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Case Report

Right-sided congenital diaphragmatic hernia: Case report

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Right-sided congenital diaphragmatic hernia is a rare clinical condition that often presents in the neonatal period. It is important to make a differential diagnosis and decide on a clinical course for treating this condition as quickly as possible. We report a newborn with respiratory distress initially thought to be related to pneumothorax. The definite diagnosis of right-sided congenital diaphragmatic hernia was made following further imaging studies. Chest ultrasound may be helpful for preventing misdiagnosis. We also discuss the proper management of right-sided congenital diaphragmatic hernia.

Keywords: congenital; diaphragmatic; hernia

1. Introduction:

The incidence rate of congenital diaphragmatic hernia (CDH) is 1 in 2500-3000 live births [1]. This condition results from the incomplete formation of the diaphragm with herniation of abdominal organs into the thoracic cavity [2].

Classification of CDH depends on the anatomic location. The most common type is left posterolateral defect (Bochdalek hernia). Right-sided CDH presents in up to 15% of all cases [1]. In Taiwan, there are about 160,000 newborns and fewer than 10 right-sided CDH cases every year. It is important to identify the diagnostic features of this condition and to manage it properly. As the incidence rate is low, it is easy to mistake it for another clinical condition such as neonatal pneumothorax which occurs in 1-2 of every 100 live births. In this case report, we describe the clinical course of a newborn with right-sided CDH.

2. Case History:

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A female infant without significant family history was born at 37 weeks gestational age via normal spontaneous delivery (NSD). Initial development was in the less than third percentile with Apgar score of 7 to 9. She was transferred from a local hospital due to intrauterine growth retardation and suspicion of tetralogy of Fallot. Symptoms included reduced activity, subcostal retraction, and desaturation after birth.

The initial laboratory results revealed thrombocytopenia and decreased PaO2 (73.3 mmHg) without acidosis, which indicated neonate respiratory distress. On chest X- ray there were radiopaque regions in the bilateral lung fields with the presence of gas in the right pleural space (Fig.1), mimicking pneumothorax. We performed chest tapping but her condition worsened. (Fig.2). Follow-up X-ray showed iatrogenic tension pneumothorax, which was more definite than on initial x-ray. We inserted a chest tube, which relieved her symptoms (Fig.3). Once she was stabilized, chest CT showed right side diaphragmatic hernia, which contained the liver (Fig.4).

We arranged for repair of the diaphragmatic hernia the following day. After making right subcostal incision, the muscular defect was observed, measuring 4x3 cm with thin membrane. It was located

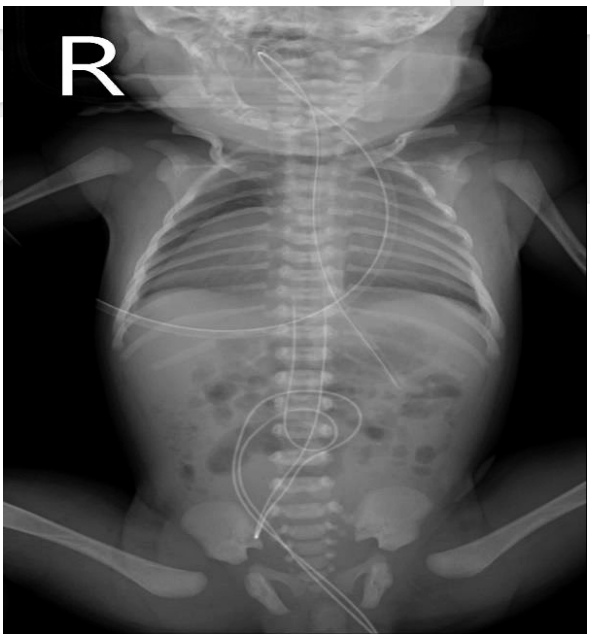

Fig. 1 Bilateral radiopaque region of lung with presence of gas in the right pleural space.

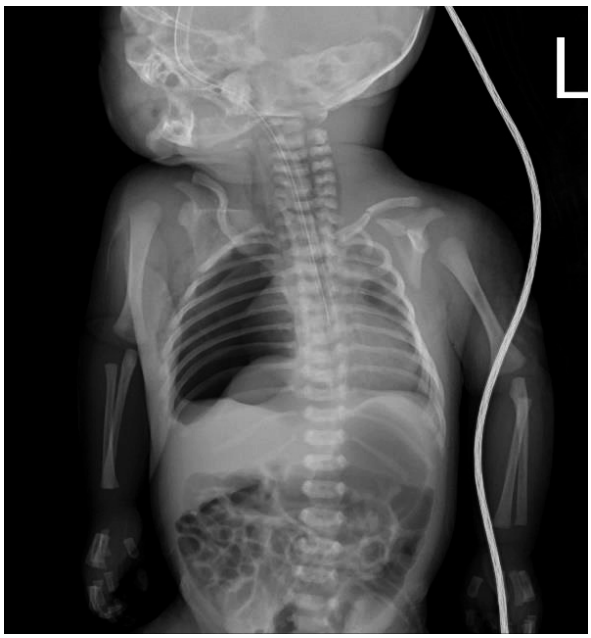

Fig. 2 After chest tapping, tension pneumothorax was impressed.

