if promptly diagnosed and treated. However, both groups exhibit similar impaired long-term neurodevelopmental outcomes.¹⁻³

The authors highlight the importance of distinguishing between SIP and NEC-related perforation for the interest of prognosis and counselling.

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