Bronchial atresia with corresponding segmental pulmonary emphysema

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This paper presents a 13-year-old boy suffering from congenital bronchial atresia of the anterior and apicoposterior segments of the left upper lobe with corresponding emphysema, and emphysema with bronchial atresia in the anterobasal segment of the left lower lobe. The aetiology of congenital localized emphysema is not always clear. In the group with a demonstrable check-valve mechanism, which permits air to enter but not to leave the lung, there is either internal stenosis or external compression of the bronchus. The condition is called idiopathic if no cause can be identified, but in some of these cases microscopical examination has disclosed alveolar fibrosis which prevents the rigid alveoli from collapsing on expiration. In these cases the check-valve mechanism exists at an alveolar level. In the small group of rare cases of bronchial atresia, air which enters through a collateral ventilation cannot be expired by the same route, and in these cases there is likewise a check-valve mechanism at an alveolar level; this is associated with the occurrence of localized emphysema, characterized by the presence of normal bronchial ramifications which arise from a bronchial sac with a blind proximal ending. The bronchial atresia is believed to occur after the 15th week of intrauterine life, probably as a result of vascular insufficiency. A preoperative diagnosis is possible on the basis of a plain chest radiograph, which shows the localized emphysema as well as an abnormal hilus shadow representing the mucus-filled blind bronchial stump; the diagnosis can be established also by a bronchogram, which shows a filling defect in the affected part of the lung. Surgical treatment is indicated.

Congenital pulmonary emphysema is a condition with which paediatricians and thoracic surgeons are now familiar.

Nelson (1932) was the first to focus attention on the serious nature of this condition in infants, in a report on a fatal case which occurred in a child aged 5.5 months. Royes (1938) was the first to mention localized lobar emphysema in an adult; its cause was partial bronchial obstruction by a mucosal flap, an incidental finding at postmortem examination of an accident victim without pulmonary complaints. Overstreet (1939) focused attention on an anomaly of the lobar cartilage in a case of lobar emphysema. Gross and Lewis (1945) described the first patient with infantile lobar emphysema, who was successfully treated by lobectomy.

Numerous cases of localized emphysema have since been described, usually in infants with acute respiratory disorders. Raynor, Capp, and Sealy (1967) and Murray (1967) report series of 120 and

of 166 cases, respectively. The difference in the number of cases and in the terminology between these two reviews must probably be ascribed to a difference in diagnostic criteria.

In many cases the aetiology is obscure; it must be assumed, however, that all cases involve a check-valve mechanism which permits air to enter but not to leave the lung. According to Stovin (1959), lobar emphysema can be found in a group of cases with internal bronchial stenosis, a group with external bronchial compression, an idiopathic group in which no stenosis is found, and a small group of unusual forms in which he includes bronchial atresia.

We have personally observed a case of segmental bronchial atresia with corresponding segmental emphysema in a boy aged 13 years. This unusual form is not often described in the abundant literature on localized emphysema. Similar cases with bronchial atresia have been described by Belsey (1958), Simon and Reid (1963), Waddell (1963), Simon, and Reid (1965), Hanna, Vattanapat, and Derrick (1969), and Vaghei (1970).

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FIG. 1. *Postero-anterior and lateral views of the chest in January 1969 show a localized pulmonary emphysema in the upper region of the left lung.*

FIG. 2. *Postero-anterior and oblique bronchographic views of the left lung in March 1969 show no filling of the bronchus of the culmen.*

CASE REPORT

V. M. R. aged 13 years was admitted to hospital on 4 February 1969 following a traffic accident in which he sustained concussion of the brain, lesions of the right forearm with paralysis of the radial nerve, and left-sided pneumothorax. After suction drainage of the pneumothorax, localized emphysema was found to exist in the upper region of the left lung.

Localized pulmonary emphysema had been detected in a school survey a few years earlier, and a recent chest radiograph was available (27 January 1969; Fig. 1). Hospital observation had been suggested at the time but had not yet taken place.

Radiographs and tomographs of the left lung confirmed the tentative diagnosis of 'congenital localized emphysema'. The left bronchogram (Fig. 2) showed adequate filling of the lower lobe bronchi and compression of the lingular branches, but no filling of the bronchus of the culmen, which gave an impression of obstruction.

