CASE REPORT PROF-1642

CONGENITAL DIAPHRAGMATIC HERNIA;
DELAYED PRESENTATION

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ABSTRACT... We report a case of late-presenting congenital diaphragmatic hernia (CDH) in a three-month-old male infant presenting to the pediatric hospital with attacks of cough and cyanosis .Although CDH is usually manifested on the first day of life, there are a number of case reports of late-presenting CDH usually presenting with respiratory or gastrointestinal symptoms. In this case report, we have focused on the anesthetic management of late-presenting CDH. **Text:** Congenital diaphragmatic hernia is usually manifested on the first days of life as respiratory distress and a scaphoid abdomen. The abnormality is herniation of the abdominal viscera through a defect in the diaphragm, most commonly the foramen of Bochdalek on the left side[1]. These infants often have a dramatic presentation[3], in contrast, late onset or late-presenting CDHs present outside the neonatal period with variable signs and symptoms[4]. We describe a case of late-presenting CDH that referred to the pediatric hospital with attacks of cough and cyanosis.

Key words: Congenital diaphragmatic hernia

CASE REPORT

A three-month-old male infant referred to pediatric hospital with attacks of cough and cyanosis which had started since 45 days ago.

On physical examination he was febrile, with no evidence of respiratory distress and cyanosis except for a mild substernal retraction. Air entry was reduced on the left side chest with the shift of heart apex beat to right.

Chest radiography showed intestinal loops in the left hemithorax with mediastinal shift to right (figure 1). Arterial blood gases analysis showed: PH =7.39 Pco_2=40, HCO_3=23.9, BE=-0.8, O_2 sat=90 (1%).

The infant came to the operating room with the diagnosis of diaphragmatic hernia. His weight was 6.5 kg. Pulse oxymetery showed 96% and respiratory rate was 35 breaths per minute.

After administration of atropine 0.12 mg and fentanyl 12 microgram and preoxygenation, we anesthetized the baby with halothane and oxygen so that his spontaneous breathing was maintained. After intubating the trachea with an uncuffed orotracheal tube number 3.5 , we maintained anesthesia by halothane and oxygen and atracurium. During the two hours of surgery we infused 255 milliliters of 4.5% saline & dextrose10%). We assumed no deficit replacement because it had been replaced before the surgery by crystalloids.

On laparotomy, there was a large posterolateral defect in the left hemidiaphragm with the stomach,

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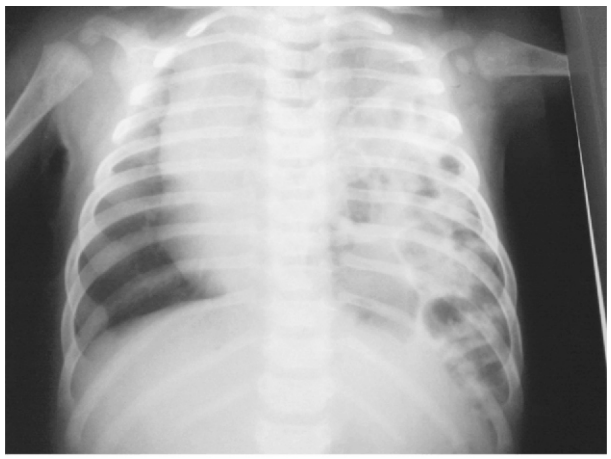

Fig-1.

proximal bowel, colon and spleen in the left hemithorax that were reduced and diaphragm was repaired (figure 2). There was also malrotation of the gut which was corrected.

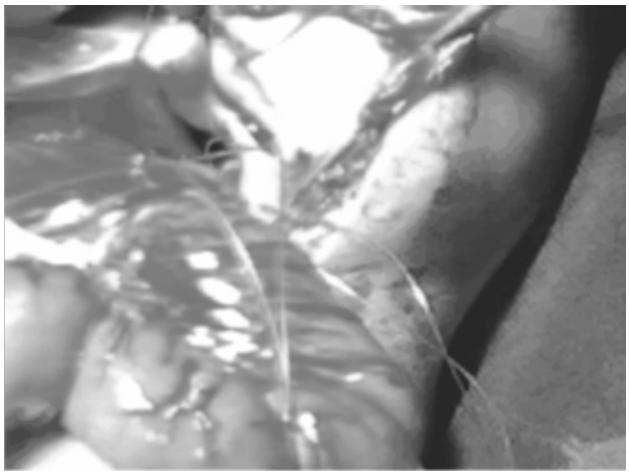

Fig-2.

During surgery, the patient was ventilated by pressure control mode and the hemodynamic conditions were stable. After reducing the viscera and repairing the defect, we had good ventilatory sounds on the left hemithorax and the peak airway pressure that was needed to ventilate the lungs did not change. The apex beat came back to its normal place.

