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Congenital Lobar Emphysema

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Summary. Congenital lobar emphysema is reported occurring in a 10 day old, low birth weight infant. Following resection, gross examination of the affected lobe revealed collapsed but unobstructed bronchi, suggesting a cartilaginous defect. The specimen was insufflated with formalin and the bronchial tree dissected out. Histological study including comparison with a normal pulmonary lobe, confirms that abnormalities of cartilage may be important in the pathogenesis of congenital lobar emphysema.

Key words: Emphysema – Congenital lobar – Cartilage defect.

Introduction

Congenital lobar emphysema is characterized by overdistention and air-trapping in the inflated lobe, compression atelectasis of the remaining lung, and displacement of the mediastinal contents with extension of the emphysematous lobe across the midline (De Luca et al., 1973). This condition becomes evident during early infancy, presenting with persistent progressive respiratory distress, and is frequently fatal unless surgically treated. It was rarely mentioned in the literature prior to 1945 when the first instance of successful treatment by lobectomy was reported (Gross and Lewis, 1945). Congenital lobar emphysema must be distinguished from regional obstructive emphysema in infants, a condition which is related to acute respiratory infection, and in which air trapping with bullous change sometimes occurs. The disease may result from external compression of a lobar bronchus, or from obstruction of the bronchial lumen by a mucous plug; however, in series of case reports (Murray, 1967; Janovski et al., 1969), the etiologic factor most often suspected was maldevelopment of the bronchial cartilage.

In a review of 166 reported cases, Murray (1967) noted that the structural

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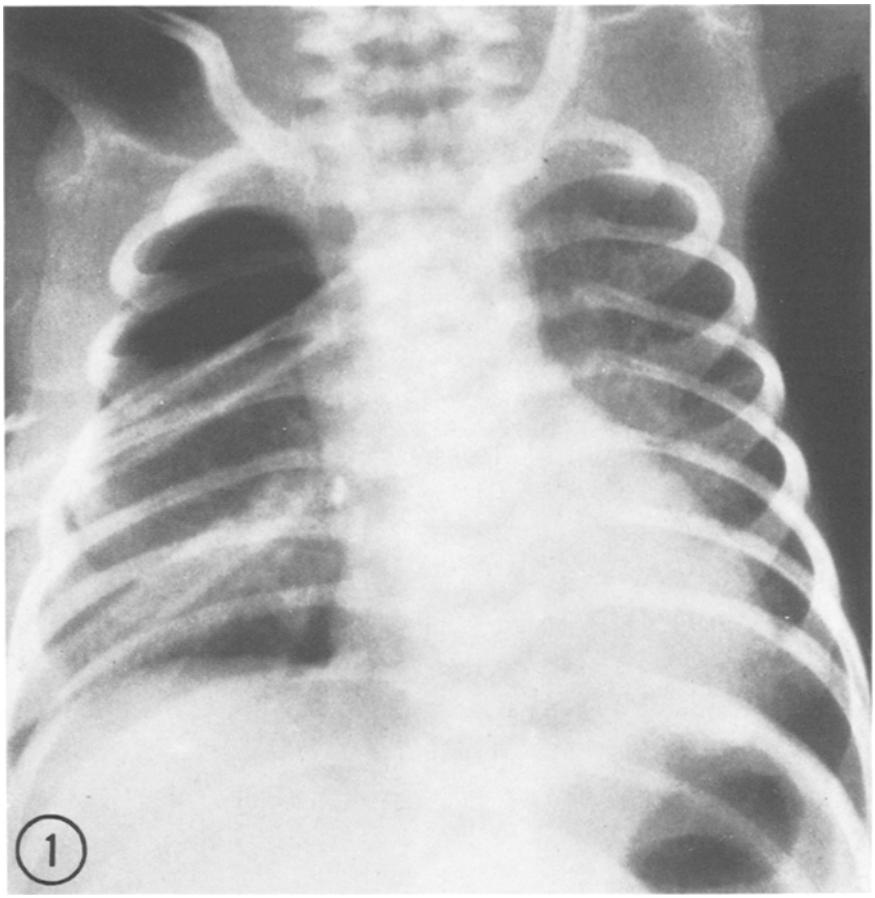

