Cystic Pulmonary Hamartoma Simulating Posterolateral Diaphragmatic Hernia

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SUMMARY
Posterolateral diaphragmatic hernias and cystic hamartomatous malformations of the lung can cause acute respiratory distress in the immediate postnatal period. The clinical and radiological features of these two conditions can be similar.

Owing to misdiagnosis, the surgical approach was inappropriate in 2 of 26 patients diagnosed as having posterolateral diaphragmatic hernias on admission. During the same period, 5 cases of congenital cystic lung disease were treated surgically. Three of these patients were admitted in severe respiratory failure within 48 hours of birth, and in 2 of these cases the wrong diagnosis was made. The clinical features of these two easily confused congenital anomalies are discussed, and the main differentiating points on the chest radiographs are presented.

At the onset of symptoms and signs of respiratory distress, the newborn, under ideal circumstances, is cared for in an intensive care area of a maternity hospital by a neonatologist. To establish the cause of the pulmonary disorder, an urgent chest radiograph is required. Often the films obtained are of poor diagnostic quality, as they are taken hurriedly by a mobile machine because of the severe respiratory insufficiency. In these instances, when a surgically correctable cause is found, the incorrect interpretation of the radiographic appearance may lead to a wrong surgical exploration, for example, an abdominal exploration instead of a thoracic approach.

CASE REPORT
The patient, born after a normal pregnancy and delivered vaginally, was referred to a paediatric surgeon when 48 hours old with a diagnosis of diaphragmatic hernia. On admission, severe respiratory insufficiency was present, and there were signs of a large left-sided intrathoracic air-filled mass causing displacement of mediastinal structures. The abdomen did not appear scaphoid.

A chest radiograph confirmed the presence of an air-filled lesion in the left thorax (Fig. 1a). Cystic structures of varying sizes were seen in the left lower hemithorax. The left diaphragm was not visible, but a normally sited intragastric air/fluid level was noted. Because of the severity of the respiratory distress, the baby was intubated and mechanical ventilation was instituted. This only partially alleviated the insufficiency. To resolve the uncertainty about the presence of a congenital diaphragmatic hernia, a small amount of contrast material was introduced via a nasogastric tube and a further radiograph was obtained (Fig. 1b), on which the intrathoracic air-filled cyst-like structures appeared to have increased in size. The distribution of the contrast material in the upper gastro-intestinal tract was unhelpful, and a congenital diaphragmatic hernia was therefore diagnosed and a laparotomy performed. No intra-abdominal or diaphragmatic abnormality was found. A thoracotomy then followed and a grossly abnormal cystic lower lobe of the left lung was detected and resected. Histologically, the resected pulmonary tissue had the appearance of a cystic adenomatoid malformation.

COMMENT
Most paediatric surgeons prefer a transabdominal approach when repairing a posterolateral diaphragmatic hernia in the newborn. This has the added advantage of exposing the abdominal cavity and its contents and gives the surgeon the opportunity to correct a congenital rotational anomaly of the intestine which is often present. The herniated bowel on reduction can be accommodated in the abdominal cavity, as this approach also allows manual stretching of the anterior abdominal wall. By contrast, cystic pulmonary dysplasias are resected through a thoracotomy incision.

Over an 18-year period, 36 patients with congenital posterolateral diaphragmatic hernia have been treated by the paediatric surgical service at the Red Cross War Memorial Children's Hospital. Twenty-six of these patients were seen within 24 hours of birth, and all presented in severe respiratory failure. An inappropriate operation was performed in 2 of the 26 patients owing to misinterpretation of the chest X-ray film, intrapulmonary air-filled cysts being confused with intestinal intraluminal gas. In both instances, cystic adenomatoid malformation of the lung was the underlying cause of the respiratory failure.

During the same period, 5 patients with congenital cystic lung disease were treated surgically. Of these, 3 presented within 48 hours of birth with severe respiratory insufficiency. The misdiagnosis occurred in 2 of these 3 cases.

