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Perioperative management of a neonate with Cantrell syndrome

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Abstract
Cantrell syndrome is a congenital malformation with a pentalogy characterized by defects involving the abdominal wall, lower sternum, anterior diaphragm, and diaphragmatic pericardium, as well as congenital cardiac anomalies. We recently managed anesthesia in a patient with this syndrome and herein report our experience. The patient was a 14-day-old male neonate, who had been diagnosed with Cantrell syndrome, including ventricular septal defect, left ventricular diverticulum, abdominal wall defect, omphalocele, and sternal hypoplasia. Surgical interventions to close the ventricular septal defect, resect the left ventricular diverticulum, and close the omphalocele were scheduled. After cardiac surgery, the hernial contents were returned to their original compartment and, subsequently, an attempt was made to suture the abdominal wall. However, blood pressure fell markedly and the attempt was discontinued. The chest was left open postoperatively and the patient was transferred to the intensive care unit (ICU), during which time circulatory and respiratory management was very complex. Issues requiring particular attention in the management of anesthesia for patients with this syndrome include complications of diverse cardiac malformations, pulmonary hypertension, pulmonary hypoplasia, and respiratory and circulatory failure associated with increased intraabdominal pressure due to primary closure of the omphalocele. Accordingly, extreme caution must be taken to restore respiratory and circulatory control.

Key words Perioperative management · Cantrell syndrome · Neonate

Introduction

Cantrell syndrome is a congenital malformation with a pentalogy characterized by defects involving the abdominal wall, lower sternum, anterior diaphragm, and diaphragmatic pericardium, as well as congenital cardiac anomalies [1]. We recently managed anesthesia in a patient with this syndrome and herein report our experience.

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Case report

The patient was a 14-day-old male neonate, who had been diagnosed with Cantrell syndrome, including ventricular septal defect, left ventricular diverticulum, abdominal wall defect, omphalocele, and sternal hypoplasia.

History of present illness

The patient was born by spontaneous vaginal delivery at 40 weeks and 5 days of gestation (body weight, 3888 g) at another hospital. The Apgar score was 8 points at 1 min and 8 points at 5 min. Immediately after delivery, an omphalocele was noted and the baby was transferred as an emergency case to the neonatal intensive care unit (NICU) at our hospital the same day.

Conditions present upon hospitalization

Blood pressure was 65/35 mmHg, pulse rate was 180 beats·min^{-1} (bpm), and peripheral oxygen saturation (Sp_{O_2}) was 97%. Neither cyanosis nor tachypnea was noted. Hepatomegaly was found and pulsation was confirmed at the site of the omphalocele (Fig. 1). Neither peripheral hematology nor blood chemistry tests revealed abnormalities. Cardiomegaly (cardiothoracic ratio [CTR], 55%) and signs of congestion in both lungs were noted in chest radiography findings (Fig. 2). A ventricular septal defect II (12.6 × 13.6 mm), high-flow pulmonary hypertension (PH), left ventricular diverticulum, and patent ductus arteriosus were noted in echocardiography findings. A three-dimensional computed

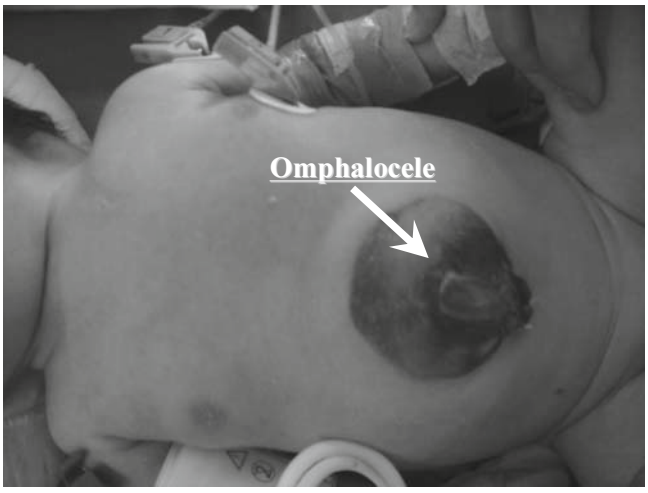

Fig. 1. An omphalocele was found. Pulsation was confirmed at the site of the omphalocele

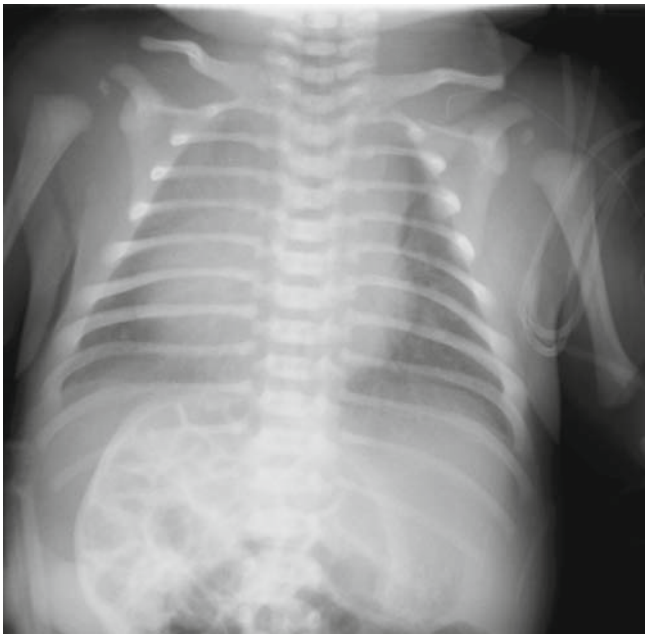

Fig. 2. On chest radiography, cardiomegaly (cardiothoracic ratio, 55%) and signs of congestion in both lungs were noted

