A Case of Congenital Pulmonary Lymphangiectasis

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Introduction

IT has been suggested that congenital pulmonary lymphangiectasis can be expected in one case out of every 170 post-mortem examination on stillbirths and newborn babies (Leader, B.M.J. 1972, 1 : 395). The condition is characterised by intercommunicating, thin-walled, fluid containing cystic spaces of varying sizes, lined by endothelium situated mainly in the sub-pleural, peri-bronchial and interlobular connective tissue.

The following is a case of congenital pulmonary lymphangiectasis associated with diaphragmatic hernia. This, I believe is the first reported case in Malaysia.

Case report

B/O LAY was a full-term Indian male infant weighing 2,400 g. delivered normally after a labour induced by ARM. The Apgar score at 1 minute was 5/10. Intubation and intermittent positive pressure respiration was done.

On examination, his general condition was poor. There was central cyanosis with laboured grunting respiration. Chest expansion was decreased on the left side with poor breath sounds. The apex beat was 150/min., regular and localised on the right chest. Heart sounds were normal and no murmurs were heard. The other systems were all clinically normal. An immediate chest X-ray examination revealed a left sided diaphragmatic hearnia. The diaphragmatic hernia occured through the Foramen of Bochdalek which was repaired surgically.

Post-operatively, the patient was ventilated using the Engstrom ventilator. The next day he was weaned off the respirator and nursed in an incubator with 70% oxygen. However, about 10 hours later, he suddenly became cyanosed because of a right tension pneumothorax. A right chest tube was inserted and connected to an underwater seal. He was then put back on intermittent positive pressure respiration.

Following this he had repeated episodes of cyanosis despite the above treatment. Serial chest X-rays were done which showed improvement of the right pneumothorax, although 24 hours later a left tension pneumothorax was seen. A left chest tube was then inserted and connected to an underwater seal. Repeat chest X-rays did not show any improvement. The patient remained cyanosed and expired 10 hours later.

Necropsy

At post-mortem examination performed 30 hours after death, the body weighed 2,300 g., had a crown-heel length of 47 cm and a crown-rump length of 32 cm. No air was obtained by needling the pleural cavities. The left lung (8.1 g.) was hypoplastic, collapsed and haemorrhagic. Numerous elongated thin-walled cysts containing serous fluid were present in the left upper lobe; they measured 0.2-0.6 cm in maximum dimensions. A few smaller cysts were seen in the left lower lobe (Fgi. 1). No cysts were seen in the right lung (18.2 g.), which was firm and atelectatic with some sub-pleural haemorrhages.

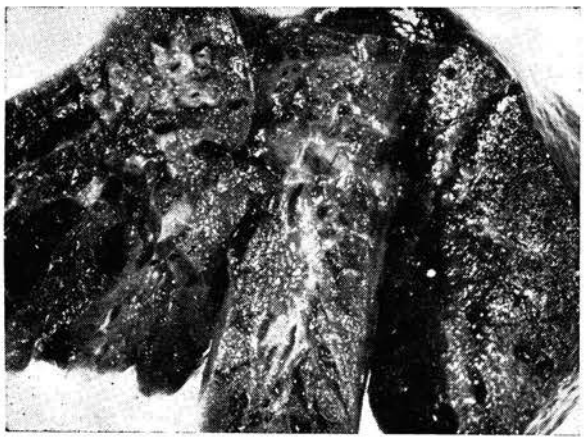

Fig. 1
Lower lobe of left lung showing numerous cystic spaces of varying sizes.

The heart (13.7 g.) had a dilated right atrium and right ventricle. The formen ovale was probed patent and its orifice measured 0.6 cm when stretched. The superior vena cava, inferior vena cava, portal vein, aorta and its major branches were normal in appearance and distribution. The foramen of Bochdalek in the diaphragm was repaired by a dacron patch graft (2.5 cm x 1.5 cm). There were numerous petechial haemorrhages in the thymus and pericardium. A haemorrhage 0.5 cm in diameter in the left adrenal gland and an extensive haemorrhage in the left choroid plexus were present. The liver, spleen and kidneys were unremarkable.

Histology

The cystic spaces in the left lung were structurally dilated lymphatics, lined by flattened endothelium which was desquamated in places (Fig. 2). The cystic spaces were mainly sub-pleural, interlobular, peri-vascular and peri-bronchial in location (Fig. 3). Atelectasis was marked with some associated over-distension of contiguous alveoli. Many of the alveolar ducts were lined by well-formed eosinophilic hyaline membrane. There was an area of necrosis with an infiltrate of neutrophils and macrophages in the left lower lobe. There were extensive haemorrhages in the alveolar spaces.