Unfortunately, no bronchoscopic examination was made.

For this congenital anomaly of the left lung, which on the occasion of a traumatic injury had given rise to pneumothorax as a complication, surgical treatment was suggested and carried out on 24 March 1969 through a left thoracotomy. It disclosed localized emphysema of the three upper segments of the left upper lobe (culmen) and in the anterobasal segment of the left lower lobe. These segments were a pale pink colour, felt like foam-rubber and protruded immediately after the thoracotomy (Fig. 3). Dissection of the hilus of the upper lobe and of the lower lobe

FIG. 3. Operative view of the left lung discloses a localized pulmonary emphysema in the three upper segments of the left upper lobe and in the anterobasal segment of the left lower lobe.

revealed a normal arterial and venous blood supply to the affected segments; however, careful dissection failed to reveal a bronchus or even a fibrous connection of the affected segments with the bronchial tree. The emphysematous segments were resected after ligation and severance of the vessels. These segments retained their air volume and floated on water. The lingular cut surface, via which the air supply took place, showed a very large air leakage. In order to save the lingula, this was closed upon itself; an attempt to expand the compressed lingula was successful.

Pathological anatomical examination of the resected emphysematous segments disclosed no bronchi in the hilar cut surface. The pulmonary tissue had a smooth surface, was greyish-red and showed no anthracosis.

Examination of thin sections cut after fixation showed that both resected specimens contained immediately below the hilus, a cystically distended thin-walled bronchus which ramified into moderately distended smaller bronchi, filled with accumulated mucus. The wall of the cystically distended bronchus consisted in part of cartilage and in part of a thin layer of connective tissue which was covered by one or several layers of cylindrical epithelium.

Since the bronchial wall was not demonstrable in the hilus, it may be concluded that the proximal part of the bronchus had a blind ending.

The smaller bronchi were markedly distended and filled with mucus; they had one or several layers of cylindrical epithelium and a thin wall which consisted of collagenous tissue.

The alveoli showed moderately diffuse emphysema without lesions of the interalveolar septa.

There were no inflammatory lesions, and the vessels were normal.

There was only very slight postoperative air leakage, and drainage was therefore discontinued after 48 hours. Persistent atelectasis of the lingula then occurred so that no complete filling of the thorax was achieved. A bronchoscopic follow-up disclosed marked oedema of the left main bronchus for which corticosteroid therapy was started. This gradually led to improved expansion, and the patient was discharged on 30 April 1969.

A radiograph five months after the operation revealed complete expansion of the remainder of the left lung with complete filling of the left hemithorax (Fig. 4).

DISCUSSION

Congenital lobar emphysema is the term most widely used (Robertson and James, 1951). Other terms used are lobar emphysema (Fischer, Potts, and Holinger, 1952); infantile lobar emphysema (Leape and Longino, 1964); obstructive emphysema (Lewis and Potts, 1951); lobar obstructive emphysema (Sloan, 1953); regional obstructive emphysema (Thomson and Forfar, 1958); con-

FIG. 4. Postero-anterior and lateral views of the chest 5 months after surgery show complete expansion of the remainder of the left lung with complete filling of the left hemithorax.

genital obstructive emphysema (DeBord and Sibilsky, 1954); progressive infantile emphysema (Ehrenhaft and Taber, 1953); tension emphysema (Nelson and Reye, 1954); lobar tension emphysema (Binet, Nezelof, and Fredet, 1962); congenital localized emphysema (Campbell, Bauer, and Hewlett, 1961); localized hypertrophic emphysema (Shaw, 1952); localized pulmonary hypertrophic emphysema (Williams, 1952); and congenital hypertrophic emphysema (Hamilton and Gillespie, 1958).

Since none of these exactly describes the situation which prevailed in our case, we prefer the description 'bronchial atresia with corresponding segmental pulmonary emphysema'.

The aetiology can be explained when an internal or an external cause can be identified. In other cases there is no unequivocal aetiological factor.

