TENSION EMPHYSEMA IN INFANTS*

BY

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The condition of obstructive emphysema confined to one lobe of a lung has aroused considerable interest because of the lack of uniformity in the pathological conditions found in specimens removed either at operation or necropsy.

Lobar emphysema is usually confined to one of the upper lobes or the right middle lobe. The affected lobe is grossly over-distended, causing mediastinal displacement and if this condition occurs in the neonatal period and is not relieved by lobectomy it may be rapidly fatal. In other cases a history of wheezing and of some difficulty in breathing over a period of months may be obtained. Any respiratory infection will cause an increase in the symptoms and the diagnosis is usually made by *x*-ray examination during a hospital admission necessitated by one of these attacks.

The commonest abnormality found in our cases was a deficiency of cartilage in the bronchial wall allowing the bronchus to collapse and producing an obstruction to the expulsion of air from the affected lobe during expiration. It is the purpose of this report to present a series of nine cases of lobar emphysema, in seven of which the same pathological condition was found in the affected lobe and to suggest that this may be the essential abnormality in the production of the condition.

Case Reports

Case 1. N.C., a boy, was born on June 8, 1953. It was noted soon after the birth of this infant that the apex beat was on the right side. Radiological examination confirmed the diagnosis of mediastinal displacement, and a diagnosis of tension cyst on the left side was made. A needle introduced into the left side of the chest and connected to an underwater drain failed to give relief and the baby died in a few hours. Necropsy revealed emphysema of the left upper lobe, the bronchus to the lobe being unduly soft and compressible. Sections of the bronchus showed that its walls were entirely devoid of cartilage. The left main bronchus and the bronchus to the lower lobe were normal.

Case 2. The demonstration of this abnormal bronchus by the pathologist recalled to his mind a similar case which had occurred in 1939. A search of the records showed that this was the case of an infant, F.A., of 10 months, who began having attacks of cyanosis and dyspnoea at the age of 4 weeks. The child died in one of these attacks soon after admission to the hospital. Necropsy disclosed a grossly emphysematous right middle lobe which had herniated across the midline and occupied about one-third of the left side of the chest. The bronchus to the middle lobe was soft to palpation and on section showed an absence of cartilage in its wall.

Case 3. D.C., a girl, had a history of 'panting' since the age of 2 months, of bronchitis at 5 months and of poor development following this attack. She was thought to have a lung cyst and at the age of 17 months was subjected to lobectomy of the grossly distended right upper lobe. She made an uneventful recovery and a section of the upper lobe bronchus showed mural hypoplasia with almost complete absence of cartilage (Fig. 1).

Case 4. M.B., a girl, of 5 weeks, was admitted to hospital on June 27, 1953, during the height of an acute respiratory tract infection. A diagnosis of tension emphysema was made and an urgent operation to remove the left upper lobe was performed as her condition was deteriorating. She survived the operation but died six hours later. Examination of the specimen removed at operation showed that the bronchi after division were incompletely surrounded by cartilage but not entirely devoid of it. At necropsy the stump of the left upper lobe bronchus was sectioned and found to be deficient in cartilage. The bronchi to the other lobe were normal.

Case 5. T.M., a boy, had a history of cyanosis and difficulty in breathing 12 hours after birth. The symptoms continued and at the age of 4 months radiological examination suggested the presence of tension emphysema. The right upper lobe was removed in July, 1953, and he recovered from the operation. Examination of the specimen showed that the bronchus was deficient in cartilage and in all supporting structures. It was thought that the artery to the lobe was causing pressure on the bronchus and contributing to the obstruction. In other parts of the removed lobe some sections showed inadequate mural development while others showed the structure of normal bronchi.

These five cases have been reported previously (Nelson and Reye, 1954).

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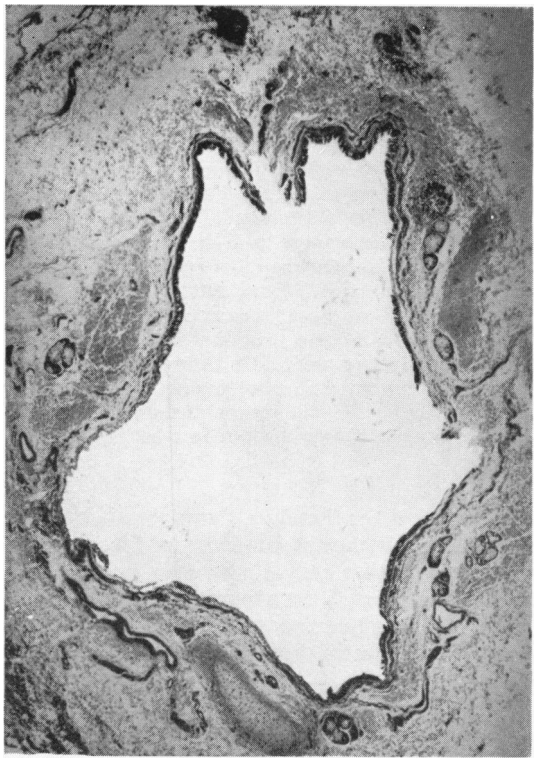

FIG. 1.—Section of affected bronchus in Case 3. There is only one small bar of cartilage seen in the section.

