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CASE REPORT

VACTERL Association Complicated with Right-sided Congenital Diaphragmatic Hernia

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We describe a neonate with VACTERL association and right-sided congenital diaphragmatic hernia (CDH). Such coexistence is rare. The lack of symptoms during the early neonatal period, the absence of bowel loops herniated into the right thoracic cavity, and an unfinished surgery led to clinical and radiological diagnostic difficulties. Respiratory distress occurred when the patient was 2 months old. Chest radiology plain film revealed typical findings of right-sided CDH. The diagnosis was confirmed after surgical exploration.

1. Introduction

VACTERL association is an acronym that includes vertebral defects, anal atresia, cardiac defects, tracheoesophageal (TE) fistula, renal anomalies, and limb defects. The incidence of VACTERL association is one in 10,000—40,000 births.[1] Patients diagnosed with VACTERL association have three or more of these features. Other less frequent

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defects are prenatal and postnatal growth deficiency, laryngeal stenosis, ear anomaly, large fontanels, defects of ribs, and external genitalia anomaly.[2] Otherwise, the incidence of congenital diaphragmatic hernia (CDH) is one in 2000—5000 births.[3] Approximately 30—40% of patients with CDH have additional congenital anomalies, mainly of the heart, central nervous system, and genitourinary system. Coexistence of VACTERL association and CDH has been reported, but these are mostly left-sided hernias.[4] Right-sided CDH complicated with VACTERL association is very rare. In a review of the literature, few case reports were found.[5,6] We report a neonate with such coexistence, and the late-presenting symptoms of CDH made the diagnosis more difficult. We believe our case report makes a useful contribution to the literature.

2. Case report

A 2830-g male infant was born at 36^{+5} weeks' gestational age to a G2P0A1 mother by cesarean section due to prolonged labor. His Apgar score was 9 and 10 at 1 minute and 5 minutes, respectively. Prenatal sonography had shown the following: (1) a so-called double vessel sign, which occurs when the azygos vein is seen next to the aorta and the inferior vena cava is not seen; (2) aberrant hepatic vasculature, and left atrial isomerism (LAI) was suspected. The baby presented to our ward with respiratory distress. Physical examination revealed no gross dysmorphic features except for an imperforate anus with a perineal fistula. A nasogastric tube was not successfully passed from the nose to the stomach. Supplemental oxygen was provided and treatment with antibiotics was initiated.

Plain film X-ray revealed haziness of bilateral lung fields and a right lower thoracic radio-opaque lesion (Figure 1). Transient tachypnea of the newborn (TTNB) due to retention of the lung fluid was considered. Echocardiography showed situs solitus with a thin membrane attached laterally to the left atrial appendage, dividing the left atrium into two chambers, which indicated cor triatriatum. There was no obvious obstruction of pulmonary venous return. There was no LAI. Computed tomography (CT) of the chest and abdomen showed marked focal dilatation of the upper esophagus with a blind end as well as evidence of a TE fistula between the trachea and mid-esophagus. There was a large mass measuring 3.4 cm × 4.7 cm in the right lower posterior lung (Figure 2A). X-rays of the spine and kidney sonography were normal. There were no limb defects. VACTERL association was diagnosed.

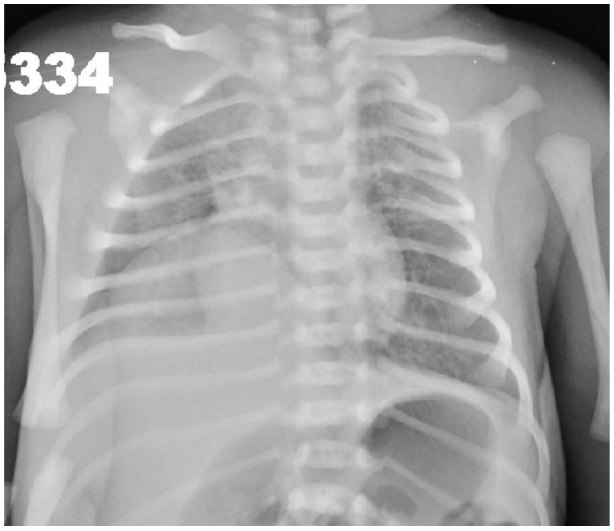

Figure 1 Uneven haziness of the bilateral lung fields with a prominent right lower thoracic radio-opaque lesion.

On the 2nd day of life, thoracoscopy repair of the esophageal atresia was done. A total of three ports (3 mm, 5 mm, and 3 mm) were placed along the right third, fifth, and fourth intercostal spaces, respectively, to perform the operation. The surgeons ligated and divided the TE fistula. The proximal esophageal pouch was mobilized to gain enough length to create the new anastomosis. Just prior to the accomplishment of anastomosis, the patient became bradycardic and hypoxic. After resuscitation, his condition stabilized. The operation was then converted to an open, right-sided thoracotomy and anastomosis was accomplished. A posterior sagittal anorectoplasty was performed at the same time. The surgeons were not able to explore the mass in the right lower posterior lung due to the event of hypoxemia necessitating resuscitation and prolonged surgery.

The patient was extubated 3 days later, and feeding was attempted on postoperative Day 10. He had no respiratory distress after recovery from the surgery. A series of examinations was arranged to evaluate the right lower thoracic mass. Abdominal sonography showed a suspicious subphrenic hypoechoic mass from the liver, but diaphragmatic eventration with elevation of the liver could not be ruled out. Chest CT with CT angiography was arranged. A well-defined mass was observed in the right lower hemithorax. This lesion appeared to be attached to the adjacent liver. A feeding artery from branches of the hepatic artery to the mass was seen after intravenous injection of contrast medium in the early arterial phase (Figure 2B). The radiologist diagnosed a focal eventration of the right hemidiaphragm rather than a diaphragmatic hernia. The patient was discharged without respiratory symptoms. Regular follow-up was suggested.

One month after discharge, the patient returned for follow-up abdominal ultrasound. The mass remained. Six days later, he was brought to our emergency room due to shortness of breath. Chest X-ray showed several cystic-like lesions in the right lower hemithorax and blurring of part of the right hemidiaphragm (Figure 3). Right thoracotomy was performed under the impression of right-sided CDH. A posterolateral diaphragmatic hernia defect (3 cm) was identified with herniated organs of the colon and liver. No evidence of right lung hypoplasia was found during the surgery. No sac was seen. The defect was repaired with a little tension. However, respiratory acidosis persisted and he could not be weaned off the ventilator due to coexisting respiratory syncytial virus infection until postoperative Day 7. Feeding increased gradually. In a stable condition, the patient was discharged on postoperative Day 14.

The patient had fair weight gain and an adequate urine amount. The cardiologist arranged cardiac catheterization when he was 3 months old. Contrast injection to the right atrium showed mild dilatation of the right ventricle and