Case Report

Pulmonary hypoplasia with hepatic and renal anomalies in a dead fetus

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ABSTRACT

Pulmonary hypoplasia is a developmental malformation characterized by incomplete development of lung tissue. During routine fetal autopsy of an apparently normal female dead fetus of 36 weeks gestation presented completely hypoplastic left lung, partially hypoplastic right lung, right-sided shift of heart, right-sided shift of trachea, left-sided diaphragmatic hernia through which an extra lobe from left lobe of liver extended into the left half of thoracic cavity. Left kidney was iliac in position.

KEY WORDS: Congenital diaphragmatic hernia, extra lobe of liver, hypoplasia

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INTRODUCTION

Pulmonary hypoplasia (PH) is a developmental abnormality of the lung with decrease in size and weight of lung. The cause and severity of the lesion depends upon timing of intrauterine insult during lung development and presence of other associated developmental anomalies of cardiac, genitourinary, gastrointestinal, renal and musculoskeletal systems.[1-3] With the advancement of medical knowledge for other pulmonary conditions, there is gradual decrease in the neonatal morbidity and mortality. Immaturity in the development of respiratory passages and lung can lead to increased prevalence of respiratory illnesses in neonatal to senile age groups and are clinically challenging to the neonatologists, pediatricians, chest physicians and geriatricians alike. PH has been described as the most common anomaly in infants who die in the neonatal period. PH can result from several causes, and it can often be suspected on the basis of previous obstetric history or ultrasound findings during the current pregnancy.[4] Appropriate medical or surgical intervention is required to treat the infants with PH.

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CASE REPORT

The present case is a 36 weeks spontaneously delivered still born female fetus weighing 1.7 kg with 32 cm crown–rump length (CRL) of a second gravida with a previous obstetric history of still born baby. During routine thoracic and abdominal autopsy the fetus presented a left-sided diaphragmatic hernia of 1 inch diameter. In the thoracic cavity, right-sided shift of heart and trachea and a grossly reduced left lung compared to right lung was observed [Figures 1 and 2]. The trachea and its bifurcation in to principal bronchi was normal [Figure 2] with a clearly visible lumen. Beyond principal bronchi the bronchial tree could not be differentiated.

The left lung was thin, quadrilateral in shape, without lobation and nearly 1/10th of normal size for the gestational age. The right lung presented three lobes but the middle lobe and part of upper lobe are thin and fibrosed [Figure 2]. On microscopic examination, entire left lung and the middle lobe and the lower part of upper lobe of right lung contained small amount of lung tissue and more of connective tissue. Microscopic appearance of left lung and fibrosed parts of right lung correspond to pseudoglandular stage of lung development with undifferentiated intrapulmonary bronchi and alveoli [Figures 3a-b]. Rest of the right lung presented terminal sac stage of development which is normal for the gestational age.

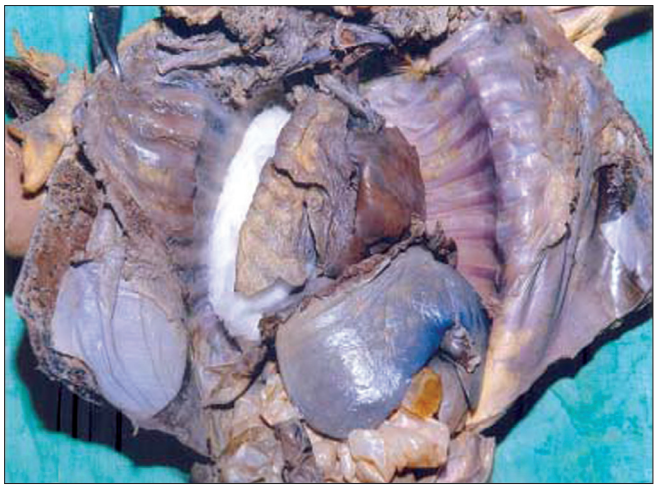

Figure 1: Showing empty left thoracic cavity with left hypoplasia

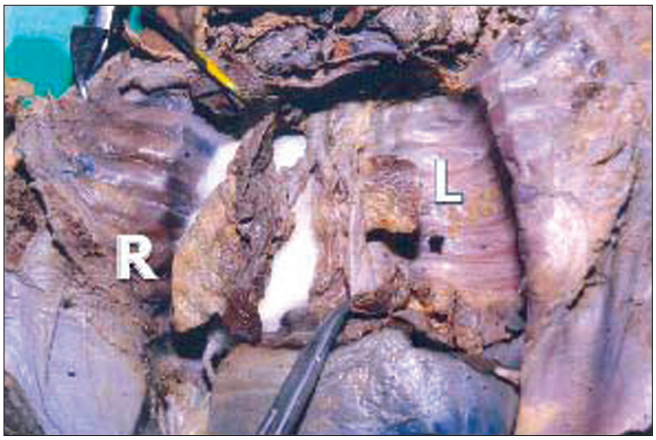

Figure 2: Showing thoracic cavity after removal of heart and left-sided diaphragmatic hernia through which an accessory left lobe entered inside the thoracic cavity

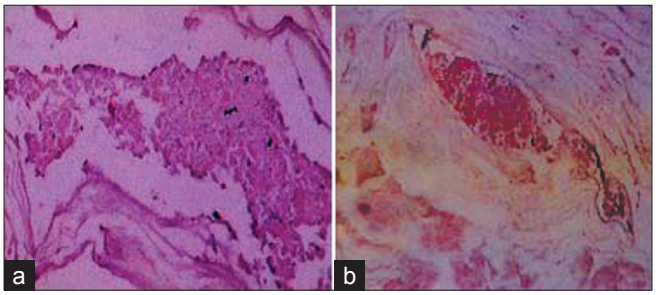

Figure 3: (a) and (b) – fibrosed parts of left lung correspond to pseudoglandular stage of lung development with undifferentiated intrapulmonary bronchi and alveoli

