Congenital lobar emphysema presenting at late childhood: A rare case report

Sir,

Congenital lobar emphysema (CLE) is a rare congenital abnormality that is characterized by overdistension and air trapping of one or more lobes of lung, leading to a compression atelectasis of ipsilateral lung and subsequently of contralateral lung. Similar to other developmental lung anomalies, this condition is usually diagnosed in the neonatal period or during early childhood. However, we report a case of CLE that was detected in late childhood.

A 15-year old boy presented with progressive exertional breathlessness since 5 months. There was no other significant history. Patient denied any history of recurrent chest infection. According to his mother, the achievement of developmental milestones was normal but he did not gain adequate weight. His immunization history was up-to-date. On examination, he was of thin built with 13.7 kg/m^2 BMI. There was no cyanosis, clubbing, oedema, icterus, or lymphadenopathy, although pallor was present. Vitals were within the normal limit. On inspection, right hemithorax appeared enlarged. Movement of chest was restricted on the right side with mediastinum shifted to the left. Breath sound, vocal fremitus and vocal resonance were grossly reduced on the right side along with a resonant percussion note. Liver was palpable at 4 cm below the costal margin. A clinical diagnosis of right-sided pneumothorax was made on admission.

But chest X-ray in PA view [Figure 1] revealed hyperlucency of the entire right lung with leftward mediastinal shift without any visible visceral pleural lining. Attenuated but maintained vascularity was noted in the hyperlucent right lung. ABG analysis and other blood investigations were normal except Hb-10.4 gm/dl. Electrocardiography showed only left axis deviation without any evidence of left ventricular hypertrophy. Two-dimensional echocardiography did not reveal any cardiac abnormality. Spirometry showed a mixed ventilatory pattern with poor bronchodilator reversibility. Computed tomographic scan of thorax [Figure 2] showed hyperlucent and hyperexpanded right middle and lower lobe with collapsed right upper lobe along with mediastinal shift to left. Pulmonary vasculature on the right side appeared attenuated and displaced. Herniation of lung tissue was noted in retrocardiac space and in anterior mediastinum. The left lung was normal except for compression atelectasis. No evidence of pneumothorax or intraluminal foreign body was found. Subsequent fibreoptic bronchoscopy [Figure 3] showed a hypoplastic right upper lobe bronchus along with a normal trachea and left main bronchus.

Thereby, a final diagnosis of congenital lobar emphysema of right middle and lower lobe with right upper lobe bronchial hypoplasia was made on the basis of radiological and bronchoscopic findings. Patient was treated conservatively because of clinically less severe disease. The patient had been advised for a close monitoring and regular follow-up. He is currently being followed up since last one year and no clinical deterioration has been noted so far.

CLE is an uncommon congenital anomaly of respiratory system with a prevalence of one per 20,000 to 30,000 deliveries and the incidence is estimated to be one in 70,000

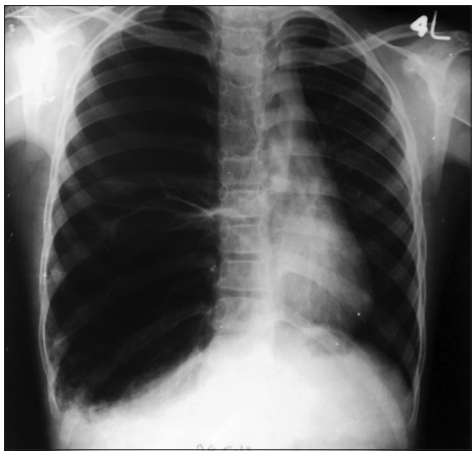

Figure 1: Chest X-ray shows hyperlucent and hyperexpanded right lung with mediastinal shifting to the left side along with attenuated but maintained vascularity of the right lung