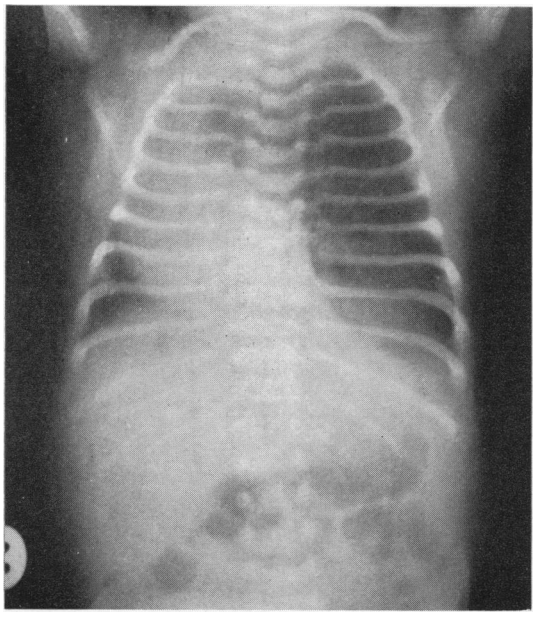

FIG. 2.—Emphysema of the left upper lobe with herniation across the midline in Case 6. Lung markings can be seen in the emphysematous lobe.

Case 6. The mother of J.C., a boy, had noticed a persistent wheeze from birth. At the age of 2 months he developed a respiratory infection and after this breathing became more difficult. He was admitted to hospital on September 6, 1953, at the age of 4 months after severe spasms of coughing causing cyanosis. Emphysema of the left lung with herniation of the lung into the right hemi-thorax was demonstrated by radiological examination (Fig. 2). He made a good recovery from this attack, but a considerable degree of mediastinal displacement persisted for some months, during which he was kept under observation. In May, 1954, the emphysematous left upper lobe was removed. Examination of the specimen did not disclose any cause of the obstruction, the bronchi being normal.

Case 7. D.B., a girl, was admitted to hospital on July 15, 1954, on the day of her birth. She was slow to breathe after delivery and cyanosed, and although her colour improved after removal of mucus by suction, she continued to have cyanotic turns. Her condition was improved by the administration of oxygen and she was kept in an oxygen tent. Examination of the chest showed that the right side moved less than the left, but breathing was very shallow. Radiological examination by fluoroscopy suggested that both lungs were emphysematous and that diaphragmatic movements were deficient. On July 16 radiographs suggested that both lungs were emphysematous, the left more so than the right, and on July 20 the same appearances were seen. Her condition continued to improve with the administration of oxygen and on July 29th further emphysema had occurred with mediastinal shift to the right. As there was no clear evidence that emphysema was confined to one lobe and as her condition continued to improve, it was thought wise to defer lobectomy. During the next two months her nursing care was a constant anxiety. She was tube fed for a considerable time as the effort of feeding exhausted her but gradually she was able to feed from a bottle, at first in the oxygen tent and later without this aid. By October she was in good condition and left thoracotomy was performed and an emphysematous upper lobe removed. The lower lobe was collapsed but inflated well at the end of the operation. The bronchus to the removed lobe was unduly soft and sections through the main bronchus and its primary divisions showed that they were supplied with cartilage but less liberally than is normal and quite large segments contained only small and immature cartilaginous bars.

On the day after operation mediastinal shift to the right was still present and a tension pneumothorax was aspirated. Two days later the mediastinum was central, the lower lobe expanded and she made an uneventful recovery.

Her subsequent history is interesting. In May, 1955, she was admitted to hospital again with cough and difficulty in breathing after a respiratory infection. Radiological examination showed well-marked emphysema of the remaining left lower lobe with mediastinal shift to the right. Since this period she has had five other attacks with wheezing and cough but they are less severe and she recovers rapidly.

Case 8. The birth of M.G., a boy, was normal but by the tenth day he had developed some cyanosis which persisted. He wheezed occasionally but had no alarming symptoms and progressed well. In August, 1954, when 1 month old, radiological examination showed emphysema of the right middle lobe with some mediastinal shift, and although he continued in good health further radiographs taken in January, 1955, showed that there had been a progressive increase in the degree of emphysema, which now produced more mediastinal displacement. On March 17, the right middle lobe, which was grossly emphysematous, was removed and he made a good recovery. Although the bronchus to the lobe was thin-walled the sections showed no abnormality in either the large or small bronchi or bronchioles.