In a newborn baby with a suspected congenital posterolateral diaphragmatic hernia, radiological confirmation is sought. The radiograph shows a continuum of gas shadows extending from the abdomen into the hemithorax and displacing mediastinal structures. The gas-filled intrathoracic intestine has an X-ray appearance identical to that normally seen on the abdominal survey.
films (Fig. 2). Unless the diaphragmatic shadow on the side of the hernia is visualized, which is usually not the case, the correct diagnosis is easily missed. Since the radiological appearance is not always the classic one, the differential diagnosis of pulmonary cystic disease must always be considered. Because of the rarity of this anomaly, this possibility is seldom considered. When the respiratory insufficiency is not too severe, a less hurried approach with better diagnostic assessment of the newborn with respiratory distress can take place. Errors in diagnosis are less likely under these circumstances, but a hurried evaluation done under emergency conditions may result in misdiagnosis in a few cases.

On clinical grounds alone, it may be impossible to differentiate between congenital lung cysts and a congenital posterolateral diaphragmatic hernia. Both of these conditions can cause respiratory distress, which may begin at or shortly after birth. As with many other causes of respiratory distress in the newborn, lung cysts are usually associated with a normal or even distended abdomen. The empty abdomen, scaphoid in appearance owing to the displacement of abdominal viscera into the thoracic cavity, may be a differentiating clinical sign but is often difficult to interpret. The local chest signs do not differentiate the two conditions from each other as the displacement of mediastinal structures, hyperinflation of a hemithorax with hyper-resonance on percussion, and diminished or absent breath sounds on auscultation are common to both. Intrathoracic bowel sounds on auscultation are a sign of doubtful clinical value because seldom noted.

So, too, the pathophysiology of the respiratory insufficiency that develops in these patients is similar. In diaphragmatic hernia, the presence of intrathoracic intestine during fetal life leads to a varying degree of underdevelopment and hypoplasia of the ipsilateral as well as the contralateral lung. Immediately after birth the volume of the misplaced intestine increases when it is distended and ballooned by intraluminal gas. This results in distortion and compression of surrounding lung and mediastinal structures and further mechanical impairment of pulmonary function.

Developmental defects of pulmonary tissue cause respiratory distress when a lung, or more commonly a segment of lung, is involved and ventilation is mechanically impaired by large air-filled cysts. It is in these instances where the diagnostic confusion can quite understandably occur.
Extremely variable X-ray appearances are seen in patients with posterolateral diaphragmatic hernias, ranging from incomplete or partial herniation of intestinal contents through the diaphragmatic defect seen especially where a hernial sac is present, or with only partial air filling of the intrathoracic intestine, to complete filling of the whole hemithorax. These factors lead to the confusing appearances. The respiratory distress in the patients with incomplete or partial herniation is often of a milder nature and of delayed onset. Time is therefore on the diagnostician's side and adequate films or radiological studies using intestinal contrast may be obtained.

On the other hand, poor air filling of intestine may be associated with strangulation of the herniated bowel. Here the radiograph showing the abdominal gas pattern may demonstrate proximal obstructive dilatation of the intestinal tract. Airless intestine or solid intra-abdominal viscera that have herniated into the thoracic cavity may further confuse this picture. The spleen, the lobes of the liver and an extrapulmonary sequestration can all give rise to opacities which may lead to difficulties in interpretation (Fig. 3). In right-sided diaphragmatic hernias a weeping liver surface partially herniated into the chest may add a further confusing element, with the development of a hydrothorax complicating the herniation. Various lesions of the bronchopulmonary system may cause abnormal cystic areas within the pleural cavity in early life, and it is often difficult to know whether a pulmonary cyst is congenital or acquired. Cystic lung disease simulating a diaphragmatic hernia in presenting symptoms and signs in the newborn period is usually congenital in type and pathologically classified as a pulmonary hamartoma.

