Necrotizing enterocolitis following the surgical repair of a left congenital diaphragmatic hernia

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A B S T R A C T
Necrotizing enterocolitis (NEC) is an important cause of mortality and morbidity in neonatal intensive care units. It often occurs in preterms with a low birth weight, associating infectious and vascular phenomena in a context of immune system immaturity. This leads to the alteration of the intestinal wall and potentially lethal complications, requiring medical support and surgical management. In full term infants, NEC is less common and an infectious hypothesis is usually suspected given the epidemic distribution of cases. Here, we report on a rare case of NEC following the surgical repair of a congenital diaphragmatic hernia.

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Necrotizing enterocolitis (NEC) is a severe disease affecting intestines, especially in premature children, potentially leading to peritonitis and death [1]. The association of infectious, vascular phenomena and the immaturity of the immune system are often reported to explain its multifactorial pathogenesis [2–4]. In full term neonates, an infectious cause is usually suspected. However, other etiologies can be identified, such as intestinal abnormalities, congenital heart disease or any perinatal stress, which could affect mesenteric blood flow [5,6]. It has actually been shown that NEC could occur after various types of surgical procedures and that it must be included in the differential diagnosis in case of post-surgical degradation [7]. These cases, however, remain seldom, and need to be reported to help neonatologists identifying this diagnosis. Here, we report on our experience concerning an unusual NEC, following the surgical repair of a left congenital diaphragmatic hernia.

1. Case report

A 33-year-old pregnant woman, without any medical or surgical history, had her baby diagnosed at 23 weeks of gestation with a left congenital diaphragmatic hernia (CDH) containing stomach and intestinal structures. Ultrasound exam identified no residual left lung, a right mediastinal deviation and no alteration of the right pulmonary tissue, except a 20 ml volume (a third of the expected volume). The lung-to-head ratio was estimated at 2.2 using an MRI scan. The rest of the ultrasound exam was normal. The follow-up was realized in our prenatal diagnostic department, without further complications, including obstetrical, pediatric, cardiologic and surgical council and management. A full term girl was born at 38.5 weeks of gestation by vaginal delivery, helped with obstetrical forceps due to the necessity of extraction, for abnormal fetal cardiac rhythm, after a spontaneous pregnancy labor initiation.

The newborn presented a poor adaptation to extrauterine life (APGAR 2, 5, 7 and 8 at 1, 3, 5 and 10 min following delivery), was intubated 2 min after birth following intrarectal sedation by pentobarbital, and ventilated allowing a hemodynamic stabilization (heart rate > 100 bpm) with an increase of the oxygen inspired fraction to 0.6, secondary lowered to 0.38. It was immediately transferred to the neonatal intensive care unit (NICU) for high frequency oscillatory ventilation (HFOV): mean airway pressure (MAP) at 11 cm H2O, and a fraction inspired of oxygen from 0.25 to 0.3. Initial medical examination confirmed the persistence of the ductus arteriosus. Echocardiography confirmed an isolated left asymmetry of the pulmonary auscultation and a weight of 3500 g. A left CDH was confirmed by chest X-ray. Vascular access was provided via an umbilical venous catheter. Echocardiography confirmed the persistence of the ductus arteriosus.

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Initial hemodynamic and respiratory stability allowed a rapid surgical management, realized at 48 h of life under general anesthesia. Hemodynamic and respiratory stability was maintained throughout surgery using a 35 mL/kg physiological serum vascular expansion and a 5 μg/kg/min dobutamine infusion. A sub-costal approach was realized. Surgery revealed a left posterolateral CDH containing the small intestine, colon, stomach and spleen in an intra-thoracic position, due to a large posterior diaphragmatic defect. It was reported as a C defect according to the Congenital Diaphragmatic Hernia Study Group classification [8]. After examination and abdominal reintegration of the CDH contents, a Gore-tex patch was sutured due to the extent of the diaphragmatic hernia. There was no alteration of the abdominal organs and they were reintegrated without elevation of abdominal pressure. Abdominal sutures were realized above a patch, without consequences on the inferior member saturations in oxygen. We did not observe any modification of respiration or hemodynamic parameters at the time of abdominal suture.

A central venous catheter (CVC) was placed in the left subclavian vein. HFOV was continued (MAP = 10 cm H2O, oxygen from 0.35 down to 0.25). Nitric oxide (NO) was introduced at 10 ppm, based on echocardiography findings of pulmonary hypertension. Post-operative blood tests were normal. Sedation was obtained by IV infusion of 2 μg/kg/h of fentanyl and 40 μg/kg/h of midazolam. A fast was maintained, and total parenteral nutrition was provided.

