Infantile Lobar Emphysema in Association with Congenital Heart Disease

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Three cases of congenital heart disease (CHD) are presented; all infants developed the unusual complication of lobar emphysema (LE). In all cases delay in reaching the correct diagnosis resulted in two of the infants undergoing a second operation to resect the emphysematous lobe. The chest radiograph in all cases was dominated by the heart disease and the radiological signs of LE were initially missed. The radionuclide ventilation/perfusion lung scan was used in two of these infants to provide conclusive proof and to emphasise the clinical significance of the condition. Lobar emphysema in association with CHD should be recognised early and lobectomy performed at the same time as surgical correction of the heart defect.

Infantile lobar emphysema is a relatively uncommon, but well recognised cause of infantile respiratory distress

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Fig. 1 - Chest radiograph showing a large heart with pulmonary plethora. In the right mid zone there is an increased transradiancy. The right hemidiaphragm is flattened medially. The compressed right lower lobe is seen medially in the right lower hemithorax.

CASE REPORTS

Case 1. A full term female infant delivered by forceps delivery was healthy at birth, and for 5 weeks until feeding difficulties developed. At 4 months of age she was admitted to hospital with increasing feeding difficulties. A chest radiograph was interpreted as showing atelectasis of the right upper and lower lobes. The sweat test, immunoglobulins and a barium swallow were all normal. She was treated with antibiotics. At 8 months of age she was noted to have a systolic murmur with a loud
interpreted the chest radiographs as showing middle lobe emphysema (Fig. 1) which was confirmed by \(^{99}\text{Tc}\)-MAA macroaggregate perfusion and \(^{133}\text{Xe}\) ventilation lung scan (Figs 2 and 3). Cardiac catheterisation revealed a 3:1 left to right shunt at ventricular level through an infundibular ventricular septal defect (VSD), for which she underwent elective surgical closure. The lobar emphysema, was considered functionally unimportant. Postoperatively the child required continuing assisted ventilation and could not be weaned off the ventilator. Two weeks following the initial operation a thoracotomy and right middle lobe lobectomy were performed when marked chondromalacia of the middle lobe bronchus was noted. The postoperative course was uneventful, with early spontaneous unassisted respiration. A follow-up cardiac catheterisation a year following discharge from hospital demonstrated normal right heart haemodynamics and proximal pulmonary arteries of normal calibre which did not impinge on the bronchial tree. The follow-up chest radiograph showed re-expansion of the compressed right upper and lower lobes (Fig. 4).

**Case 2.** A 37 week gestation female infant born by spontaneous vaginal delivery, with a birth weight of only 1.4 kg had an Apgar of 2 at one minute and 5 at 5 min. She developed tachypnoea, was intubated and placed on intermittent positive pressure ventilation. She was a very small infant with a low hair line and residual skin folds suggesting Turner's syndrome. There was dextrocardia and auscultation suggested a patent ductus arteriosus. She had poor peripheral pulses except in the left arm. An ECG showed right atrial hypertrophy; echocardiography revealed situs solitus, atrio ventricular and ventriculo atrial concordance, a secundum atrial septal defect, no ventriculo-septal defect, a left superior vena cava draining into the coronary sinus, enlargement of the right atria, right ventricle and pulmonary artery with a moderate patent ductus arteriosus and coarctation of the left sided aorta. These findings were confirmed on cardiac catheterisation. The coarctation was repaired surgically when the child was 11 days old. The postoperative course was stormy. The child developed massive consolidation of the left lung with a pleural effusion as well as a transradiant area in the right lower zone (Fig. 5). Despite drainage of the effusion the child continued to require artificial ventilation. An emergency \(^{99}\text{Tc}\)-MAA perfusion scan demonstrated reduced perfusion in the right middle lobe, and normal perfusion...
in the right upper and lower lobes (Fig. 6). No krypton was available for a ventilation scan. The child continued to deteriorate; a thoracotomy was performed and a right middle lobectomy was performed for middle lobe emphysema. The postoperative course was again stormy with the child requiring artificial ventilation most of the time. Later, when continuous ventilation was required, a decision was made to close the atrial septal defect surgically. Postoperatively the child rapidly deteriorated and died; the congenital heart condition was felt to be the major contributory factor.

**Case 3.** A female infant born at 34 weeks gestation, birth weight 2.3 kg showed the stigmata of Down’s syndrome. A laparotomy on the day following delivery confirmed a periampullary duodenal atresia. A duodenostomy was performed and a number of Ladd’s bands were divided. Postoperatively the infant required intermittent positive pressure ventilation for apnoea and hypoxia coupled with cardiac failure. A systolic murmur was noted at this stage and the diagnosis of a ventricular septal defect and patent ductus arteriosus (PDA) was made. The chest radiograph was interpreted as showing generalised pulmonary plethora (Fig. 7). Progressive transradiancy developed in the right mid zone suggesting middle lobe emphysema (Fig. 8). Thoracotomy was undertaken one week following the first operation. The pulmonary artery was banded, the PDA was ligated and a right middle lobectomy was carried out. The pulmonary artery (PA) pressure was noted to equal systemic pressure. The postoperative course was uncomplicated with early weaning off the ventilator. At 17 months of age cardiac catheterisation showed persistent proximal pulmonary hypertension (main PA mean 33 mmHg, aorta mean 62 mmHg, distal PA mean 9 mmHg). A VSD demonstrated on cineangiography was closed at 18 months of age.

