CONGENITAL LOBAR EMPHYSEMA

BY

R. H. WHITE-JONES and L. J. TEMPLE

From Whiston County Hospital, Lancs., and Broadgreen Hospital, Liverpool

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Regional obstructive emphysema is not uncommon and is due in most cases to obstruction of a bronchus by secretions, tumour, foreign body or glands. Cohen (1943) described such a condition in association with a bronchial carcinoma and Spivek (1936) mentioned a similar picture in connexion with tuberculous glands. In such cases, however, the obstruction is usually progressive and leads finally to collapse of the affected lobe.

Maxwell (1940) drew attention to the sign, which had been described by Chevalier Jackson, of an expiratory wheeze heard over an emphysematous lobe indicating an incomplete bronchial obstruction, and mentioned a case of a child aged 2½ years who had progressive emphysema which subsided before bronchoscopy could be undertaken.

Little has appeared in the English literature on the subject of lobar emphysema in infancy, the early onset of which suggests a congenital basis. The recent publication of several reports in the American literature of successful lobectomy for obstructive emphysema in infants has made the recognition and treatment of the condition of great interest and importance and suggests that it is more frequent than has hitherto been realized.

The symptoms of the condition are fairly characteristic and consist of increasing attacks of dyspnoea and cyanosis. A history of such attacks calls for a radiograph of the chest which should be repeated a few days later if there is nothing apparent on the first film. The increasing emphysema of the affected lobe will gradually become more obvious both clinically and radiologically.

The purpose of this article is to describe a case of congenital lobar emphysema in a newborn baby, which was also associated with a diaphragmatic abnormality, and to review the papers that have so far been published.

Case Report

J.N., a first child, was born at Whiston County Hospital on December 12, 1951. His birth weight was 7 lb. 6 oz. The delivery was normal and the condition of the baby was satisfactory. No artificial respiration

or other resuscitative measures had to be used. Two days after delivery he had a cyanotic attack and examination at that time showed diminished air entry over the right lung suggesting atelectasis. He improved following the administration of oxygen. Six days after delivery, cyanotic attacks recurred and dyspnoea and cyanosis became more constant. Radiographs of the chest showed what appeared to be collapse of the right upper lobe with slight mediastinal displacement to the right. During the next few days the symptoms became more pronounced and gradually the percussion note over the left upper lobe became more and more hyper-resonant and the air entry into the right lung diminished. Further x-ray examination showed an obvious difference from the first film, for now there was a marked herniation of the left upper lobe across the midline leading to compression of the right upper lobe (Fig. 1). It will be noted that at this time there was no evidence of the diaphragmatic hernia which appeared subsequently.

A provisional diagnosis of a congenital cyst of the

FIG. 1.—Radiograph showing marked emphysema of left upper lobe with herniation across the midline.

lung was made, though some doubt was felt about the accuracy of this as lung markings appeared to be visible and the shadow did not have the circular appearance of a cyst. As the symptoms were by now becoming severe, surgical intervention was clearly indicated and the baby was transferred to the Thoracic Unit at Broadgreen Hospital, where on January 8, 1952, when the infant was 3 weeks old, bronchoscopy was undertaken.

Operative Findings. The infant was anaesthetized with open ether and bronchoscopy carried out. The smallest size bronchoscope was used and the trachea, carina, main bronchi and the opening of the upper lobe bronchi were clearly seen and were normal. There was no flattening and no valvular structure was seen. It was not possible to pass the bronchoscope any further than the opening of the upper lobe bronchi.

It was decided, therefore, to proceed immediately to thoracotomy. Anaesthesia presented something of a problem, as previous experience with congenital cystic disease had shown the dangers of the usual methods employed when the lung is inflated by the anaesthetist's bag. In these cases the cyst continues to expand with each compression of the bag with, as a result, increasing difficulty in oxygenation until the chest is opened and the affected bronchus clamped. In this instance, therefore, a non-cuffed endotracheal tube was used and anaesthesia maintained with nitrous oxide and oxygen only. The left sixth rib was removed and the pleura opened. The upper lobe was voluminous and had the appearance of a pink soufflé. There were no adhesions, although there was a small area of congenital fusion to the lower lobe. The lower lobe itself was foetal in appearance. The change in the upper lobe appeared to affect the whole of the lobe and no normal lung tissue was noted in it. The upper lobe bronchus was clamped and upper lobectomy was carried out by the usual dissection technique. Some air leaked from the raw surface where the two lobes had been fused but in spite of this the lower lobe now expanded to a much greater volume than it had formerly occupied. The chest was closed with drainage.

