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A Case Series of Congenital Lobar Emphysema: Antenatal Diagnosis and Postnatal Management

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Abstract: ***Congenital lobar emphysema is a congenital abnormality caused by overinflation of a pulmonary lobe. This commonly presents in the antenatal and neonatal period. Respiratory distress in newborn poses a challenge and more so is the diagnosis of this condition, as one of its rare cause, only possible with high index of suspicion and appropriate investigations. Once diagnosis is made, treatment is by emergency lobectomy, and cure is possible in such cases. We present here 5 such cases in infants, which were treated by surgery.***

Keywords: Respiratory distress newborn, congenital lobar emphysema, lobectomy

1. Introduction

Congenital lobar emphysema (CLE) is a life-threatening congenital malformation leading to respiratory distress in early life and if undetected and treated can lead to severe morbidity and even mortality. Of late, the antenatal diagnosis of pulmonary malformations forms the significant group of diagnosis and the rest may present in perinatal or infantile period with respiratory distress or respiratory infections. Diagnostic modalities antenatally used are serial ultra sonography and MRI of foetus, after birth chest radiography and HRCT of chest are useful. The natural history of the disease is variable. Five newborns and infants in our study required emergency surgery. The risk of normal adjacent lung compression, infection and malignant degeneration making the resection necessary in these children.

Aims

To study the diagnostic and therapeutic challenges posed by Congenital lobar emphysema in prenatal and perinatal period

2. Material & Methods

Five cases of Congenital lobar emphysema with antenatal diagnosis were studied. The babies were antenatally & perinatally studied from period of antenatal diagnosis to 2 years after births All of them presented with poor feeding, intolerance to feeds, respiratory distress and two with cyanosis, were studied here. The antenatal presence of fluid filled lungs and mediastinal shift lead to confusion in the diagnosis. On examination at birth, air entry was reduced on affected sides with shift of mediastinum to the opposite side. Blood investigations were normal. Chest x - rays showed inappropriate findings of hyperinflation of left upper lobe in four cases and right upper lobe in one case, with mediastinal shift to opposite side & collapse of opposite upper lobe in these cases. To confirm this CT scan of chest was done in all cases, which showed hyperinflation affecting upper lobe with anterior mediastinal herniation of the affected lobe with mediastinal shift and compression atelectasis of ipsilateral lower lobe, and contralateral upper lobe in all the four cases. Echocardiography was done in all the 5 cases and was normal. All babies underwent posterolateral thoracotomy under general anaesthesia, and the affected lobe which classically popped out of the incised wound, was pale in colour and bulbous and was resected with vessels first and lastly the bronchus, closed with nonabsorbable sutures

3. Results

The mean operative time was two hours. One patient required packed cell volume transfusion intraoperatively. Two patients were put on ventilator support. One patient developed hypercarbia and took longer time to be weaned off ventilator support i. e, for 4 days. The histopathology in all the patients revealed changes of congenital lobar emphysema. The mean postoperative hospital stay was of ten days.

Postoperative period was uneventful, with feeds started on the next day, intercostal drainage tubes removed on day five to seventh day and discharged a day or two later. All patients recovered well. One patient had recurrent respiratory infections till 6 months of age. All other patients are doing well on follow up period.

4. Discussion

Congenital lobar emphysema is a rare congenital anomaly characterized by over inflation of a lung lobe with no functional continuity with ventilation, and is also called poly alveolar lobe.1 Air trapping in emphysematous lobe might be due to polyalveolar lobe, dysplastic cartilages, endobronchial obstruction or extrinsic compression.1 This condition is diagnosed before or at birth in 25% of cases, by 1 month in about 50% and sporadically after 6 mths.1 Male to female ratio is 2: 1, associated conditions include congenital heart disease commonly PDA, VSD in 15% of cases. The diagnosis of poly alveolar lobe disease may present a diagnostic challenge and needs a high index of suspicion in antenatal period with serial scanning and diagnosis of exclusion in some cases. At birth they present with respiratory distress and is sometimes confirmed. In one study of 10 patients, three patients were mistakenly diagnosed as pneumothorax and intercostal drains were inserted in the emergency department. Chest x - ray shows hyperlucent lobe with decreased bronchovascular markings with herniation and compression of the other lobe, opposite lung and mediastinal shift.2 CT scan was routinely advised in some studies, confirms the diagnosis by showing a hyperlucent, hyperexpanded lobe with substernal lobar herniation and compression of the

remaining lung & mediastinal shift to the opposite side.[2, 3, 4] MRI can be used to evaluate vascular supply and distribution to the involved lobe, but is not routinely employed, but diagnosis is certain when done antenatally and postnatally. USG can show a large, fluid - filled lobe with mediastinal shift or herniation of lung lobes beyond midline. A prenatal diagnosis of CLE is made by USG by increased echogenicity and reflectivity differentiating it from CCAM and absent systemic blood supply from BPS.5 Ventilation - perfusion scan can diagnose CLE by delayed contrast uptake and washout of xenon isotope from affected lobe and poor blood supply.1 CLE almost always involves one lung lobe; left upper lung lobe in 40 - 50%, right middle lung lobe in 30 - 40%, right upper lung lobe about 20% and lower lung lobes in around 1%[1]. Even though observation and conservative management is followed by few in asymptomatic cases the best treatment once diagnosis is certain is by emergency or planned thoracotomy and lobectomy of the affected lobe, and the outcome of surgery is good in most of the cases.[6, 7] Long term lung growth and function after lobectomy for congenital lobar emphysema is also good in these children.1

5. Conclusions

Congenital lobar emphysema presents a great challenge for early diagnosis in prenatal period, follow up till birth and treatment perinatally. Respiratory distress in a neonate or early infancy, should invoke one to rule out this disease, because if undetected may lead to severe morbidity and mortality and similarly to the contrary if diagnosed timely and operated upon, is a curative for life. These congenital lung malformations are now being increasingly diagnosed antenatally, hence need serial follow up, watchful waiting and may necessitate at most times the emergent surgical resections in the post natal period.

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