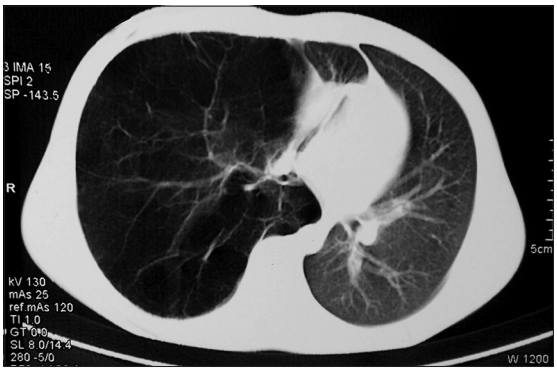

Figure 2: CT scan of thorax showing marked overinflation of right middle and lower lobe along with collapsed right upper lobe with mediastinal shifting to the opposite side. There is attenuation of vascularity on the right side and herniation of right lung tissue to anterior mediastinum and retrocardiac space

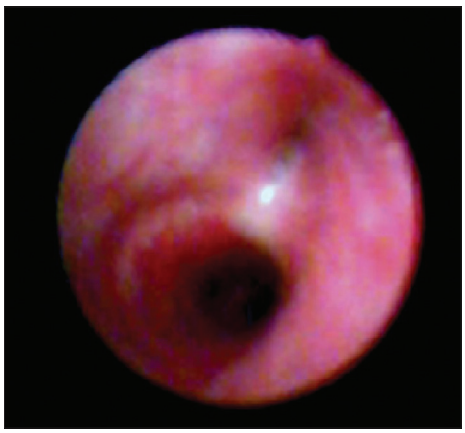

Figure 3: Fibreoptic bronchoscopy showing hypoplasia of right upper lobe bronchus, visible as a slit-like opening

to 90,000.[1] This disorder mostly presents within few hours after birth to six months. Usually CLE involves a single lobe; however, multilobar as well as bilateral involvement has also been recorded.[1] The most common affected lobe is the left upper lobe, followed by the right upper lobe and right middle lobe.[2] But multilobar involvement including a lower lobe is very uncommon and no such case was recorded among 30 cases as mentioned in a case series by Ozçelik *et al*.[2] Such multilobar involvement (including lower lobe) was found in only three out of 28 cases as mentioned in another case series by Lincoln *et al*.

Breathlessness is the most common symptom associated with cough, wheezing, grunting or recurrent infection. Patients who present later in life usually have history of recurrent chest infections and frequent antibiotic use, which was not the scenario in this case. Exact aetiology is not known and no apparent cause is found in 50% cases.[3] Congenital cartilage defect is found in 25% of cases that causes bronchial collapse at expiration, leading to overinflation of the alveoli. Other bronchial obstructions (redundant mucosal fold, mucus plugging, bronchial hypoplasia/stenosis etc.) are responsible for remaining 25% of cases. In our patient, although hypoplastic bronchus was present in association with emphysematous right middle and lower lobe, it was not related causally and we could not find such an unusual association in the literature.

Clinically CLE may mimic pneumothorax but they can be differentiated from the chest X-ray and CT finding. This differentiation is absolutely essential as because insertion of an intercostal tube in CLE cases in suspicion of pneumothorax may be deleterious to the patient. Bronchoscopy is done to look for any congenital or acquired tracheobronchial abnormality and to exclude foreign body obstruction. Cardiological evaluation must be done as concomitant congenital heart disease and CLE has been found in 12-20% cases in the literature.

Treatment of CLE is mainly surgical. Lobectomy is the standard surgical procedure that allows expansion of compressed normal lung tissue. Long-term outcome was found to be excellent with complete cure in over 85% of cases.[4] Recently endoscopic parenchymal sparing resection has been performed in a case of CLE with mild symptoms.[5] But the conservative approach has also been tried among patients with milder disease presenting later in life. As several literatures described the successful conservative approach with stringent follow-up, we opted for conservative management in our patient as his symptoms were less severe.[6] Also right middle and lower lobectomy was unlikely to produce any improvement in our patient as he had hypoplastic right upper lobe bronchus and atelectasis of right upper lobe was not due to compression.

In conclusion, CLE presenting in late childhood or adulthood for the first time may lead to a diagnostic challenge to the physician and it can be detected only if a high diagnostic suspicion is maintained. The diagnosis and its differentiation from pneumothorax are important, so that unnecessary intervention for pneumothorax can be avoided and the patient can be taken up for a more definitive surgical management. Asymptomatic and less severe patients may be considered for a non-operative conservative approach with strict advice regarding regular follow-up.

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