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A Case Report of Hyaline Membrane Disease with Rare Complications*

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Hyaline membrane disease (HMD) was first described in 1903. Today, this disease is one of the major causes of mortality in the neonatal units.

The incidence of HMD is difficult to determine from the available literature, since the diagnostic criteria vary from institution to institution.[1,2] The incidence of this disease increases with decreasing maturity.[3] There is higher incidence in Cesa rean section delivered babies and infants of diabetic mothers. This disease is rare in fullterm infants.

This paper presents a fullterm baby who had rare complications of HMD, pneumomediastinum, pneumothorax, and pulmonary pseudocyst formation probably resulting from the progression of intrapulmonary interstitial emphysema (IIE).

Case Report

Baby T (HCH: 704704) was a full term male baby delivered by repeated Cesarean section following an uneventful pregnancy with a birth weigt of 3530 g. The one minute Apgar score was 10. The mother was 29 years old, gravida 3, para 0. The gestation period for the first child was 42 weeks and for the second 34 weeks. Both of the infants were delivered by Cesarean section and they both died immediately after the delivery. The cause of the death in both cases is unknown.

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Our case, baby T. was transported to neonatal unit of the Children's Hospital from obstetrics clinic of Hacettepe University because of respiratory distress immediately after birth. During the following hours, he had cyanosis, tachypnea, grunting, sternal and intercostal retractions, nasal flaring. At the 10th hour of the life, his chest radiograph showed mild pneumothorax and homogeneous alveolar opacification of the right lung, marked pneumomediastinum and air bronchogram in the lower lobes of the left lung retrocardiacally (Figure 1).

The patient was given oxygen and alkali therapy was applied for metabolic acidosis. At the 18th hour of the life the second chest radiograph showed that the amount of the air had decreased in the mediastinum (Figure 2). During the next few hours, the patient's general condition was poor, and he died at the 32th hour of his life.

At post-mortem examination, the main gross pathological findings were in the lungs and the mediastinum. In the areolar tissue of the mediastinum there were many air bubbles varying from 0.3 to 0.8 cm in diameter. Pulmonary pseudocysts localized peripherally beneath the visceral pleura in the middle lobe of the right lung. These pseudocysts occurred

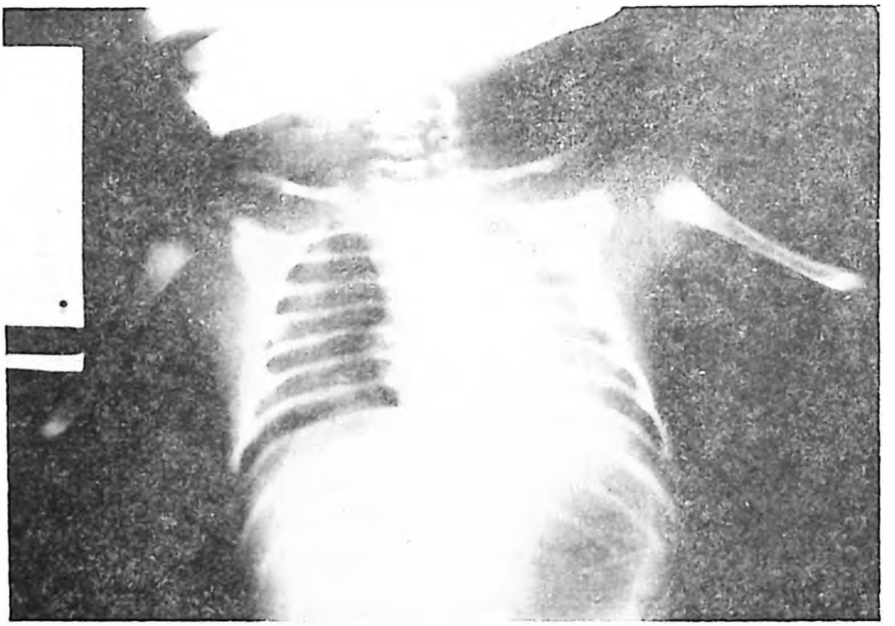

Figure 1

The first chest radiogram: Marked penumomediastinum and homogeneous alveolar opacification of the right lung.

