CASE REPORT

Pneumoperitoneum in a neonate weighing less than 500 g. What do we really know about it?

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SUMMARY

Pneumoperitoneum in preterm infants is a surgical emergency as it is usually indicative of intestinal perforation. Rare cases of idiopathic pneumoperitoneum have been described in the literature, the underlying causes and pathophysiology of which remain uncertain. We present a case of pneumoperitoneum in an extremely preterm infant with severe growth restriction. This occurred while she was receiving high frequency oscillatory ventilation. She had respiratory distress syndrome with pulmonary interstitial emphysema. The pneumoperitoneum occurred in isolation. Despite the insertion of two surgical drains and two exploratory laparotomies in which no obvious intestinal perforation was noted, the free air in the abdomen reaccumulated. A decision was made to manage it conservatively. She was successfully extubated on the fourth week of life and the pneumoperitoneum resolved spontaneously. She was discharged home on day 136 of life. This case highlights our limited understanding of the intricate physiology of extremely low birthweight preterm neonates.

BACKGROUND

The presence of free air in the abdomen, also known as pneumoperitoneum, is a surgical emergency because it is caused by intestinal perforation in more than 90% of the cases. In neonates, it is usually caused by necrotising enterocolitis or spontaneous perforation of a viscus.^{1 2} However, rare non-surgical causes of pneumoperitoneum have been reported. Abdelmohsen *et al*³ subdivided these into five major categories: pseudopneumoperitoneum, thoracic, abdominal, gynaecological and idiopathic. We describe the case of a growth restricted extremely preterm neonate who developed an idiopathic pneumoperitoneum.

CASE PRESENTATION

An extremely preterm female infant was born at 27+4 weeks gestational age with a birth weight of 420g by emergency caesarean section due to antenatal bleeding and placenta praevia. This was an in vitro fertilisation quadruplet pregnancy (two monochorionic twin pairs); which was complicated with early onset selective intrauterine growth restriction for one set of twins. Laser separation was performed at 19 weeks' gestational age between this female twin and her sister. She was severely growth restricted and noted to have reversed end diastolic flow on antenatal scanning. A

full course of antenatal steroids was given prior to delivery.

She was intubated at birth and received surfactant. On admission to the neonatal unit, she was commenced on synchronous intermittent positive pressure ventilation with a target tidal volume of 5 mL/kg in FiO₂ 0.3. An initial chest X-ray showed bilateral ground glass opacity and a left lower lobe consolidation. She was noted to be profoundly anaemic and received two O negative blood transfusions. She required inotropes from day 1 of life in view of hypotension. Her initial cranial ultrasound scan revealed a left-sided resolving intraventricular haemorrhage with a dilated ventricle and parenchymal cystic changes indicative of antenatal ischaemic injury. She remained very sick during her first week of life with persistent hypotension, sepsis, acute renal failure and had a pulmonary haemorrhage.

On day 13 of life, she was commenced on high frequency oscillatory ventilation in view of increasing oxygen requirements and high peak inspiratory pressures. Chest X-rays showed parenchymal lung changes consistent with bilateral pulmonary interstitial emphysema. On the same day, a discoloured and distended abdomen was noted; however, there was no evidence of abdominal perforation or necrotising enterocolitis on an X-ray taken at that time. On day 15 of life, she had worsening abdominal distension and a repeat abdominal X-ray showed a radiolucent area in the upper abdomen (figure 1A). A lateral decubitus view confirmed a pneumoperitoneum (figure 1B). A Penrose drain was inserted as she was felt to be too unstable to tolerate a laparotomy. The air reaccumulated in less than 24 hours post-insertion of the drain and her clinical condition deteriorated with increasing oxygen requirements and a worsening metabolic acidosis (figure 2A). A second Penrose drain was resited on day 17 of life as she remained unstable. Once again, she developed abdominal distension and a repeat X-ray confirmed reaccumulation of air in her abdomen (figure 2B). She was switched to conventional ventilation and a laparotomy was performed on day 18 of life. During the laparotomy, the entire large and small bowel were carefully examined. There was no evidence of perforation, but a short 2 cm segment of haemorrhagic dilated mid-small bowel was excised and a defunctioning ileostomy was formed. She acutely deteriorated postoperatively with increasing