Fig. 1. Marked radiolucency was present in the right upper lobe region on chest X-ray

changes and pathogenesis of congital lobar emphysema are not clearly defined. He recommends that the excised lung be fixed in the inflated state and states that bronchial whole mount preparations are essential for proper study. The later technique, described by Stovin (1959) requires the dissecting out of short lengths of main segmental bronchi which are then incised along their length, opened out flat, stained with toluidine blue, dehydrated and mounted in thick Canada balsam. In some cases of congenital lobar emphysema, there is deficiency of cartilage in the lobar bronchus.

In other instances, such as the case to be described, excised lungs have remained hyperinflated even when the lobar bronchus is adequately patent. This implies a more generalized defect in the bronchial system and its investigation requires extensive dissection of the bronchial tree. If this is done a thorough histologic study of the bronchial tree can be made. In the following report each of the microscopic sections was photographed and the photomicrographs were reconstructed to resemble the original dissection. This method enables

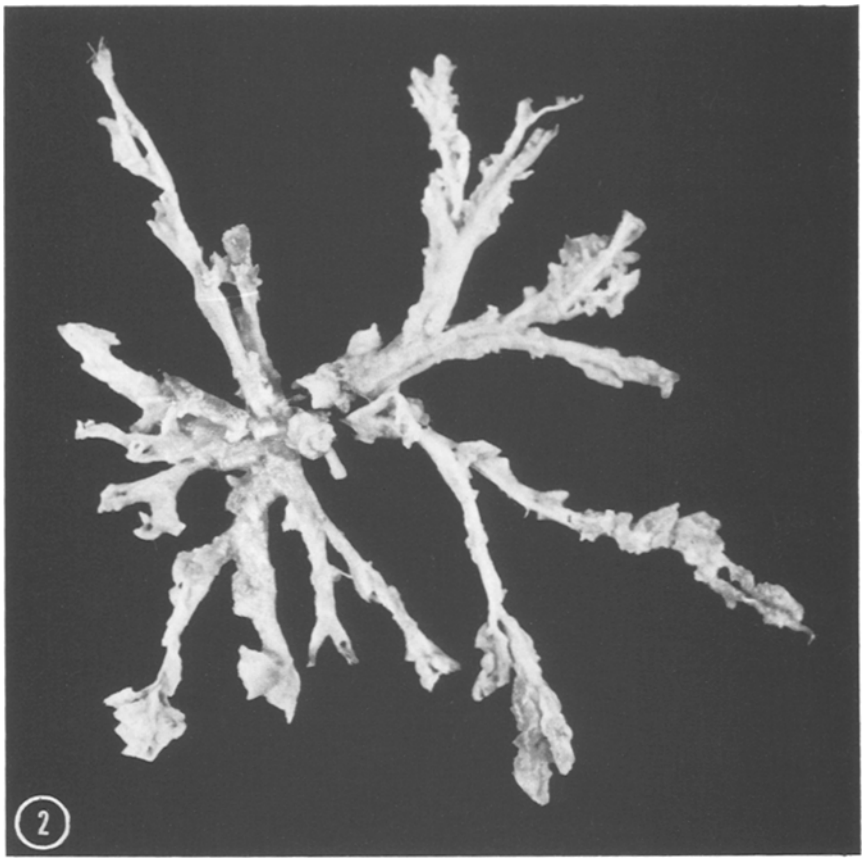

Fig. 2. The bronchial system was dissected out from the affected right upper lobe. Individual bronchi were abnormally flaccid. Magnification ×2.5

the pathologist to survey the distribution and organization of the cartilaginous elements.

