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CASE REPORT

Bubbles in the heart as first sign of gastric pneumatosis

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Abstract This case report describes a 5-week-old boy with an unusual presentation of gastric pneumatosis caused by suspected necrotizing enterocolitis (NEC) after cardiogenic shock. Postnatally, a pulmonary atresia with intact ventricular septum was supplied by a modified Blalock–Taussig shunt. On transthoracic echocardiography in week 5, primarily performed to rule out pericardial effusion, air bubbles have been apparent in the right atrium. Intracardiac air found on echocardiogram is generally associated with an indwelling venous catheter but—as exemplified by this case report—may also occur in the setting of NEC. An abdominal radiograph showed an isolated gastric pneumatosis, which is an unusual location of NEC. It is speculated that air moved through the connecting veins to the right atrium, the pneumatosis located in the gastric wall being a prerequisite to this pathophysiology. *Conclusion*: The suspected NEC was located in the stomach enabling the intramural air to pass through connecting veins to the right atrium. The first specific sign of NEC in our case was air bubbles in the right atrium on echocardiography.

Keywords Children · Necrotizing enterocolitis · Gastric pneumatosis · Intracardiac air

Introduction

Necrotizing enterocolitis (NEC) occurs primarily among preterm infants with an incidence inversely proportional to the gestational age at birth. About 10 % of the cases involve term infants with reduced intestinal perfusion due to different reasons, such as shock. Because of its singular mesenteric blood supply, the small bowel is predisposed to ischaemic events [8]. Gastric involvement in NEC is unusual, probably because of the generous blood supply in the stomach, and is usually associated with a widespread disease [3, 5, 8, 10]. In the following, we report a case of suspected NEC after cardiogenic shock, located in the stomach, with an unusual presentation involving air bubbles in the right atrium on echocardiography as the first specific sign.

Case report

A male infant was born vaginally at 39 weeks of gestation with a birth weight of 3,500 g after an uneventful pregnancy. The newborn adapted well with Apgar 6/8 at 1 and 5 min. Because of cyanosis with pulse oximetry oxygen saturation of 90 %, echocardiography was performed at the first day of life and revealed pulmonary atresia with intact ventricular septum. At the second day of life, transcatheter radiofrequency perforation and balloon valvuloplasty of the pulmonary valve was performed, followed by a modified Blalock–Taussig (BT) shunt at the 11th day of life because of persistent cyanosis. The patient could be discharged from the hospital at the age of 19 days.

At the age of 5 weeks, there was a sudden deterioration with tachypnea and refusal of drinking. At the time of

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presentation, the infant was in cardiogenic shock with a minimal pH of 6.7 and a lactate concentration >20 mmol/l. An echocardiogram revealed a pericardial tamponade. An immediate puncture was done, with release of 70 ml of haemorrhagic fluid. Afterwards, the patient was transferred to our unit.

At the time of arrival, a circular pericardial effusion could be seen again on echo. On operative exploration, there was no obvious bleeding. After surgery, the patient was cardiopulmonarily stable, he was weaned from mechanical ventilation, extubated at the first postoperative day, and started on enteral feeds. Pulse oximetry oxygen saturation was around 90 %, and diastolic blood pressure was around 30 mmHg, compatible with runoff over the modified BT shunt.

Some hours later, a progressive, isolated tachypnea was noted. Recurrent pericardial effusion was suspected. On echo, there were no signs of pericardial effusion, but a large amount of air bubbles localized at the right atrium could be seen (Fig. 1). Due to the restrictive right ventricle physiology, there was a right-to-left shunt across the patent foramen ovale with air bubbles in the left heart and systemic circulation (as shown in the animation of Online Resource 1; Echocardiography.avi). An indwelling central venous line (positioned in the superior vena cava via right internal jugular vein) has been initially suspected as potential entrance of intracardiac air bubbles. Upon removal of the central line, intracardiac air bubbles persisted, and in turn, NEC was suspected.

An abdominal radiograph was obtained, which showed an isolated gastric pneumatosis (Fig. 2). An abdominal ultrasound made shortly after the radiograph exhibited portal venous air. On cranial ultrasound, there was no indication of intracranial air. Laboratory investigations revealed thrombocytopenia and a mild metabolic acidosis.

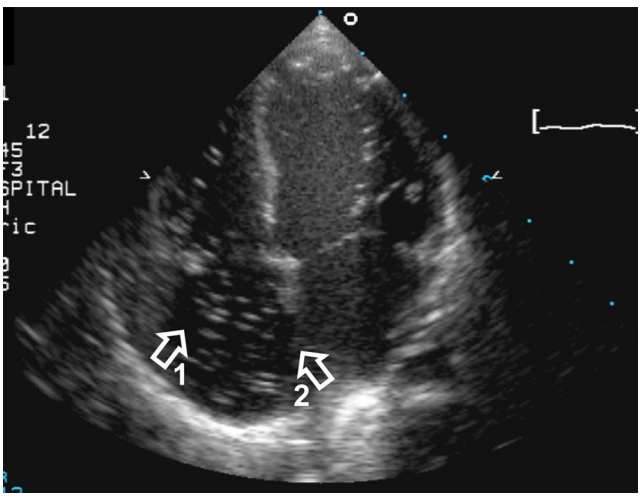

Fig. 1 Two-dimensional transthoracic echocardiography (four-chamber view) demonstrating air bubbles in the right atrium (*arrow 1*) and small right ventricle. Due to restrictive right ventricular physiology, there are also air bubbles passing the patent foramen ovale (*arrow 2*) to the left atrium and left ventricle