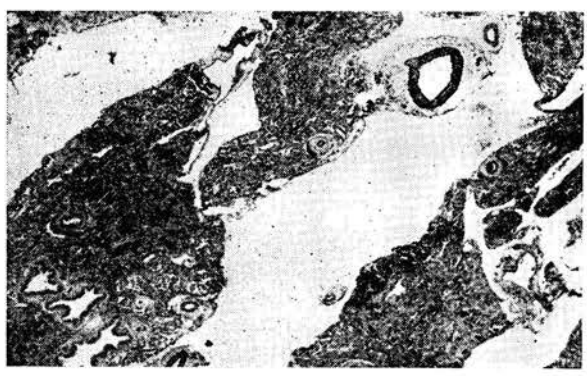

Fig. 2
Lower power magnification of the cystic spaces in the left lung. (Elastic Van Gieson)

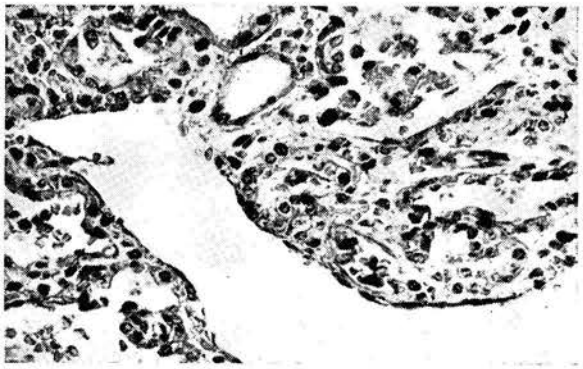

Fig. 3
High power magnification showing the endothelium lined spaces. (Elastic Van Gieson)

Sections of the right lung showed that the lymphatics were also dilated, though not forming macroscopically visible cysts. There was only mild atelectasis. Eosinophilic hyaline membrane was not well-formed. A few of the blood vessels contained fibrin thrombi. There were scattered sub-pleural and alveolar haemorrhages.

Sections of the brain showed infiltrations of moderate numbers of neutrophils in the meninges. There were multiple foci of extramedullary haemopoiesis in the liver with marked splenic congestion.

Discussion

Congenital pulmonary lymphangiectasis was first described by Virchow in 1856 and since that time most of the records are accounts of single cases. However, Laurence (1955) reported 3 cases and in 1959 a further 7 cases. In 1971, N.E. France and R. J. K. Brown reported a series of 11 new cases.

From these and similar reports, these infants develop respiratory distress and cyanosis soon after birth and is usually not relieved by high concentrations of oxygen. As a rule they die within 2 days although a few cases have survived longer. One child (Javett et al. 1963) diagnosed by lung biopsy at 7 weeks was still alive 5 years later (Noonan et al. 1970), though subjected to attacks of wheezing and respiratory distress precipitated by respiratory tract infections.

In the case now recorded, the clinical presentation was that of a diaphragmatic hernia and the condition post-operatively was complicated by bilateral pneumothorax. The left pneumothorax was not relieved even with intercostal drainage suggesting the presence of a broncho-pleural fistula.

The pathogenesis of congenital lymphangiectasis is unknown. Giammalvo (1856) suggested that there was delay in linkage of isolated lymphatic spaces. However, it has been shown by post-mortem injection experiments (Noonan et al. 1970) and re-construction of serial sections (Laurence 1959) that the spaces are part of an intercommunicating network of abnormal vessels.

Lymph vessels grow during the 9th week of foetal life and normally at the 14th week there are large lymphatic vessels lying in the abundant connective tissue dividing the lung parenchyma into distinct lobules. However by the 20th week, a reduction of both the lymphatic vessels and connective tissue occurs. Laurence (1955) postulated that, due to a failure of the normal regression of the lymphatic vessels and connective tissue, growth of these two elements continues.

Shortland-Webb et al. (1966) postulated that intrauterine obstruction to venous flow might result in increased lymphatic drainage with consequent retention of the large lymphatics of early foetal life.

Cardiovascular malformations have been quite frequently reported in association with congenital pulmonary lymphangiectasis. Total anomalous pulmonary venous drainage was most frequently found in association with pulmonary lymphangiectasis (France et al. 1971, Noonan et al. 1970). Varieties of the hypoplastic heart syndrome were also recorded with dilated lymphatics. In both these conditions, obstruction to the pulmonary venous flow in utero leading to increased lymphatic drainage is a possibility, thereby causing the retention of the large lymphatics of the early foetus (France et al. 1971, Noonan et al. 1970).

J. R. Esterly and E. O. Oppenheimer (1970) reported that 13 of their 22 cases of asplenia syndrome had dilated lymphatics and lymphatic proliferation in association with other various anomalies of the pulmonary veins and are thus more likely to have an increased lymphatic drainage.

Summary

A case of congenital pulmonary lymphagiectasis associated with diaphragmatic hernia is described. The pathogenesis of this condition is briefly discussed. Its association with cardiovascular malformations especially total anomalous pulmonary venous drainage is noted.

Acknowledgement

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