tomography (3D-CT) examination (Fig. 3) revealed that the left ventricular diverticulum was located in front of the left hepatic lobe.

Preoperative course during hospitalization and operative details

Milk feeding was started and body weight increased satisfactorily. On the 8th hospital day, the patient was transferred to a general ward, though his respiratory rate gradually increased to more than 50 breaths·min^{-1}.

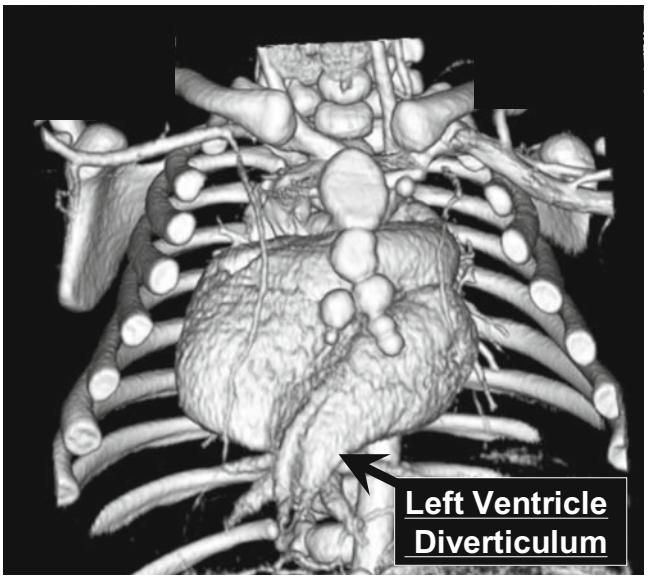

Fig. 3. Three-dimensional computed tomography (3D-CT) examination. The left ventricular diverticulum was located in front of the left hepatic lobe

On the 13th hospital day, chest radiography revealed an enlarged shadow of the heart (CTR, 70%) and on the 14th day, the respiratory rate was increased to around 70 breaths·min^{-1} and echocardiography indicated worsening of the high-flow PH. These findings indicated the need for heart surgery as soon as possible. On the 15th hospital day, surgical interventions to close the ventricular septal defect and resect the left ventricular diverticulum were scheduled. Anesthesia was induced with midazolam (1 mg), fentanyl (5 μg), and pancuronium (0.4 mg) for tracheal intubation, then maintained with air, oxygen, sevoflurane, fentanyl (25 μg/kg in total), and pancuronium (0.5 mg·kg^{-1} in total). Immediately before the patient was weaned from extracorporeal circulation, we administered dopamine (6 μg·kg^{-1}·min) and milrinone (0.5 μg·kg^{-1}·h^{-1}·min^{-1}). At that time, blood pressure was around 60/40 mmHg and heart rate was 140 bpm. Figure 4 shows a photograph of the operative field. Following the completion of cardiac surgery, an operation to close the omphalocele was started. The hernial contents were returned to their original compartment and, subsequently, an attempt was made to suture the abdominal wall. However, the patient's blood pressure fell markedly and the attempt was discontinued. The chest was left open postoperatively and he was transferred to the ICU.

Postoperative course

Immediately after the surgery, oxygenation capability declined, as indicated by a Pa_{O_2}/fractional inspired oxygen ($F_{I_{O_2}}$) (P/F) ratio of 58 mmHg, and treatment

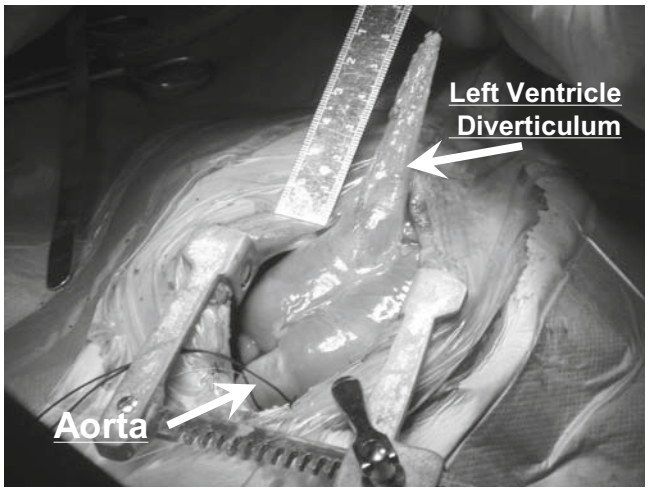

Fig. 4. Photograph of the operative field. The left ventricle diverticulum was found. Surgical interventions to close the ventricular septal defect and resect the left ventricle diverticulum were performed

with NO inhalation was started. Following the NO treatment, S_{PO_2} remained in the range of 80% to 90%, then P/F ratio recovered to 107 mmHg the next day. Accordingly, continuous hemodialysis was started on postoperative day 7. However, his blood pressure gradually decreased and S_{PO_2} was in a range below 60%, indicating the occurrence of respiratory and circulatory failure. Treatment with catecholamine did not improve blood pressure and S_{PO_2}; thus, veno-arterial extracorporeal membrane oxygenation (VA-ECMO) was started on postoperative day 8. Although intensive care was continued, the patient died on postoperative day 27.

