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Congenital Lobar Emphysema

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Abstract: ***Background****-Congenital Lobar Emphysema (CLE), Congenital lobar emphysema (CLE) is a rare malformation of lung development which may be the cause of respiratory insufficiency of the suckling child. It is caused by the hyper inflation of the lung lobe with compression of the normal lung parenchyma and contra lateral displacement of the mediastinum [1].Over distension and hyperinflation of the pulmonary lobe is secondary to partial bronchial obstruction [2]. Most common affected lobe are upper lobe followed by middle lobe but any lobe may be affected. There is inspiratory air entry but collapse of the narrow bronchial lumen during expiration. The bronchial defect results in lobar air trapping.*

Key words*: congenital lobar emphysema(CLE)atelectasis,hyperinflation,lobectomy,case report.*

I. Introduction

Congenital Lobar Emphysema (CLE) was first defined in 1932 by Nelson(1)is a rare developmental anomaly of the lower respiratory tract characterized by hyperinflation of one or more pulmonary lobes. Pressure effects to adjacent organs can result in severerespiratory distress in early infancy.. CLE is a rarely encountered congenital developmental anomaly of the lungs and it is characterized by compression of adjacent lung tissue and displacement of the mediastinum towards the opposite side together with hyperinflation of one lung lobe. The prevalence of congenital lobar emphysema has been reported to be 1/20,000-30,000 (2). Airway obstruction is the most common cause of the disease, but the cause of the disease cannot be detected in 25% of the patients. It manifests with progressive dyspnoea and tachypnea a few days after birth in more than half of the cases (3-5). It is more prevalent among males (6) with an M/F ratio of 3/1. A consolidation area is seen on the chest x-ray of new-borns with respiratory distress, performed just after birth. The diagnosis is usually made clinically, radiologically and scintigraphycally. The traditional treatment of the disease is surgical (lobectomy) (7); however, conservative treatment is also being recommended in the recent years (8). A few cases, which were diagnosed at adult age, have been reported in the literature (5). Herein, we presented a patient with congenital lobar emphysema diagnosed in adult age, in which bronchial asthma treatment was started, but replaced with anxiolytic medications as asthma treatment provided no benefit.

II. Case Presentation

A 2 month old female baby weighing presented in pediatric opd with difficulty breastfeeding and fatigability since two days. Mother also noticed that the baby was fast breathing. Antenatal ultrasound scan was done at 34 weeks which revealed cystic , septate lesion in right hemithorax s/o congenital cystic adenomatous malformation.General examination was normal. There was no malnutrition. Tachypnoea was present, other vitals were stable. Respiratory examination showed mild tachypnoea,symmetrical chest expansion, Apex beat was felt at left 4th intercoastal space at mid clavicular line, bilateral subcoastal retractions, normal vesicular breath sounds with decreased breath sounds heard in upper zone of leftlung with no adventitious sounds. Other systemic examination was normal.Right Posterolateral Thoracotomy was performed for right upper lobe congenital lobar emphysema. The massive emphysematous lobe was compressing the middle and lower lobes and had separate blood supply. Lobectomy was performed. The procedure went uneventful.

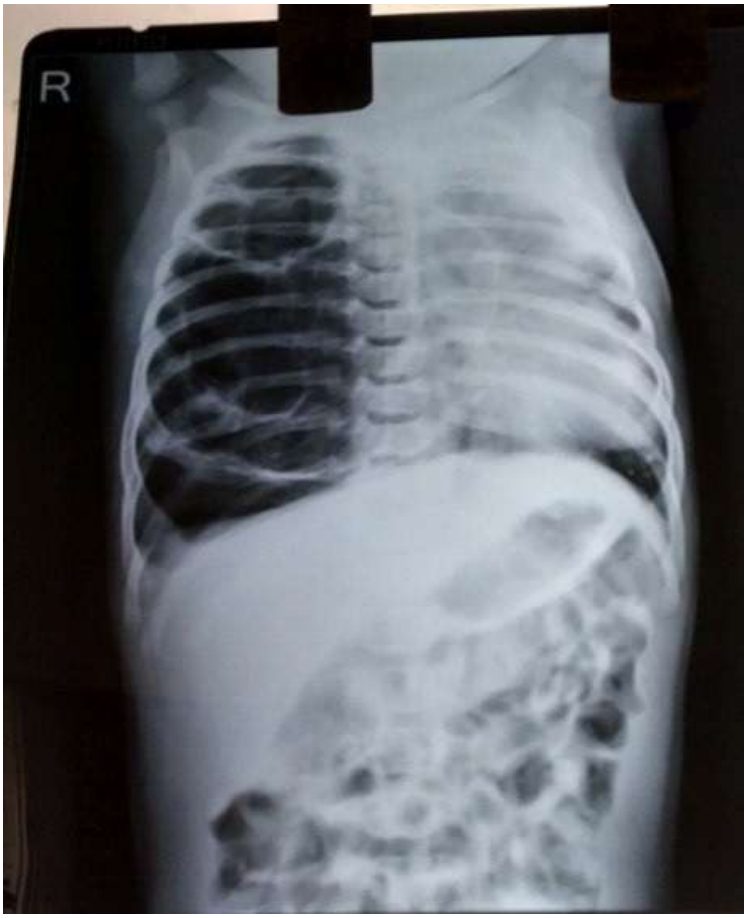

Figure.1 chest xray revealing increased radioluency of right lung lobe with mediastinal shift.