The fact that in six of these eight cases the common pathological condition of diminution or absence of the cartilaginous support of the main bronchus to the lobe was found suggests that this is the essential cause of the condition. But in Cases 6 and 8, in which the emphysematous condition of the lobe was identical with that of the other cases, no abnormality was noted in the bronchi of the removed specimen. In Case 4 it was possible to examine both the lobe removed at operation and also the stump of the bronchus remaining. Although the deficiency was noted in the bronchi of the removed lobe it was even more apparent in the proximal bronchus. In other specimens it has been noted that there is an abrupt change from abnormal to normal bronchial structure in the more distal part of the lung, and it is suggested that section of the bronchus to remove an emphysematous lobe may be made through an area which is distal to the affected segment of bronchus, and that this may account for the failure to demonstrate the condition in the two cases.

All these children had symptoms from the neonatal period and in all cases a grossly emphysematous lobe was demonstrated producing mediastinal displacement. But there was a considerable variation in the severity of the symptoms and to some extent it is possible to correlate this with the pathological findings. In Case 1 the child died rapidly and there was a complete absence of cartilage in the bronchus. In other cases, in which small amounts of cartilage were present, the patients recovered from the acute condition and were cured by lobectomy.

That this abnormality of bronchial structure may be found in the presence of a much more benign condition is shown by the following case.

Case 9. S.I., a boy, was noted to have a persistent cough from the age of 6 months. He was subject to attacks of wheezing and had frequent attacks of bronchitis. He came under the care of Professor Dods at the age of 7 years when a radiological examination of the chest revealed appearances that were interpreted as being due to a cystic condition of the right upper lobe. A bronchogram was carried out and a large area in the upper half of the right lung field failed to fill with 'lipiodol' but crowded and compressed the upper lobe bronchi and to a lesser extent those of the middle lobe. The child was still suffering from cough and was dyspnoeic on slight exertion and unable to keep up with other children. In June, 1954, an emphysematous lobe was removed from the upper part of the right lung. This was an accessory lobe; its blood supply came from the artery to the upper lobe, but it did not show the gross degree of emphysema seen in the other cases. Examination of the specimen showed deficient cartilage in several of the bronchial branches. It was evident that there had been sufficient tension in this lobe to prevent the entry of 'lipiodol' when the bronchogram was performed, but not enough to cause herniation across the midline or mediastinal displacement which was a constant feature in other cases.

Discussion

A perusal of the literature shows that it has been common to mistake the condition of lobar emphysema for a tension cyst as was done in the earlier cases in this series, but a careful study of the radiographs will usually demonstrate fine lung markings through the affected area if one is alive to the possibility.

It has usually been found that the condition is confined to one of the upper lobes or the right middle lobe, but in Case 7 of this series there is definite evidence that the left lower lobe produced the typical picture of emphysema after the upper lobe had been removed, and it is not at all certain that there was not some degree of emphysema on the right side also.

Although the clinical picture is well recognized the method of its production is not so well established. In about half the reported cases a bronchial wall deficient in cartilage has been demonstrated. Various explanations have been put forward to explain the others. These include a flap of bronchial mucosa; a patent ductus overlying and compressing the bronchus; and abnormal flaccidity of the bronchial wall without absence of cartilage.

In the early neonatal period a grave emergency may arise as in Case 1. This raises the question as to whether any treatment is available to tide the patient over this period. In my case, under-water drainage through a needle gave no relief but more recently Korngold and Baker (1954) have treated two infants successfully by aspiration first of the affected lobe and later of the tension pneumothorax that resulted. Both these infants were relieved of the emphysema and had no recurrence in the next six months. It is difficult to understand how aspiration could relieve more than a lobule of the affected lobe unless the needle were introduced into a bronchus of some size, but the method offers some prospect of

tiding these infants over a dangerous period, although condemned by Sloan (1953) and others. Most writers are agreed that the only prospect of cure lies in removal of the affected lobe, and although there is no doubt that many of these infants may recover from acute attacks and aspiration may be a means of assisting this, there seems little place for conservative treatment when the lobe is permanently over-distended and produces mediastinal displacement over a long period.

In submitting the view that deficient cartilage formation is the underlying cause of the condition under discussion, I would set out the following considerations: (1) Deficient cartilage formation has been shown to be a direct cause of tension emphysema. (2) Deficiency may be of varying degrees; sometimes the deficiency is so great that the patient manifests symptoms immediately after birth. If the deficiency is of a lesser order some event such as an infection may produce mucosal changes and retention of secretion which will precipitate a crisis. (3) Sometimes, as we have found, the deficiency of cartilage varies in different parts of a bronchus. Here the possibility that tension emphysema will be caused is still present, but the cartilage deficiency may be hard to demonstrate in specimens removed at operation. (4) The demonstration of absence of supporting cartilage in the bronchus in a high proportion of cases would accord with my conception that this is the essential factor and the other abnormal conditions found are contributing causes.

I am grateful to my colleagues for referring cases, and to Mr. E. S. Stuckey for allowing me to include Case 5 in the series. Dr. Douglas Reye has been responsible for the pathological reports and has maintained a keen interest in the subject.

References

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