Discussion

It has been reported that Cantrell syndrome occurs in approximately 5.5 of every 1 million births [2]. The embryologic mechanism of this syndrome is considered to be abnormal development of the ventral mesoderm at 3 to 4 weeks after conception [3]. Cardiac malformations frequently noted in the syndrome include ventricular septal defects, atrial septal defects, pulmonary artery stenosis, and tetralogy of Fallot [1], with poorer prognosis related to the severity of these congenital malformations.

Therapeutic strategies for individual defects in Cantrell syndrome should not be performed separately, but rather in parallel, and determined on the basis of whether (1) an omphalocele or abdominal wall rupture exists, (2) the heart is completely prolapsed, (3) the size of the diaphragmatic defect is large and a diaphragmatic hernia has occurred, and/or (4) malformations of the heart and great vessels are severe [4]. Regarding left ventricular diverticulum, Lowe et al. [5] have reported that a spontaneous rupture occurs very frequently, and can be explained by an increase in pressure inside the diverticulum as a result of a difference in the phase of contraction between the left ventricle and the diverticulum. Orsmond et al. [6] have recommended diverticulum resection if any of the following conditions exist: (1) cosmetic repair is performed for extracardiac malformations, (2) there is an increased risk of damage, and (3) intracardiac malformations are repaired. Because there is a risk of rupture, we think that the diverticulum should be resected whenever possible. However, because left cardiac failure is likely to intensify, as seen in the present patient, a two-stage left ventricular diverticulum resection might considered.

The issues requiring particular attention in the management of anesthesia for Cantrell syndrome include, among others, overlapping complications of different cardiac malformations, changes in hemodynamics due to compression of the exposed heart, acute respiratory and circulatory failure associated with increased intraabdominal pressure when the contents of the omphalocele are returned to their original compartment, loss of fluids and reduced body temperature due to exposure of viscera, and control of electrolytes [7]. In addition, whether repeated and frequent anesthesia is needed during radical surgery for an omphalocele, for palliative and radical operations for congenital cardiac anomalies, for plastic surgery within the precordial region, and for various tests and/or examinations is a challenging issue [7].

It has been demonstrated that patients with Cantrell syndrome and a concurrent omphalocele alone can tolerate an intraabdominal pressure of up to 25–35 cmH_2O [8]. On the other hand, patients with the syndrome as well as cardiac malformations are vulnerable to circulatory failure, as noted in our patient, and therefore require careful management. Treatment of abdominal malformations should be planned according to their extent and type. An omphalocele that has not ruptured or that is sufficiently epithelialized does not require an urgent operation, though it should be repaired early, because it is easy to treat in a young patient [9]. For adequate control of increased or decreased pulmonary blood flow due to circulatory failure, it is necessary to carefully adjust the respiratory rate, maximum intraairway pressure, positive end-expiratory pressure, the concentration of inhaled oxygen, and any other relevant parameters.

Many neonates with the syndrome develop pulmonary hypoplasia concomitantly [10]. Because the frequency of intrapericardial gastric hernia is relatively low, the cause of the pulmonary hypoplasia is not regarded as a secondary change due to compression, but rather as a genetic abnormality [11].

The occurrence of pulmonary hypertension (PH) has also been reported in patients with heart disease with decreased pulmonary blood flow, and Cantrell syndrome might be related to increased pulmonary vascular resistance [12]. In the present patient, PH occurred suddenly postoperatively. It is speculated that the hypertension may have resulted from preoperative increased pulmonary vascular resistance as well as additional acute left cardiac failure associated with the left ventriculectomy. Thus, for the perioperative management of patients with the syndrome, it is important to ensure pulmonary blood flow. Also, in our patient, oxygenation capability deteriorated as well. Treatment with NO inhalation and magnesium did not improve the oxygenation capability and, finally, VA-ECMO was performed, which failed to save his life.

Conclusions

We performed perioperative management for a neonate with Cantrell syndrome. Issues requiring particular attention during the management of anesthesia for this syndrome include, among others, complications of diverse cardiac malformations, PH, pulmonary hypoplasia, and respiratory and circulatory failure associated with increased intraabdominal pressure due to the primary closure of an omphalocele. Accordingly, extreme caution must be taken to restore respiratory and circulatory control. Moreover, in patients for whom postoperative management is expected to be difficult, a two-stage operation might be considered.

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