Post-operatively the infant's condition was far more satisfactory than before operation and it was no longer necessary to administer oxygen. Some air continued to leak from the left lower lobe, however, and after the withdrawal of the tube on the second post-operative day a needle was inserted from which the air still escaped until the sixth post-operative day. At this time the lower lobe was filling more than half the hemithorax. There was no further leakage and the child remained a good colour, feeding well, and in a completely satisfactory condition. Thereafter, successive films showed further expansion of the left lower lobe. On examination of the specimen in the theatre no abnormality whatsoever could be seen in the bronchus which had been divided flush with the left main bronchus leaving no stump.

Pathological Report. An aerated upper lobe showing some emphysematous blebs under the pleura: on section, the lobe contains several emphysematous cysts, the largest

of them being about 1 cm. across and there is also moderate diffuse emphysema. Areas of intensified emphysema are present which appear to communicate

FIG. 2.—Section of the left upper lobe showing marked emphysematous changes.

FIG. 3.—Antero-posterior radiograph four months after operation showing the translucent areas at the right base suggesting herniation of the bowel through the diaphragm.

with the hilar connective tissue. No bronchial abnormality was seen but dissection was extremely difficult.

Microscopically there are no inflammatory changes (Fig. 2).

Post-operative Progress. A routine post-operative radiograph showed that the mediastinum had returned to normal, and a further film four months after operation showed a satisfactory state of affairs but now several translucent areas at the right base were visible suggesting herniation of the bowel through the diaphragm (Fig. 3). This was especially obvious in the lateral view (Fig. 4). The baby was thriving, however, and a barium meal was not undertaken until nearly a year after the operation. This confirmed that a fairly large part of the ileum and the mid-transverse colon was herniated through the diaphragm and was lying in the anterior mediastinum (Fig. 5). The baby continues to thrive and now at the age of 2 years has no symptoms referable to the lungs or the alimentary tract. In particular he has no undue liability to respiratory infections. In view of this, surgery for repair of the diaphragmatic defect is being postponed until the child is bigger.

Discussion and Review of the Literature

Isolated cases of emphysema involving one lobe in infants or young children have been reported in

FIG. 4.—Lateral view of Fig. 3 showing the well marked herniation of the bowel.

FIG. 5.—Barium meal, 11 months after birth, showing a large part of the ileum and the mid-transverse colon herniated through the diaphragm.

the American literature. Caffey (1940) reviewed this condition in association with infection and described seven cases, some of which were relieved by bronchoscopic aspiration of mucus. Overstreet (1939) drew attention to the fact that lobar emphysema could occur due to a congenital defect and he described a case of an infant aged 1 month with obstructive emphysema of the left upper lobe who was found at necropsy to have deficient cartilage rings in the affected bronchus. Shaw (1952) described a case of localized hypertrophic emphysema involving the right middle lobe and another in which the left upper lobe had been affected. He noted that the bronchi to the affected lobes were flaccid. In his discussion he raised the question whether the condition could be due to vigorous artificial respiration, especially high pressure oxygen. This point is interesting in view of the modern use of endotracheal insufflation of oxygen in resuscitating cases of asphyxia neonatorum. Interstitial emphysema and pneumothorax have been known to follow trauma at birth but such emphysema is usually transient and in any event is unlikely to single out one lobe. Shaw