When dysmorphic pulmonary tissue is the cause of severe respiratory insufficiency, either a large enough amount of pulmonary tissue is primarily involved by the dysplasia or segmental lesions have undergone air filling with a subsequent increase in tension and in the volume of the abnormal tissue. Bilateral cystic lesions do not cause diagnostic confusion, since bilateral congenital diaphragmatic hernias are extremely rare. However, segmental lesions give rise to large cystic lesions associated with gross mechanical compression of surrounding intra-thoracic structures, thereby compromising pulmonary function.

Another condition, although rare, which may cause confusion is congenital pulmonary lymphangiectasis. This may occur as part of generalized lymphangiectasis or secondary to pulmonary venous obstruction, but is more usually seen as a primary developmental defect of the lung. In the latter instance the lesion may be unilateral, when it must be distinguished from the rarely encountered pulmonary lymphangioma. The radiological features of this abnormality are shown in Fig. 4.

Fig. 3. Left congenital posterolateral diaphragmatic hernia with prolapse of spleen, left lobe of liver and partial air filling of the intrathoracic intestine. The inclusion of the abdomen in the study aided this diagnosis in a 12-hour-old neonate.

Fig. 4. Congenital pulmonary lymphangiectasis of the right lung in a patient 48 hours old. Note the honeycomb appearance in the mid and lower zones of the right chest due to multiple cystic cavities, some containing air/fluid levels.
Cystic adenomatoid malformation of the lung is a hamartomatous lesion involving pulmonary tissue. During fetal life cystic areas within the abnormal tissue are filled with fluid. In some cases, resorption of this lung fluid may be delayed in the immediate postnatal period and shows initially as areas of abnormal density on chest X-ray films (Fig. 5a). The classic air-filled cysts are encountered once the fluid has been absorbed and air trapping within the cystic spaces occurs (Fig. 5b and c).

Congenital cystic lung disease and diaphragmatic hernia can be differentiated from each other by a critical analysis of the radiological features of each.

Gas-filled intrathoracic intestine usually conforms to the following radiological criteria (Fig. 2):

1. The cyst-like structures are polygonal in shape. As the intraluminal gas is usually under only moderate tension, distortion of the walls of the cyst by surrounding structures readily occurs.
2. The intestinal wall is seen as a well-defined structure and appears of similar thickness at all sites.
3. The air-filled cyst-like shadows are all of similar size.
4. Intracystic air/fluid levels are absent. In rare instances, when the stomach is lying in the thorax, a gastric air/fluid level may be detected. This may be verified by means of a radio-opaque nasogastric tube demonstrating the intrathoracic position of the stomach.
5. The greatest concentration of the air-filled structures is found in the lower part of the thoracic cavity.

The following features typify the radiological appearance of hamartomatous pulmonary cysts (Fig. 1a):

1. The cysts are spherical in shape. This is a result of progressive air entrapment and the development of high intracystic air pressures.
2. The walls of the cyst are thin and often not clearly defined.
3. Areas of compression collapse of intervening and surrounding normal lung tissue may give rise to opacities distributed irregularly throughout the pathological area.
4. Air/fluid levels within the cystic cavities may be present but are very rarely encountered.

CONCLUSION

When congenital pulmonary cystic disease has to be differentiated radiologically from posterolateral congenital diaphragmatic hernia, radiographs of good diagnostic quality are necessary. This is especially so when the clinical condition of the baby does not allow fuller radiological investigation. It is important to include the abdomen as well as the chest in X-ray films. The pattern of intra-abdominal gas may aid in the differentiation between the two conditions, but the finer distinguishing points are only evident after a careful evaluation of the radiographic appearances of the air-filled cystic areas within the chest cavity. Thus a careful and unhurried appraisal of the chest radiographs should indicate the correct diagnosis and thus prevent an inappropriate surgical approach.

REFERENCES