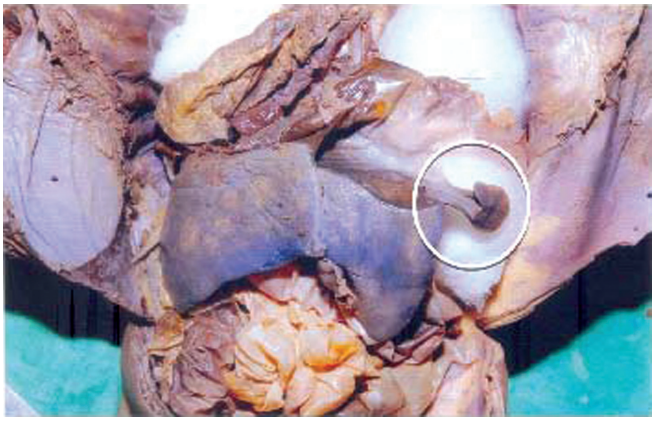

Figure 4: Showing liver with an accessory left lobe

In the abdominal cavity, a third hepatic lobe [Figures 2 and 4] in the form of a stalk of 5.0 cm length was extending from the left lobe of liver into the thoracic cavity passing through the diaphragmatic defects with a terminal circular dilatation of 2.0 cm width. Terminal dilatation of third lobe projecting into the left thoracic cavity presented normal histological features of liver, while the stalk connecting it with the liver presented fibrous tissue. The left kidney was iliac in position, while the right kidney was in normal position.

DISCUSSION

According to Sultana *et al*,[5] PH occurs in all cases of congenital diaphragmatic hernia (CDH) and reported an incidence of 7.8% based on 165 newborn fetal autopsies. According to Harrison *et al,*[6] the incidence of CDH is 1 in 2200 live births with a 50% mortality rate in prenatally diagnosed cases. The worldwide incidence of PH is approximately 13%, with a range of 9–28% in United States.[3] Incidence of PH in live births is 1 in every 10000–12000 (Moore *et al*. 1992). The reported incidence of varying degree of PH in neonatal autopsies varies from 7 to 26%.[7] If unilateral, left lung is more commonly involved than the right lung. Development of the bronchial tree takes place at about 26th to 31st day of intrauterine life.[2,8] Categorized maldevelopment of lung into three groups:

Group 1: Agenesis, in which there is complete absence of lung tissue.
Group 2: Aplasia, with rudimentary bronchus and without lung tissue.
Group 3: Hypoplasia with normal pulmonary tissues that is under-developed.

Monaldi classified maldevelopment of lung into four groups.[2] They are
Group 1: No bifurcation of trachea.
Group 2: Only rudimentary main bronchus.
Group 3: Incomplete development after division of main bronchus.
Group 4: Incomplete development of subsequent bronchi and small segment of the corresponding lobe.

In the present case, right lung presented the features of Group 3 of Boyden and Group 3 of Monaldi classification, while the left lung presented Group 3 of Boyden and Group 4 of Monaldi.Hypoplasia of the lung may be regarded as primary (idiopathic) or secondary. Primary hypoplasia occurs without any cause. Secondary PH is frequently associated with adverse intrauterine influences that cause fetal lung compressions which may be intrathoracic or extrathoracic.[9] The intrathoracic causes

are diaphragmatic defects, intrathoracic tumors, while extrathoracic causes are oligohydramnios, renal agenesis or chronic elevation of hemidiaphragm.[3] The cause of PH in the present case is secondary hypoplasia due to diaphragmatic hernia. An important finding in association with PH is its coincidence with other congenital anomalies. Anencephaly, diaphragmatic hernia, cardiac lesions, hepatic anomalies, abnormalities of the thumb, deformities of the thoracic spine, urinary tract abnormalities, renal anomalies and pleural effusions have been reported and these associated anomalies may be due to their close proximity.[10]

The basis for variations in presenting features, morphological and histological findings may be related to severity and cause of hypoplasia as well as to the timing of the etiologic events that led to anomaly.[4] According to Areechon and Reid[1] the size of diaphragmatic hernia and maturity of fetus affect alveolar development. In the present case, branching of the intrapulmonary conducting system is arrested due to the diaphragmatic hernia and the alveolar passages remained in pseudoglandular phase with increased amount of connective tissue. Because of the diaphragmatic hernia and pressure of the developing extra lobe and intrathoracic pressure changes, branching of intrapulmonary bronchi and alveolar development were arrested between 7 and 16 weeks of intrauterine life. Cause of death in the present case could be due to immaturity of the lung that is not compatible for the survival.

CONCLUSION

In the present case, the histological finding of pseudoglandular phase in lung development before 16 weeks of development suggests that a correction of congenital diaphragmatic hernia may increase the chances for aeration and growth of hypoplastic lung and total normal alveolar development. PH due to congenital diaphragmatic hernia (CDH) can be prevented by prenatal diagnosis by ultrasonography and fetoscopic tracheal occlusion to treat CDH. This type of abnormalities can also be prevented by adopting few preventive measures like preterm delivery before 32 weeks followed by postnatal ventilator resuscitation and surgical intervention to close diaphragmatic defect and inflation of lungs. This procedure, if successful will decrease the chances of PH as size of diaphragmatic defect and maturity of fetus both affect lung development. The association of left-sided unascended kidney with the CDH and PH was not reported in the literature.

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