postulated a chondromalacia of the bronchial tree and noted that in his cases the cartilaginous rings of the bronchi were indeed softer than normal. He assumed therefore that air could enter the lung on inspiration, but during expiration the bronchial walls collapsed due to the weakness of the rings and so retained the air in the distal portion of the lobe affected. A curious feature of this theory appears to be that only one lobe is usually affected by the emphysema. In all the cases reported hitherto lobectomy has lead to a cure without, so far, any recurrence in the other lobes. In a recent publication Sloan (1953), however, quotes four patients who had residual symptoms even after lobectomy and in these the theory of flaccidity of the bronchial tree is reasonable. In two of his cases the left upper lobe was affected and in the other two, the right middle lobe. Lobectomy was successful in each case. The youngest patient was aged 6 days and the oldest 8 months. Each of the lobes showed marked generalized emphysema but no infection and no specific bronchial obstruction was present. In two of the infants there was radiographic evidence of emphysema in other portions of the lung following operation.

Other cases in association with defects of the anterior mediastinum have been reported. Ochsner, De Bakey and Murray (1939) reported a complete absence of the anterior mediastinum visualized at operation for a diaphragmatic hernia in an 18-month-old child but there was no abnormality of the lung. Gross and Lewis (1945) reported an absent anterior mediastinum in a 4-year-old child who was operated on for obstructive emphysema of the right upper lobe caused by absence of the cartilaginous rings in the affected bronchus. The lobe was removed and the anterior mediastinum was reconstructed by suturing the pericardium to the periosteum of the back of the sternum. That a defect in the anterior mediastinum could directly cause lobar emphysema was shown by Lewis and Potts (1951) who reported the case of a 4-month-old boy who had been well until 9 weeks of age when typical symptoms developed. At operation the right middle lobe had herniated through a defect in the anterior mediastinum leading to a kinking of the bronchus and obstructive emphysema of the affected lobe. No definite abnormality of the right middle lobe bronchus could be found.

Williams (1952) reported a case of a 7-month-old infant presenting as asthma. The child was treated as such but finally at the age of 17 months was operated on with a diagnosis of congenital cyst of the lung. At operation a large emphysematous right middle lobe was found but no bronchial abnormality was noted.

Leahy and Butsch (1949) also reported a case involving the left upper lobe in which lobectomy was successful. They also drew attention to the fact that a congenital diaphragmatic hernia itself may be a cause of respiratory distress due to increased pressure on the lungs and mediastinum and that this may necessitate surgical intervention in the neonatal period.

Apart from these isolated cases, no author had produced a series and it seemed therefore that the condition must be rare. However Robertson and James (1951) quoted five cases which had occurred in their practice over a period of two years and suggested that, in fact, the condition might be more common than had hitherto been realized. They found a different set of causes for the bronchial obstruction. In one of their cases a valve-like fold of mucous membrane was demonstrated which apparently allowed air to enter the lobe but, by acting as a valve, impeded the exit. In another case no mucosal fold was demonstrated but the authors suggested that it lay in the stump of the bronchus left behind at operation. In a third case a different reason was postulated, for this time a large vein was found hooked round the right upper lobe bronchus leading to constriction. In a further case a patent ductus arteriosus attached low down on the pulmonary artery was thought to be responsible for the pressure on the bronchus.

Fischer, Potts and Holinger (1952) reported a further series of five cases. In two of these the obstruction was due to a patent ductus arteriosus and in two others to mucosal folds associated with hypoplasia of the bronchial cartilages. In the other case no definite cause was found. Lobectomy was performed with success in every case.

In all the cases reported, with the exception of those of Sloan, complete recovery appears to have been the rule, though if the theory of weakness of the cartilages of the bronchial tree be accepted it is puzzling to know why other lobes are not affected either at the same time or subsequently. Sloan's cases are in marked contrast to all the others in that though they did well in the immediate post-operative period with complete relief of the immediate respiratory embarrassment, the infants seemed more prone to respiratory infections and had more marked symptoms with each attack than would have been anticipated in a normal infant. Sloan suggested that the extra mucus in the bronchi on these occasions was sufficient to increase the obstruction already potentially there due to the flaccid rings. He concluded that it appeared that the whole bronchial tree was involved in the abnormality, and it is interesting to note that Shaw also observed

extreme flaccidity of the whole bronchial tree, though his cases, unlike those of Sloan, had no residual disability.