Case History

A 10 day old premature infant developed respiratory distress with grunting and retraction of the thoracic cage. Pregnancy, labor, and delivery had been without complication. The child was born after 35 weeks gestation and weighed 4 lbs 6 oz at birth. The infant was noted to be a poor feeder and failed to gain weight during the first ten days of life. During feedings grunting and circumoral cyanosis were repeatedly observed. The examining physician described a pink, but small infant, active and tachypneic, with slight nasal flaring, and moderate suprasternal and infracostal retractions. The right chest appeared more expanded than the left. Normal breath sounds were heard over the left chest. Breath sounds were heard quite well at the right apex, not so well at the right base and poorly over the right mid-chest region. An X-ray (Fig. 1) revealed marked hyperaeration of the right upper lobe with herniation across the midline, compression atelectasis of the right middle lobe and part of the right lower lobe, and shift of the mediastinal contents

Fig. 3. Dissected bronchial tree from the right upper lobe of a (stillborn) mature infant. The montage has been reconstructed from micrographs of individual segments. The cartilage bars have been marked with India ink

towards the left side. Lobectomy was recommended. At operation the surgeons remarked that the right upper lobe bronchus was limp and atretic. Postoperative progress was satisfactory.

Method

A cannula was placed in the lobar bronchus and the lobe was inflated with 10% formalin siphoned from a receptacle which was placed 30 cms above the specimen as described by Whimster (1970). When suitably fixed it was examined under a dissecting microscope, and commencing with the right upper lobe bronchus the bronchial tree was gradually dissected away. A "control" right upper lobe was obtained during autopsy of a stillborn infant of mature gestation. After a similar dissection was performed, each bronchial tree was pinned out on cardboard and re-immersed in formalin. Later the specimens were photographed. From each of the photographic prints a map was made, and the bronchial tree divided into segments of suitable size for histologic preparation. After each individual segment was embedded in paraffin, 15 micron sections were cut and stained with hematoxylin and eosin. The sections were then photographed. From the prints the original dissection was reconstructed using the map previously described. The bronchial cartilages were marked with India ink and the montage was rephotographed. The "control" specimen was similarly processed.

Pathology

The resected specimen measured $5 \times 5 \times 3$ cm and was uniformly soft, crepitant, and pale pink. Following removal of a suture from the proximal end of the