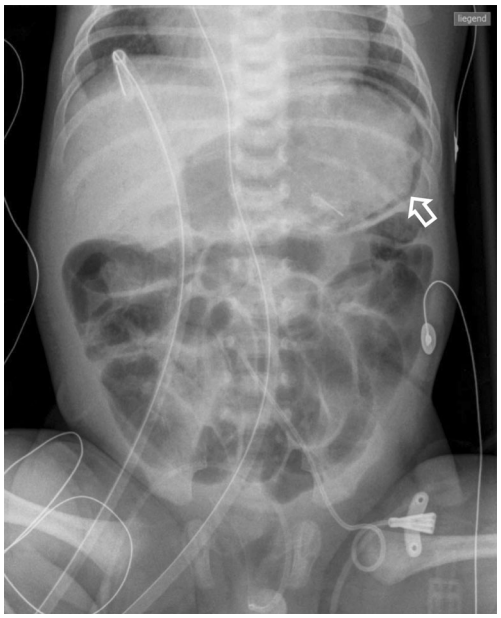

Fig. 2 Abdominal radiograph showing gastric pneumatosis (*arrow*)

The patient was treated with meropenem while being maintained on exclusive parenteral nutrition. Subsequent cranial magnetic resonance imaging 10 days after the cardiogenic shock showed a discrete restriction of diffusion in the white matter fronto-parietal on both sides without any signs of embolization.

Discussion

This case illustrates an unusual presentation of suspected NEC (intracardiac air bubbles on echo) with an unusual location of pneumatosis (stomach). To our knowledge, there is only one other case report of spontaneous intracardiac air resulting from NEC [1] (Pubmed search has been performed without any restrictions using relevant combinations of the following terms: intracardiac air, air contrast heart, air bubbles echocardiogram, necrotizing enterocolitis, gastric pneumatosis, children). Spontaneous intracardiac contrast is known in context with indwelling central venous lines. In NEC, there is a formation of intramural air in the intestine, which enters the portal venous circulation. Only in the case of a patent ductus venosus can portal venous air transverse the liver and pass into the heart. The ductus venosus closes in 76 % of healthy neonates before day 7 and in all infants before day 18 [4]. A reopening of the ductus venosus has not been observed [4]. At the time of diagnosis, our infant was already 5 weeks old; therefore, the ductus venosus was certainly closed, and on abdominal ultrasound, it was not detectable anymore. Therefore, we speculate that the air moved through the short gastric

veins, esophageal veins, azygos vein, and finally through the superior caval vein to the right atrium, the pneumatosis located in the gastric wall being a prerequisite to this pathophysiology.

Intracardial spontaneous air contrast, in particular in combination with right-to-left intracardiac shunting, raises concern for possible ischaemic adverse events secondary to embolization [1]. Both cerebral ultrasound and cranial MRI did not show any signs of embolization, since the restriction of diffusion in the white matter was not limited to a well-defined vessel region. We interpreted the tachypnea in our patient as a result of the infection and not of a pulmonary air embolization.

NEC is the most common neonatal gastrointestinal emergency [7]. The risk of developing NEC is inversely related to the gestational age at birth with extremely premature neonates at substantially greater risk than those born at full term. Aside from prematurity, no single factor or combination of factors had been found consistently to predispose a neonate to NEC [6]. But in term infants, NEC occurs almost exclusively in neonates already admitted for some other diagnoses [2]. Foremost among these are congenital heart disease (particularly duct-dependent pulmonary circulation), polycythaemia, and early onset sepsis [2]. The authors speculate that these disorders predispose to NEC because of a reduced mesenteric perfusion followed by overfeeding using cow's milk formulas [2]. In our case, there was certainly an insufficient perfusion of the gastrointestinal tract during the cardiogenic shock and probably a significant runoff over the modified BT shunt. Because of the good short-term recovery, nutrition was started early and constituted an additional risk factor.

Pneumatosis intestinalis can occur throughout the gastrointestinal tract, but is mostly diagnosed in the small bowel [8]. Gastric involvement in NEC is unusual [5], probably due to the generous blood supply of the stomach, where ischaemic and vascular events are less likely to occur as in the small bowel with its singular mesenteric blood supply [8]. In addition, infection of the stomach wall is rare as the gastric mucosal barrier, aided by the generous blood supply and acidity, prevents invasion by infectious agents [10]. Therefore, gastric pneumatosis has been attributed to widespread and severe disease requiring urgent surgical intervention [3]. In our case, the pneumatosis intestinalis was radiographically limited to the stomach. In one case report, it was hypothesized that vomiting combined with the predisposition to mucosal damage was the cause of gastric pneumatosis [9]. Our infant did not vomit and had no intestinal obstruction. Therefore, we do not have an explanation for this particular location.

Because of missing evidence of perforation and a clinically stable situation, there was no need of laparotomy, and the outcome was satisfactory.

In conclusion, our case demonstrates NEC (suspected; no histological confirmation) related to cardiogenic shock in a 5-week-old infant. The NEC was located in the stomach, enabling the intramural air to pass through connecting veins to the right atrium. The first specific sign of NEC in our case was air bubbles in the right atrium on echocardiography.

Conflict of interest All authors declare no conflict of interest.

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