A curious feature of the condition as a whole is that only the upper lobes or right middle lobe are involved. No cases affecting the lower lobes have been reported. One explanation that has been given is that the lower lobes are more adequately compressed during expiration by the collapse of the thoracic cage and the rising of the diaphragm and that this pressure overcomes any potential obstruction in the lower lobe bronchi. The upper lobes tend to empty more passively.

Robertson and James (1951) reviewed the differential diagnosis, which appears to lie between atelectasis, interstitial emphysema, agenesis, congenital cyst, lobar emphysema secondary to the viscid secretions of bronchitis, foreign body and diaphragmatic hernia pressing on the lung. As they pointed out, in atelectasis there is retraction of the mediastinum with elevation rather than depression of the diaphragm and the atelectatic lung will appear more radio-opaque than the lung compressed by the emphysematous lobe. The lung opposite to the atelectatic lobe never has the ballooned and herniated appearance of the emphysematous lobe. Interstitial emphysema with spontaneous pneumothorax usually has a history of violent respiratory efforts either actively in the form of asthma or passively in the form of pressure anaesthesia. In this connexion it is important to remember the danger of too high an oxygen pressure when an endotracheal tube is passed in the resuscitation of infants suffering from asphyxia neonatorum. Salmon, Forbes and Davenport (1947) reported the necropsy findings of six infants dying shortly after birth with pneumothorax and pulmonary emphysema. There was no bronchial abnormality and the condition was thought to be due to excessive pulmonary ventilation secondary to difficult birth and artificial respiration leading to rupture of the lung alveoli.

In interstitial emphysema, surgical emphysema may be palpable in the neck, and, if the infant survives, the condition is short-lived in contrast to cases of lobar emphysema where the symptoms tend to be progressive.

In agenesis lung markings are completely absent and of course the diaphragm will be elevated on the affected side.

Congenital cysts may present a very difficult differential diagnosis, though in these cases there tends to be a definite ring shadow on radiography. Albert and Potts (1953) have recently mentioned six cases—all emergencies—during a two-year period.

They described two of them. The symptoms were very similar to those of lobar emphysema, namely cough, moderately rapid breathing and tendency to cyanosis. The real difference was only apparent at operation. All the patients, the youngest of whom was 5 days old, recovered.

Lobar emphysema secondary to viscid secretions of bronchitis should present no problem, occurring as it does in relation to the infection, but it is well to bear in mind that even in these cases there may be some degree of flaccidity of the bronchial tree as an underlying abnormality.

In the case of foreign bodies, the history and the results of bronchoscopic examination should solve the problem but in any event this accident is unlikely to happen in a young infant unless some thick mucus be aspirated at delivery. It is improbable that such will remain as a plug partially blocking one bronchus—especially an upper lobe bronchus.

A diaphragmatic hernia may cause pulmonary embarrassment by pressure on the lungs but x-ray examination will settle the difference. It is interesting to note that the very large diaphragmatic hernia in our case has never caused any symptoms.

The case which is described here follows the pattern of the others reported though we were unaware of any theories of aetiology at the time of operation and no abnormality of the bronchial tree could be detected but, as the pathologist mentioned, dissection was extremely difficult. The case appears to be distinctive in that there is an associated diaphragmatic defect which has yet to be repaired. No abnormality of the anterior mediastinum was observed at operation, and it is difficult to postulate any direct connexion between the two conditions except on the basis that one congenital defect in the body tends to be associated with another.

Summary

A case of congenital lobar emphysema occurring in a newborn infant is described. A diaphragmatic hernia as an associated defect was also present.

The importance of early recognition of the condition is emphasized as these cases tolerate lobectomy well. The operation is a life-saving measure.

The symptoms are fairly constant and characteristic and it is suggested that when an infant, previously well and with no evidence of infection, has attacks of dyspnoea and cyanosis, congenital lobar emphysema should be considered.

The literature is reviewed and the theories of aetiology mentioned.

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