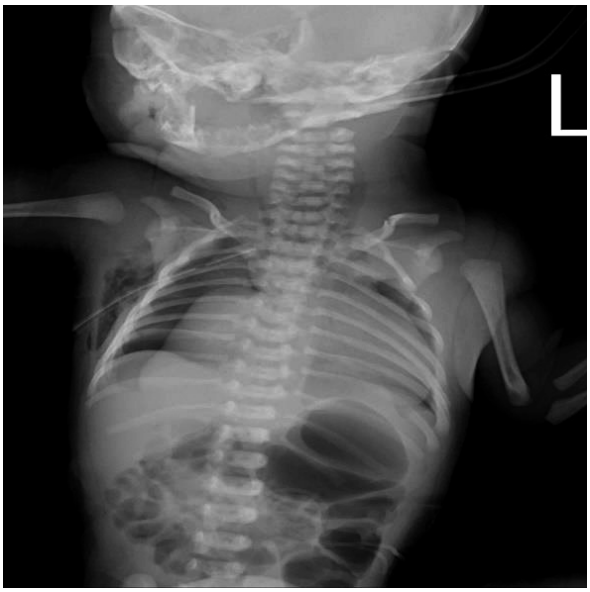

Fig. 3 Clinical condition improved after insertion of chest tube.

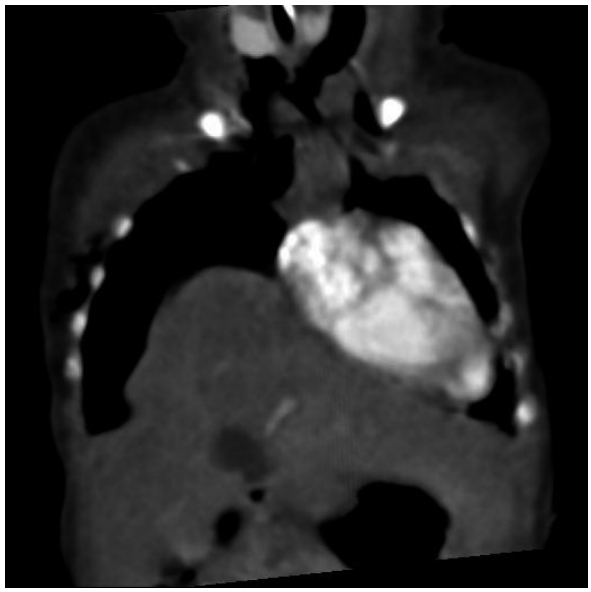

Fig. 4 Right-sided diaphragmatic hernia containing the liver.

in the anterior region of the right-side diaphragm with liver migration. After pulling the liver back into the abdominal cavity, the defect was repaired with 4-0 silk interrupted sutures and reinforced by Dexon mesh. There was good movement of the right-side diaphragm on postoperative X-ray.

3. Discussion:

CDH arises from a developmental defect of the diaphragm, leading to protrusion of abdominal organs into the thoracic cavity and abnormal development of both the abdominal and thoracic cavities. It occurs in 1 out of every 2500-3000 live births [1].

Compared with left-sided CDH, right-sided CDH is less common (15%) and sometimes accompanied by liver herniation. It may also result in higher mortality and morbidity [3] as many of the defects are large [4]. However, other studies have demonstrated that high mortality rate of right-sided CDH is not associated with the size of the defect but, rather, pulmonary hypertension and the requirement for pulmonary vasodilator and support [1].

It is also important that CDH be correctly and promptly identified and managed. Factors for evaluation include assessment of lung volume, liver herniation, pulmonary hypoplasia, and observed-to-expected lung-head ratio on sonography. Observed-to-expected lung-head ratios of $< 45\%$ and $\leq 25\%$ have been reported to predict poor outcomes for right-sided CDH and left-sided CDH, respectively [3].

On chest X-ray, there may be bowel loops or air-filled stomach in the hemithorax on the same side as the CDH with mediastinum shifting, confounded by features of pneumothorax. Considering that the incidence of neonatal pneumothorax is around 1.3 per 1000 live births, which is higher than that of CDH, it is a priority consideration in infants presenting with acute respiratory failure [5]. However, misdiagnosis may worsen the infant's condition, as in our case.

Ultrasound may be helpful for proper diagnosis, especially for right-sided CDH [6]. The CDH ultrasound pattern includes partial absence of the pleural line or hyperechoic line of the diaphragm and presence of parenchyma of a solid organ (liver or spleen).

In this case, iatrogenic tension pneumothorax led to worsening of the condition of this infant due to misinterpretation of CDH as pneumothorax. For infants presenting with emergent respiratory failure and features of pneumothorax, CDH should be considered. Moreover, clinical features should be evaluated carefully before performing an invasive procedure and chest ultrasound images should be obtained as they may be helpful for diagnosis of CDH.

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