At the end of surgery, the muscle relaxant's effect was antagonized by neostigmine 0.04mg/kg and atropine 0.02 mg/kg. The patient had good and effective respirations and we extubated the trachea when he was fully awake with good hemodynamic and respiratory factors.

The patient was sent to the pediatric intensive care unit. On the first day after surgery he had a very good hemodynamic and respiratory condition. Arterial blood gases analysis showed PH=7.33, Pco_2=40,HCO_3 =20.7,BE=-4.8,O_2 sat=98%. In the chest radiography there was normal lungs with good expansion and the two hemidiaphragms were normal.(figure 3).

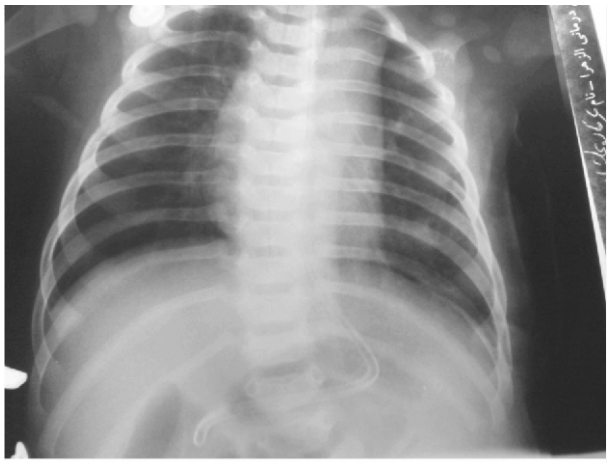

Fig-3.

The child went home after one week without any complication.

DISCUSSION

Congenital diaphragmatic hernia (CDH) occurs in approximately 1 of 2500 newborn infants[5].

Although CDH is more likely manifested on the first days of life, there are a number of case reports of late presenting congenital diaphragmatic hernias in the literature. There is little consensus in the age definition of late presenting CDH, although many authors have used between 1 and 2 months as the lower age limit[2,6,7,8]. Most cases of late presenting CDH occur in the first few years

of life[6,7], but cases are reported with some frequency well into adolescence. With the exception of age, however, the epidemiology of late presenting CDH seems to be similar to that of neonatal CDH. It demonstrates a 2:1 male-to-female ratio and a strong left sided predominance[6,7,9].

Although the diagnosis of late-presenting CDH is usually easily made by chest radiograph, the symptoms preceding diagnosis can be different. Congenital diaphragmatic hernia study group retrospectively reviewed 79 cases of late-presenting CDH collected during 1995-2005. In these cases, presenting symptoms are respiratory in 43%, gastrointestinal in 33%, both in 13% and none in 11%[6].

Respiratory symptoms were wheezing, cough and respiratory distress. Our patient had cough and cyanosis attacks as presenting respiratory symptoms.

Anesthesia concerns include (1) hypoxemia and hypotension caused by overdistention of the stomach and herniation across the midline (2) hypoxemia due to primary pulmonary hypoplasia (3) hypoxemia due to pulmonary hypotension (4) pneumothorax of the contralateral lung during attempts at high-pressure ventilation and (5) systemic hypotension caused by kinking of major blood vessels, particularly those of the liver. In general anesthesiologist's ability to control arterial carbon dioxide tension ($Paco_2$) reflects the severity of the lung pathology and therefore survival. An inability to reduce $PaCO_2$ is associated with a poor prognosis. Extracorporeal membrane oxygenation (ECMO) and the use of nitric oxide have reduced the mortality associated with this condition, and the urgency for surgical intervention has diminished and given way to a desire to stabilize the infant and minimize stress. It is common practice to postpone surgery on infant with a diaphragmatic hernia until their condition has been stabilized for several days. In some centers the closure is accomplished at the bedside in the neonatal intensive care unit[1].

In our case, the patient was hemodynamically stable with a good blood gases condition. We maintained spontaneous ventilation before intubation because we were concerned about overdistension of the stomach and herniation across the midline. We didn't administer nitrous oxide for maintaining anesthesia as its diffusion into the loops of bowel presenting in the chest may result in overdistension of those loops with subsequent compression of lungs[10]. During intraoperative mechanical ventilation, airway pressures maintained less than 25 to 30 cmH_2O to minimize the risk of pneumothorax[10]. Prolonged postoperative ventilation is required for almost all neonates with CDH[10], but our patient had good spontaneous ventilation and SaO_2 was adequate, so we extubated the patient at the end of the surgery and ventilation and arterial blood gases analysis were good in the ICU. It can be due to good lung maturation in our patient.

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