Post-surgical management showed a favorable evolution leading to a decrease of ventilation parameters (MAP = 9 cm H2O, oxygen = 0.25, NO = 5 ppm), a hemodynamic stability (3.5 μg/kg/min dobutamine infusion). Continuous enteral feeding (anti-regurgitation milk) was initiated 96 h after surgery, at the resume of bowel function. In particular, we observed a conserved diuresis, a non-tender abdominal palpation and the absence of lower extremities oedema. Unfortunately, complications appeared 48 h later with respiratory difficulties leading to a necessary increase of ventilation parameters for both oxygenation and carbon dioxide epuration (frequency = 13 Hz and peak to peak at 52), of NO to 10 ppm to counter pulmonary hypertension. Hemodynamic stability was maintained with a 3.5 μg/kg/min dobutamine infusion. Chest X-ray revealed a left pleural effusion, with a right mediastinal deviation. Monogen milk was then initiated for continuous enteral feeding at 96 h post surgery with small amounts of antireflux milk following the standard operating procedures of the NICU. HFOV was continued throughout the procedure (MAP = 10 cm H2O, oxygen from 1 to 0.35). Postoperative blood tests were normal and sedation assured by a 2.3 μg/kg/h fentanyl and 60 μg/kg/h midazolam infusion.

This second surgery was followed by 2.5 weeks of positive evolution with the removal of the dobutamine hemodynamic support and the reduction of HFOV parameters, except for the development of oedemas probably connected to pulmonary hypertension. Continuous enteral feeding was progressively reintroduced at 15 days post-surgery. A sudden degradation then appeared with an increase of jejunostomy outflows, the necessity to reintroduce hemodynamic support to counter pulmonary hypertension and the development of an inflammatory syndrome rapidly connected to a Staphylococcus epidermidis sepsis, leading to the administration of an initially large antibiotic therapy (vancomycin, metronidazole, amikacine and vancomycine), secondary reduced to vancomycin infusion after bacterial identification. The CVC was still in place for parenteral nutrition and drug administration.

This sepsis would, unfortunately, remain uncontrolled with the development of an exudative enteropathy, the necessity to increase the hemodynamic support (1 μg/kg/min norepinephrine infusion) in connection with vascular expansion to insure a medial arterial pressure around 40 mm Hg. The next 48 h led to a multiple organ dysfunction syndrome and death at 33 days after birth. The autopsy revealed acute and chronic NEC lesions (perforations, fibrosis, and stenosis mainly) of variable stages, acute bronchopneumonia lesions in both lungs, sub-diaphragmatic fibro-inflammatory alterations due to the initial surgery with spleen and common bile duct infarcts. Multiple septic thromboses were identified in pulmonary and mediastinum vessels.

2. Discussion

NEC is a known complication occurring in neonates, but also in adults [9]. However, it has been rarely reported in full term neonates. The originality of this case is the initial surgery, which concerned the repair of a left CDH. This abnormality is not identified in previous series as a major source of post-operative NEC, the leading cause being neurosurgery [7]. The occurrence of the different complications seems to follow the time pattern described previously, with a NEC development 5 days post-surgery at a post-natal age of 7 days. Contrary to the observations made in preterm neonates, the NEC expansion concerns both small and large intestines as reported in post-operative NEC. In this case, the peri-operative description of the intestines during the CDH repair does not show any sign of suffering, nor the abdominal reintegration seems to be responsible for the secondary development of NEC. The hemodynamic stability obtained during surgery and in the immediate post-surgical management in the NICU is also an element against ischemic phenomena to explain this abdominal degradation. However, it would have been useful to monitor the intra-abdominal pressure or the oxygen supply to organs using near-infrared spectroscopy to eventually exclude any chronic compression phenomena or acute abdominal compartment syndrome. The initiation of continuous enteral feeding at 96 h post surgery with small amounts of antireflux milk following the standard operating procedures of our department, based on a perfect clinical exam of the abdomen, cannot be identified as a satisfactory explanation. Another hypothesis might be the pulmonary hypertension, leading to perfusion deficiencies and thus diffuse ischemic phenomena on the entire intestines that would explain the NEC development affecting the small and large bowels. A pulmonary sepsis, clearly identified by the autopsy, might also be the initial step of this process. However, the symptom progression does not plead for this hypothesis. It seems that this post-surgical NEC results from a
multi-factorial sequence, involving various phenomena, and leading to an extended intestine suffering.

The causes and consequences of this post-surgical complication remain unclear. NEC must be considered as a potential diagnosis in the case of abdominal degradation after any surgery and at any term.

**Conflict of interest**

None to disclose.

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