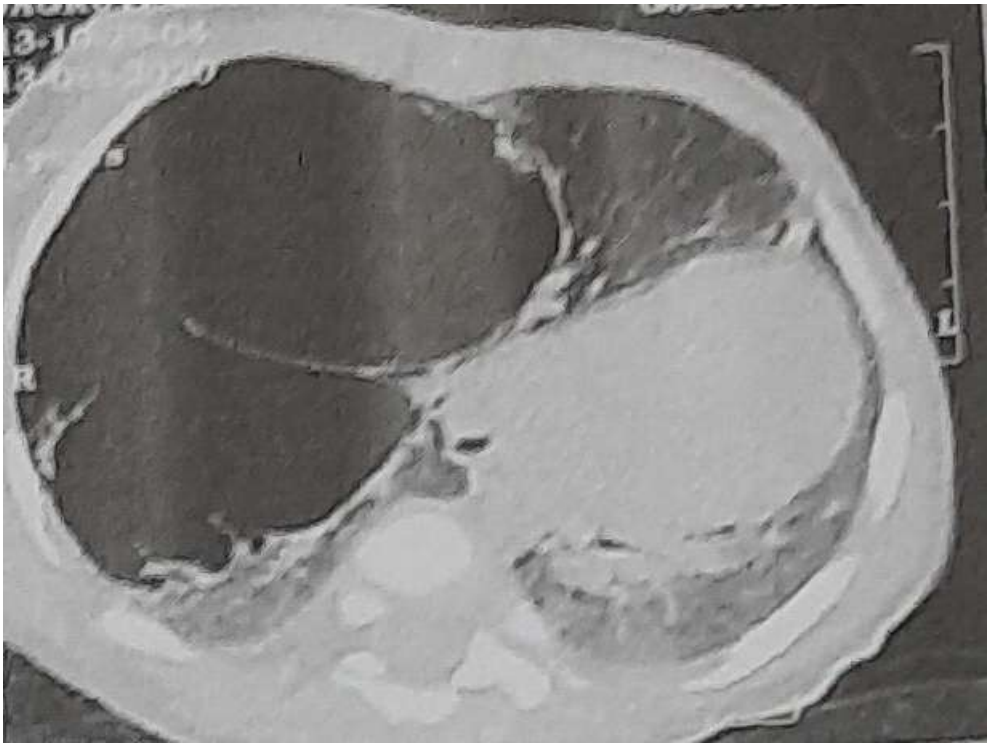

Figure.2 CT scan showing severe bullous changes in the right upper lung with mediastinal shift

All the routine blood investigations were normal.Radiographic images of chest showed hyperlucency of right upper Zone with herniation of right upper lobe tracheal shift and mild shift of the mediastinum to the left. CT scan showed Emphysema of right upper lobe of lung Management- With right thoracotomy approach, lobectomy was done. Surgery went uneventful.Appropriate post-operative care was given. Infantrecovered well with good left lung expansion.

III. Discussion

CLE is a rare disease of the lung with an incidence of about 1 per 20,000 to 30,000 births (4). Low incidence and co-infection often cause misdiagnosis and delayed treatment. The left upper lobe and the middle lobe are the most commonly affected lobes, followed by the right upper lobe and the right lower lobe, and CLE affecting two ipsilateral lobes is extremely rare. The etiology of 50% CLE remains unclear (5). Congenital cartilage defect accounts for 1/4 of the causes and is also the most common cause (2). It is presumed that the bronchial cartilage defect may lead to excessive inflation and air trapping on expiration due to the ball-valve effect. This causes lung lobar hyperinflation (5). The clinical manifestations are mainly respiratory dysfunction and recurrent pulmonary infection. The age at the onset of respiratory symptoms varies considerably, ranging from neonatal period to adulthood. Respiratory dysfunction is predominant within 6 months after birth; however, it is also reported in patients older than 6 months J1 . Adult patient may be asymptomatic (7) and are often diagnosed with CLE incidentally. Recurrent pulmonary infection is the most common manifestation in children and adults. CLE usually has no specific clinical manifestations, which poses diagnostic and therapeutic dilemmas(3) . In clinical practice, it should be differentiated from many other lung diseases. Prenatal ultrasonography (1) and magnetic resonance imaging can be used to differentiate CLE from other congenital pulmonary lesions in the uterus at an early stage, such as bronchopulmonary sequestration, congenital cystic adenomatoid malformation, congenital diaphragmatic hernia, and bronchogenic cyst . Some studies have reported that chest X-ray facilitates the diagnosis of CLE (2). The CLE on chest X-ray presents hyperinflation of the affected lobe with the compression of normal lung parenchyma and the displacement of contralateral mediastinum. Regardless, it is often confused with compensatory emphysema caused by atelectasis or pneumothorax based on clinical presentations, which affects its diagnosis and then delays its treatment in the early stage(4). Chest CT is important for confirmed diagnosis (2,10). The flexible bronchoscopy can be used to evaluate for alternative causes of lung hyperinflation including vascular compression, foreign bodies, inspissated mucus, airway anatomical anomalies and endobronchial tumors. Bronchoscopy also can be used to exclude foreign body aspiration and identify candidates for conservative treatment. The resection of the affected lung lobe is recommended for children with severe respiratory symptoms. Surgery is also safe for patients with cardiac dysfunction . The earlier the presentation is, the greater is the need for surgery(4) . The mortality is low in children with CLE undergoing surgery . Asymptomatic patients or patients with mild respiratory symptoms in older children should be closely monitored and receive conservative treatment (2). Eigen et al. found that lung growth and function in a small group of children with CLE treated conservatively were no different from that of children treated surgically by lobectomy(7).

IV. CONCLUSION

The high index of suspicion is required to diagnose congenital lobar emphysema and it should always be considered as a differential diagnosis in neonates on infants presenting with respiratory distress. As both surgical and conservative management are possible,protocol should be individualised according to the clinical presentation and status of the child along with status of the family.

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