CONGENITAL LOBAR EMPHYSEMA, A HISTOPATHOLOGICAL STUDY

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ABSTRACT

Congenital lobar emphysema is a very rare congenital cystic malformation of the lung that can cause acute respiratory distress early in life. This study reviews 19 cases of CLE over a period of 5 years at a Tertiary Pediatric Referral Center. The cases were studied with special emphasis on Histopathological findings of the lobectomy specimens, particularly bronchial cartilage abnormalities, radial alveolar count and their predictive value in pathogenesis of CLE. This condition can mimic other causes of respiratory distress in the neonatal period. Early diagnosis and effective surgical management is curative.

KEYWORDS

CLE, Histopathology, Bronchial Cartilage, RAC.

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INTRODUCTION

CLE is an uncommon, but potentially life threatening cause of respiratory distress.⁽¹⁾ It is characterised by over distension and air trapping in the inflated lobe across the midline. CLE becomes evident usually in early infancy presenting with persistent progressive respiratory distress and is frequently fatal unless surgically treated.

CLE has no uniform etiology. It can be caused by extrinsic or intrinsic factors. Intrinsic factors include cartilaginous deficiency, bronchial stenosis, redundant bronchial mucosa or mucous plugs. Extrinsic causes include compression of bronchus from outside by vascular rings or lymph nodes.^{(2),(3)} Bronchial atresia and defective bronchial cartilage are common causes.^{(3),(4),(5)} Exact cause of CLE is unknown in 50% of the cases.⁽⁶⁾

We report our experience in 19 cases of CLE discussing their clinical features along with histology. Our study attempts to suggest etiopathogenesis of CLE through bronchial cartilage abnormalities and evaluate the extent of emphysema by Radial Alveolar Count.

MATERIALS AND METHODS

Nineteen surgically resected cases of CLE were studied over a period of 5 years at a Tertiary Pediatric Referral Hospital. The clinical history was reviewed and the x-ray and CT scan findings were correlated in all cases.

The microscopic analysis included a detailed histopathological examination of the slides for the following findings,

1. Acinar dilatation.
2. Size of the alveoli.
3. Bronchial atresia/stenosis.
4. Bronchial cartilage abnormalities.
5. Radial alveolar count.

Normal lung tissue of a perinatal autopsy case was used as control. RAC was counted by drawing a perpendicular from geometric centre of bronchiole to nearest connective tissue septum on 10 fields in each case.⁽⁷⁾

A photograph of the histopathology slide was taken and the measurements were done on a computer by drawing a perpendicular on the photograph. Ten fields were counted in each case and average was calculated.

RESULTS

Nineteen cases of CLE were diagnosed in the 5-year period. Significant male preponderance was noted with 13 out of 19(68%) cases being male children. Most of the patients presented in early infancy with 10 patients presenting between birth and 2 months of age. Earliest presentation was at 16 days and the oldest child was 20 months old. Most common lobe involved was left upper lobe in 9(75%) cases and the other lobes of left lung remained uninvolved in the study. Shortness of breath was the commonest clinical presentation in 47% of the cases. Cough, fever and feeding difficulties were the other chief complaints.

Cyanosis was seen only in one case, which was associated with congenital heart disease. X-ray findings showed hyperinflated lobe in 60% of the cases with associated mediastinal shift. CT scan confirmed the findings. Histopathological findings of dilatation of airspaces, the hallmark of any emphysema was seen in all the cases (Image 1). Alveoli were found to be 4-6 times enlarged in 7 cases and 8-10 times enlarged in 5 cases out of 19 (Table1). Stenotic bronchi were seen in 7 cases and dilated bronchi were noted in 3 cases. (Image 2) Coming to cartilage abnormalities, absence of bronchial cartilage was seen in 14 cases. Hypoplastic cartilage in 4 cases and normal looking cartilage plate in one case. (Image 3) RAC of <8 was seen in 8 cases >8 in 11 cases. (Image 4).