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Figure 1 Pneumoperitoneum on day 15 of life: (A) AP view and (B) lateral decubitus view.

oxygen requirements. A significant reaccumulation of free air was again identified on X-ray (figure 2C). Concerns were raised about a missed intestinal perforation. A relook laparotomy was performed on day 19 of life, including inspection of the posterior wall of the stomach; yet, still no underlying cause for the air leak was found. Postoperatively, she continued to have significant abdominal distension with X-rays confirming the presence of a pneumoperitoneum (figure 3A). The decision to treat conservatively was made by medical and surgical teams, and it was agreed further surgical interventions would not be in her best interests.

OUTCOME AND FOLLOW-UP

She continued to be ventilated on conventional synchronous intermittent positive pressure ventilation until day 27 of life when she was successfully extubated to nasal continuous positive airway pressure. Following extubation, an improvement in the pneumoperitoneum was observed (figure 3B). She had good clinical progress and enteral feeding was increased slowly. She was progressively weaned off respiratory support and was self-ventilating in air with no respiratory support by day 77 of life. A closure of her ileostomy was performed on day 126 of life and she reached full enteral feeding soon after that. She was discharged home on day 136 of life on demand bottle-feeding.

DISCUSSION

We described a case of idiopathic pneumoperitoneum in a severely growth restricted extremely preterm infant. To date, this is the smallest preterm infant with pneumoperitoneum described in the literature (table 1). The free air in the abdomen persisted over a prolonged period and surgery excluded a bowel perforation. In our case, the patient had no evidence of a pneumothorax or pneumomediastinum; thus, mechanical ventilation,

Figure 2 Reaccumulation of free air following the insertion of Penrose drain 1 (A), Penrose drain 2 (B) and first laparotomy (C).

Figure 3 Pneumoperitoneum persists following second negative laparotomy (A) with spontaneous resolution postextubation (B).

high airway pressures and large tidal volumes in non-compliant lungs with pulmonary interstitial emphysema were felt to be the cause. The pneumoperitoneum only seemed to improve once the baby had been extubated to continuous positive airway pressure.

In the paediatric population, non-surgical pneumoperitoneum occurs in 1%–3% of mechanically ventilated infants, depending on the mode of ventilation.² Leonidas *et al*⁴ studied 222 mechanically ventilated infants and observed that of the nine infants who developed pneumoperitoneum, four of them were non-surgical. Zerella *et al*⁵ distinguished the ‘medical’ pneumoperitoneum from the ‘surgical’ pneumoperitoneum in his 10 critically ill infants with respiratory disease and no evidence of an intestinal perforation.

Gupta *et al*⁶ argue that the entity of benign pneumoperitoneum should be recognised and laparotomy avoided. Al-lawama *et al*⁷ describe the case of a neonate who developed major post-operative complications including a grade 3 intraventricular haemorrhage and a patent ductus arteriosus following an unnecessary laparotomy for a benign pneumoperitoneum. For this reason, they propose criteria to differentiate between the two. They argue that neonates with low birth weights <1500 g, gestational age <32 weeks, on oral feeds, more than 5 days old, with signs of peritonitis and who have been noted to have dysmorphic features, congenital anomalies and an antenatal suspicion of gastrointestinal anomalies should be suspected to have developed a gastrointestinal perforation and should undergo a laparotomy.⁷ In our case, the patient had significant risk factors to suspect necrotising enterocolitis and in our experience many babies with spontaneous intestinal perforation do not exhibit classic signs, despite the operative findings including peritoneal contamination. In our current practice, if the neonate is fit for a general anaesthetic and the presence of free air is confirmed radiologically, then a laparotomy is performed. Neonates who are unfit to undergo surgery are treated with the insertion of an abdominal drain followed by a laparotomy once the patient becomes more stable. Other relative indications in which we would perform a laparotomy in premature neonates with suspected necrotising enterocolitis are in cases of failed medical management, persistent mass with evidence of obstruction and clinical deterioration despite maximal medical therapy.

