Bowel sounds in the chest of a 7-day-old baby: what's going on?

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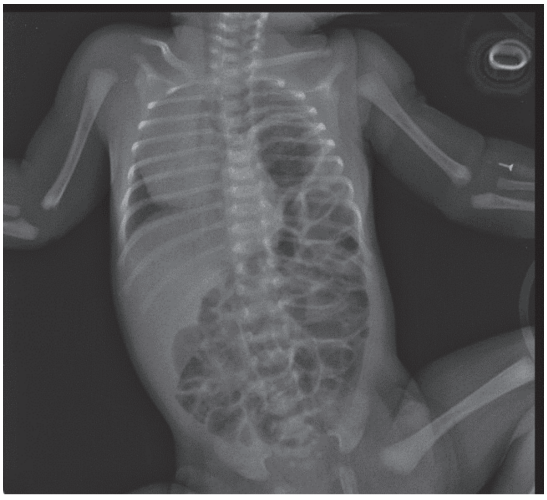

A 7-day-old female neonate presented with feeding difficulties and respiratory distress. She was born at term by vaginal delivery, weighed 2625 gr, with Apgar scores 9 and 10 in the first and fifth minutes of her life. Her mother did make complete prenatal care, and she reported smoking 10 cigarettes a day during the pregnancy. The baby was born well and evolved without respiratory distress, with good reactivity, a positive neonatal reflex, the emission of meconium in the first 24 hours, and breastfeeding with valid suckling. The baby was discharged at home in good condition. Three days later, she presented with feeding difficulties, and she was admitted to our institute. At admission, the heart rate was 180 bpm, the respiratory rate was 70/minute, and the oxygen saturation was 93% in room air. At the clinical examination, subcoastal retractions and inspiratory stridor with cyanosis were evident. A right-sided heart auscultation and bowel sounds in the chest were detected. We decided to perform an X-ray of the chest and abdomen.

Question

Based on the X-Ray imaging and the clinical history, which is the correct diagnosis?

A Kartagener syndrome
B. Bochdaleck hernia
C. Morgagni hernia
D. Massive thymic hyperplasia

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Answer

A diagnosis of Bochdaleck hernia (BH) was made, and the baby was immediately intubated to perform a quick decompression of the chest with abdominal straps and prevent pulmonary hypertension. Echocardiography was performed to establish the presence and severity of pulmonary hypertension and shunting. In our case, the exam showed a mesocardiac heart, with the stomach and the spleen in the left chest and no shunting detectable. BH is a life-threatening condition. It is a congenital diaphragmatic hernia caused by a developmental defect of the diaphragm that allows abdominal viscera (intestines, spleen, stomach, kidney, and liver) to herniate into the chest.[1] Because herniation occurs during a critical period of lung development, clinical manifestations of BH result from the pathologic effects of the herniated viscera on lung development. With the rising severity of lung compression, there are corresponding decreases in bronchial and pulmonary arterial branching, resulting in increasing degrees of pulmonary hypoplasia. As a result, it is very common to have misplaced heart or bowel sounds in the chest, as well as some clinical findings of splanchnic organs in the thorax.[2] Although symptoms often occur early at birth, some cases can show a late presentation, making the diagnosis very intriguing. In our case, the first evaluation was completely negative, and the first symptoms appeared on day 7. For this reason, the clinical examination after birth and in the following days constitutes a milestone in recognizing rare diseases with unusual presentations. Another important aspect is the maternal pregnancy history. Interestingly, some studies have reported an association between smoking and BH, possibly due to the observed lower levels of vitamin A measured in the cord blood of congenital diaphragmatic hernia infants exposed to cigarette smoking.[3,4] It could reflect an alteration in the retinoic acid pathway, which has been identified as a primary mechanism in mouse models.[4] This underlines the importance of a careful collection of anamnesis to better understand various conditions related to the newborn and the mother. Finally, this case showed the importance of a comprehensive clinical approach even when there is a negative prenatal diagnosis. The screening ultrasound examinations are arguably lacking in sensitivity and prognostic value.[5] Ultrasound is currently the gold standard diagnostic test for congenital diaphragmatic hernia.6 Routine prenatal ultrasound identifies less than two-thirds of hernia pregnancies, such as displaced bowel loops, stomach or abnormal cardiac axis, mediastinal shift, and polyhydramnios.[6] In addition, in some cases, herniation of abdominal viscera into the thorax takes place presumably just at delivery through a small diaphragmatic defect; thus, prenatal diagnosis can result negative.[7] Therefore, the birth-first evaluation can be negative, as it was in our case. Typical signs of displaced organs in the chest appeared at further investigations, pointing out the need for a deeply complete evaluation in the following days. This underlines the neonatologist's important role in identifying and treating potentially life-threatening disorders. In our case, the best treatment for BH is surgical repair to prevent pulmonary hypertension, which remains the major cause of mortality. Other treatments include isotonic fluid support, inotropic agents, and inhaled nitric oxide.[8] The timing of surgery is dependent on the cardiorespiratory status of the patient. The mortality and morbidity of BH are related to the severity of lung hypoplasia and pulmonary hypertension. Outcome studies have reported survivance rates of 80 percent at tertiary centers. Survivors are at risk for respiratory infection, gastroesophageal reflux, failure to thrive, recurrence, neurodevelopmental delay, and musculoskeletal deformities.[9]

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