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 × 10⁹/l; platelets 1026 × 10⁹/l. Erythrocyte sedimentation
rate 48 mm in the first hour; blood urea 32 mg/100 ml (5·3 mmol/l); alkaline phosphatase 42·5 KA units. Serum proteins, electrolytes, calcium, phosphate were all normal and test for syphilis non-reactive. IgA was 2·05 g/l, IgG 15·0 g/l, IgM 4·25 g/l.

In view of the high platelet count, steroids were not used (Pickering and Cuddigan, 1969). Complete clinical recovery occurred by 9 months and by 15 months there was no radiological evidence of hyperostosis. Radiographic examination of the parents was unremarkable. There was no family history of the disease.

Case 2. An Indian boy aged 2 months was admitted with a 6-day history of fever and swelling of the face. He was pale and irritable. Lower jaw bones felt thick and were tender to palpation. There was a hard and tender swelling at the junction of the outer and middle thirds of the left clavicle.

X-rays showed no abnormalities on admission but a week later hyperostosis of the mandible and left clavicle were obvious (Fig. 2).

Investigations showed Hb 8 g/dl; packed cell volume 29%; white blood count 20·4 × 10⁹/l; platelets 850 × 10⁹/l. ESR 40 mm in the 1st hour (Westergren); serum calcium 8·8 mg/100 ml (2·2 mmol/l); serum phosphate 2·8 mg/100 ml (0·9 mmol/l); alkaline phosphatase 22 KA units; blood urea 20 mg/100 ml (3·32 mmol/l); test for syphilis negative; serum proteins, serum electrolytes normal. IgA was 1·02 g/l, IgG 9·25 g/l, IgM 2·7 g/l.

Discussion

These two cases had typical clinical, haematological, and radiological features of Caffey's disease (Caffey, 1957). There was no family history of the disease and
extensive radiographic survey in both families failed to provide evidence of the disease.

Temperley et al. reported a case with raised immunoglobulins in 1972. There have since been few reports. Both our cases showed raised immunoglobulins. Particularly remarkable were the IgA and IgM levels. We believe that a virus infection during intrauterine or early neonatal life may be the cause of infantile cortical hyperostosis.

Summary

Two cases of infantile cortical hyperostosis are reported. Both had raised immunoglobulins. Particularly remarkable were the IgA and IgM levels, a finding infrequently reported.

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References


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Case report

A male infant was born at 40 weeks’ gestation weighing 2040 g. His mother was aged 39, para 2 + 0, and had 2 healthy children born at term. During this pregnancy she had severe pre-eclamptic toxaemia and it was thought that the fetus was small-for-dates for much of the pregnancy. There was no history of rubella contact. After spontaneous rupture of membranes, fetal distress necessitated lower segment caesarean section and the infant required resuscitation with intermittent positive pressure ventilation from birth. Grunting was noticed at 15 minutes and he was transferred to the special care baby unit. On admission he was cyanosed in 50% oxygen with grunting, tachypnoea, nasal flaring, and intercostal recession but had a lusty cry.

Blood pressure was 60/25 mmHg, heart sounds and peripheral pulses were normal. Chest x-ray and electrocardiogram were normal at this time and initial arterial blood gas measurements were F1O2 50%, Pao2 53 torr, Paco2 51 torr, and pH 7.15. Despite increases in ambient oxygen, constant positive airways pressure, and then intermittent positive pressure ventilation, the blood gas tensions remained virtually unchanged. At age 18 hours the baby was remarkably active and the lungs were easily inflated with low pressures; on auscultation, there was good air entry. 24 hours after birth a marked bradycardia occurred which was persistently unresponsive to inflation of the lungs with 100% oxygen and the Pao2 was 10 torr with Paco2 73 torr. The baby died at age 26 hours.

Necropsy findings. Abnormalities were restricted to the respiratory and cardiovascular systems. The upper airways were normal. The lungs weighed 16 and 12 g, right and left respectively (expected combined weight 47 g) and were small but well aerated. The heart weighed 22 g (expected weight 16 g). There was marked right ventricular dilatation with hypertrophy of the wall. The foramen ovale was valvular. The ductus arteriosus was widely patent, measuring 0·6 cm diameter. Valves were healthy and there were no abnormal communications.

Histological examination of the lungs showed areas of collapse associated with mucus plugging of small bronchi and bronchioles. The large branches of the pulmonary artery showed generalised endothelial proliferation and localised, thicker endothelial cushions containing fine elastic fibres. In many medium-sized pulmonary arteries, localised nodular lesions protruded into the lumen reducing it to a crescentic slit. They arose from the vessel wall and were covered by endothelium. Immediately

Peripheral pulmonary artery stenosis

We report an infant with persistent cyanosis from birth, who was found to have anomalies of the small pulmonary arteries.