**DISCUSSION**

Since the first successful lobectomy for lobar emphysema in 1945 (Gross and Lewis, 1945) a large number of cases have been reported. No aetiologial factor can be
Fig. 6 - The $^{99}$Tcm MAA perfusion lung scan in the right posterior oblique projection shows a well perfused right lower lobe and a very poorly perfused right middle lobe. The left lung is normal.

Fig. 7 - The chest radiograph one day following abdominal surgery shows loss of the cardiac outline and overinflated lungs. The minor fissure is clearly seen with the right middle lobe being more transradiant than the upper lobe.

Fig. 8 - Three days later the compressed right upper lobe is more evident as the transradiant right middle lobe is now larger. The right lower lobe vessels are clearly seen through the overinflated middle lobe. The plethora persists in the left lung.

identified in the majority of patients (Cottom and Myers, 1957). Only relatively recently has the association between lobar emphysema and congenital heart disease been fully appreciated (Jones et al., 1965). In a collective series of 166 cases of infantile lobar emphysema, 14% were associated with congenital heart disease (Murray, 1967). The commonest abnormality was a patent ductus arteriosus or a ventriculo-septal defect (Stranger et al., 1979). The common finding in all patients with congenital heart disease was a left to right shunt and pulmonary hypertension. Lobar emphysema is rarely seen with other congenital cardiac abnormalities.

Pulmonary arteries, particularly when dilated proximally within the pulmonary hila may readily compress a lobar bronchus where the vessels cross the bronchus (Stranger et al., 1979). Anatomical factors explain the increased frequency of infantile lobar emphysema of the left upper (40%) and the right middle lobes (34%) in congenital heart disease with pulmonary hypertension (Hendren and McKee, 1966). However, this distribution is found in the absence of pulmonary hypertension, albeit in slightly differing proportions (Stranger et al., 1979).

The patency of airways has been shown to be a balance between factors tending to collapse the airway (the intrathoracic pressure), and the factors opposing it (the rigidity of the tracheal and bronchial walls, well as the intraluminal pressure) (Campbell, 1967). If the wall rigidity is reduced by cartilage thinning due to external pressure from a dilated pulmonary artery then bronchial collapse is favoured. With the developing emphysema, the intraluminal pressure is also reduced, exacerbating the situation.

The radio-isotope ventilation-perfusion lung scan is helpful in establishing the diagnosis of infantile lobar emphysema when it is in doubt. An important finding is a reduction in ventilation and perfusion in the emphysematous lobe, while the adjacent apparently collapsed lobes on the chest radiographs appear normally perfused and ventilated. If $^{31}$Kr gas is available then both ventilation and perfusion scans should be carried out. In the first case when the child was about to undergo cardiac surgery, the senior medical staff noted the 'hypertranslucent' area within the lung, but its clinical significance was not appreciated. Only when the child could not be weaned off the ventilator and the emphysematous lobe continued to enlarge was the importance of the abnormality accepted. In the other two cases, there was an initial reluctance of numerous senior medical staff to acknowledge the importance of the radiological observation. In the second case a
perfusion scan was required to prove its significance. In the third case the child was under intensive care in a hospital without nuclear medicine facilities; following discussion and in the light of the previous two cases the surgeon undertook a lobectomy at the time of cardiac surgery.

Infants with lobar emphysema and congenital heart disease usually present with the CHD as the major clinical problem, and thus the lobar emphysema may be easily overlooked (Leape et al., 1970). There has been some debate as to the appropriate management of these infants, in terms of whether the cardiac defect should be corrected first, in the hope that the pulmonary abnormality may regress, or whether lobectomy should be performed in the first instance, with subsequent correction of the cardiac defect, or indeed whether both should be treated simultaneously (Rudolph, 1969). There is evidence that artificial ventilation at the time of, and following cardiac surgery, may make the lobar emphysema worse. Two of the cases described in this report demonstrate the development of lobar emphysema and respiratory distress following correction of the congenital cardiac lesion. In both cases lobectomy later resulted in prompt resolution of the respiratory distress. Failure to recognise lobar emphysema in the clinical context of CHD may result in prolonged respiratory distress and ventilation dependence in the postoperative period.

CONCLUSION

Infantile lobar emphysema is a relatively uncommon condition, but it is a well-recognised cause of infantile respiratory distress. The close association it has with CHD makes its early diagnosis essential. Ventilation and perfusion scintigraphy have an important role when the diagnosis is in doubt. The respiratory distress may compromise recovery from corrective cardiac surgery. The emphysematous lobe requires resection.

REFERENCES