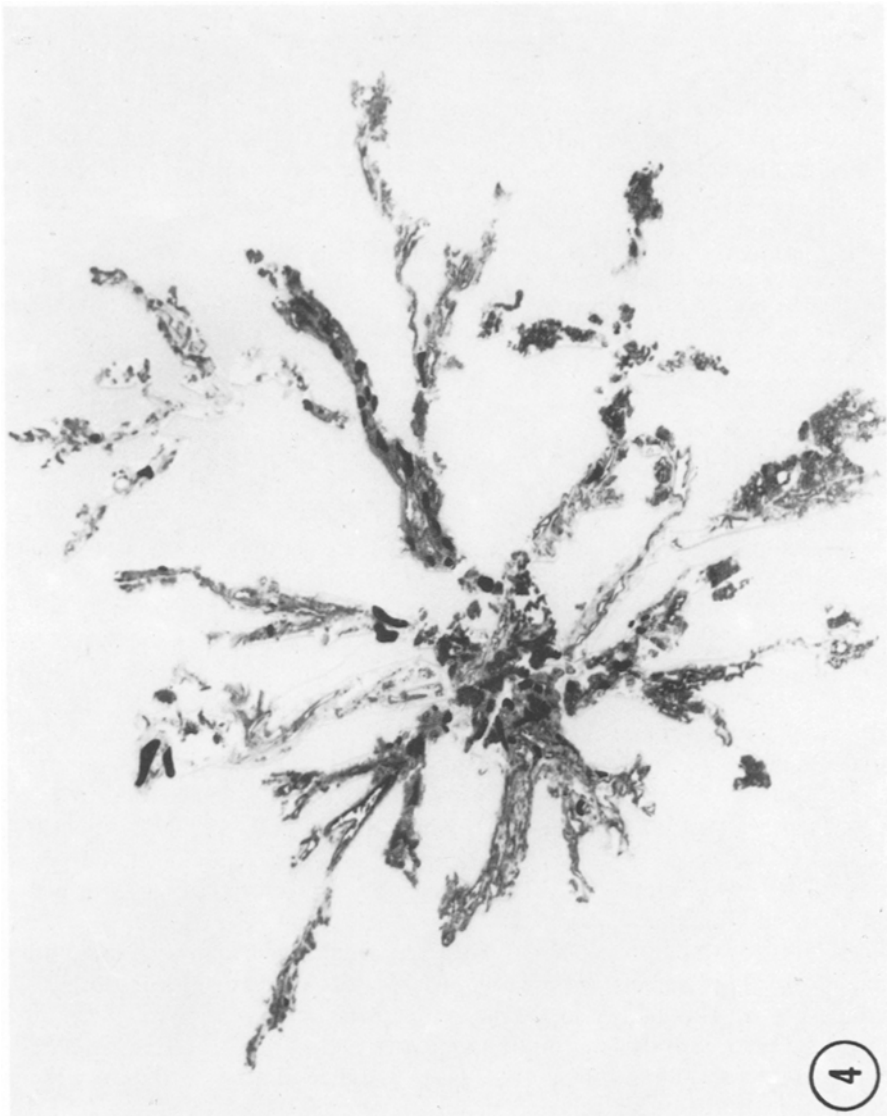

Fig. 4. Montage of photomicrographs of the affected bronchial tree from an infant with congenital lobar emphysema. Cartilage bars have been marked with India ink. Note the reduced size and abnormal orientation of the cartilage. Magnification ×3.5

right upper lobe bronchus, the bronchial lumen was observed to be patent but the lobe did not deflate. After dissection out of the fixed specimen, the bronchial system (Fig. 2) appeared limp and collapsed, unlike that from the control specimen. Microscopic examination of the pulmonary parenchyma revealed uniform marked distension of the right upper lobe alveoli and terminal bronchioles. Sections of the patient's bronchi (Fig. 4) showed that cartilage was present, however, when compared to the control (Fig. 3), the individual cartilaginous bars appeared to be irregularly shaped and their distribution within the bronchial tree seemed disorderly. Qualitatively, however, the cartilage from the affected lobe did not appear abnormal on microscopic examination.

Discussion

Histologic evaluation of the bronchial tree in an infant with congenital lobar emphysema suggests that maldevelopment of bronchial cartilage, resulted in a bronchial defect which permitted irreversible hyperinflation of the affected lobe. The method of examination described here may help in the study of future cases.

Lobar emphysema in infants has been ascribed to the following factors:

1. Vascular anomalies or other pathologic processes which cause external pressure on the bronchi (Lupascu et al., 1965);

2. Intrabronchial obstruction; i.e., mucous plugs (Harwitz et al., 1966; Murray et al., 1967), aspiration of barium Marie et al., 1965) and;

3. Absence or nondevelopment of cartilaginous rings, or redundant folds of bronchial mucosa giving rise to checkvalve mechanism (Horanyi et al., 1964).

However, Murray (1967) states that only cartilaginous deficiency has been so consistently present, 67% of the obstructive group, and thus, this anomaly warrants further study in the development of congenital lobar emphysema.

Overstreet (1939) was the first to suggest that congenital lobar emphysema could be the result of a cartilaginous defect. In the literature abnormalities of cartilage have been variously reported as: present throughout the lobe (Colton and Myers, 1957; Nelson, 1957), occurring as a focal lesion in a major bronchus (Nelson and Reye, 1954) or primarily involving the peripheral bronchioles (High and Arey, 1956; Shefts, 1961). Different authors describe the cartilage as being flaccid, immature, hypoplastic, deficient, or completely absent. Stovin (1959) who described the technique of bronchial whole mount preparations referred to "spidery cartilage—oriented longitudinally". Qualitative defects of cartilage have not been described in the literature by histologists, however, the histochemical studies of Binet and associates (1962) revealed a deficiency of chondroitin and diminished hyaluronic acid in the bronchial cartilage of children with congenital lobar emphysema.

Congenital lobar emphysema has been reported concurrent with extrapulmonary cartilaginous defects. Moore (1962) describes a patient with chondroectodermal dysplasia who had a bronchial malformation. Eiken (1961) reported on congenital lobar emphysema in a patient with chondrodystrophy. In a series of 23 reported cases, Janovski et al. (1969), cardiovascular deformities, either patent ductus arteriosus or ventricular septal defects, were found in 15% of patients. The patient reported here has been clinically diagnosed as having patent ductus arteriosus. Survey of the literature suggests that bronchomalacia sufficient to produce congenital lobar emphysema can occur despite the presence of significant amounts of cartilage which histologically is not dissimilar from normal cartilage.

