measurements of plasma glucagon (McQuarrie et al., 1950; Wagner et al., 1969; Gotlin and Silver, 1970). In Vidnes's (1976) case of persistent neonatal hypoglycaemia due to glucagon deficiency, verified by measuring plasma, treatment with zinc-protamine-glucagon was successful: after 2 months the dosage of glucagon had to be gradually increased as in our case, probably because of antibody production.

Neonatal hypoglycaemia due to glucagon deficiency may be extremely dangerous for the neonate. During glucose need it is glucagon, through a mechanism that remains to be elucidated, that converts the liver to a 'ketogenic mode' and thereby maintains a level of hepatic ketone production necessary to meet cerebral fuel needs (Unger and Orci, 1976). The ketones are an important alternate substrate for the neonatal brain. Ketogenesis in our patient was not adequately studied because the fasting period was limited.

Summary

In a newborn baby, suffering from persistent severe hypoglycaemia with convulsions glucagon deficiency was shown. Treatment with zinc-protamine-glucagon injection twice daily resulted in normal blood glucose levels. Motor development is delayed.

We thank Mr J. B. C. van Veen (Novo Industries AS, Amsterdam) for the zinc-protamine-glucagon preparation, and Dr T. J. Benraad (Department of Medical Biology) for insulin and growth hormone measurements.

References


At 43 hours the infant collapsed after endotracheal aspiration. Clinically there was decreased air entry in the left hemithorax, but needle aspiration failed to show the presence of intrathoracic air, although chest x-ray had shown a collapsed left lung with extrapulmonar air. With an inspired pressure of 30 cm H₂O and F₁O₂ of 1.0 he improved over the ensuing hour. Gradually the F₁O₂ and inspired pressure were lowered and he was extubated on day 5. After extubation, tachypnoea (60–80/min) persisted and F₁O₂ of 0.4 was needed. Repeat chest x-rays showed multiple cystic radioluencies localised to the left upper lobe.

At age 11 days oxygen therapy stopped, but tachypnoea persisted and on day 22 supplementary oxygen was re-started. Chest x-ray now showed cystic changes in the left upper lobe with mediastinal shift to the right, and collapse of the contralateral lung. Increasing dyspnoea led to a decision in favour of thoracotomy. At operation on day 24 the left upper lobe was grossly distended and consisted entirely of many small cysts; it was excised.

The resected lobe measured 10 × 8 × 6 cm. Mediastinal and external surfaces were studded with subpleural blebs up to 1 cm diameter. The cut surface showed a honeycomb network of cystic spaces containing air (Fig.). The lingula was unaffected. The bronchi down to the fifth division and beyond were normal, with normal amounts of mural cartilage. Microscopically (Fig.) the upper lobe consisted of collapsed, compressed lung tissue displaced by empty cystic spaces with thin fibrous walls, lined by foreign body giant cells containing no neutral fat. No cyst was lined by anything resembling bronchial or bronchiolar epithelium. The cysts occupied mainly the subpleural and interlobular regions. In some fields the walls of the cysts were formed by the peribronchial connective tissue suggesting that they may have been greatly dilated lymphatic spaces. There was no demonstrable communication between bronchi or bronchioles and the cysts. Alveolar tissue immediately adjacent to the cysts showed some fibrosis, but there was no significant inflammation.

Postoperatively, the infant rapidly recovered normal respiratory status. He next presented at 2 months of age with acute bronchiolitis; respiratory syncytial virus antigen was detected by immunofluorescence in nose and throat mucus; he recovered in 5 days. At 6 months he is well with no respiratory symptoms.

**Discussion**

Pulmonary interstitial emphysema is a frequent complication of HMD. It is usually bilateral and resolves spontaneously. In some cases the interstitial pulmonary air dissects along peribronchial and perivascular spaces leading to pneumomediastinum or pneumothorax (Thibeault et al., 1973). Pulmonary interstitial emphysema has been increasingly reported following IPPV (Fletcher et al., 1974). Our case is unusual as the interstitial emphysema was localised to one lobe and progressed to develop tension causing mediastinal shift to the opposite side and compression of the contralateral lung. The lesion was unilateral from the start and followed collapse of the affected lobe. Presumably, ball-valve obstruction led to alveolar rupture, with dissection of air into extra-alveolar spaces, then into the interlobular septa, and possibly into the peribronchial lymphatics. Possibly endotracheal aspiration and/or the insertion of a needle blind into the thoracic cavity were factors in our case.

Other conditions must be considered in the differential diagnosis. Congenital lobar emphysema is usually associated with demonstrable flaccidity of bronchi of which the commonest cause is cartilage...
deficiency. In our case the cartilage content of the bronchi was normal and the collapsed lung separated by large cysts differed completely from the over-distended alveoli of emphysema. Congenital cystic lung was excluded by the complete preservation of bronchi and bronchioles in the collapsed lung tissue, the normal relationship of bronchi to vessels and lung alveoli, and the localisation of the cysts to the interlobular connective tissue regions.

An unusual feature was the presence of giant cells lining the cysts; their cause is unknown, but no significant amounts of neutral fat were demonstrable suggesting the reaction was not stimulated by free fat. Multinucleate cells are sometimes seen lining lymphatic spaces in other conditions and it is possible that the cysts represented greatly dilated lymphatic channels, resulting from rupture of air into the pulmonary lymphatics.

Conservative methods of treatment for localised pulmonary interstitial emphysema have been suggested, such as giving 100% inspired oxygen, or vigorous therapy with tracheobronchial suction along with vibration and percussion (Leonidas et al., 1975). Roberton (1976) used an Argyle thoracentesis catheter to aspirate from the centre of the cystic area. Brooks et al. (1976) reported the successful use of selective bronchial intubation in 4 premature infants. We chose to treat our infant surgically as the tension was progressive and life-threatening, and the condition was confined to one lobe.

Summary

An infant with hyaline membrane disease treated with intermittent positive pressure ventilation developed pulmonary interstitial emphysema localised to one lobe after collapse of the affected lobe. The development of tension and further symptoms necessitated lobectomy, after which the infant became totally asymptomatic.

Microscopy of the resected lobe showed the unusual feature of giant cells lining the air-containing cysts. The presence of these multinucleate cells suggested the cysts may have represented greatly dilated lymphatic channels resulting from rupture of gases into the pulmonary lymphatics.

References


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Infantile cortical hyperostosis with raised immunoglobulins

Infantile cortical hyperostosis (Caffey and Silverman, 1945) is characterised by fever, irritability, swelling of the soft tissues, and cortical hyperostosis of the underlying bones. The disease usually occurs before the fifth month of life. In most cases there is complete recovery, though recurrences and crippling deformities have been reported (Blank, 1975). The cause remains unknown. We report 2 cases, both with raised immunoglobulins.

Case reports

Case 1. A Negro boy aged 3 months was admitted with a 7-day history of fever, refusal to feed, and swelling of both forearms. He was pale and irritable with both forearms grossly swollen. The swelling was hard and tender, but there was no warmth or redness. Movement of the arms was limited. The face was broad, with hard and tender swelling confined to the lower jaw.

Radiographic examination confirmed infantile cortical hyperostosis. There was marked diaphyseal hyperostosis of the radius and ulna of both forearms (Fig. 1). Cortical hyperostosis was also present in the mandible and all the long bones.

Investigations showed Hb 8 g/dl; packed cell volume 26%; white blood count 25 × 10^9/l; platelets 1026 × 10^9/l. Erythrocyte sedimentation...
Pulmonary interstitial emphysema requiring lobectomy. Complications of assisted ventilation.

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