Spontaneous intestinal perforation or necrotizing enterocolitis?
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See also related COTM March 2002
This 580 g female infant was delivered at 24 4/7 weeks of gestation by emergency Cesarean section secondary to placental abruption. The mother had received antenatal corticosteroids at 24 0/7 weeks of gestation. Apgar scores were 2, 5 and 6 at 1, 5 and 10 minutes, respectively, and arterial cord-pH was 7.30. She was intubated in the delivery room and surfactant was administered. Umbilical arterial and venous catheters were placed. Refractory arterial hypotension was treated with dopamine and stress doses of hydrocortisone (35 mg/m2/day in three divided doses). Indomethacin (0.1 mg/kg/dose every 24 hours for 3 doses) was given for prophylaxis of intraventricular hemorrhage. Enteral feeding (gut priming) was started on the first day of life.

On the 7th day of life, the abdomen was distended, but abdominal X-ray was normal. Thrombocytopenia and an increased immature-to-total granulocyte ratio were present. Antibiotics were started (imipenem/vancomycin). On the 9th day of life, abdominal X-ray showed free intraperitoneal air without evidence of pneumatosis or portal venous gas (Fig. 1) and a peritoneal drain was placed in the NICU. Because of persistence of thrombocytopenia and an increased C-reactive protein, laparotomy was performed on the 12th day of life.

At laparotomy, extensive necrosis of the stomach (Fig. 2) and multiple intestinal perforations adjacent to
Babygram on the 9th day of life: pneumoperitoneum without pneumatosis or portal venous gas.
Laparotomy on the 12th day of life: extensive necrosis of the stomach (S; the nasogastric tube (NGT) can be seen through the distroyed stomach wall (L: liver; U: umbilicus).
the necrotic stomach were found. Since there were no valid surgical options, life support was withdrawn after detailed discussion with the parents and the patient died.

Intestinal perforation is a major and life-threatening complication in VLBW infants. Necrotizing enterocolitis (NEC) is probably the most frequent cause of intestinal perforation in premature infants. Localized, spontaneous intestinal perforation (SIP) is a separate clinical entity from NEC.

NEC is a multifactorial disease that is diffuse and is associated with prematurity, fetal inflammatory response syndrome, infectious agents and enteral feeding. Multiple areas of coagulative and hemorrhagic necrosis with disruption of the mucosal architecture and submucosal pneumatosis define NEC histology.

SIP, on the other hand, is a localized disease limited to the area of perforation with isolated mucosal ulceration and inflammation, submucosal edema and serosal inflammation. Although SIP can involve the stomach, duodenum and colon, perforation of the distal small bowel is the most common location (1,2). Recent data suggest an increasing incidence of SIP, probably due to a combination of greater awareness of the condition and increased survival of VLBW infants (1).
In our case, the combined use of indomethacin and corticosteroids were discussed as potential etiological factors. However, in the indomethacin IVH prophylaxis trial (3), the incidence of SIP and NEC were not affected by the prophylactic administration of indomethacin. Similarly, in a retrospective review of 40 neonates with gastrointestinal perforations, Hwang found no association between the use of indomethacin and intestinal perforations (4). This was further supported by a recent Cochrane metaanalysis (5). On the other hand, use of dexamethasone has been shown to increase the risk of gastrointestinal bleeding and intestinal perforations (6). Whether the use of stress doses of hydrocortisone produces similar adverse effects is unknown.

Can SIP and NEC be distinguished clinically? In a retrospective review of a cohort of infants who had undergone laparotomy, Hwang found no statistical difference in the timing of perforation (4). In contrast, Okuyama did observe differences in gestational age (23.8 +/- 1.8 versus 27.0 +/- 2.5 weeks), birth weight (635 +/- 134 versus 883 +/- 256g) and age at onset (7.3 +/- 2.7 versus 14.4 +/- 14.4 +/- 7.9 days) between SIP and NEC in a small retrospective review (7). All infants had undergone laparotomy. The results of two additional studies were similar, but the diagnosis of SIP was only implied by radiological evidence of pneumoperitoneum without other radiological features suggestive of NEC (1,8). In the series of Hwang,
two neonates with histologically proven SIP had pneumatosis on abdominal X-ray (4).

Recent reports support the use of peritoneal drainage as the primary therapy for intestinal perforation in VLBW infants and possibly as the definitive treatment for most premature infants with SIP (8,9). Infants with evidence of clinical deterioration after peritoneal drainage require subsequent laparotomy. Laparotomy after failed treatment with peritoneal drainage is associated with poor outcome (4,8).

Because our patient was fairly stable and had no evidence of pneumatosis or portal venous gas, we believe that SIP is a more likely diagnosis than NEC.


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