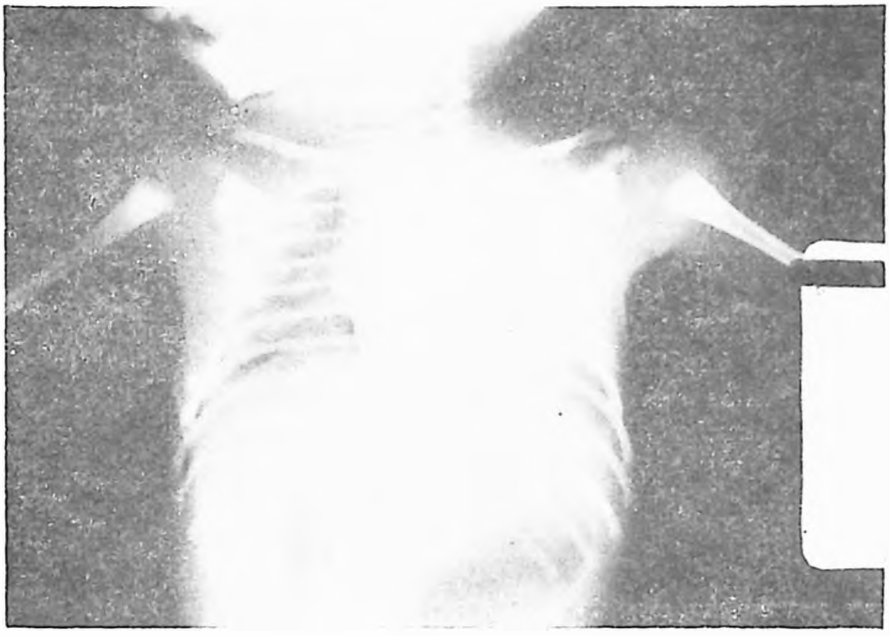

Figure 2
The second chest radiogram: Decreased air in mediastinum is seen.

Figure 3
Pulmonary pseudocysts localized peripherally beneath the visceral pleura in the middle lobe of the right lung.