Following surgical exploration, it was clear that our patient had developed a non-surgical pneumoperitoneum. The mechanism which has been proposed by many authors is that of air leaks from ruptured alveoli of non-compliant lungs, tracking

Table 1 Reported cases of idiopathic pneumoperitoneum in the literature ordered by birth weight

Reference	Gestation (weeks)	Birth weight (g)	Age at onset (days)	Clinical signs	MV	Laparotomy	Survival
Our case	27	420	15	AD, RD	Y	Y	Y
Gummalla <i>et al</i> ¹	23	560	19	AD, RD, P	Y	N	N
Bakal <i>et al</i> ⁹	26	640	15	AD, RD, P	Y	N	Y
Zerella <i>et al</i> ⁵	U	1020	U	RD, P	Y	N	N
Zerella <i>et al</i> ⁵	29	1050	5	AD, RD, P	Y	N	N
Zerella <i>et al</i> ⁵	30	1190	6	RD	Y	N	N
Zerella <i>et al</i> ⁵	29	1200	11	P, PM	N	N	Y
Zerella <i>et al</i> ⁵	U	1220	U	RD, P	Y	N	Y
Zerella <i>et al</i> ⁵	U	1280	U	RD, PM	Y	N	N
Zerella <i>et al</i> ⁵	U	1300	U	RD, PM	Y	N	N
Vohra <i>et al</i> ¹⁰	30	1300	2	AD, RD	Y	Y	Y
Wang <i>et al</i> ⁸	30	1660	2	AD, RD	Y	Y	Y
Khan <i>et al</i> ¹¹	38	1700	12	AD	N	N	Y
Abdelmohsen <i>et al</i> ³	34	1750	5	AD, RD, S	U	Y	N
Zerella <i>et al</i> ⁵	U	1890	U	RD, P, PM	Y	N	N
Khan <i>et al</i> ¹¹	34	1900	12	AD	N	N	Y
Zerella <i>et al</i> ⁵	U	1920	U	RD, P, PM	Y	N	Y
Gupta <i>et al</i> ⁶	36	2100	2	AD, RD	N	N	Y
Zerella <i>et al</i> ⁵	U	2155	U	RD, P, PM	Y	N	N
Al-lawama <i>et al</i> ⁷	34	2280	2	AD, RD	Y	Y	Y
Shah <i>et al</i> ¹²	36	2300	5	AD	N	Y	Y
Bedi <i>et al</i> ¹³	U	2400	0–1	AD, RD	U	Y	Y
Porter ¹⁴	U	2900	2	RD	N	Y	Y
He <i>et al</i> ¹⁵	37	2950	4	AD, RD	N	N	Y
Pati <i>et al</i> ¹⁶	40	3000	3	AD	N	N	Y
Al-Salem ¹⁷	40	3300	2	AD, S	N	Y	Y
Karaman <i>et al</i> ²	30	U	3	AD, RD	Y	N	N
Karaman <i>et al</i> ²	37	U	1	RD, P	Y	N	Y

AD, abdominal distension; P, pneumothorax; PM, pneumomediastinum; MV, mechanical ventilation; N, no; N/A, not applicable; RD, respiratory distress; S, sepsis; U, unknown; Y, yes.

downwards along sheaths of vessels in the mediastinum and anatomical defects in the diaphragm, resulting in a collection of free air in the abdomen.^{6,8} While this is a plausible explanation that can be applied to this case, one can argue that knowledge on the physiology and anatomy of extremely premature growth-restricted infants remains very limited and further research is required.

Learning points

- Although pneumoperitoneum in neonates is most frequently caused by an intestinal perforation, there are other possible medical causes of it.
- Our case demonstrates that there is limited knowledge on the physiology of extremely preterm and severely growth restricted neonates and suggests mechanical ventilation, with large tidal volumes and high airway pressures, as a possible cause of free air in the abdomen.
- Further research into the complex physiology of such a rare cohort of neonates that survive the first few days of life is required before being able to make this assumption.

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