In the present case the affected bronchial tree was uniformly flaccid on gross examination and the bronchial cartilage, though histologically unremarkable, was abnormally oriented. This correlates with the observations of Stovin (1959) suggesting that disorientation of bronchial cartilage may be relevant. Overstreet's theory of a cartilaginous defect is still the most tenable explanation in many cases of congenital lobar emphysema. The case reported here supports

this view, and the method of study may be of use in pathologic examinations of similar specimens in the future.

References

Binet, J., Nezeloff, C., Fredet, J.: Five cases of lobar tension emphysema. Dis. Chest. **41**, 126–133 (1962)

Colton, D., Myers, N.: Congenital lobar emphysema. Brit. med. J. **1957I**, 1394–1396

Eiken, M.: Congenital obstructive emphysema in infants. Acta pediat. (Uppsala) **50**, 17–24 (1961)

Gross, R., Lewis, J.: Defect of the anterior mediastinum. Surg. Gynec. Obstet. **80**, 549–554 (1945)

Harwitz, A., Selvage, I., Orbeton, E.: Hypertrophic lobar emphysema secondary to a paratracheal cyst in an infant. J. thorac. cardiovasc. Surg. **51**, 412–416 (1966)

High, R., Arey, J.: Lobar emphysema in infants. Amer. J. Dis. Child. **92**, 498–500 (1956)

Horanyi, J., Szots, I., Halasy-Nagy, F.: Über eine durch Knorpelanomalie verursachte hochgradige zystische Lungenblähung im Säuglingsalter. Thoraxchirurgie **11**, 710–713 (1964)

Janovski, N., Balacki, J., Keenan, F.: Congenital segmental lobar emphysema. Med. Annu. DC **38**, 249–254 (1969)

Luca, F. de, Wesselhoeft, C., Frates, R.: Congenital lobar emphysema documented by serial roentgenograms. J. Pediat. **82**, 859–862 (1973)

Lupaşcu, I., Barcan, F., Bărbuneanu, Niţeanu: Traiect aberant al arterei pulmonare stîngi cu enfizem consecutiv. Pediatrica **14**, 539–547 (1965)

Marie, I., Henednet, A., Durand, C.: Emphyseme lobaire chez un nourrison en rapport avec un pneumopathie a corps etrangers contenant du barium. Role d'une inhalation accidentelle de sulfate de barium après examen radiologie de l'estomac. Ann Pédiat. **12**, 722–729 (1965)

Moore, T.C.: Chondro-ectodermal dysplasia with bronchial malformation and neonatal lobar emphysema. J. thorac. cardiovasc. Surg. **46**, 1–10 (1963)

Murray, G.: Congenital lobar emphysema. Surg. Gynec. Obstet. **124**, 611–625 (1967)

Murray, G., Talbert, J., Haller, J.: Obstructive lobar emphysema of the newborn. Documentation of the "mucous plug syndrome" with successful treatment by bronchotomy. J. thorac. cardiovasc. Surg. **53**, 886–890 (1967)

Nelson, T.: Tension emphysema in infants. Arch. Dis. Childh. **32**, 38–41 (1957)

Nelson, T., Reye, D.: Tension emphysema: Surgical emergency in infants. Med. J. Aust. **2**, 342–343 (1954)

Overstreet, R.: Emphysema of a portion of lung in the early months of life. Amer. J. Dis. Child. **57**, 861–870 (1939)

Shefts, L.M.: Quoted in Campbell, D., Bauer, A., Hewlett, T.: Congenital localized emphysema. J. thorac. cardiovasc. Surg. **41**, 575–586 (1961)

Stovin, P.: Congenital lobar emphysema. Thorax **14**, 254–262 (1959)

Whimster, W.: Techniques for the examination of excised lungs. Human Pathology **1**, 305–314 (1970)

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