Internal causes are reported by Raynor *et al.* (1967) to be involved in 38% of cases. They usually consist of anomalies of the cartilage rings—absence, hypoplasia or abnormal softness (bronchomalacia). A survey of the literature shows that these anomalies have been described by Overstreet (1939), Fischer *et al.* (1952), Shaw (1952), Sloan (1953), Bolande, Schneider, and Boggs (1956), Holzel, Bennett, and Vaughan (1956), Mathey, Binet, and Galey (1956), Cottom

and Myers (1957), Nelson, T. Y. (1957), Stovin (1959), and Moyson, Desmul, and Cremer (1962).

Other possible internal causes are the presence of a mucosal flap (Royes, 1938) and hypertrophy of bronchial mucosa resulting in the formation of a fold which serves as a valve during inspiration (Robertson and James, 1951; Fischer *et al.*, 1952). According to Raynor *et al.* (1967), this mucosal fold is present in 13% of cases but is often overlooked in the resected specimen because it often coincides with the site of bronchial section. Mention has been made also of bronchial kinking secondary to herniation through an anterior mediastinal defect (Gross and Lewis, 1945; Lewis and Potts, 1951) and of the presence of a cartilaginous septum in the bronchus (Drift, Waldeck, and Van Rooden, 1956).

External causes are reported by Raynor *et al.* (1967) to be involved in 8% of cases. External compression of a main bronchus or a lobar bronchus may have a vascular cause: patent ductus arteriosus (Fischer *et al.*, 1952—2 out of 6 cases; Bolande *et al.*, 1956; Cottom and Myers, 1957); aneurysm of a pulmonary vein (Robertson and James, 1951); or an abnormal pulmonary artery (Potts, Holinger, and Rosenblum, 1954; Morse and Gladding, 1955; Belsey, 1958; Contro, Miller, White, and Potts, 1958a,b; Sherman, 1959).

Cottom and Myers (1957) reported a congenital heart defect in four out of six cases of lobar emphysema. The two conditions are best treated jointly, if possible, first the pulmonary and then the cardiac anomaly.

The term idiopathic emphysema is used if neither an internal stenosis nor an external compression of the bronchus is found. In the majority of cases (about 55% according to Raynor *et al.*, 1967) pathological examination fails to disclose a bronchial cause of the emphysema.

The condition has been ascribed to rupture of the alveoli due to excessive inflation at postpartum artificial respiration (Robertson and James, 1951; Shaw, 1952; Ehrenhaft and Taber, 1953). The occurrence of a bulla might so be explained, but it is not clear why the emphysema remains confined to one lobe or to a few segments.

Mayer and Rappaport (1953) believed that there must be predisposing factors in the pulmonary parenchyma for emphysema to occur, because normal alveoli do not rupture when exposed to physiological pressures. Bolande *et al.* (1956) established an increase in collagenous tissue in the alveolar wall, giving rise to 'alveolar fibrosis'. During inspiration the fibrous alveoli distend, but they are too rigid to collapse at expiration, and emphysema results. However, it is not certain that this alveolar fibrosis is the cause; it can be a secondary occurrence. Leape and Longino (1964) likewise reported this 'fibrous dysplasia' in several of their patients.

Zatzkin, Cole, and Bronsther (1962) suggested a division into hypertrophic and obstructive forms, and maintained that the designation 'congenital hypertrophic lobar emphysema' should be reserved for cases in which the alveolar anomaly is regarded as the causative factor.

The unusual forms of lobar emphysema include those associated with localized bronchial neuromuscular dysfunction (Williams, 1952; Sloan, 1953; Holzel *et al.*, 1956). A second unusual form is that associated with bronchial atresia, as described by Belsey (1958: 1 case); Simon and Reid (1963: 3 cases); Waddell *et al.* (1965: 2 cases); Hanna *et al.* (1969: 1 case); Vaghei (1970: 2 cases) and the case presented here.

In the cases with bronchial atresia the air supply to the corresponding segments takes place through alveolar communications with the normal lung tissue. Air thus inspired cannot be evacuated by the same route; this is prevented by a check-valve mechanism which can be localized at an alveolar level. Localized pulmonary emphysema results. The concomitant presence of alveolar

fibrosis has been demonstrated only in the case of Hanna *et al.* (1969).