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Image 1: Acinar Dilatation in CLE

Image 2: Bronchial Stenosis and Bronchial Dilatation in CLE

Image 3: Hypoplastic and Absent Bronchial Cartilage in CLE

Image 4: The Radial Alveolar Count Method of Emery and Mithal

DISCUSSION

Congenital lobar emphysema is a very rare condition. Exact etiology of CLE is unknown. Several factors have been proposed, but congenital deficient cartilaginous support of the involved bronchus is present in half of the cases. Other rare identifiable causes include extraluminal and compression of the affected bronchus by abnormal blood vessel or congenital lung cyst, congenital bronchial stenosis or redundant mucosal flaps. All of this leads to ball valve effect that permits inflation of the effected lobe during periods of negative intrathoracic pressure, but collapses and obstructs effected bronchus during expiration. Ultimately, this leads to air trapping and over expansion of effected lobe of the lung.⁽⁸⁾

Congenital lobar emphysema typically effects children below 6 months of age and is more common in males than females. The youngest case of CLE presented at 16 days in our study and the oldest at 20 months of age. Most cases of CLE present very early, though few cases may present late with infections. Most of the patients were male. Left upper lobe was involved in 75% of the cases.

Presenting complaints, sex and age distribution are in line with all the previous literature.^{(9),(10),(11),(12),(13),(14),(15),(16)} Only one out of 19 cases had congenital heart disease. Left upper lobe was most commonest affected. Multilobar involvement was not seen in our study. Surgical resection of the hyperinflated lobe is the mainstay of treatment for CLE where respiratory symptoms are severe or progressive. Chest X-ray and CT scan are the usual preoperative investigations and show hypertranslucent lobe with attenuated vessels.

Bronchoscopy is performed for assessment of bronchomalacia, demonstrate impression of extrinsic mass and exclude endobronchial causes. Bronchoscopy was not performed in any of the cases in our study. Lobectomy was performed in all the cases. No deaths were reported postoperatively. Among the 5 variables taken for histopathological examination, dilatation of alveoli was seen in all the cases confirming the diagnosis of emphysema. Degree of alveolar enlargement correlated with age at presentation and degree of illness. Late presentation had greater degree of alveolar enlargement and vice versa. Coming to cartilaginous abnormalities absent cartilage was noted in 14 cases, hypoplastic cartilage was seen in 1 case.

Intrinsic obstruction may be due to cartilaginous deficiency or bronchial stenosis. Our findings were compared to a similar study by Stark et al. (Table 2). These histopathological findings point to etiological diagnosis. In 1960 Emery and Mithal introduced the radial count method, which assesses the complexity of the terminal respiratory unit (Acinus). Measurements in that study were made from a respiratory bronchiole to the edge of the acinus. A respiratory bronchiole was defined as a bronchiole lined by epithelium in one part of the wall.

From the centre of such a bronchiole, a perpendicular was dropped to the edge of the Acinus (Connective tissue septum or pleura), and the number of alveoli cut by this line was then counted. Ten such counts were made from each of 19 cases and the mean for each case was estimated. Results were expressed as average figures in different age groups. This study documented increase in RAC in inflated lobes, we attempted to evaluate the degree of raise in RAC in emphysema to assess the degree of emphysema. Higher RAC counts were noted and were significantly higher than expected age in younger infants. Normal development of the lung consists of an increase in number of alveoli from birth till adulthood. RAC therefore is more useful in younger infants.

The etiology of CLE is also attributed to increased number of alveoli within each lobe.⁽¹⁷⁾ Hence, increase in RAC is of predictive value in etiopathological diagnosis and suggests polyalveolar lobe.⁽¹⁸⁾

SUMMARY

Surgical treatment of CLE is total lobectomy. The histopathological parameters chosen for the diagnosis of congenital lobar emphysema were found to be very useful, adjunct to the routine description. Bronchial cartilage abnormalities and RAC predict etiopathogenesis and extent of emphysema. These 5 histopathological parameters should be included in the detailed histopathological report of CLE.

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