Answers to Medical Quiz

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1. Cystic shadow at the base of the left chest, diaphragmatic contour is not seen, shift of mediastinum to the right side and gasless abdomen.

2. The most likely diagnosis is left congenital diaphragmatic hernia.

3. The other two possible differential diagnoses in this patient are cystic adenomatoid malformation and bronchopulmonary sequestration.

DISCUSSION

The three basic types of congenital diaphragmatic hernia include the posterolateral Bochdalek hernia, the anterior Morgagni hernia, and the hiatus hernia. The left-sided Bochdalek hernia occurs in approximately 85% of cases[1,2]. Left-sided hernias allow herniation of both the small and large bowel and intraabdominal solid organs into the thoracic cavity. In right-sided hernias (13% of cases), only the liver and a portion of the large bowel tend to herniate[1,2]. Bilateral hernias are uncommon and usually fatal. Both sexes are affected equally. Most infants with congenital diaphragmatic hernia manifest at birth with respiratory distress and cyanosis, although a later presentation is possible[1].

The diagnosis of congenital diaphragmatic hernia is based on prenatal ultrasonography findings in approximately one half of affected infants[3]. Infants may have a prenatal history of polyhydramnios. Postnatal diagnosis of diaphragmatic hernia often can be made by history and physical examination in which infants frequently exhibit a scaphoid abdomen, barrel-shaped chest, and signs of respiratory distress (retractions, cyanosis, grunting respirations)[4].

In left-sided posterolateral hernia, auscultation of the lungs reveals poor air entry on the left, with a shift of cardiac sounds over the right chest and the presence of bowel sounds in the chest. In patients with severe defects, pneumothorax signs (poor air entry, poor perfusion) may also be found. Plain chest x-ray is usually required to confirm the diagnosis[4].

Treatment of this condition requires endotracheal intubation and mechanical ventilation in all infants who present in the first hours of life[1,5]. If the diagnosis is known at the time of delivery, avoiding bag-and-mask ventilation in the delivery room is essential because the stomach and intestines become distended with air and further compromise pulmonary function[5]. A nasogastric tube should be placed as soon as possible to provide stomach decompression. Medical therapy is directed to stabilize blood pressure, circulating volume, pulmonary distress, and hypoxemia. This therapy may extend to ten days or more and may require extracorporeal membrane oxygenation[6,7]. Surgical repair is needed after stabilization and the open abdominal approach is the most preferable to pediatric surgeons. Open thoracotomy and thoracoscopic technique are other options in selected cases[4,8]. The overall survival rates for infants after congenital

diaphragmatic hernia repair range from 40-90%[4,8]. The survival rate depends on the degree of pulmonary hypoplasia and pulmonary hypertension[6,11].

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