Unlike other patients with lobar emphysema, who usually show pronounced respiratory discomfort and are presented for treatment as neonates or young infants, patients with bronchial atresia are older and usually asymptomatic. The patient described by Belsey (1958) was 6½ years old and had only a one-year history of an unproductive cough; the patients discussed by Simon and Reid (1963) were 19, 25 and 18 years old, and only the last-mentioned patient showed recurrent pulmonary infections; the patients of Waddell *et al.* (1965) were 25 and 16 years old and showed no symptoms; the patient described by Hanna *et al.* (1969) was a 7-year-old boy without symptoms; the patients of Vaghei (1970) were 13 and 36 years old and the condition was an incidental finding in these patients. Our patient was a boy, aged 13, and the condition was treated after a traumatic pneumothorax had occurred.

The sites affected in our patient were both the left upper and the left lower lobes. The left upper lobe was affected in the cases reported by Belsey (1958), Simon and Reid (1963), and Waddell *et al.* (1965). The middle lobe was affected in the case reported by Hanna *et al.* (1969), and the left and right upper lobes were affected in the cases reported by Vaghei (1970).

Fischer *et al.* (1952) reported that localized emphysema is most likely to affect the upper lobe and the middle lobe, and they ascribed this to the stronger influence of the diaphragm and lower ribs on the lower lobes during expiration. This explanation is plausible for the idiopathic group, in which the anomaly should be at an alveolar level but not for the group with obstruction or atresia for bronchial anomalies are most common in the upper lobe bronchi (Holzel *et al.*, 1956) and the vascular anomalies with their invariable localization in the upper mediastinum can exert an influence only on the upper lobe bronchi. According to Boyden (1955), most congenital bronchial anomalies occur in the left upper lobe, and they result from an embryological instability rather than from mechanical causes.

Bucher and Reid (1961) ascribed the occurrence of bronchial atresia to a disorder in the vascularization, not at the time of development of the lobar bronchus but at a later stage—after the 15th week of intra-uterine life, when the bronchial tree has already attained its full ramification. The atresia therefore, develops upon a previously normal air passage, and the distal bronchi consequently show normal development. The corresponding seg-

mental or lobar emphysema is characterized by the presence of normal bronchial branches, arising from a bronchial sac with a blind proximal ending.

According to Waddell *et al.* (1965) and Vaghei (1970), bronchial atresia with corresponding emphysema can be diagnosed on the basis of standard chest radiographs, which reveal a butterfly shadow in the hilus, suggestive of a mucus-filled bronchial sac, and emphysema in the corresponding segments or lobe.

Unless the patients are severely ill and dyspnoeic infants, bronchoscopy and bronchography can supply useful information. In older children, in particular, the possibility of an intra-bronchial foreign body must be taken into consideration; and this possibility must be ruled out before a decision in favour of surgery can be made. In the rare cases of bronchial atresia, the absence of a bronchus or the presence of a vestigial depression can be established.

Bronchography gives useful information also in all cases involving bronchial anomalies, internal stenosis or external compression. Unfortunately, we omitted bronchoscopy in our case and obtained only a bronchogram. This revealed a filling defect of the culmen, which was ascribed to internal bronchial stenosis caused by a cartilaginous or mucosal anomaly, with localized emphysema as a result. The presence of bronchial atresia with pulmonary emphysema was not suspected. In retrospect, we advise preoperative bronchoscopic examination. When the diagnosis is dubious, diagnostic aspiration must be avoided because it may give rise to a pneumothorax which only aggravates the respiratory discomfort.

There can be no doubt about the surgical indication in infants with localized emphysema and pronounced respiratory discomfort due to compression of intact lung parts; but the treatment of an incidentally found emphysema in an older child is open to discussion. We believe that surgery is indicated in these cases also, unless they involve post-infectious pneumatoceles which will absorb spontaneously. In our patient a complication—pneumothorax caused by a blunt chest injury without costal fractures—led to establishment of the diagnosis and operation.

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