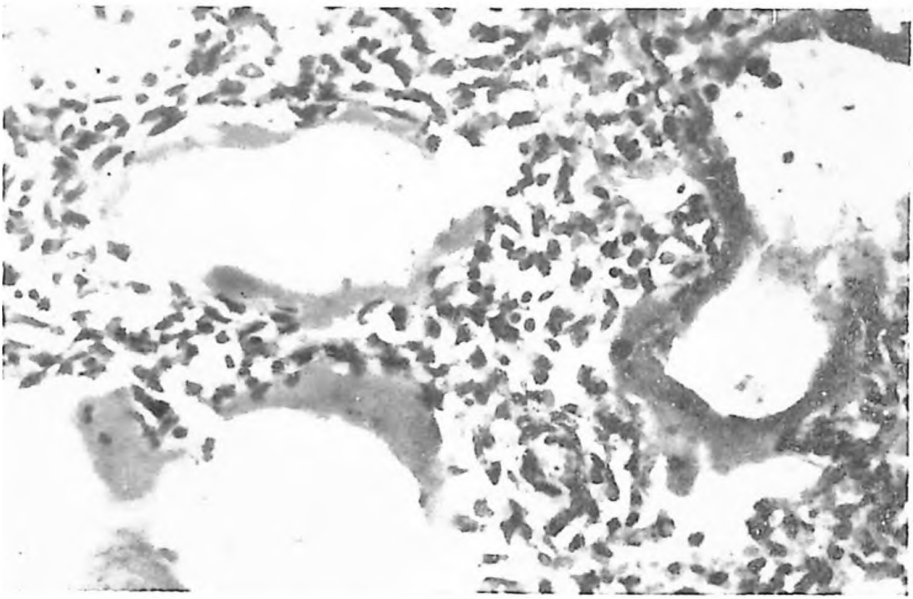

Figure 4
Lung: The hyaline membranes are seen lining air spaces. X&E, X 400

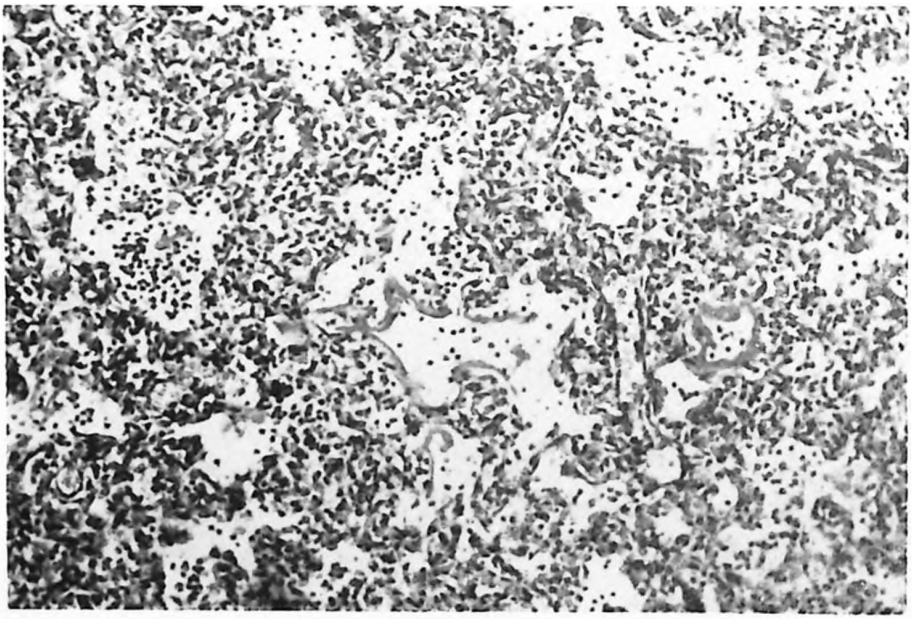

Figure 5
Lung: Hyaline membrane disease and bronchopneumonia H&E, X 100

singly or in groups and their diameters varied from 0.8 cm to 3 cm. The lungs appeared normal in size, but firm, airless, and gray-purple in color Their cut surfaces were homogenous and gray-purple in color. Histologic examination of the lungs revealed atelectasis, bronchopneumonia, and homogeneous hyaline eosinophilic staining material lining many of the dilated terminal bronchioles and alveolar ducts. (Figures 3, 4, 5).

Discussion

The use of assisted ventilation and intensive supportive therapy in severe HMD has lowered the mortality rate somewhat, but has added iatrogenic complications including interstitial emphysema, pneumothorax, pneumomediastinum, pneumopericardium, and pneumoperitoneum.[3-7] There were no iatrogenic complications in our case since he died before the application of continuous positive airway pressure.

Probably the most popular current hypothesis regarding the etiology of HMD is that the disease is basically due to a deficiency of surfactant, a pre-dominantly phospholipid complex which coats the alveolar lining cells, enhancing aeration and lowering the surface tension within the the alveoli.[2-8] The alveolar atelectasis, eosinophilic staining hyaline membranes and dilation of the terminal airways are the important pathological findings in HMD. Sometimes, an airblock phenomenon leads to overdistention and rupture presumably of the alveolar ducts and non atelectatic alveoli in this disease. The interstitial air is located along pulmonary vessels. The roentgenologic appearance of the interstitial air consists of the coarse, short, quite black, non-branching linear radiolucencies in the lung periphery and it should not be confused with the air-bronchogram in HMD. This IIE may be responsible for many of the complications of HMD. If IIE progresses centrally along the vascular sheaths, the air may disect into specific anatomic compartments producing singly or in combination pneumothorax, pneumomediastinum or pneumopericardium. The observation of IIE should alert the clinician to observe the neonate closely for the development of pneumothorax and pneumomediastinum. It is possible that the absence of observable IIE in our patient with pneumomediastinum and pneumothorax is related to the collapse of the lung. Further extention of air superiorly through the mediastinum leads to extrathoracic air collection. If the IIE collects in the interlobular septa or peripherally beneath the visceral pleura pseudocysts may develop as we have observed that it had on the postmortem examination of the present case. Spontaneous resolution of these pseudocysts usually occurs, but occasionally the pseudocysts may enlarge to the point of rupturing into either the pleural space or the mediastinum.[9] The small

cyst-like radiolucencies range in diameter from 1 to 4 mm and although usually round, may appear oval or slightly lobulated. They may resemble bubbles of air in the lung. They should not be confused with the pneumotoceles of Staphylococcal pneumonias. The latter do not appear so early in the neonatal period. Five of 84 cases with HMD in Campbell's series developed pseudocysts.[10] In the newborns, pneumomediastinum and pneumothorax due to IIE are rare complications of HMD.[11, 12] Why IIE develops in some,but not in all neonates with HMD is unknown. It has been suggested that in HMD pneumomediastinum is very often associated with pneumothorax and the latter occurs most often in infants with birth weights over 2000 g. Presumably this is because they can develop greater intrathoracic pressure changes that can result in rupture of overdistended alveolus.[12]

HMD is rare in fullterm babies. In these babies, although HMD has been attributed to the maternal diabetes mellitus or Cesarean section,[13] elective Cesarean section does not effect the incidence of HMD. An increased incidence of HMD has been demonstrated if the operation is performed because of maternal bleeding due to placenta previa, abruptio placenta or toxemia of pregnancy, conditions which might be expected to cause fetal asphyxia.[14] At present, the role of maternal diabetes mellitus in the pathogenesis of HMD is controversial. Many infants of diabetic mothers have been delivered prematurely by early Cesarean section to prevent intrauterine mortality. Some maternal and fetal conditions are associated with delayed maturation of fetal lung.[15] These include diabetes mellitus (classes A, B, C), chronic non-hypertensive glomerulonephritis, and certain cases of hydrops fetalis. In these conditions maturation of the L/S ratio may not occur until after 37 weeks of gestation. These maternal factors should be investigated in this full term case.

Summary

A full-term baby with HMD associated with rare complications is presented and the mechanism of the formation of these complications is discussed.

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